MAKING THE GRADE: INVESTIGATING ACADEMIC OUTCOMES OF CHILDREN WITH CLEFT LIP AND PALATE

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DISCLOSURE STATEMENT

- Neither I nor any member of my immediate family has a financial relationship or interest with any proprietary entity producing health care goods or services related to the content of this CME activity.
- My content will not include discussion/reference of any commercial products or services.
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THANK YOU CINDY

OBJECTIVES

- Illustrate trends in academic outcomes for children with orofacial clefts
- Evaluate potential factors that contribute to observed academic deficits
- Propose an intervention to improve outcomes

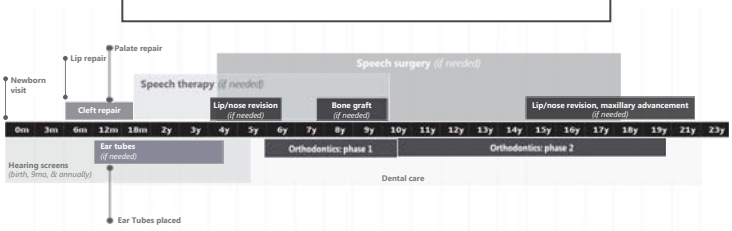
OROFACIAL CLEFTS: THE BASICS

- CLP: 1 in 700 births
- CP: 1 in 2000 births
- Genetic and environmental factors
- Isolated vs Syndromic

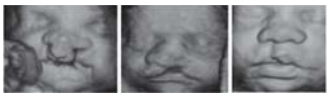
Functions of the palate

- FEEDING
- HEARING
- SPEECH

TIMELINE OF CARE



PRENATAL DIAGNOSIS



Will I feel comfortable taking my baby in public before lip repair?

Will my baby have other problems?

Will other people bully my child because of the cleft?

Is this because of something I did wrong?

Does this mean my child will have learning problems?

IMPACTS OF OROFACIAL CLEFTS

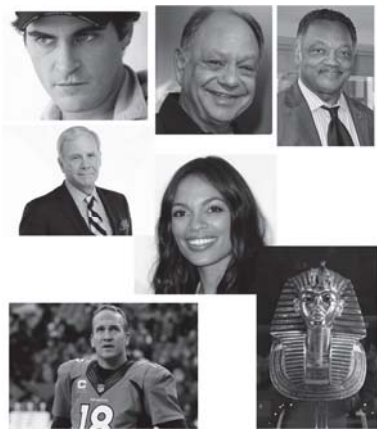
"There appears to be an innate human tendency to associate craniofacial malformations with abnormal cognitive development."

Cunningham, 2007

ACADEMIC OUTCOMES

Key Point:

This does **not** mean that every child with an orofacial cleft will have academic deficits.

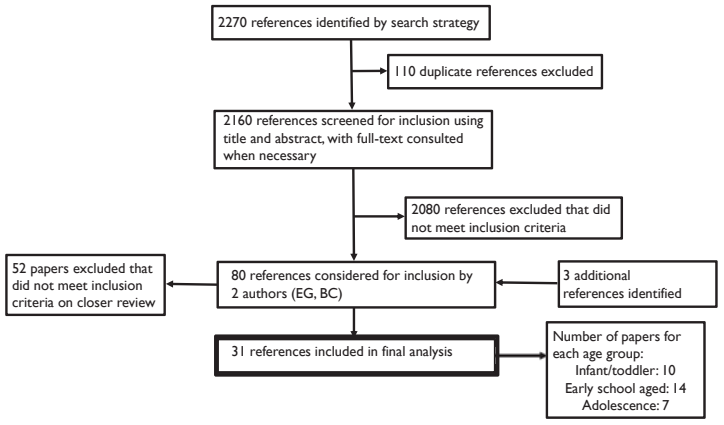


SYSTEMATIC REVIEW

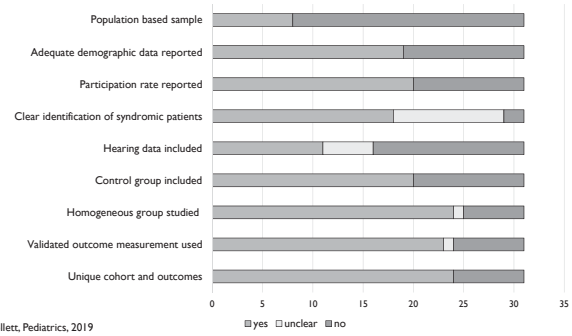
- Search strategy
  - Medline, Embase, PsychInfo, CINAHL
  - 1980-2017
- Search terms
  - Cleft palate, cleft lip, orofacial cleft
  - Terms to include neurodevelopmental and academic outcomes

SYSTEMATIC REVIEW

- Inclusion
  - Patients <25 years with orofacial clefts
  - Measures of neurodevelopmental outcomes
  - English language
  - Middle to high income economies
- Exclusion
  - <10 cases
  - Qualitative studies



SYSTEMATIC REVIEW: QUALITY SCORES



Gallagher and Collett, Pediatrics, 2019

SYSTEMATIC REVIEW: SUMMARY OF FINDINGS

	INFANT	TODDLER
AVERAGE	EXPRESSIVE LANGUAGE	RECEPTIVE LANGUAGE (CLP) EXPRESSIVE LANGUAGE (CLP) 2-WORD PHRASES
AVERAGE RANGE, BELOW CONTROLS		DUTCH BAYLEY
BELOW AVERAGE	MOTOR SKILLS RECEPTIVE LANGUAGE BAYLEY	EXPRESSIVE LANGUAGE (CP) RECEPTIVE LANGUAGE (CP) BAYLEY FINE MOTOR

SYSTEMATIC REVIEW: SUMMARY OF FINDINGS

	INFANT	TODDLER	EARLY SCHOOL
AVERAGE	EXPRESSIVE LANGUAGE	RECEPTIVE LANGUAGE (CLP) EXPRESSIVE LANGUAGE (CLP) 2-WORD PHRASES	WORKING MEMORY EARLY READING IQ (CP, uCLP)
AVERAGE RANGE, BELOW CONTROLS		DUTCH BAYLEY	EARLY READING
BELOW AVERAGE	MOTOR SKILLS RECEPTIVE LANGUAGE BAYLEY	EXPRESSIVE LANGUAGE (CP) RECEPTIVE LANGUAGE (CP) BAYLEY FINE MOTOR	IQ (bCLP) READING COMPREHENSION SCHOOL ATTENDANCE GRADES STANDARDIZED TEST SCORES SPECIAL EDUCATION LEARNING DISABILITIES

SYSTEMATIC REVIEW: SUMMARY OF FINDINGS

	INFANT	TODDLER	EARLY SCHOOL	ADOLESCENT
AVERAGE	EXPRESSIVE LANGUAGE	RECEPTIVE LANGUAGE (CLP) EXPRESSIVE LANGUAGE (CLP) 2-WORD PHRASES	WORKING MEMORY EARLY READING IQ (CP, uCLP)	SCHOOL ATTENDANCE
AVERAGE RANGE, BELOW CONTROLS		DUTCH BAYLEY	EARLY READING	IQ
BELOW AVERAGE	MOTOR SKILLS RECEPTIVE LANGUAGE BAYLEY	EXPRESSIVE LANGUAGE (CP) RECEPTIVE LANGUAGE (CP) BAYLEY FINE MOTOR	IQ (bCLP) READING COMPREHENSION SCHOOL ATTENDANCE GRADES STANDARDIZED TEST SCORES SPECIAL EDUCATION LEARNING DISABILITIES	SUSTAINED ATTENTION GRADES STANDARDIZED TEST SCORES GRADUATION CERTIFICATE SPECIAL EDUCATION LEARNING DISABILITIES GRADE RETENTION

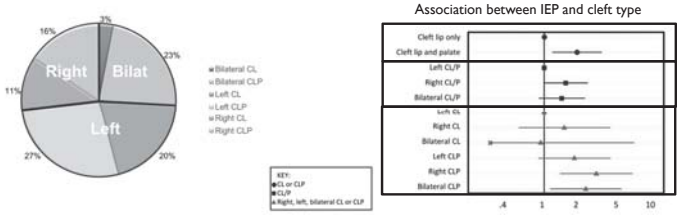
SYSTEMATIC REVIEW

- Quality of studies was variable
- Several high quality studies clearly show academic deficits
- Deficits were present in a range of domains and ages
- Future studies should include more rigorous review of participants
- Children with orofacial clefts are at risk for neurodevelopmental deficits and should be monitored and supported

ACADEMIC DEFICITS

- Evaluating neurodevelopmental outcomes is complex
- Functional and psychosocial impacts of orofacial clefts
- Many potential factors
  - Intrinsic
  - Extrinsic

INTRINSIC DIFFERENCES



Gallagher et al. Associations between laterality of orofacial clefts and medical and academic outcomes. American Journal of Medical Genetics. 2017.

PARENTAL BONDING

- Normally, adult gaze focuses on an infant's eyes before 6 weeks, then includes more time on the mouth when infant starts to vocalize.
- Maternal eye contact predicts mother-infant relationship 1 year later and has been linked to developmental outcomes.
- Maternal gaze was shifted when infant had a cleft lip
  - Gaze towards infant's body
  - Gaze towards facial areas other than eyes or mouth.

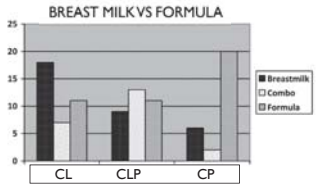
DePascalis et al. Maternal gaze to the infant face: Effects of infant age and facial configuration during mother-infant engagement in the first nine weeks. 2017, Infant Behavior and Development.

FEEDING/NUTRITION

- Does breastfeeding have a positive impact on cognition and behavior for children?
  - Nutritional benefits support neural maturation and may impact language development
  - Some studies found better neurodevelopmental outcomes after exclusive breast milk feeding
  - More recent studies have been less clear
  - Skin-to-skin contact may help with bonding and subsequently behavior

AAP policy statement, 2012
Girard et al. Breastfeeding, Cognitive and Noncognitive Development in Early Childhood: A Population Study. Pediatrics, 2017.

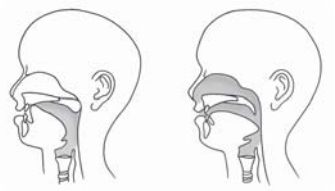
FEEDING/NUTRITION



- 5 year retrospective review of children with cleft palate
- Breast milk feeding (ever) was 29.5%
- CDC report = 81%
- Lower z-scores for weight and weight for length

Kaye et al. Initial Nutritional Assessment of Infants With Cleft Lip and/or Palate: Interventions and Return to Birth Weight; Cleft Palate-Craniofacial Journal, 2017.
Gottschlich et al. A Retrospective Study Identifying Breast Milk Feeding Disparities in Infants with Cleft Palate; Journal of the Academy of Nutrition and Dietetics, 2018.

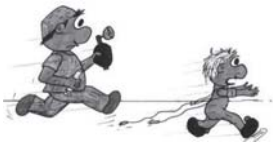
SPEECH OUTCOMES



VELO:
VPI Effects on Life Outcomes

- Speech Limitations
- Swallowing problems
- Situational difficulty
- Emotional impact
- Perception by others
- Caregiver impact

EXPOSURE TO ANESTHESIA



- In non-cleft populations, few clear differences have been identified in developmental outcomes after anesthesia.
- Danish study: neurodevelopmental outcomes of CL, CLP, CP
- CL had higher scores, CP lowest scores
- Cleft type, not number of surgeries, was associated with lower outcomes.

Hu et al. Association between Exposure of Young Children to Procedures Requiring General Anesthesia and Learning and Behavioral Outcomes in a Population-based Birth Cohort. Anesthesiology. 2017.
O'Leary et al. Influence of Surgical Procedures and General Anesthesia on Child Development Before Primary School Entry Among Matched Sibling Pairs. JAMA Pediatrics, 2018.
Sun et al. Association Between a Single General Anesthesia Exposure Before Age 36 Months and Neurocognitive Outcomes in Later Childhood. 2017, JAMA.
Clausen et al. Oral Clefts and Academic Performance in Adolescence: The Impact of Anesthesia-Related Neurotoxicity, Timing of Surgery, and Type of Oral Clefts. 2017, Cleft Palate-Craniofacial Journal.

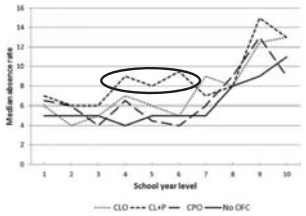
SCHOOL ABSENCE

Population-based cohort in Western Australia

400 cases, 1800 controls

Quantifying school absence for children with orofacial clefts

Impact of school absence on test scores



- Higher absence rates for CLP in grades 4-6
- No difference in high school
- Higher absences associated with lower standardized test scores
- Children with CP had lower scores regardless of absence rates

Bell et al., School Absence and Its Effect on School Performance for Children Born with Orofacial Clefts; Birth Defects Research, 2017.

PSYCHOSOCIAL OUTCOMES

Psychological adjustment to cleft lip and/or palate: A narrative review of the literature

Nicola Marie Stock[a]* and Kristin Billaud Feragen[b]

[a]Centre for Appearance Research, University of the West of England, Bristol, UK; [b]Senter for sjeldne diagnoser (Centre for Rare Disorders), Oslo Universitetssykehus HF, Oslo, Norway

(Received 8 September 2015; accepted 12 January 2016)

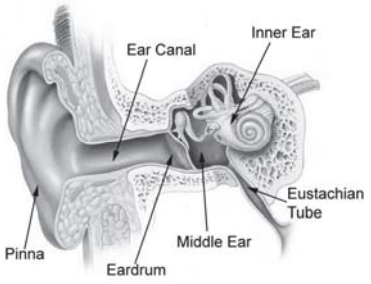
- Narrative review of 148 quantitative and qualitative studies, 2004-2015
- 5 domains of adjustment:
  - Developmental trajectory
  - Behavior
  - Emotional Well-being
  - Social Experiences
  - Satisfaction with Appearance and Treatment
- Contradictory results in all areas but overall impact of cleft seems low

HEARING LOSS

Conductive hearing loss

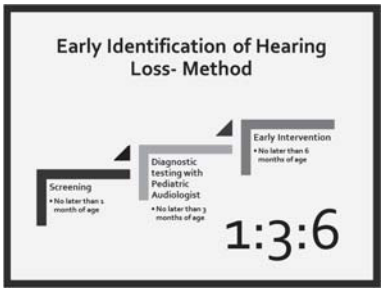
Chronic middle ear effusions

Chronic otitis media

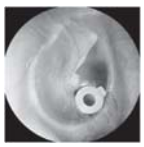


Early Hearing Detection & Intervention

a program of the American Academy of Pediatrics



HEARING LOSS



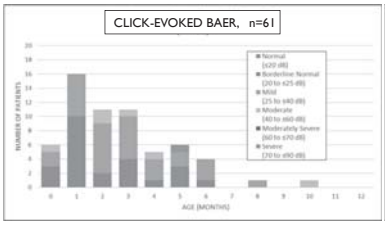
Tubes placed with palatoplasty

What is the degree of hearing loss before palate repair for infants with cleft palate?

- Retrospective chart review
- Cleft palate ± cleft lip
- DOB 2008-2015
- Palate repaired at SCH before age 3 years
- Audiograms in AudBase

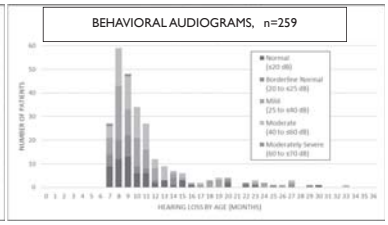
DEGREE OF HEARING LOSS BY AGE

CLICK-EVOKED BAER, n=61



49.2%: mild hearing loss
11.5%: moderate or greater hearing loss

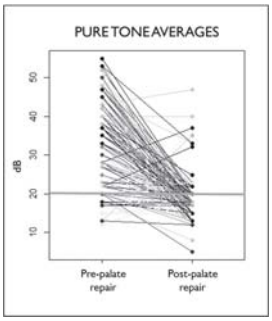
BEHAVIORAL AUDIOGRAMS, n=259



32.0%: mild hearing loss
30.5%: moderate or greater hearing loss

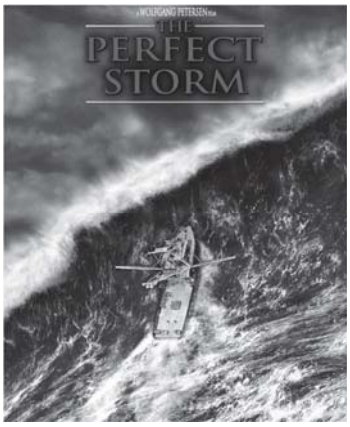
Gallagher et al. In progress.

HEARING AFTER PALATE REPAIR



4.6%: mild or greater hearing loss

Gallagher et al. In progress.



Potential targets for intervention:

- Parental bonding
- Breastmilk feeding
- Hearing loss

- Home language environment

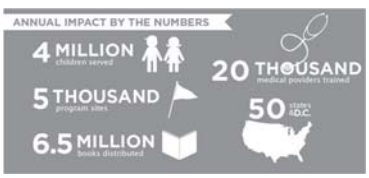
INTERVENTION STUDY

Can we change the home language environment?

INTERVENTION STUDY

Can Reach Out and Read be used to positively impact the Home Language Environment?

Reach Out & Read®
where great stories begin®



www.reachoutandread.org

EVIDENCE FOR REACH OUT AND READ

- Improves home literacy environment
  - Frequency of shared reading
  - Availability of books in the home
  - Reading becomes a favorite shared activity
- Increases scores on testing
  - Receptive and expressive language
  - Literacy scores at school entry
  - Low socioeconomic settings
  - English and non-English-speaking children

CRANIOFACIAL REACH OUT AND READ

- 2012: partnered with national ROR
- Developed a list of books by age and specific speech sounds
- Follow ROR model but also demonstrate how to use books to practice speech

MEASURING THE HOME LANGUAGE ENVIRONMENT

LENA device

- 16 hours of recordings at home
- Sorts child vs others, TV, radio
- Software analyzes and provides
  - Adult Word Count
  - Child Vocalizations
  - Conversational Turns

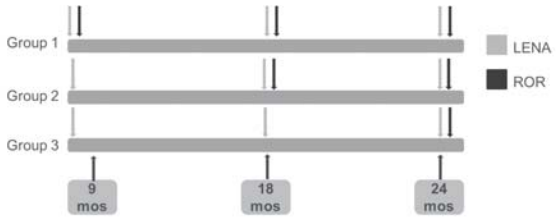
LENA ROR STUDY

- Feasibility study
  - Recruitment
  - Protocol implementation
- Study population
  - Goal: 60 children with clefts
  - 9 months (±2 months)

LENA ROR STUDY

- Inclusion
  - CL, CLP, CP
  - SCH Craniofacial Center
  - English or Spanish-speaking
- Exclusion
  - Syndrome with known delays
  - Brain malformation, seizure
  - Profound hearing loss
  - Hypotonia
  - Hospitalized >6 weeks
  - State custody, adopted

STUDY DESIGN



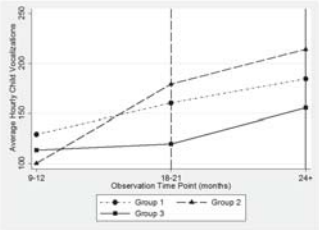
BASELINE CHARACTERISTICS OF PARTICIPANTS

- 78 approached, 27 enrolled
- Consent rate: 35%

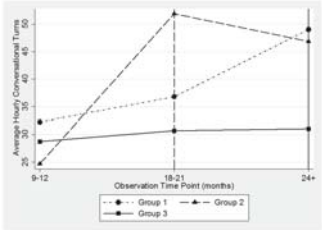
- Combined recordings with others from a different study to increase pre-intervention recordings

Characteristic	N or mean	% or SD
Total	38	100.0
Age, months	11.5	4.4
Cleft type		
Cleft lip	9	23.7
Cleft palate	14	36.8
Cleft lip and palate	15	39.5

CHILD VOCALIZATIONS



CONVERSATIONAL TURNS



- Improvement in slope of the curve after intervention?
- Home language environment is a modifiable target of an intervention
- Feasibility study, need larger sample size
- Future plans: multicenter randomized trial with reading intervention and coaching

IMPROVING ACADEMIC OUTCOMES

MODIFIABLE TARGETS	POTENTIAL INTERVENTIONS
BONDING	COACHING
HEARING LOSS	AMPLIFICATION
BREAST MILK	PARENT SUPPORT
HOME LANGUAGE	READING

QUESTIONS?

- Thank you!
  - Craniofacial Center
  - Seattle Children's Hospital Academic Enrichment Fund
  - Research collaborators
  - Patients and families

Improving Practice Efficiency to Deliver High-Quality Preventive Services

Greg Blaschke, MD, MPH, FAAP
Shirley R. Kuse Professor of Pediatrics
Division Head, General Pediatrics
OHSU Doernbecher Children's Hospital

Agenda

- **Introduction & Background**
- **Implementation & Practice Workflow**
- **Using Tools**
  - **Maternal Depression**
  - **Development/Autism Screening**
  - **Social Determinants of Health**
- **Adolescent Well Visits**
- **Resources**

Faculty Disclosure: Greg Blaschke, MD, MPH, FAAP

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I do not intend to discuss an unapproved/investigative use of a commercial product/device in my presentation.

I am one of the contributors/reviewers of the *Bright Futures: Guidelines for Health Supervision of Infants, Children, and Adolescent,* 3rd and 4th Editions.

I acknowledge that today's activity is certified for CME credit and thus cannot be promotional. I will give a balanced presentation about well-child care using the best available evidence to support my conclusions and recommendations.

Faculty Disclosure: Edward Curry, MD, FAAP

In the past 12 months, I do not have any financial disclosures.

I do not intend to discuss an unapproved/investigative use of a commercial product/device in my possession.

Change in Practice

Participants will be able to:

- ❑Review clinical content in *Bright Futures Guidelines,* 4th Edition
- ❑Identify office systems-based strategies to maximize flow and efficiency for health promotion
- ❑Use pediatrician-tested strategies and Bright Futures tools to improve the quality of preventive services delivered in the clinical setting
- ❑Identify opportunities to tailor and apply Bright Futures/AAP recommendations with available tools and resources

Bright Futures

…is a set of principles, strategies and tools that are theory - based, evidence - driven, and systems - oriented, that can be used to improve the health and well-being of all children through culturally appropriate interventions that address the current and emerging health promotion needs at the family, clinical practice, community, health system and policy levels.

Bright Futures

Bright Futures is the health promotion/disease prevention part of the medical home.

At the heart of the medical home is the relationship between the clinician and the family or youth

The Periodicity Schedule and the Bright Futures Guidelines

The Periodicity Schedule tells you what to do in well- child visits, while the *Bright Futures Guidelines* tell you how to do it—and how to do it well.

Bright Futures Guidelines, 4th Edition

Part 1: Health Promotion Themes
- **12** chapters highlighting key health promotion themes
- New themes: **Social determinants of health**; Media use; Children and Youth with Special Health Care Needs

Part 2: Health Supervision Visits
- Rationale and evidence for screening recommendations
- 32 age-specific visits (including prenatal visit)
- 5 health supervision priorities for each visit
  - Designed to focus visit on most important issues for child that age
  - Includes: **social determinants of health,** health risks, developmental issues, positive reinforcement

Health Promotion Themes: 4th Edition

- Promoting Lifelong Health for Families and Communities
- Family Support
- Promoting Health for Children and Youth with Special Health Care Needs
- Healthy Development
- Mental Health
- Healthy Weight
- Healthy Nutrition
- Physical Activity
- Oral Health
- Adolescent Development
- Promoting the Healthy and Safe Use of Social Media
- Safety and Injury Prevention

Components of a Bright Futures Visit

- History
- Surveillance of development
- Review of systems
- Physical examination
- Screening
- Immunizations
- Anticipatory guidance

- **Tasks**
  - Disease detection
  - Disease prevention
  - Health promotion
  - Anticipatory guidance
- **Duration**
  - Approx. 18 minutes

What's New about the 4th Edition?

- Social determinants of health are embedded in many visits
  - ✓ Strengths and protective factors make a difference
  - ✓ Risk factors make a difference
- Features updated milestones of development and developmental surveillance questions
- Provides new clinical content about the latest recommendations and provides guidance on implementation
- Includes updates to several adolescent screenings including cervical dysplasia; depression; dyslipidemia; hearing; vision; tobacco, alcohol, or drug use

Screenings Updated from the 3rd Edition

- ***Adolescent hearing screening:***
  - **3rd Edition:** Selective audiometry based on risk assessment at all Adolescent Visits
  - **4th Edition:** Universal audiometry (once during the Early, Middle, and Late Adolescence Visits)
- ***Adolescent tobacco, alcohol, or drug use assessment:***
  - **3rd Edition:** Selective based on risk assessment for alcohol and drugs
  - **4th Edition:** Tobacco, alcohol, or drugs – universal administration of an assessment tool at all Adolescent Visits
- ***Cervical dysplasia:***
  - **3rd Edition:** Selective based on risk assessment at all Adolescent Visits
  - **4th Edition:** Universal beginning at the 21year visit in the 4th Edition

New Screenings Since the 3rd Edition

- ***Bilirubin screening:*** Universal at the Newborn Visit.
- ***Maternal depression screening:*** Universal at the 1 Month through 6 Month Visits.
- ***Oral health:*** Universal fluoride varnish at the 6 Month (first tooth eruption) through 5 Year Visits, in addition to Selective fluoride supplementation at the 6 Month through 12 Month and 18 Month through 16 Year Visits.
- ***Dyslipidemia screening:*** Universal once between the 9 and 11 Year Visits, in addition to the Universal dyslipidemia once between the 17 and 21 Year Visits carried over from the 3rd Edition.
- ***Depression screening:*** Universal for adolescents, annually beginning at the 12 Year Visit.
- ***Human immunodeficiency virus (HIV) screening:*** Universal once between the 15 and 18 Year Visits.

Bright Futures Tool and Resource Kit, 2nd Edition

The toolkit consists of 2 main sections:

Core Forms

These are the key documents to carry out each Bright Futures visit:

- Previsit Questionnaire
- Visit Documentation Form
- Bright Futures Parent-Patient Handouts

Supporting Materials

- Screening and Assessment Tools
  - ❖ Medical Screening Reference Tables
  - ❖ Commonly Used Screening Instruments and Tools
- Additional forms that accompany the Visit Documentation Form
  - ❖ Initial History Questionnaire
  - ❖ Medication Record
  - ❖ Problem List
  - ❖ Problem Visit
- Supplementary AAP Education Handouts

Core Tools: Integrated Format

EXAMPLE Tools

- Previsit Questionnaire
  - Surveillance tool allows healthcare professional to gather pertinent information without using valuable time asking questions

- Documentation Form
  - To document all pertinent information and fulfill quality measures

- Parent/Patient Educational Handout
  - Provides parental education for all Bright Futures Priorities at each visit

Implementation & Practice Workflow

How Does *Bright Futures* Help You?

For health care professionals:
With Bright Futures, health care professionals can accomplish 4 tasks in 18 minutes. The tools and resources help clinicians to structure visits and create practice processes to better address patient needs.

For AAP Chapters:
Provides resources to assist members in following the Guidelines and sharing best implementation practices. Bright Futures serves as the basis for quality improvement projects

***For public health professionals*:**
Provides a roadmap for structuring visits and sharing health information with the community; helps identify priorities for funding and provides recommended standardized developmental assessments.

***For families*:**
Provides resources and educational materials specific to each well-child visit. Bright Futures recognizes the strengths that families and parents bring to the health care partnership.

Implementing *Bright Futures into Daily Practice*

How it gets done in your practice setting in partnership with your patients and parents

You and your team are the experts

Implementing *Bright Futures into Daily Practice*

Can it be done?

YES!

Office-Based Systems Components

- ❑ Utilize a preventive services prompting system
- ❑ Utilize a recall/reminder system
  - ❖ To address immunizations and well child visits
- ❑ Utilize a system to track referral
  - ❖ Paper-based or electronic
- ❑ Utilize a system to identify children with special health care needs
- ❑ Link families to appropriate community resources
- ❑ Utilize a strength-based approach and shared decision-making strategy

Questionnaires

- ❑ Paper
- ❑ Electronic
  - At the visit in the waiting or exam room
  - At home (via email or patient portal)
- ❑ Make appointment time 15 minutes earlier

- ❑ **Practice support and nursing staff in charge of how this happens:**
  - Have a staff session to reinforce importance and contribution
  - Train how to distribute
  - Develop a scoring system
  - Develop a system to alert the healthcare professional to know "when ready to proceed"
  - Help parents/youth with literacy or language differences
  - Have all tools and supplies ready
  - Shift some responsibilities from the clinician to non-clinician staff where appropriate

What Can You Get From a *Bright Futures* Previsit Questionnaire?

Here are examples of what you can learn about how your patient and family are doing…

- ❑ Parental/youth concerns and questions for this visit
- ❑ Surveillance of patient/family strengths
- ❑ Surveillance of major changes in family
- ❑ Medical risk assessment (unique for each age/visit) such as:
  - TB, Lead, Anemia, STIs, Cholesterol
  - Vision and Hearing
- ❑ Oral health risk assessment
  - Dental home/fluoride H_2O
- ❑ Developmental surveillance for young children
- ❑ Strengths/developmental surveillance for school aged children & adolescents
- ❑ Expanded anticipatory guidance questions such as:
  - Social Determinants of Health
  - Caring for infant/child/adolescent
  - Patient's emotional well-being
  - Safety

This surveillance tool also alerts the patient/family that they will be universally screened for topics based on their age/stage (eg, child development, autism, depression, etc.).

Case Studies

Using the Tools Through Case Studies

- Maternal Depression (1 Month Visit)
- Child Development/Autism (18 Month Visit)
- Social Determinants of Health (3 Year Visit)

Strategies for Implementing Adolescent Well Visits



Adolescent Generalities

Adolescents are special! (like newborns, 5 year old, preteen)	Start with strengths and practice building rapport	Need to destigmatize, and do universal screening
Adolescents are 'hyper aware/ in tune' with environmental clues and may 'read things in' when not intended	Have an office action plan for things we fear: • Pregnancy • Suicide • Addiction • Violence	Visits are part of transition planning • Becoming responsible for own health over time • Letters for parents re: screeners • Letters for adolescents re: confidentiality, consent and disclosure

Adolescent Generalities

Practice is contextual - modify to community, epidemiology, setting (rainbow flags help)	Best to be obvious and talk out loud (no hidden agenda) *"I ask all my patients these questions"*	Plain language • consent = giving permission • confidentiality = telling others only if… • disclosures = can happen unintentionally (open record, billing, reminders)
For 10 years and over, completing screening together promotes understanding	Flow: together, separate, together • Parent concerns and ability to promote understanding and discussion	

Adolescent Generalities

State laws vary	Break confidentiality/disclosures • Talk with permission • Generally when needed (no contraindications) • Parents involvement improves outcome • They don't need to know all (or sometimes any) details

Suggestions

- ✓ Convert Sports PE and explore further if complaint doesn't = PE
- ✓ Normalize asking questions
- ✓ Do NOT ignore any concerning statement
- ✓ Use motivational interviewing
- ✓ Use tools!
- ✓ No such thing as Negative screen (thanks for answering, who could you talk about…XYZ)
- ✓ Encourage longer appointments
- ✓ Visit lasts over entire time in clinic (use team)
- ✓ Continuity and longitudinal care (not everything in 1 visit)

Adolescent Well Visits

STRATEGIES

- Schedule a longer visit
- Have an adolescent-friendly space
- Move from non-threatening questions to more sensitive topics
- Explain why you're asking the questions
- Clearly define confidentiality
- Remember surveillance is not screening and vice versa

Confidentiality

PRACTICAL POINTS

1. Parent and patient together at beginning of visit

2. Parent and patient separated during sensitive questions and physical exam

3. Parent and patient together to review assessment

Adolescent Well Visits

STRATEGIES

- Avoid medical jargon – speak simply
- Ask sensitive questions in the third person (particularly for younger adolescents)
- Use open-ended questions whenever possible
- Treat all comments seriously
- Keep the tone non-judgmental Avoid "Why?"

Adolescent Well Visits

STRATEGIES

- Explore the adolescent's issues
- Treat all comments seriously
- Use clarification, reflection, and interpretation as strategies
- Be aware of nonverbal communication
- Don't chart during the interview

Adolescent Well Visits

STRATEGIES

- Consider the adolescent's developmental stage, culture, ethnicity
- Reassure when the adolescent seems uncomfortable
- Encourage regular and open communication with parents

Confidentiality Sample Script

"There are some things I talk about with everyone your age. I keep this information private from your parents if you don't want to share it with them. If I hear something that sounds dangerous to you or someone else, I may need to tell your parents about that. I encourage everyone your age to talk to their parents about important things, but if you don't feel ready, you can talk about those things here."

Caution

- Have an adolescent office action plan
  - Suicide? imminent or past?
  - Pregnancy
  - Addiction
  - Disclosure of violence
- Use your full team and partners

Adolescent Previsit Questionnaires

EXAMPLE

Summary

- Interview the adolescent patient alone.
- Explain to patients what you can and cannot do confidentially.
- Explain limits of confidentiality.
- Implement policies to protect confidentiality and inform staff.
- Involvement of the family is optimal.

Questions?

Establishing a Workflow: Review

Workflow – 1 Month Visit Example

Workflow Needs to be Job-Specific, not Person-Specific

- ❑ Starts with initial entry point to medical office
  - Receptionist provides age appropriate Previsit Questionnaire
    - Pre-formatted age specific packet (1 Month Packet example)
      - 1 Month Previsit Questionnaire
      - Maternal Depression screening tool
      - Parental Educational Handout
    - Parent would complete questionnaires/screening tools in waiting area
  - Medical assistant on rooming child would make sure questionnaire is completed
    - MA attaches questionnaire to chart or enter the results into the EHR
  - Physician would review either paper copy or EHR
    - Would document intervention in chart
  - Completion of visit medical assistant would provide appropriate parent handout

EXAMPLE

Community Linkage Tips from the Practices

- ❑ Systems measure
  - Do you have someone in your office or clinic who is in charge of liaisons with community organizations and updates to accessible list of community resources for parents?
- ❑ Consider hiring a care coordinator, or use current staff with skills in this area
- ❑ Use community liaisons in the practice to handle referrals, communicate with specialists, and coordinate services/resources for families
- ❑ Consider hosting "mixers" with potential referral sources in the community to establish relationships
- ❑ If you have set it up, everything related to a difficult situation goes better

Team-Based Approach

You don't have to do all this alone!

- ➢ Multiple health supervision visits, thus multiple opportunities
- ➢ Sharing and delegation of tasks
- ➢ Practice change management resources can be found on the following websites:
  - o Bright Futures
  - o STAR Center
  - o National Resource Center for Patient/Family-Centered Medical Home
  - o AAP Quality Improvement

Billing & Coding

- ❑ When standardized screening tools are administered, scored, and interpreted as part of preventive service visit, each screening can be individually coded for billing purposes.
- ❑ Example:

HEALTH RISK ASSESSMENTS	
CPT® Codes	
96160	Administration of patient-focused health risk assessment instrument (eg, health hazard appraisal) with scoring and documentation, per standardized instrument
96161	Administration of caregiver-focused health risk assessment instrument (eg, depression inventory) for the benefit of the patient, with scoring and documentation, per standardized instrument

Source: https://www.aap.org/en-us/Documents/coding_preventive_care.pdf

Accessing Screening Tools

➔ https://toolkits.solutions.aap.org/ss/screening_tools.aspx

Pediatric Preventive Coding Resources

Coding at the AAP Website

- One stop shop for all coding related resources from the AAP
- Includes ICD-10-CM information and all topic-specific coding fact sheets
  - ***Coding for Pediatric Preventive Care*, 2019 Booklet**
  - available at: https://www.aap.org/en-us/Documents/coding_preventive_care.pdf
- AAP Coding Hotline aapcodinghotline@aap.org for all your coding and payer questions and issues!!

MCH/Title V Connection

Title V MCH Services Block Grant National Performance Measures

Breastfeeding
Safe Sleep
Developmental Screening
Physical Activity
Bullying
Adolescent Well-Visit
Medical Home
Transition
Preventive Dental Visit
Smoking

mchb.tvisdata.hrsa.gov/PrioritiesAndMeasures/NPMDistribution

CHIPRA 2019 Core Measures

Weight Assessment and Counseling for Nutrition and Physical Activity
Chlamydia Screening in Women Ages 16-20
Childhood Immunization Status
Screening for Depression and Follow-Up Plan: Ages 12-17 (CDF-CH)
Well-Child Visits in the First 15 Months of Life
Immunizations for Adolescents
Developmental Screening in the First Three Years of Life
Adolescent Well-Care Visits
Access to Primary Care Practitioners

medicaid.gov/medicaid/quality-of-care/performance-measurement/child-core-set/index.html

Education in Quality Improvement for Pediatric Practice (EQIPP)

- EQIPP courses help you identify and close gaps in your practice using practice tools.
  - Bright Futures - Infancy and Early Childhood Course
  - Bright Futures - Middle Childhood and Adolescence Course

Website Resources

- Resources and tip sheets
- Resources for families, states and community health programs
- Implementation strategies and stories from practices, states, and communities that use Bright Futures

brightfutures.aap.org

Bright Futures Tools

Below are some tools and resources available to assist with implementation of the 4th Edition:

- *Bright Futures Guidelines*, 4th Edition – Introductory Webinars
  - Available at: https://brightfutures.aap.org/materials-and-tools/Pages/Bright-Futures-Webinars.aspx
- *Bright Futures Tool and Resource Kit,* 2nd Edition – Overview (narrated PPT)
  - Available at: https://brightfutures.aap.org/materials-and-tools/Pages/Presentations-and-Handouts.aspx
- Screening and Priorities for each age/stage
  - Available at: https://brightfutures.aap.org/materials-and-tools/Pages/Presentations-and-Handouts.aspx
- Medical Screening Reference Tables
  - Available at: https://brightfutures.aap.org/materials-and-tools/tool-and-resource-kit/Pages/Medical-Screening-Reference-Tables.aspx

Changes in Practice: Recap

Participants can:

- ❑ Review clinical content in *Bright Futures Guidelines*, 4th Edition
- ❑ Identify office systems-based strategies to maximize flow and efficiency for health promotion
- ❑ Use pediatrician-tested strategies and Bright Futures tools to improve the quality of preventive services delivered in the clinical setting
- ❑ Identify opportunities to tailor and apply Bright Futures/AAP recommendations with available tools and resources

Questions?

References

- ❑ Duncan P, Pirretti A, Earls MF, Stratbucker W, Healy JA, Shaw JS, Kairys S. Improving delivery of Bright Futures preventive services at the 9- and 24-month well child visit. *Pediatrics*. 2015;135(1)e178-e186. Available at: http://pediatrics.aappublications.org/content/135/1/e178
- ❑ Lannon CM, Flower K, Duncan P, Moore KS, Stuart J, Bassewitz J. The Bright Futures Training Intervention Project: implementing systems to support preventive and developmental services in practice. *Pediatrics.* 2008;122(1)e163-e171. Available at: http://pediatrics.aappublications.org/content/122/1/e163
- ❑ Hagan JF, Shaw JS, Duncan PM, eds. *Bright Futures: Guidelines for Health Supervision of Infants, Children and Adolescents*, 4th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2017.
- ❑ Shaw JS, Hagan JF Jr, Shepard MT, Curry ES, Swanson JT, Janies KM, eds. *Bright Futures Tool and Resource Kit.* 2nd ed. Itasca, IL: American Academy of Pediatrics; 2019

How to Obtain *Bright Futures* Materials

Visit the *Bright Futures* Web site: **brightfutures.aap.org**

To order the *Bright Futures* Guidelines and Toolkit, go to **shopAAP.org**

Sign up for the Bright Futures eNews and other alerts at **brightfutures.aap.org/Pages/contactus.aspx**

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Neonatal Hyperbilirubinemia Updates

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DOERNBECHER CHILDREN'S Hospital OHSU

Ilse Larson, MD, IBCLC

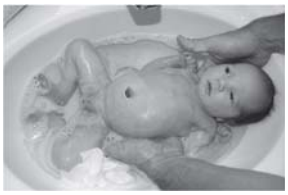
With gratitude to:
- Ellen Laves, MD, Carrie Phillipi, MD, PhD, and Mina Tahai, MD (for many of the slides)
- Tom Newman, MD, MPH (for all the learnings)

Learning Objectives

1. Review the basic pathophysiology of neonatal hyperbilirubinemia
2. Understand the AAP's clinical practice guidelines for hyperbilirubinemia in newborns ≥35 weeks
3. Review outcomes of guidelines implementation and emerging data about the possible risks associated with phototherapy
4. Discuss Northern California Neonatal Consortium Consensus Guidelines for Screening & Management

Neonatal Jaundice

60% of healthy newborns will have clinical jaundice



Why Newborns?

- **Increased bilirubin production** (↑Hgb & short RBC lifespan)
- **Limited bilirubin-binding capacity (l**ow serum albumin)
- **Decreased conjugation (↓**glucoronysyl- transferase activity)
- **Decreased excretion** leading to reabsorption in the bowel (bowel flora, intestinal motility, stool frequency, caloric intake, and feeding frequency)

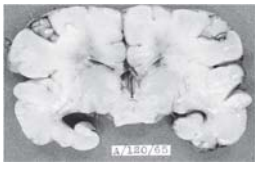
What's the significance?

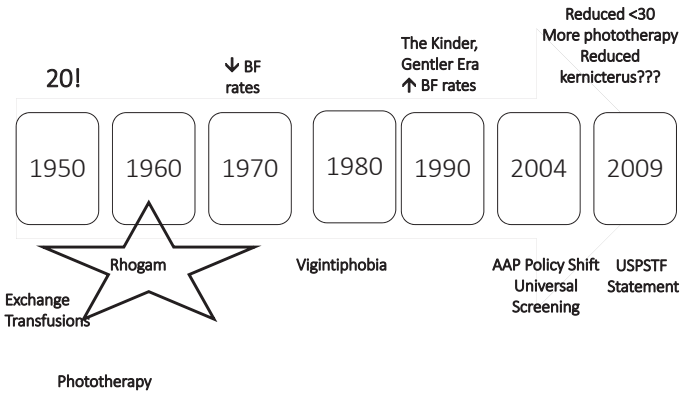
Acute bilirubin encephalopathy:

- Lethargy→stupor
- Hypotonia→ hypertonia→ retrocolis-opisthotonis
- Poor feeding, shrill cry

Kernicterus (chronic bilirubin encephalopathy):

- Extrapyramidal signs (athetosis), severe delays/MR
- Sensorineural hearing loss
- Gaze palsies
- Dental dysplasia

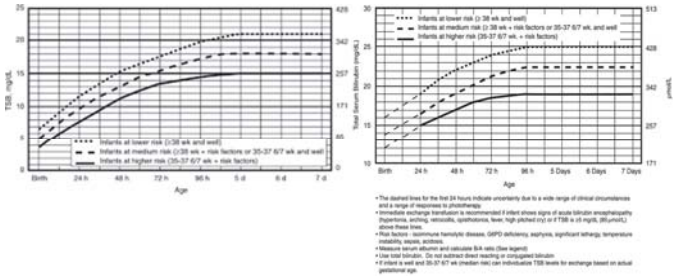




2004 AAP Guidelines

1. Promote and support successful breastfeeding.
2. Establish nursery protocols for the identification and evaluation of hyperbilirubinemia.
3. Measure the total serum bilirubin (TSB) or transcutaneous bilirubin (TcB) level on infants jaundiced in the first 24 hours.
4. Recognize that visual estimation of the degree of jaundice can lead to errors, particularly in darkly pigmented infants.
5. Interpret all bilirubin levels according to the infant's age in hours.
6. Recognize that infants at less than 38 weeks' gestation, particularly those who are breastfed, are at higher risk of developing hyperbilirubinemia and require closer surveillance and monitoring.
7. Perform a systematic assessment on all infants before discharge for the risk of severe hyperbilirubinemia.
8. Provide parents with written and verbal information about newborn jaundice.
9. Provide appropriate follow-up based on the time of discharge and the risk assessment.
10. Treat newborns, when indicated, with phototherapy or exchange transfusion.

AAP Guideline Graphs



Effect of Universal Screening

Kuzniewicz et al. 2009 38,182 infants.

10.6% were born at facilities with universal bilirubin screening.

Compared with infants born at facilities that were NOT screening:

- 62% lower incidence of TsB levels over the AAP threshold (0.17% vs 0.45%; P < .001),
- Had twice the rate of inpatient phototherapy (9.1% vs 4.2%; P < .001), and
- Had slightly longer birth hospitalization lengths of stay (50.9 vs 48.7 hours; P < .001).

Effect of Universal Screening

- Only 56% of those who received phototherapy had TsB above threshold, compared with 70% in facilities without universal screening.

Is this the right approach?

Phototherapy NNT

Newman, et al 2009

281,898 AGA infants born ≥35 weeks' gestation at 12 Northern California Kaiser hospitals from 1995 to 2004.

- 22,547 with a TsB within 3 mg/dL of the AAP phototherapy threshold
- Used multiple logistic regression to estimate the efficacy of hospital phototherapy in preventing the bilirubin level from exceeding the 2004 guideline's exchange transfusion threshold within 48 hours.

NNTs (95% CI)

Gestational Age, wk	Age at Qualifying TSB: <24 h	Age at Qualifying TSB: 24 to <48 h	Age at Qualifying TSB: 48 to <72 h	Age at Qualifying TSB:≥72 h
Boys				
35	14 (7–40)	26 (14–57)	83 (36–190)	171 (70–426)
36	10 (6–19)	19 (12–39)	59 (31–101)	122 (68–236)
37	16 (10–28)	29 (20–58)	95 (52–168)	196 (100–407)
38	35 (14–100)	67 (31–215)	222 (107–502)	460 (196–1352)
39	74 (31–244)	142 (62–554)	476 (197–1385)	989 (373–3607)
40	106 (44–256)	204 (98–487)	682 (367–1294)	1419 (634–3755)
≥41	148 (54–428)	284 (127–780)	953 (366–3017)	1983 (676–8408)
Girls				
35	21 (12–49)	40 (21–86)	126 (50–267)	261 (105–585)
36	15 (11–26)	28 (20–51)	90 (43–146)	186 (102–347)
37	23 (16–39)	44 (31–75)	145 (73–243)	300 (146–671)
38	53 (23–134)	102 (43–236)	339 (154–730)	705 (314–2016)
39	113 (58–342)	217 (103–713)	729 (272–1730)	1516 (614–4520)
40	162 (75–400)	312 (164–704)	1046 (491–2136)	2176 (922–6107)
≥41	226 (92–702)	435 (183–1140)	1461 (510–4842)	3041 (888–11096)

Table 4: Newman et al *Pediatrics*. 2009 May ; 123(5): 1352–1359. doi:10.1542/peds.2008-1635

In the Setting of Universal Screening, do Infants Exceed Exchange Transfusion Levels?

Flaherman et al., 2012 ~ 18,000 newborns (2005-2007) in the KP Northern California Hospitals after the implementation of universal screening

- 22 infants (14 infants <38 weeks) exceeded exchange transfusion threshold
- Only 1 received an ET
- **No documented sequelae**

In the Setting of Universal Screening, do Infants Exceed Exchange Transfusion Levels?

- Screening TsB was at least "high-intermediate risk" for all 22 infants and "high-risk" for all ≥38 weeks.
- 4 outcomes may be attributable to incomplete adherence to AAP guideline
- 13 might have been prevented by better adherence to AAP *follow-up* guideline

BUT…

- Re-testing would have required 2166 additional bilirubin tests to prevent (at most) 13 outcomes

Jaundice Outcomes

Wickremasinghe, et al 2015

- **SNHL**: Only bilirubin levels ≥10mg/dl above exchange transfusion thresholds (or ≥ 35 mg/dl) were associated with a significantly increased risk

Wu, et al 2015

- **Cerebral Palsy** consistent with kernicterus occurred only in infants with 2+ risk factors for NT and TsB >5mg/dl above exchange transfusion threshold

Vandborg, et al 2012

- No significant difference in **development** at age 1-5 years (ASQ) in infants with a peak serum bilirubin over 25mg/dl

Who Gets Kernicterus?

Kuzniewicz et al 2014: Kaiser Northern California.

525,409 infants ≥35 weeks gestation between 1995-2011

- 47 infants identified with TsB ≥30 (8.6 per 100,000 births)
- Median follow up 7.9 years

TABLE 3 Characteristics of Infants With CBE

Case no.	Gestational Age, wk	Peak TSB, mg/dL	Seizures	SNHL	CP	G6PD Activity, U/g Hb	Sepsis	Coombs Test
1	35	38.2	No	Yes	No	Not tested	No	Negative
2	38	40.7	Yes	Yes	No	0.8	No	Negative
3	36	49.1	Yes	Yes	Yes	7.6	Yes	Negative
4	36	48.5	Yes	Yes	Yes	6.6	No	Negative

Are there risks of phototherapy?

Does Phototherapy affect Breastfeeding?

Waite, et al 2016: small reduction in breastfeeding rates at 12 months and in exclusivity at 1, 2, and 4 months

TABLE 2. RATES OF ANY BREASTFEEDING AND EXCLUSIVE BREASTFEEDING BY MONTH FOR PHOTOTHERAPY EXPOSED AND PHOTOTHERAPY UNEXPOSED INFANTS

	Breastfeeding rate in phototherapy exposed, N = 220	*Breastfeeding rate in phototherapy unexposed, N = 4,016*	
	n (%)	n (%)	*OR (95% CI)*
Any breastfeeding			
Month 1	186 (86.1)	1,813 (85.4)	1.14 (0.71–1.81)
Month 2	137 (75.3)	1,360 (74.8)	1.18 (0.78–1.78)
Month 4	108 (65.1)	1,069 (67.2)	1.03 (0.70–1.53)
Month 6	85 (54.5)	889 (59.9)	0.88 (0.60–1.27)
Month 9	66 (43.7)	695 (49.9)	0.80 (0.56–1.15)
Month 12	29 (20.7)	406 (31.4)	0.58 (0.37–0.91)
Exclusive Breastfeeding			
Month 1	81 (37.5)	1,022 (48.1)	0.69 (0.49–0.95)
Month 2	57 (31.3)	767 (42.2)	0.69 (0.48–0.99)
Month 4	29 (17.5)	462 (29.0)	0.57 (0.36–0.88)

ORs based on logistic regression adjusting for maternal age, race, maternal education, household income, gestational age, prenatal intention to breastfeed, supplemental formula use on day of life 1, and breastfeeding problems in the first 2 weeks of life.
OR, odds ratio.

Does Phototherapy lead to increased Seizure Risk?

Maimburg et al 2016

- Increased risk of epilepsy among children treated with phototherapy, the association was seen only in boys (adjusted HR 1.98, 95% CI: 1.40–2.78)

Newman, et al 2018

- Increased risk of epilepsy, adjusted hazard ratio (aHR) of 1.22 (95% CI: 1.05 to 1.42; P = 0.009)
- Boys were at higher risk of seizures overall (aHR = 1.18; 95% CI: 1.10 to 1.27) and had a higher aHR for phototherapy (1.33; 95% CI: 1.10 to 1.61)

Is Phototherapy linked to Childhood Cancer?

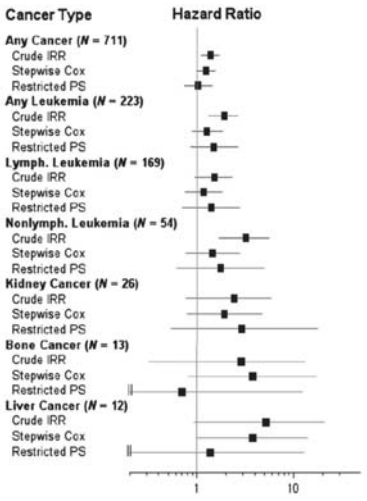
Newman et al 2016

Retrospective cohort study of 525,409 children born at ≥35 weeks' gestation between 1995-2011 at 15 KPNC hospitals

Exclusions: death, transfer, lost to follow-up at <60 days, cancer dx before 60 days

- Initial crude IRRs were uniformly positive with low p-values.
- After adjusting for confounding were no longer significant

Upper limit of the hazard ratios is most concerning for infant's with Down syndrome with the NNH being 23 at the upper limit



Is phototherapy worth even a small risk?

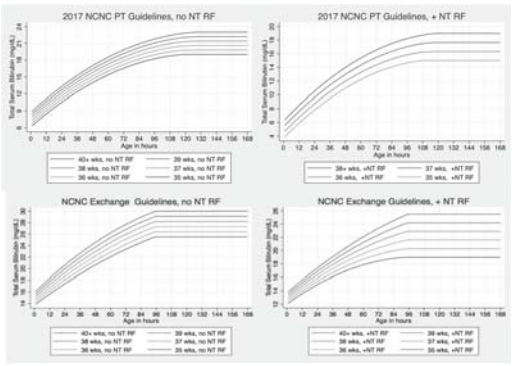
Development of the NCNC Guidelines

Based on concerns that the 2004 AAP Guideline was based on limited evidence, internally inconsistent and recommend a significant practice shift at 38 weeks gestation, the UCSF Northern California Neonatal Consortium members came together to:

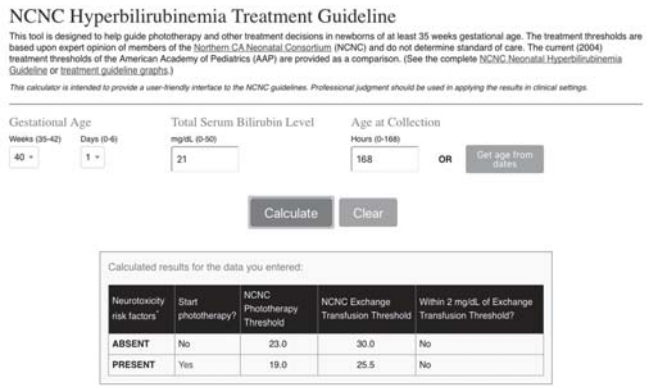
- Update hyperbilirubinemia clinical practice based on recent research
- Draw on the KP Northern California experience with updated clinical practice guidelines

Full executive summary and recommendations:
http://www.phototherapyguidelines.com/NeoHyperbilirubinemiaGuidelineFINAL_2018-0209.docx

NCNC Graphs



www.phototherapyguidelines.com

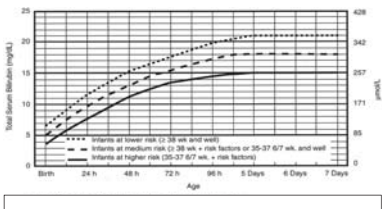


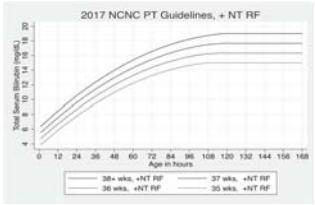
NCNC Definition of Neurotoxicity Risk Factors

Neurotoxicity risk factors include:

- Isoimmune hemolytic disease, G6PD deficiency, or other hemolytic disease
- Sepsis or suspected sepsis (sufficient to be currently on antibiotics)
- Acidosis (BE ≤ −8 meq/L or pCO2 > 50 mmHg within the last 24 hr)
- Albumin < 3.0 mg/dL
- Any clinical instability

AAP vs. NCNC (with risk factors)

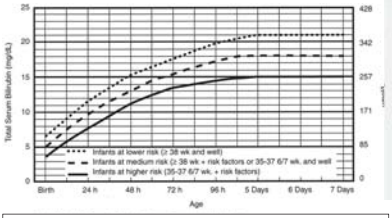


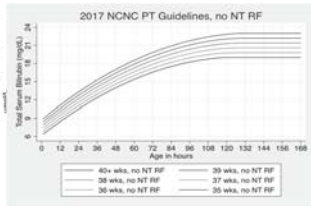


Examples for **with neurotoxicity risks**, 168 HOL infant

40 1/7 NCNC threshold = 19 (vs. 18) Exchange = 25.5 (vs. 22.5)
38 1/7 NCNC threshold = 19 (vs. 18) Exchange = 23.1 (vs. 22.5)
35 1/7 NCNC threshold = 15.2 (vs. 15) Exchange = 19.2 (vs. 19)

AAP vs. NCNC (no risk factors)





Examples for **no n**eurotoxicity risks, 168 HOL infant

40 1/7 NCNC threshold = 23 (vs. 21) Exchange= 30.0 (vs. 25)
38 1/7 NCNC threshold = 21.5 (vs. 21) Exchange 28.3 (vs. 25)
35 1/7 NCNC threshold = 19.1 (vs. 18) Exchange 25.6 (vs. 22.5)

Case Comparisons

A baby boy in clinic is noted to have jaundice at 48 hours of life. Mother is AB+/Ab-, she is expressing colostrum and exclusively breastfeeding. The baby is feeding well with appropriate output. This is mom's third baby. Infant's weight is down 7% from birth weight. There is slight facial bruising, but no cephalohematoma. TSB is obtained and is 15.5 mcg/dL.

- Let's look at AAP vs. NCNC recommendations for a 41w1d, 37w 6d and 36w2d week gestational age infant

41w1d: AAP vs. NCNC Recommendations

Calculated results for the data you entered:

Neurotoxicity risk factors*	Start phototherapy?	NCNC Phototherapy Threshold	NCNC Exchange Transfusion Threshold	Within 2 mg/dL of Exchange Transfusion Threshold?
ABSENT	No	17.0	24.5	No
PRESENT	Yes	14.0	20.6	No

For comparison purposes, here are approximate American Academy of Pediatrics' 2004 guideline phototherapy thresholds:

Neurotoxicity risk factors*	Start phototherapy?	AAP Phototherapy Threshold
ABSENT	Yes	15.0
PRESENT	Yes	13.0

37w6d: AAP vs. NCNC Recommendations

Calculated results for the data you entered:

Neurotoxicity risk factors*	Start phototherapy?	NCNC Phototherapy Threshold	NCNC Exchange Transfusion Threshold	Within 2 mg/dL of Exchange Transfusion Threshold?
ABSENT	No	15.7	22.9	No
PRESENT	Yes	13.8	19.1	No

For comparison purposes, here are approximate American Academy of Pediatrics' 2004 guideline phototherapy thresholds:

Neurotoxicity risk factors*	Start phototherapy?	AAP Phototherapy Threshold
ABSENT	Yes	13.0
PRESENT	Yes	11.0

36w6d: AAP vs. NCNC Recommendations

Calculated results for the data you entered:

Neurotoxicity risk factors*	Start phototherapy?	NCNC Phototherapy Threshold	NCNC Exchange Transfusion Threshold	Within 2 mg/dL of Exchange Transfusion Threshold?
ABSENT	Yes	14.8	21.6	No
PRESENT	Yes	12.3	18.0	Yes - see flow sheet

For comparison purposes, here are approximate American Academy of Pediatrics' 2004 guideline phototherapy thresholds:

Neurotoxicity risk factors*	Start phototherapy?	AAP Phototherapy Threshold
ABSENT	Yes	13.0
PRESENT	Yes	11.0

Next Steps??

Steps that reduce phototherapy, but are still in line with the AAP Guidelines:

→ No phototherapy under the AAP thresholds

→ Adjust around the medium risk threshold by gestational age

From the 2004 Nomograms Text:

- Use total bilirubin. Do not subtract direct reacting or conjugated bilirubin.
- Risk factors = isoimmune hemolytic disease, G6PD deficiency, asphyxia, significant lethargy, temperature instability, sepsis, acidosis, or albumin < 3.0g/dL (if measured)
- For well infants 35-37 6/7 wk can adjust TSB levels for intervention around the medium risk line. It is an option to intervene at lower TSB levels for infants closer to 35 wks and at higher TSB levels for those closer to 37 6/7 wk.
- It is an option to provide conventional phototherapy in hospital or at home at TSB levels 2-3 mg/dL (35-50mmol/L) below those shown but home phototherapy should not be used in any infant with risk factors.

Adopt the NCNC Guidelines

- Evidence base is strong
- Insider intelligence is that forthcoming AAP guidelines will not be lower than the NCNC guidelines (2020? 2021?)
- OHSU's ED, Ward, MBU, and clinics adopted the NCNC guidelines **September 9, 2019**

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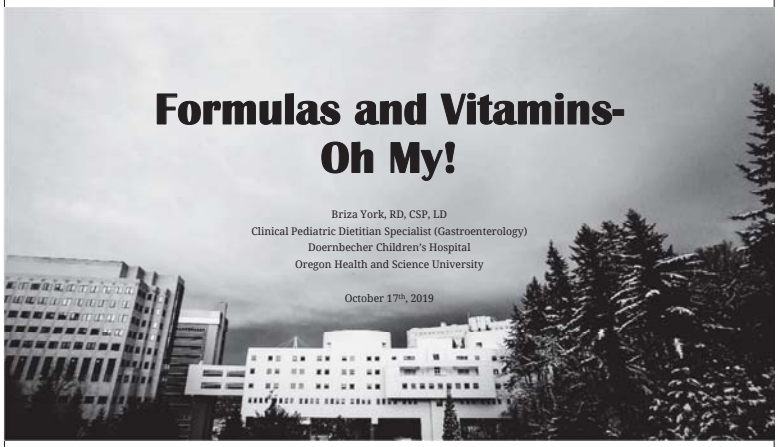


Formulas and Vitamins- Oh My!

Briza York, RD, CSP, LD
Clinical Pediatric Dietitian Specialist (Gastroenterology)
Doernbecher Children's Hospital
Oregon Health and Science University

October 17th, 2019

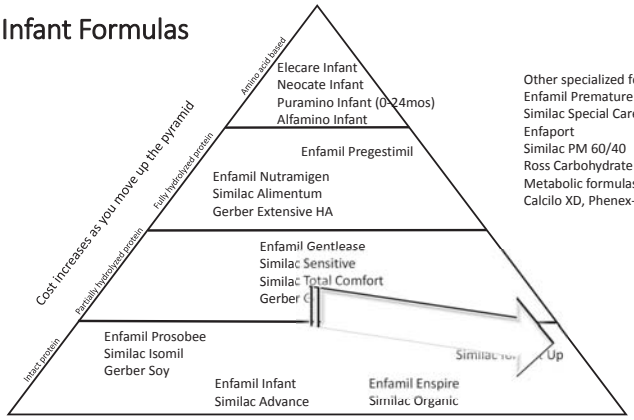
Objectives

- Understand infant and pediatric formulas and their appropriate uses
- Understand main vitamins and minerals of concern
- Review case study

Infant Formulas

- Breastmilk is best! But sometimes not available
- FDA regulated
- Standard concentration is 20 calories per ounce for majority of formulas
  - Special recipes to make formulas higher in calories if needed
  - Premature discharge formulas are 22calories per ounce standard mixing
- Main formula companies: Enfamil and Similac

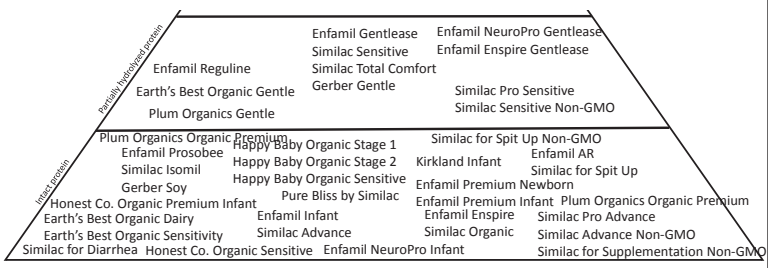
Infant Formulas



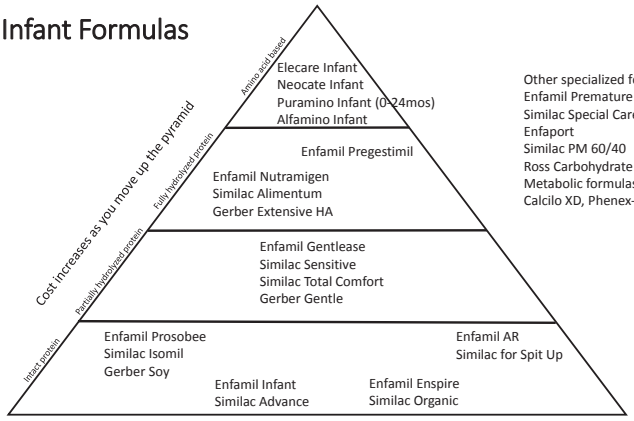
Other specialized formulas:
Enfamil Premature and EnfaCare
Similac Special Care and Neosure
Enfaport
Similac PM 60/40
Ross Carbohydrate Free Soy
Metabolic formulas (examples: Calcilo XD, Phenex-1, LMD)

The bottom half of the pyramid really looks like this…

*Not included: Non-US formulas (such as HiPP, Holle, etc)



Infant Formulas



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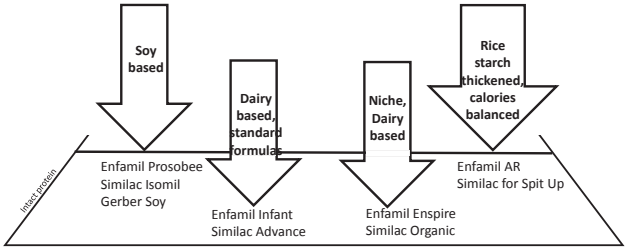
Infant Formulas



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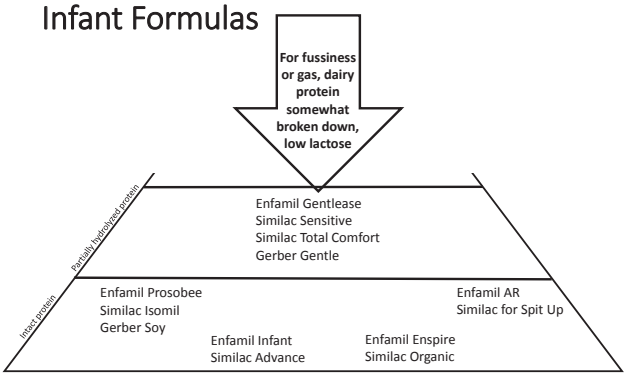

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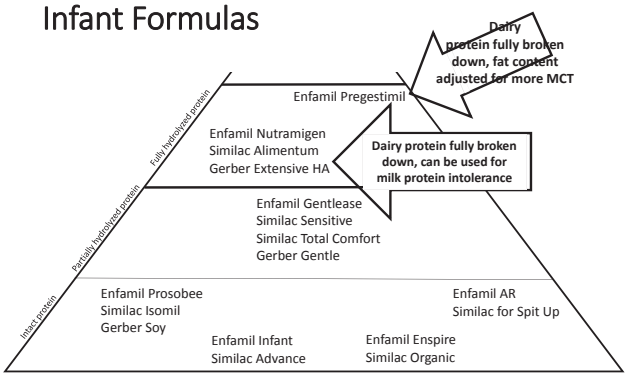

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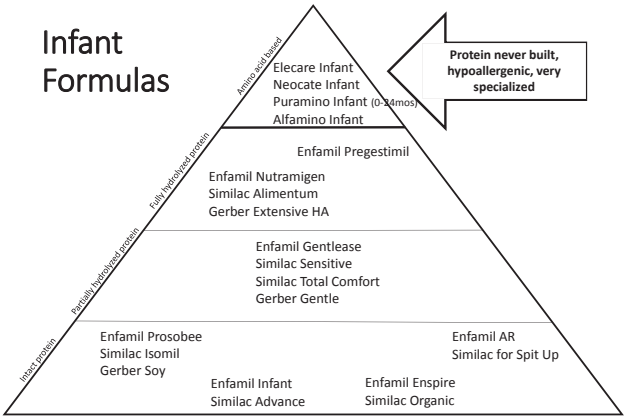

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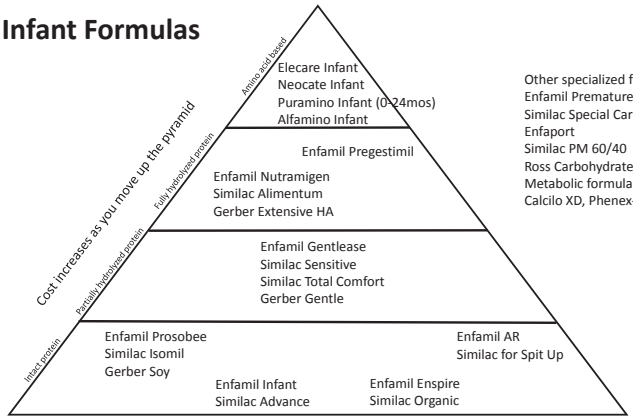

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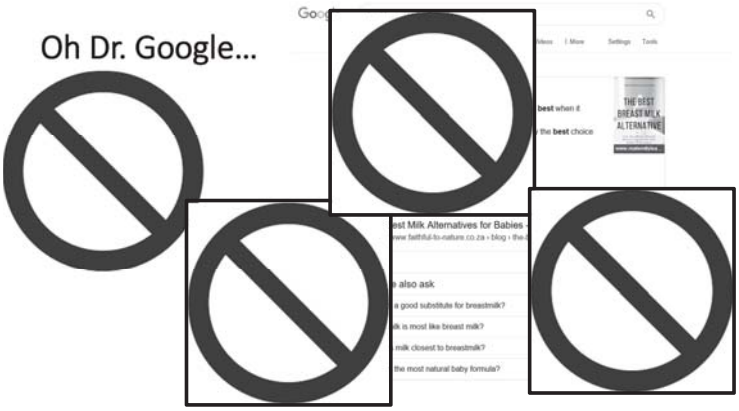
Similac Special Care and Neosure
Enfaport
Similac PM 60/40
Ross Carbohydrate Free Soy
Metabolic formulas (examples: Calcilo XD, Phenex-1, LMD)

Inappropriate Infant Milks

- Friend's breastmilk or Craigslist breastmilk
- Goat milk
- Homemade "infant formulas"
- Milk alternatives

Oh Dr. Google…



About goat milk…

- Goat milk is most similar in composition to cow's milk
- Goat milk is **NOT** like breastmilk
- Goat milk is not safe for any baby, but especially not for cow's milk protein intolerant/sensitive babies
- Homemade formulas using goat milk are NOT safe or nutritionally complete
- Raw goat milk can contain dangerous bacteria, including E. Coli, Salmonella, Listeria, Campylobacter
- If an infant is on goat milk, counsel about the dangers and send referral to Registered Dietitian

Nutrition Content Comparison

Per 100 calories	Breastmilk	Standard Infant Formula	Goat Milk
Calories per ounce	20	20	21
Protein	1.47g	2g	5.16g ↑
Calcium	46mg	78mg	194mg ↑
Folate	7µg	16µg	1µg ↓
Magnesium	4mg	8mg	20mg ↑
Potassium	73mg	108mg	296mg ↑
Sodium	24mg	27mg	72mg ↑

Nutrition Content Comparison

- Recommend Intake for Age: 1.6-2.2g/kg/day protein, 200-260mg/day of calcium, 65-80µg/day of folate, 30-75mg/day of magnesium, 400-700mg/day of potassium, and 120-370mg of sodium
- If baby drinks 800calories per day:

	Breastmilk	Standard Infant Formula	Goat Milk
Protein	12g	16g	41g ~3x more
Calcium	368mg	624mg	1,552mg ~4x more
Folate	56µg	128µg	8µg Only 12% of need
Magnesium	32mg	64mg	160mg ~5x more
Potassium	584mg	864mg	2,368mg ~4x more
Sodium	192mg	216mg	576mg ~3x more

International Formulas

- HiPP, Holle, etc are popular
- Unable to recommended at this time
- Per article: "The potential dangers are numerous. Children can fall ill or become malnourished if parents inadvertently use an incorrect formula-to-water ratio; unofficial formula vendors may not store the powdered formula properly, raising the possibility of bacterial contamination, product deterioration or loss in nutrient density; there is no system in place to notify consumers in the United States if any of these formulas are recalled; and while many European formulas contain the nutrients required in the United States, some do not. In addition, parents in the United States may not realize that European formulas labeled hypoallergenic aren't meant for children with cow's milk allergies."


T Parenting

FEEDING & NUTRITION

European Baby Formula That Is Illegally Sold in the United States Carries Risks, Pediatricians Warn

HiPP, Holle and other brands are not registered with the F.D.A., but that hasn't stopped parents from buying them from third-party vendors.

By Christina Caron and Jessica Grose

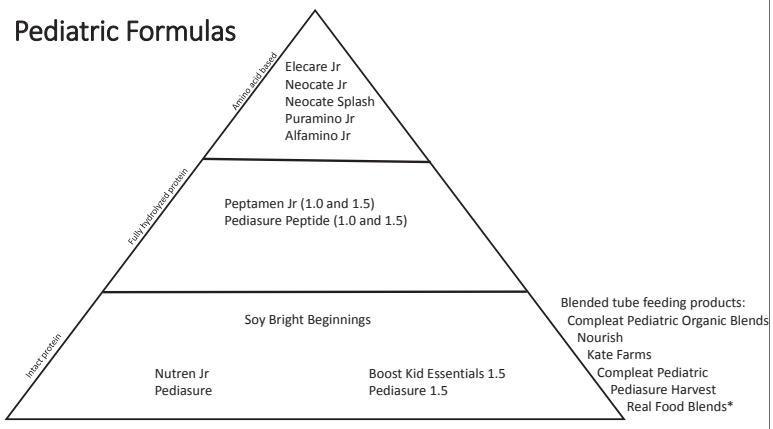
June 11, 2019

Source: https://parenting.nytimes.com/feeding/european-baby-formula

Pediatric Formulas

- Oral supplements or tube feeds
- Complete nutrition source
- Most formulas are 30 calorie per ounce or 45 calorie per ounce
- Main formula companies: Abbott and Nestle
- Blended tube feeding products are gaining in popularity

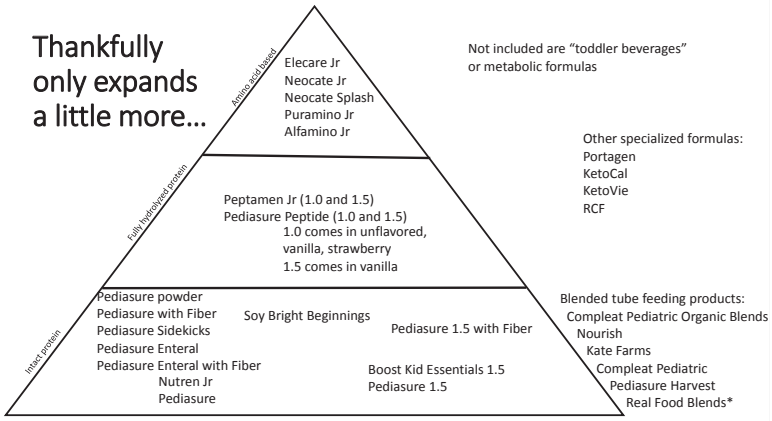
Pediatric Formulas



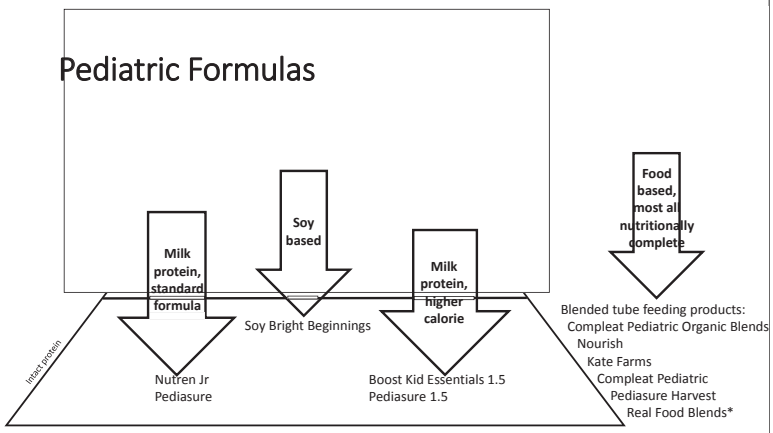
Thankfully only expands a little more…

Not included are "toddler beverages" or metabolic formulas

Other specialized formulas:
Portagen
KetoCal
KetoVie
RCF



Pediatric Formulas



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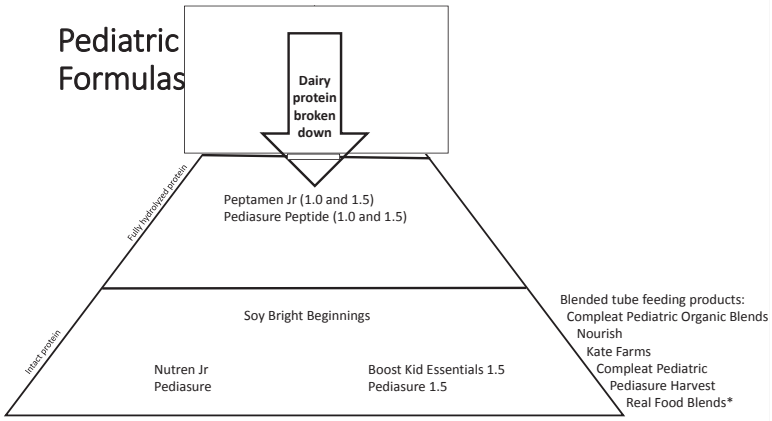

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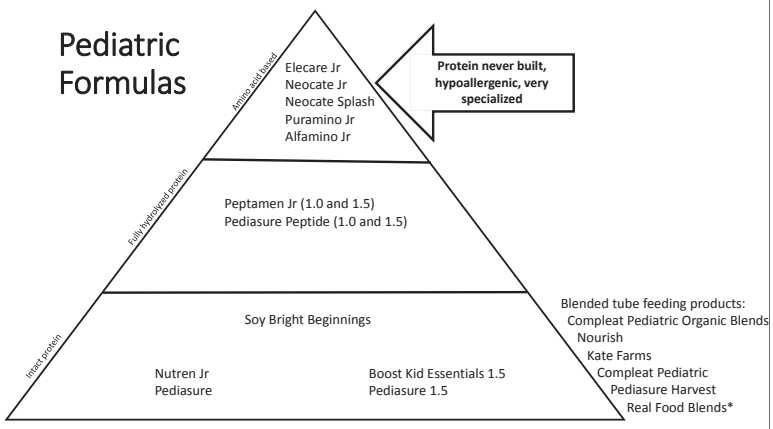

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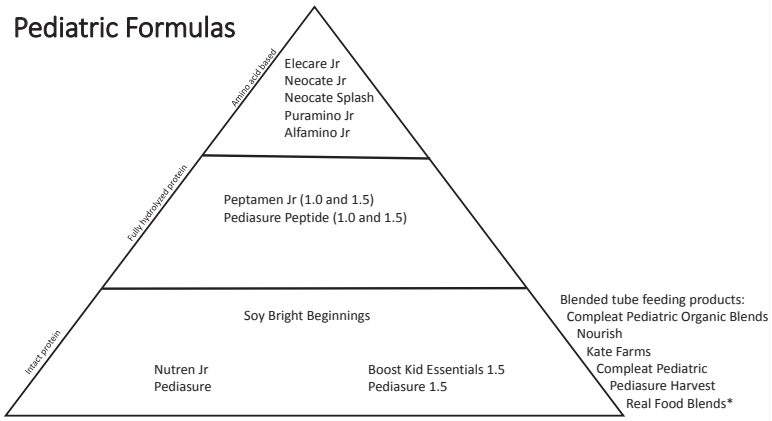


Vitamins

- Not all diets are nutritionally complete
- Malnutrition can come in many forms
- Vitamin supplements are sometimes needed
  - Limited diets due to picky eating, medical conditions, choice
  - Conditions that cause malabsorption
  - Geography
  - Increased nutrient needs, metabolic conditions

Dietary Supplement Regulation

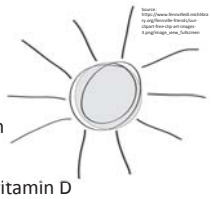
- Dietary Supplement Health and Education Act of 1994 (DSHEA)
- Manufacturers and distributors prohibited from marketing adulterated or misbranded products
  - **Manufacturers and distributors are responsible for evaluating the safety and labeling of their products**
- FDA will take action against adulterated or misbranded dietary supplements **after it reaches the market**

Source: https://www.fda.gov/food/dietary-supplements

Vitamin Supplements

- Ensure that it's age appropriate
- Not excessive
- Iron or no iron?
- Supplement specific vitamins based on lab values
  - Vitamin D (25HD Vitamin D)
  - Iron (CBC, Iron panel, ferritin)

Vitamin D



- Important for calcium absorption and bone mineralization
- Naturally in very few foods
- Breastfed infants require 400 international units daily of vitamin D
- Formula fed infants may need additional vitamin D depending on volume of formula consumed
- Older children, vitamin D should be supplemented based on lab values
  - Deficient vs. insufficient
  - Age of patient
  - Ergocalciferol (D2) or Cholecalciferol (D3)
- Recheck lab after 2-3mos of supplementing

Iron

- Important for formation of hemoglobin and other blood and muscle proteins as well as enzymes
- Food sources:
  - Heme: beef, poultry, shrimp, eggs
  - Non-heme: instant oatmeal, kidney beans, tofu, spinach
- Iron absorption is increased with vitamin C
- Calcium can decrease iron absorption
- Iron be constipating, change stool color
- Supplementation based on lab values

Elimination Diets

- Many people are on elimination diets
- Personal choice vs. experience with food vs. medical diagnosis
- These are not without risks
- Diet is easy to change on own, but should be guided to ensure adequacy
- Counsel on substitutions

Foods	Main nutrients
Cow's milk	Protein, calcium, magnesium, phosphorus, vitamins A, B6, B12, D, riboflavin, pantothenic acid (iodine in some countries)
Soy	Protein, calcium, phosphorus, magnesium, iron, zinc, thiamin, riboflavin, vitamin B6, folate
Eggs	Protein, iron, selenium, biotin, vitamin A, B12, pantothenic acid, folate, riboflavin
Wheat	Carbohydrate, zinc, selenium, thiamin, niacin, riboflavin, folic acid, iron, magnesium, dietary fiber
Peanut/tree nut	Protein, selenium, zinc, manganese, magnesium, niacin, phosphorus, vitamins E, B12, alpha linolenic acid, linoleic acid
Fish/shellfish	Protein, iodine, zinc, phosphorus, selenium, niacin Fatty fish: vitamins A, D, omega-3 fatty acids

Groetch et al, 2017

Case Study

- 14yo boy presents with fatigue
- Overall healthy and well nourished per growth charts
- Picky eater
- Blood tests found macrocytic anemia and low vitamin B12. No antibodies to intrinsic factor or tissue transglutaminase
- Given vitamin B12 injections and "dietary advice"

Harrison et al, 2019

Case Study

- Now 15yo developed hearing loss followed by vision symptoms
- MRI and ophthalmology exam were normal
- 2yrs later: progressive vision loss found to have optic neuropathy with 20/200 vision
- Neurologic exam and another MRI were normal
- Genetic tests, GI scope/biospies, Fibroscan were all normal

Harrison et al, 2019

	Result	Reference Range
Hemoglobin, g/L	148	130-160
Mean corpuscular volume, fL	**100.4**	83-100
Platelets, x10^9 cells/L	250	150-450
Creatinine, mg/dL	**0.5**	0.7-1.2
Total bilirubin, mg/dL	**1.3**	<1.2
Alk Phos µkat/L	**4.2**	1-2.7
Total protein, g/L	74	60-80
Adjusted calcium, mmol/L	2.23	2.2-2.6
CRP, nmol/L	<9.5	<57.1

Harrison et al, 2019

	Result	Reference Range
Vitamin A, µmol/L	0.8	0.8-2.2
Vitamin E, µmol/L	14.3	10.2-39
25HD Vitamin D, nmol/L	**10**	>50
Vitamin B12, pmol/L	135	132.8-664
Ferritin, pmol/L	90.8	74.2-898.9
Serum folate, nmol/L	9.2	5.7-44.3
Zinc, µmol/L	**26.8**	11-23
Copper, µmol/L	**9.8**	12-23
Selenium, µmol/L	**0.55**	0.59-1.65
Manganese, nmol/L	91.8	72.8-218.5
Homocysteine, µmol/L	**47.1**	2-14.3
Methylmalonic acid (urine), µmol/mmol	**7.2**	0.7-3.2

Harrison et al, 2019

Case Study

- Persistent macrocytosis with normal ferritin, folate, and B12
- Homocysteine and MMA levels elevated indicating functional B12 deficiency, which led to nutritional evaluation
- No alcohol or smoking
- Growth was good
- Since elementary school has avoided foods with certain textures
- Will eat French fries, chips, white bread, ham lunchmeat, and sausage
- Didn't finish previous vitamin B12 injections

Harrison et al, 2019

Case Study

- Provided supplements and referred to mental health for an eating disorder
- Vision stabilized, but did not improve
- Delayed diagnosis possibly d/t treated vitamin B12 deficiency. Homocysteine and methylmalonic acid are more sensitive indicators of functional vitamin B12 deficiency
- BMI is not the only indicator of malnutrition

Harrison et al, 2019

References

- Groetch M, Verter C, Skypala I, Vlieg-Boerstra B, Grimshaw K, Durban R, et al. Dietary Therapy and Nutrition Management of Eosinophilic Esophagitis: A Work Group Report of the American Academy of Allergy, Asthma, and Immunology. J Allergy Clin Immunol Mar/Apr 2017; 5(2), 312-324.e29
- Harrison R, Warburton V, Lux A, Atan D. Observation: Case Report: Blindness Caused by Junk Food Diet. A of Internal Medicine Sept 2019

Questions?

Thank you!

Pediatric Chronic Pain: Tips for Primary Care Providers for Prevention and Management

DATE: October 17, 2019 PRESENTED BY: Amy Holley PhD, Associate Professor of Pediatrics & Psychiatry

Disclosures

- I have nothing to disclose

Disclosures

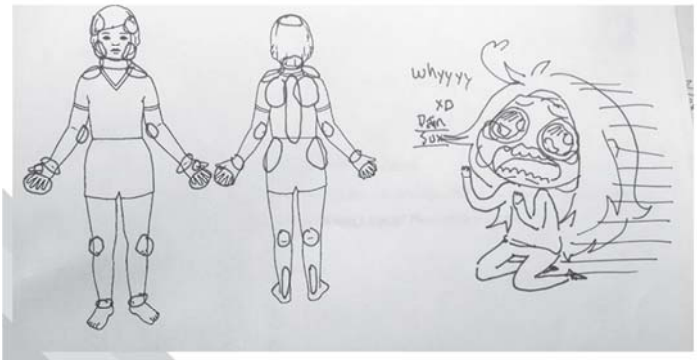
- I have nothing to disclose

Except I'm going to spend the next hour talking about pain…

Presentation Overview

- Describe prevalence and impact of pediatric pain
- Present key factors that impact pain outcomes
- Describe strategies providers can use to best support kids and their parents

Putting a face on the Numeric Rating Scale



From the medical record…

Physical findings do not explain her report of pain

Unremarkable exam. I wonder if there is some somatization

Assess for possible psychogenic component

Pain out of proportion with imaging

Pediatric Chronic Pain is Common

- 11-38% of youth
- 5-10% have moderate - severe disability
- Prevalence increases with age; peak 14-15 yrs
- Girls > than boys

King et al, Pain, 2011

Prevalence of Back and Neck Pain by Age

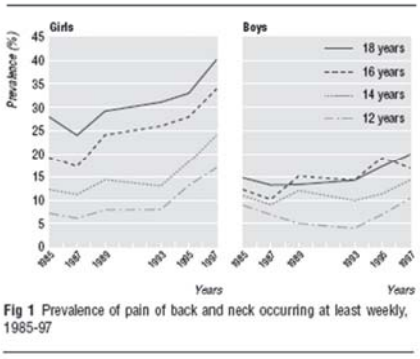


Fig 1 Prevalence of pain of back and neck occurring at least weekly, 1985-97

Hakala et al., BMJ, 2002

Impact on Children:

Missed School | Mood and Anxiety | Social Function | Physical Function | Sleep Problems

And Parents:

Missed work | Financial Stress | Emotional Distress | Changes in family roles | Stressful interactions with child

Mental Health Comorbidity

44% of youth <u>admitted for chronic</u> pain had mental health diagnosis:

- mood disorders (28%)
- anxiety disorders (18%)
- conversion and somatization disorders (6%)

26% of <u>general pediatric chronic pain sample</u> have mental health diagnosis

- <u>Increased risk for</u>:
  - anxiety disorders (OR 2.42)
  - eating disorders (OR 2.63)
  - depressive disorders (OR 2.32)
  - substance use disorders (OR 2.11)

Coffelt et al, 2013, Tegrethoff et al., 2015

Research Paper

PAIN

$19.5 BILLION

Health care expenditures associated with pediatric pain-related conditions in the United States

Cornelius B. Groenewald[a,b,*], Davene R. Wright[b], Tonya M. Palermo[a,b]

Abstract

The primary objective of this study was to assess the impact of pediatric pain-related conditions on health care expenditures. We analyzed data from a nationally representative sample of 6- to 17-year-old children captured in the 2007 National Health Interview Survey and 2008 Medical Expenditure Panel Survey. Health care expenditures of children with pain-related conditions were

Table 4

Prevalence, population estimates, total per capita, and total aggregated incremental health care costs associated with selected childhood health conditions among U.S. children (6-17 y of age).

Health condition	Prevalence, %	Population (in million)	Per capita incremental costs, USD	95% CI	Aggregated incremental costs (in billions of USD)	95% CI
Pain-related conditions	18.04	8799	1339	248 to 2447	11.8	2.18 to 21.5
ADHD	8.87	4327	2132	437 to 4181	9.23	1.89 to 18.1
Asthma	9.92	4837	1107	0 to 2537	5.35	0 to 12.3
Obesity	20.35	9923	74	−632 to 888	0.73	−6.28 to 8.81

Impact Extends into Adulthood

1/6 adult pain patients report chronic pain in childhood

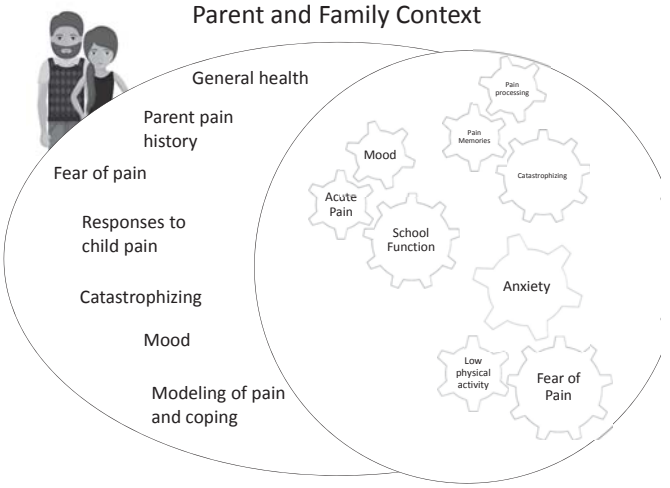
- Having pediatric pain associated with higher disability

Childhood pain increases adult risk for:

- Anxiety disorders (21.1 vs. 12.4%)
- Depressive disorders (24.5 vs. 14.1%)
- Lower household income and higher risk of unemployment
- Opioid misuse

Hassett et al., 2013; J Pain, Noel et al., 2016; Pain, Groenwald, 2019; J Pain

Parent and Family Context



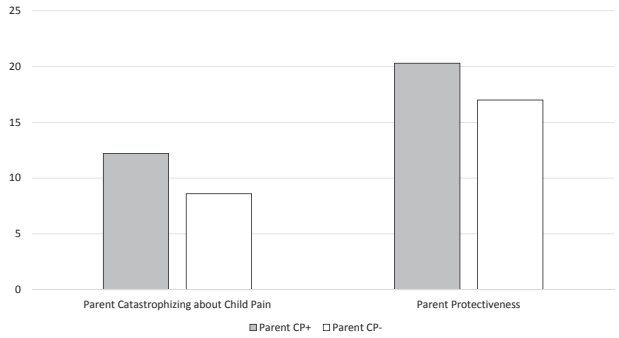
Parental Chronic Pain is Common

Who has a parent with chronic pain?

- Youth with chronic pain = 63%
- Healthy youth = 21%
- Youth seeking care for acute musculoskeletal pain = 60%
- Youth with JIA = 59% (non-arthritis pain in parents)

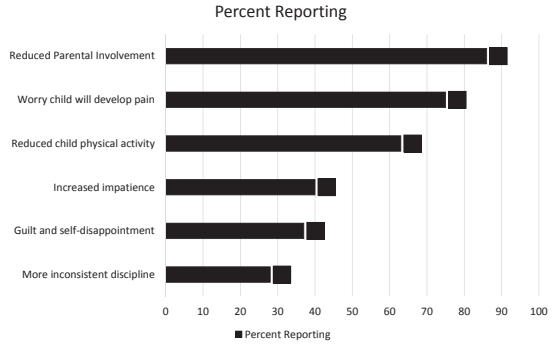
Piira & Pullukat, 2006; Campos et al., 2007; Schanberg et al., 2001; Clementi et al., 2019

Differences in Pain Responses: Parents with and without Chronic Pain



Wilson & Fales, *Clin J Pain*, 2015

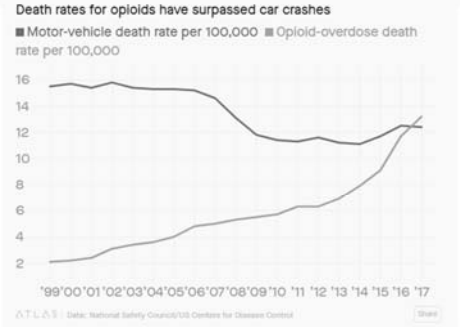
Qualitative Results: Impact of Pain on Parenting



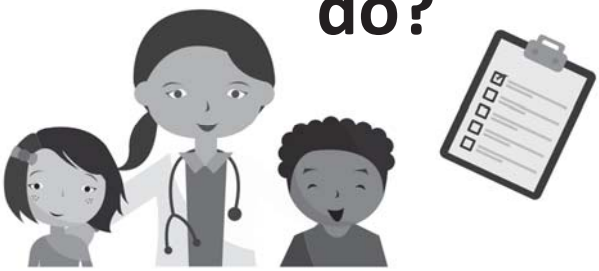
Wilson & Fales, *Clin J Pain*, 2015

The death rate for opioid use has surpassed car crashes in the US

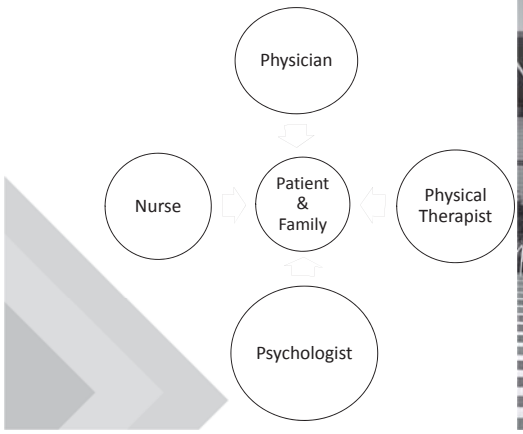
By Katherine Ellen Foley • January 15, 2019



What can we do?



The Ideal: Multidisciplinary Model

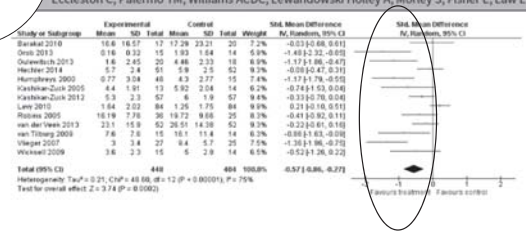


Psychological Interventions are Effective

Psychologist

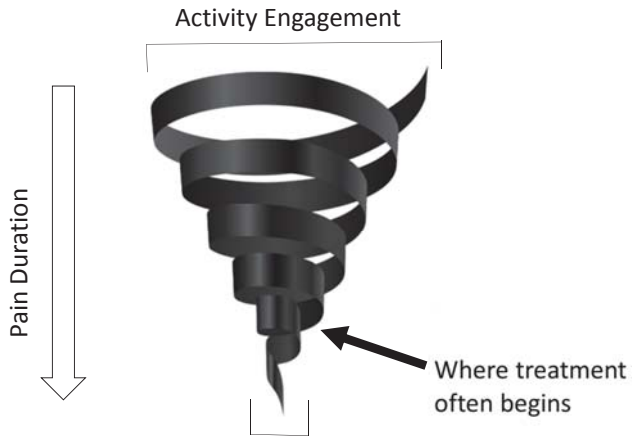
Psychological therapies for the management of chronic and recurrent pain in children and adolescents (Review)

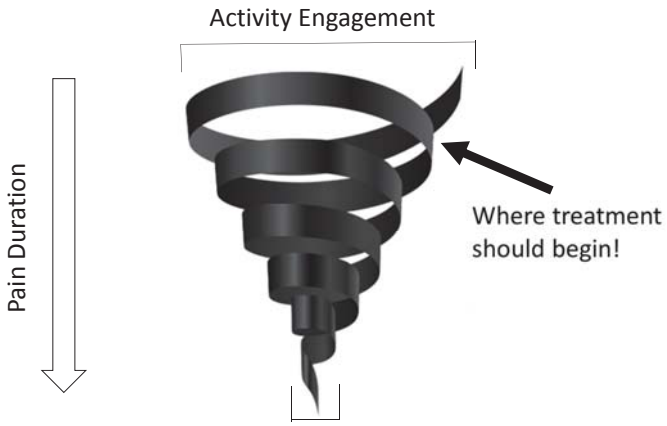
Eccleston C, Palermo TM, Williams ACDC, Lewandowski Holley A, Morley S, Fisher E, Law E



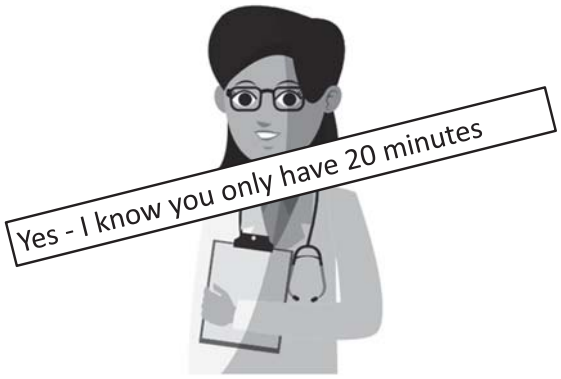
The Barriers to Care…

- **Limited availability of pediatric pain specialists**
- **Number of clinics/waitlists**
- **Transportation**
- **Insurance**
- **Provider unsure where to refer**





What can you do?



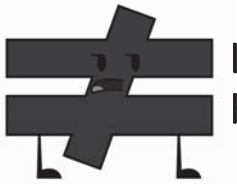
Explain Pain Neurobiology

The brain can sense pain even if imaging does not show tissue damage.

"Explain Pain", Butler & Moseley

And that….

Level of Pain



Level of Harm

Use Analogies

Chronic pain is like a car alarm

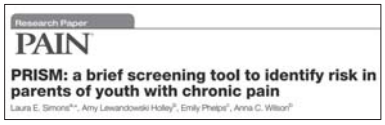
Persistent pain is like a doorbell that goes haywire

Chronic pain is a problem with the software

There is nothing wrong with the hardware in the body (e.g. bones, muscles, organs), but the software that sends messages throughout your system has a glitch

Coakley & Schechter, Pediatric Pain Letter; 2013

Assess Parent Risks/Supports


Research Paper

PAIN

PRISM: a brief screening tool to identify risk in parents of youth with chronic pain

Laura E. Simons*,+, Amy Lewandowski Holley[b], Emily Phelps[a], Anna C. Wilson[b]

Simons et al., 2018; Pain

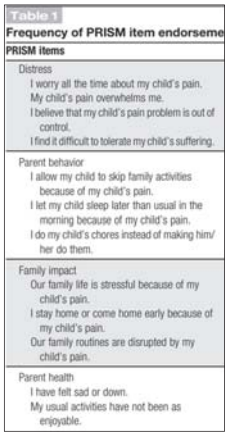


Table 1

Frequency of PRISM item endorseme

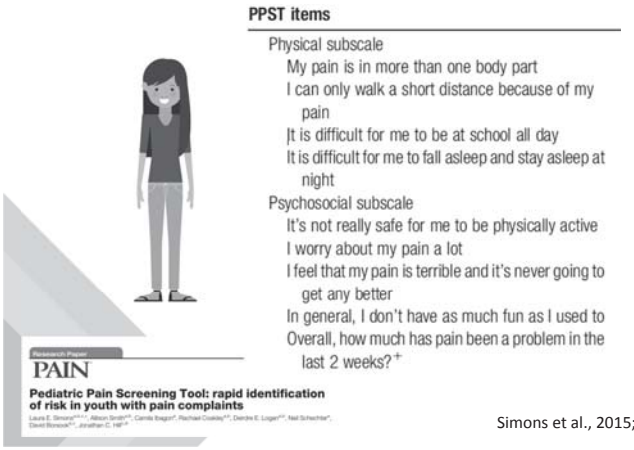
PRISM items
Distress
I worry all the time about my child's pain.
My child's pain overwhelms me.
I believe that my child's pain problem is out of control.
I find it difficult to tolerate my child's suffering.
Parent behavior
I allow my child to skip family activities because of my child's pain.
I let my child sleep later than usual in the morning because of my child's pain.
I do my child's chores instead of making him/her do them.
Family impact
Our family life is stressful because of my child's pain.
I stay home or come home early because of my child's pain.
Our family routines are disrupted by my child's pain.
Parent health
I have felt sad or down.
My usual activities have not been as enjoyable.

Risk Classification associated with Child Disability and Parent Behaviors

PRISM Risk Group	N (%)	Pain-related disability	Pain Intensity	Parent Distress	Protective Behavior	Parent Catastroph.
Low (0-3)	76 (33.2%)	16.8 (9.9)	5.9 (1.9)	8.0 (5.4)	23.8 (7.5)	22.4 (8.9)
Moderate (4-6)	66 (28.8%)	24.3 (10.5)	6.2 (1.6)	14.4 (5.9)	28.4 (8.2)	28.7 (10.2)
High (7-10	87 (38.0%)	27.4 (10.3)	6.1 (1.7)	18.1 (6.1)	31.8 (8.4)	35.9 (11.8)

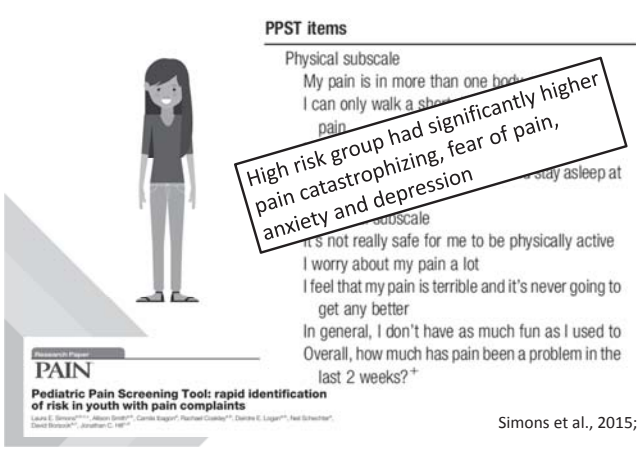
Simons et al., 2018; Pain

Assess Child Risks/Supports



Simons et al., 2015; Pain

Assess Child Risks/Supports

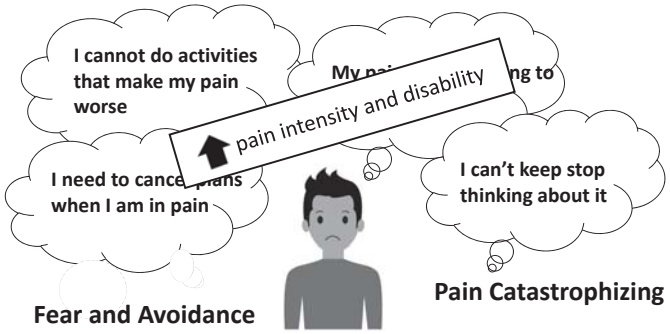


High risk group had significantly higher pain catastrophizing, fear of pain, anxiety and depression

Simons et al., 2015; Pain

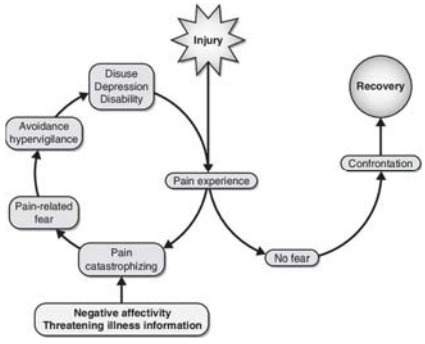
Recognize Pain Anxiety

Recognize Pain Anxiety



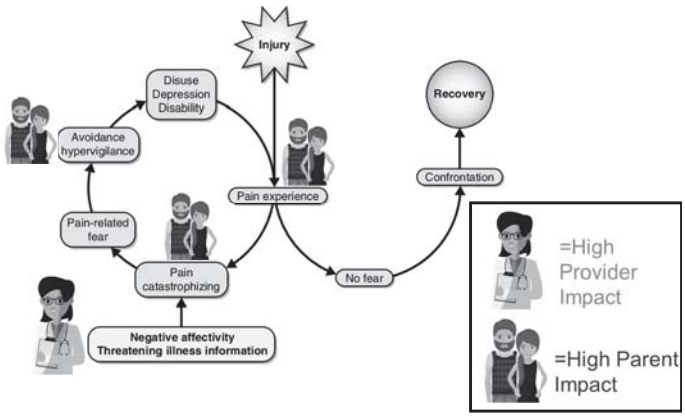
Chow et al., 2016; J Pain, Zale et al., J Pain 2013

Fear Avoidance Model



Vlaeyan, J.W.S. & Linton, S.J.; 2000, Pain

Fear Avoidance Model



Clinical Phenotyping of Youth With New-Onset Musculoskeletal Pain

A Controlled Cohort Study

Amy Lewandowski Holley, PhD, Anna C. Wilson, PhD,* Elise Cho, BS,† and Tonya M. Palermo, PhD‡*

- Fear of pain matters even in the acute pain period

T1 Predictor	B	SE	β
Step 2: Pain Intensity	.81	.76	.14
Sleep Quality	- 6.58	2.99	-.34*
Depressive Symptoms	.09	.14	.09
Trait Catastrophizing	- .17	.27	-.12
State Catastrophizing	- .27	.19	-.18
Fear of Pain	.35	.13	.51**
CPM Index	-.15	.08	.046

p<.05, ** *p*<.01 Total Model R^2 = .35, p<.001 Includes covariates: sex, age, ethnicity, BMI – all n.s.)

Holley et al., Clin J Pain, 2017

Parent Factors are Associated With Pain and Activity Limitations in Youth With Acute Musculoskeletal Pain

A Cohort Study

Michelle A. Clementi, PhD, Pari Farajl, MD,† Katrina Poppert Cordts, PhD,* Kelsey MacDougall, MA,‡ Anna Wilson, PhD,* Tonya M. Palermo, PhD,‡§ and Amy Lewandowski Holley, PhD**

- So do parent factors!

T1 Predictor	B	β
Step 2: Child Age	.17	.15
Child Sex	.77	.17
Fracture Status (yes/no)	- .36	- .08
Relation to Child	- 1.43	- .19
Parent Chronic Pain (yes/no)	1.06	.24*
Parent Somatic Symptoms	- .10	- .15
Pain Protectiveness	.76	.23*

* *p*<.05

Clementi et al., Clin J Pain; 2019

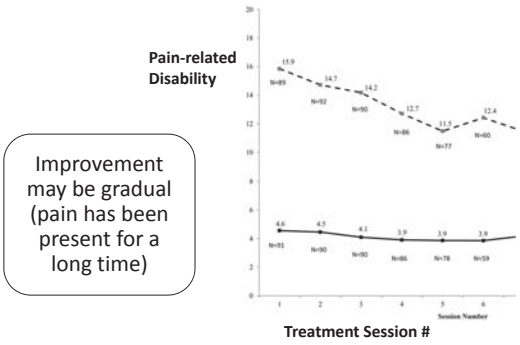
Set Treatment Expectations Early

- Treatment may have multiple components

4 flat tires: medication inflates only one

Set Treatment Expectations Early

- Function may improve before pain



Improvement may be gradual (pain has been present for a long time)

Lynch-Jordan et al. Pain, 2014

Give Parents Specific Recommendations

Limit pain check ins

Expect your child to gradually return to attending school

Reward your child for activity engagement

Explain how Parenting a Child with Chronic Pain Can be Counterintuitive

I need to let her rest so she can recover from her symptoms

I need to ask her about her pain so she knows how much I care about her

Its cruel to expect my daughter to engage in activities until her pain is gone

Know When to Refer to Behavioral Health

Who needs behavioral health interventions?

- **High fear avoidance impacting return to activity**
- **Parents who need to additional support implementing operant strategies**
- **School re-entry/504 plan development**
- **Co-occurring sleep problems**
- **Mental health assessment/treatment**

Send your patients to us!

Overview of the Parent Program

10:00–10:15 Welcome and staff introductions

10:15–10:45 Parent introductions & overview of program goals
- Group rules
- Please share about your child
- Goals of the parent program

10:45–11:00 Getting started
- Cognitive behavioral therapy
- Broadening the scope of the word "comfort"
- What does the word comfort mean to your child?
- Building long term comfort

11:00–11:30 Learning about pain
- Acute vs. chronic pain
- Why do some kids develop chronic pain?
- Pain and the brain
- Central Sensitization
- The mind body connection
- Pain and stress
- Pain and emotions

11:30–12:00 Parenting a child with pain
- Pain and child development
- First and second intuition parenting practices
- Reflective listening

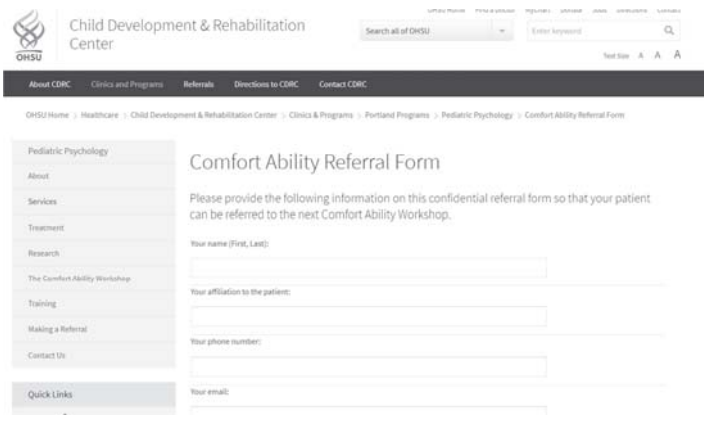
12:00–1:00 Break for lunch

1:00–1:45 Parent to parent guest speaker

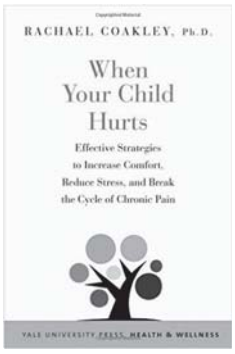
Program Goals

1. Expand your ideas of comfort
2. Understand how pain functions in the body
3. Practice relaxation skills for managing pain
4. Learn how thoughts, feelings, and actions are linked
5. Understand how stress and negativity decrease comfort
6. Try new strategies to help regulate mind and body
7. Identify active coping strategies
8. Practice how to set goals for your own recovery
9. Assemble a personalized Comfort Ability Plan
10. Review resources for continued support

You can submit online referrals through our website!
(Google search: "OHSU Comfort Ability")



We recommend this book!



https://www.amazon.com/When-Your-Child-Hurts-Strategies/dp/0300204655

Thank you!

NIH National Institute of Arthritis and Musculoskeletal and Skin Diseases

NIH National Institute on Drug Abuse

NIH NICHD

ARPPlab
Advancing Research in Pediatric Pain

QUESTIONS?

Craniofacial Medicine: Clinical Pearls and Common Cases

Emily Gallagher, MD, MPH

Doernbecher Annual Review and Update

October 17, 2019

Seattle Children's CRANIOFACIAL

Objectives

- Evaluating infant heads
- Understanding when to refer or not to refer
- Syndrome recognition

Seattle Children's CRANIOFACIAL

Evaluating infant heads

- Head size: when to worry?
  - Note relationship to other growth parameters
  - Measure parent/sibling head sizes
  - Developmental assessment
  - Few management guidelines exist!

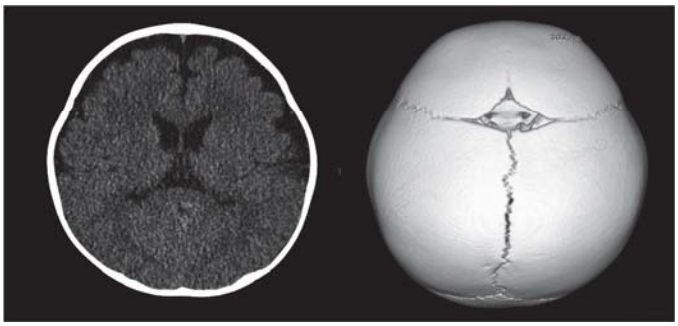
Seattle Children's CRANIOFACIAL

Fontanel size

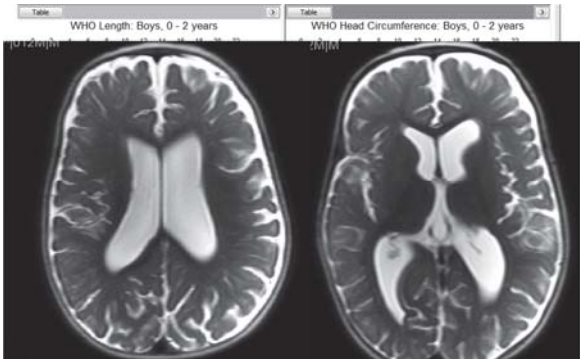
- Children with rapidly growing brains and normal bone have big fontanels
  - Hydrocephalus, benign macrocephaly
- Children with normal brains and poor bone growth have big fontanels
  - Hypothyroidism, cleidocranial dysplasia
- Children with poorly growing brains and normal bone have small fontanels
  - Primary microcephaly, hypoxic brain injury
- Children with normal brains and rapidly growing bone have small fontanels
  - Craniosynostosis, hyperthyroidism

Seattle Children's CRANIOFACIAL

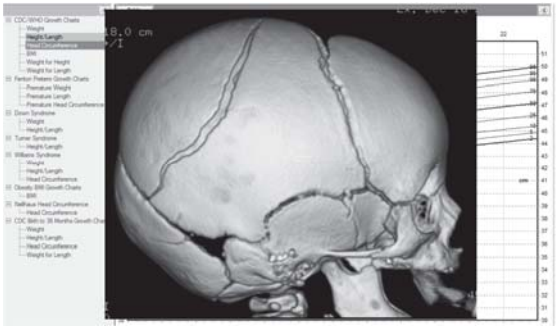
12 month old boy



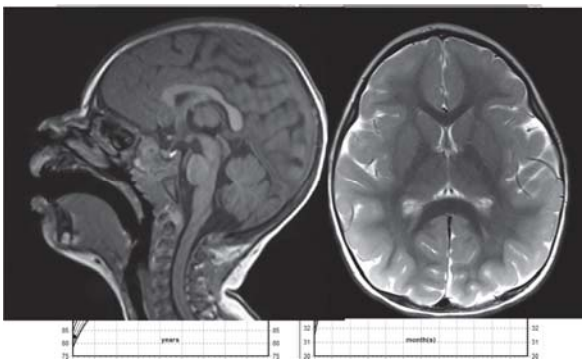
Another 12 month old boy



Previously healthy girl



2 year old girl, mild delays



Head size: when to worry?

- Macrocephaly
  - Associated with delays
  - Dysmorphic features
  - Departing normal growth curve
    - Hydrocephalus
    - Note parental head size
  - Common:
    - Benign familial macrocephaly
    - Increased extra-axial fluid
- Microcephaly
  - Hypoxic birth injury
  - CNS malformation
  - In utero exposure
    - Alcohol, drugs
  - Syndromes
  - Metabolic disorder
    - Maternal or infant
  - Congenital infection

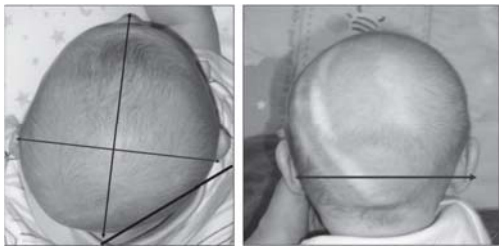
Mechanics of head shape differences

- Intrinsic: calvarial development
  - Craniosynostosis: premature fusion of infant suture
- Extrinsic: plagiocephaly
  - The Epidemic
  - Treatment: when is it "necessary"?

Deformational plagiocephaly

- Deformation of the calvaria from extrinsic forces
- Onset can be prenatal or postnatal
  - Prenatal: in utero molding or constraint
  - Postnatal: usually head position preference
- Natural history
  - Prenatal onset: spontaneous improvement
  - Postnatal onset: noticed at 1-2 months, worsens until 5-6 months

Most important views when examining a head



- Not a disease
- Parent's decision
- Emphasis on prevention
- Referral by 6 months

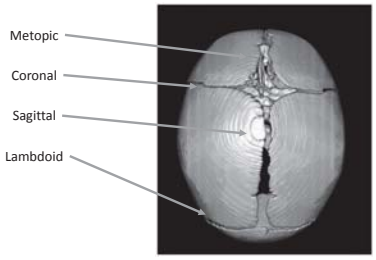
$3500

Johnny Jump Up $20-30 | Ergo $100 | Moby $50 | Bumbo $35 | Exersaucer $50 | Tummy time $0

Seattle Children's

CRANIOFACIAL

Calvarial sutures and normal closure

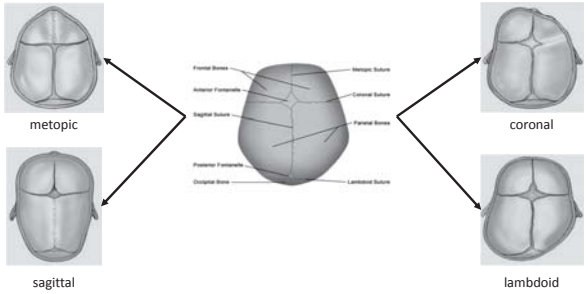


SUTURE	CLOSURE BEGINS
Metopic	3-9 months
Sagittal	22 years
Coronal	24 years
Lambdoid	26 years

Seattle Children's

CRANIOFACIAL

Single suture craniosynostosis

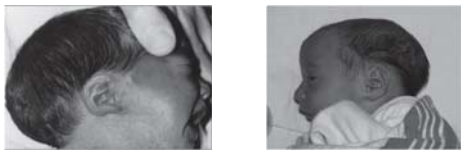


Seattle Children's

CRANIOFACIAL

A

B



Seattle Children's

CRANIOFACIAL

A

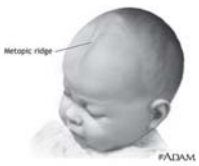
B

Seattle Children's

CRANIOFACIAL

A

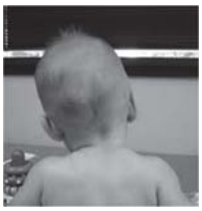
B

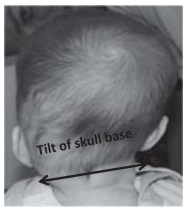


Seattle Children's

CRANIOFACIAL

A





B

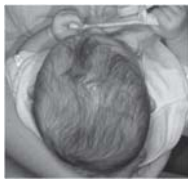
A

B

NAME THE DIAGNOSIS?

A

Metopic synostosis



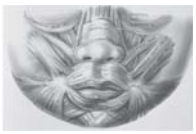
B

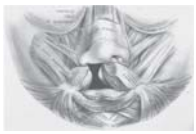
Positional plagiocephaly

C

Sagittal synostosis

Syndrome evaluation in patients with clefts





How often do patients with cleft lip and/or palate have syndromes or associated malformations?

- CLP: 15-25%
- CP: 50%

A

B

C

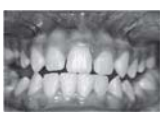
B: Midline cleft is never normal
Midline encephalocele

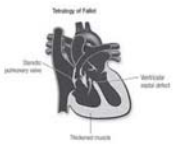
A

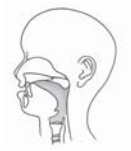
B

A: Holoprosencephaly

- Hypotelorism, depressed nasal bridge
- Midline cleft lip and palate
- Pyriform aperture stenosis
- Single central incisor







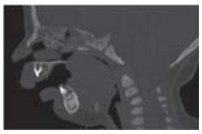
22q

22q PROGRAM

SEATTLE CHILDREN'S CRANIOFACIAL CENTER

Seattle Children's

CRANIOFACIAL

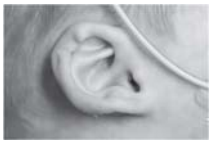


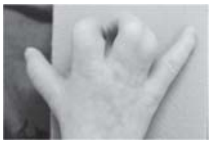
Robin Sequence
Micrognathia
Glossoptosis
Upper airway obstruction
+/- cleft palate

Stickler syndrome
~30% of children with RS

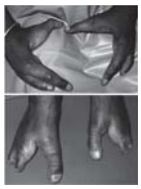
Seattle Children's

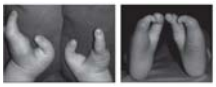
CRANIOFACIAL





TP63 Gene Mutations
- 1 gene, 6 syndromes
- Ectodermal dysplasia
- Clefting
- Sparse hair
- Risk of hyperthermia
- Cone teeth or hypodontia





Seattle Children's

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"Top Neurology Cases"

i.e. Headache, etc.

Doernbecher Annual Review

DATE: October 3, 2019 BY: Kaitlin Greene, MD
Director, Pediatric Headache
OHSU Department of Pediatrics, Division of Pediatric Neurology

Disclosures

- None

Outline:

- Case 1: Headache
- Case 2: Seizure
- Case 3: Stroke
- Discussion and Questions!

Case 1: Headache

- Goals:
  - Review indications for imaging in patient presenting with headache
  - Review diagnostic criteria for migraine and migraine with aura in children and adolescents
  - Outline approach to acute and preventive treatment of headaches
  - Be comfortable prescribing a triptan!

Case 1: Headache

- 13 year old girl presenting with worsening headaches
- When did headaches start?
  - ~~Six months ago~~ Age 8
  - Short (~1 hour), infrequent (<1x/month), typically triggered by illness or dehydration, improved with ibuprofen
  - Over the past two years, frequency gradually increased to 2x/month, then 4x/month, then to 2x/week by about 6 months ago

Case 1: Headache

Itsoktobeweird.com

- What are the headaches like?
  - Location: Mostly front, sometime back, sometimes more on one side or the other
  - Quality: Pressure (throbbing when severe)
  - Severity: Usually moderate, at least 2/month severe
- What are the associated features
  - "Sensory sensitivity": Light, sound, smell
  - Nausea when severe
  - Sees "flashes of light" for a few seconds with more severe headaches

Case 1: Headache

- PMH: None
- Family history:
  - Mom with "stress headaches" (Gets sensitive to light/noise, has to lie down)
  - Younger sister gets headaches when sick
- Medications:
  - Ibuprofen 200 mg as needed for headache
- Exam: Wt 50 kg. Normal including fundoscopic exam.

Case 1: Headache - Diagnosis

- What is the diagnosis? Migraine! With Aura?
- BUT first have to answer two questions:
  1. Are there any "red flags" to necessitate further work up?
  2. Does she meet diagnostic criteria for migraine or migraine with aura based on the International Classification of headache disorders, 3rd edition (ICHD-3)?

Case 1: Headache - Diagnosis

- Are there any "red flags"/indications for additional work up?
- "SSNOOPP"
  - Systemic symptoms (i.e. fever, rash, neck stiffness)
  - Secondary risk factors (i.e. medical co-morbidities, history of cancer, immunosuppression)
  - Neurologic signs or symptoms: focal symptoms or focal findings on exam
  - Onset: sudden, abrupt, maximum at onset ("thunderclap")
  - Older patient: age >50 (OR younger patient: age <6)
  - Progression and Prior headache history: major change in frequency, severity or clinical features, new headache type or pattern (<6 months headache history)

Dodick *Adv Stud Med* 2003

Case 1: Headache - Diagnosis

- Does she meet criteria for migraine without aura (1.1) based on the ICHD-3 [1]?
  - **A.** ≥**5 attacks** fulfilling criteria B-D
  - B. Headache attacks **lasting 2-72 hours** (untreated or unsuccessfully treated)
  - C. Headache has **at least two of** the following four characteristics
    1. Unilateral location (More often bilateral in children[2])
    2. Pulsating quality
    3. Moderate or severe intensity
    4. Aggravation by or causing avoidance of routine physical activity
  - D. During headache **at least one of** the following:
    1. Nausea and/or vomiting
    2. Photophobia and phonophobia (Can be inferred from behavior)
  - E. Not better accounted for by another diagnosis

Comment: "Migraine headache is usually frontotemporal. Occipital headache in children is rare and calls for diagnostic caution."

[1]ICHD-3 *Cephalalgia* 2018; [2]deGrauw *et al.*, *Headache* 1999

Case 1: Headache - Diagnosis

- What about occipital headaches? Is it rare? Does it call for diagnostic caution?

- Study 1: 432 children in the ED for HA[1]
  - **18/277** with discharge diagnosis (6%) had **"life-threatening headache"**
  - **3/18 occipital, 15/18** unable to localize
  - **17/18** had headaches for <2 months
  - **18/18 (100%) had objective neurological signs**
- Study 2: 150 children in the ED for HA[2]
  - **2/150** (1.3%) had occipital headache and *both* had brain tumors
  - **2/150** (1.3%) had brain tumors but did NOT have occipital headache
  - **4/4 (100%) with brain tumors had abnormal neurologic examinations**

[1]Conicella *et. al.*, *Headache* 2008; [2]Lewis and Qureshi, *Headache*, 2000

Case 1: Headache - Diagnosis

- Of children newly referred to Neurology and Headache Clinics, **6-16%** have occipital headache[1, 3]
- Children with occipital headache are more likely to get scanned BUT not more likely to find anything wrong![2, 3]
  - In children with solely occipital headache, 91% were scanned (RR 4.9, 1.2-21)
  - No significant difference in abnormal findings on MRI

[1]deGrauw *et. al.*, *Headache* 1999, [2]Bear J *et. al.*, AAN Abstract 2014, [3]Eidlitz-Marcus *et. al.*, *Pediatric Neurology* 2014

Case 1: Headache

- Occipital headache: Does it call for diagnostic caution?
  - Depends on the context!
    - In children presenting to the ED (or clinic) with NEW headache and ABNORMAL exam, caution is warranted regardless of location of headache
    - BUT in a child with a normal neurologic exam and a headache phenotype consistent with migraine, occipital head pain location alone is not necessarily associated with pathology

Case 1: Headache - Diagnosis

- What about aura? "Flashes of light for a few seconds"

1.2 Migraine with aura[1]:

A. At least two attacks

B. ≥ 1 of the following fully reversible symptoms:
   - Visual, sensory, speech/language, motor, brainstem, retinal

C. At least 3/6 characteristics:
   - Aura symptom spreads gradually over ≥5 minutes
   - ≥2 aura symptoms occur in succession
   - each individual aura symptom lasts 5-60 minutes
   - ≥ 1 aura symptom is unilateral
   - ≥ 1aura symptom is "positive"
   - aura is accompanied, or followed within 60 minutes, by headache

- Why does it matter?
  - Women with migraine with aura have a 2-fold increased risk of stroke more w/high-dose estrogen OCPs and smoking

[1]ICHD-3 *Cephalalgia* 2018

Case 1: Headache – Treatment

- Acute treatment: Decrease the duration and severity of the attack
  - Inadequate acute treatment optimization associated with a higher risk of developing chronic migraine within one year in adults[1]

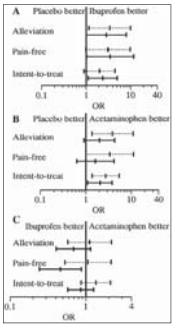
- Preventive treatment: Decrease the frequency of attacks over time
  - Consider when bothersome headache is occurring >1 day per week or >4 days per month

iStock.com

Lipton Neurology 2015

Case 1: Headache – Acute Treatment

- First-line: NSAIDs or Tylenol
  - Acetaminophen and ibuprofen both studied down to age 4[1]
    - Both superior to placebo
    - Ibuprofen 2x more likely to abort migraine at 2h
  - Consider longer-acting NSAID
    - Naproxen less likely to cause medication overuse headache and may have some preventive benefit[2,3]



1 Hamalainen Neurology 1997; 2 Lipton Neurology 2015; 3 Cady Headache 2014

Case 1: Headache – Acute Treatment

- Second-line: Triptans (5-$HT_{1B/1D}$ agonists)
  - Generally very safe and well-tolerated in children with healthy vessels!
  - Contraindications:
    - Underlying intracranial or cardiac vascular disease (including moyamoya, prior stroke, ischemic heart disease)
    - Uncontrolled hypertension
    - WPW
    - Specific aura types (hemiplegic migraine and brainstem aura)

Case 1: Headache – Acute Treatment

- **Four triptans** now FDA-approved for pediatric migraine

Triptan	Forms	Dose <40 kg	Dose >40 kg	Approval
Almotriptan	PO	6.25 mg	12.5 mg	12-17 yo (2009)
Rizatriptan	**MLT,** tab	5 mg	10 mg	**6**-17 yo (2011)
Sumatriptan/naproxen	PO (sumatriptan also NS and SQ)	10/60 mg – 85/500 mg (Sumatriptan alone: 25 mg (<40 kg) – 50 mg (>40 mg)		12-17 yo (2015)
Zolmitriptan	NS	2.5 mg	5 mg	12-17 yo (2015)

FDA.gov; Lewis *et. al., Pediatrics* 2007; Ho *et. al., Cephalalgia* 2012; Hewitt *et. al., Headache* 2013; Linder *et. al., Headache* 2008

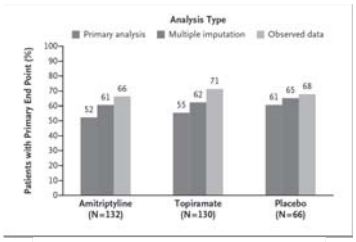
Case 1: Headache – Acute Treatment

- Triptan pearls:
  - Better to take early when pain is MILD (53% pain free at 2h)[1]
    - BUT okay to take when mod/sev (38% pain free at 2h)
  - Take with naproxen!
    - Higher 2h pain-free rate, lower 24h recurrence (adults)[2]
  - No need to re-dose
    - Safe but no evidence for better efficacy
  - Limit to <10 days per month to decrease risk of medication overuse[3]
  - Choose the formulation that makes the most sense!
    - PO, MLT, NS, SQ

1 Goadsby, *Cephalalgia*, 2008; 2 Brandes *et. al.,* JAMA 2007; 3 De Felice Ann Neurol 2010

Case 1: Headache –Preventive Treatment

- Topiramate is the ***only*** FDA-approved preventive treatment in children based on two positive RCTs
- What about CHAMP?
- Why??
  - Very high placebo-response rate, perhaps related to active co-interventions
    - Frequent visits with providers
    - Optimization of acute treatment
  - Patients with very refractory migraine or continuous headache excluded



Findings from CHAMP (Powers et al NEJM 2017)

Case 1: Headache

SPECIAL ARTICLE LEVEL OF RECOMMENDATION

Practice guideline update summary: Pharmacologic treatment for pediatric migraine prevention

Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology and the American Headache Society

Maryam Oskoui, MD, MSc, Tamara Pringsheim, MD, Lori Billinghurst, MD, MSc, Sonja Potrebic, MD, PhD, Elaine M. Gersz, David Gloss, MD, MPH&TM, Yolanda Holler-Managan, MD, Emily Leininger, Nicole Licking, DO, Kenneth Mack, MD, PhD, Scott W. Powers, PhD, ABPP, Michael Sowell, MD, M. Cristina Victorio, MD, Marcy Yonker, MD, Heather Zanitsch, and Andrew D. Hershey, MD, PhD

Correspondence American Academy of Neurology guidelines@aan.com

Neurology® 2019;93:500-509. doi:10.1212/WNL.0000000000008105

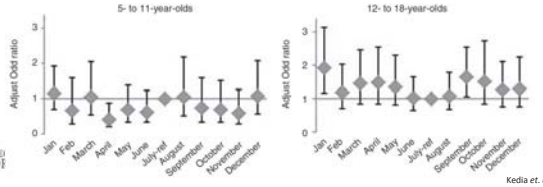
Case 1: Headache - Treatment

- Recommend discussing **lifestyle modification** and discussion of **modifiable risk factors** (Level B)
- Recommend **discussion of role or preventive treatments** in those with *frequent headaches, migraine-related disability and medication overuse* (Level B)
- Recommend **informing families of placebo response rates** in trials and that majority of preventives are not superior to placebo, with **shared decision making** about pros/cons of short-term treatment trials (Level B)

AMERICAN ACADEMY OF NEUROLOGY

Case 1: Headache – Preventive Treatment

- First-line: Lifestyle modifications!
  - "Regularity" seems to be key – regular exercise, regular meals, regular fluid intake, regular sleep
  - Among teens, significantly higher odds of presenting to the ED with headache in Jan and Sept



Kedia *et. al., Cephalalgia* 2013; CDC 2018

Case 1: Headache – Preventive Treatment

- Headachereliefguide.com

Case 1: Headache – Preventive Treatment

- Second-line: Over-the-counter medications/supplements
  - Riboflavin 200 mg BID (<40 kg: 100 mg BID)
    - Two negative RCT (very high placebo response rates)[1,2]
    - Recent positive placebo-controlled RCT[3]
  - Coenzyme Q10 100 mg BID (1-3 mg/kg/d)
    - One study in children with low CoQ10 showed decreased HA frequency with normalization of CoQ10 levels[6]
    - One RCT in children with trend toward efficacy[7]
  - Melatonin 3 mg QHS (<40 kg: 1-2 mg QHS)
    - One RCT in adolescents showed safety w/trend toward efficacy[4]
    - Uncontrolled studies showing decreased frequency[5]

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[1]Bruijn et al Cephalalgia 2010;[2]MacLennan J Child Neurol 2009;[3]Talebian Nutrients 2018;[4]Miano Neural Sci 2008; [5]Fallah Curr Drug Saf 2015; [6]Hershey Headache 2007; [7]Slater Cephalalgia 2011

Case 1: Headache – Preventive Treatment

- Third-line: Prescription medications
  - Should discuss evidence for *amitriptyline, topiramate, propranolol*
  - Should have *extended discussion of risks of medication* including concern for teratogenicity of valproic acid and topiramate

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Oskoui et al Neurology 2019

Case 1: Headache – Preventive Treatment

- What about the new anti-CGRP monoclonal antibodies?
  - Vasodilatory neuropeptide
  - Role in pathogenesis of migraine
    - Higher serum and saliva levels during migraine attacks
    - Levels decreased with triptan-induced pain relief
    - Infusion induces migraine in migraneurs only

	Site	Dose	Mode
Erenumab (Aimovig)	CGRP receptor	70 or 140 mg SQ monthly	Auto-injector
Fremanezumab (Ajovy)	CGRP molecule	225 mg monthly SQ or 675 mg SQ quarterly	Syringe/plunger
Galcanezumab (Emgality	CGRP molecule	120 mg or 240 mg SQ monthly	Auto-injector

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Russo Annu Rev Pharmacol Toxicol 2015

Case 1: Headache – Preventive Treatment

- Anti-CGRP monoclonal antibodies: expert consensus for use in adolescents

Table 1.—Suggested Indications, Contraindications, and Monitoring for the Use of CGRP mAbs in Children and Adolescents With Migraine

Indications	Contraindications	Monitoring
• ≥8 headache days per month • PedMIDAS score ≥30 • Failure of ≥2 preventive therapies (pharmacologic, nutraceutical, and/or non-pharmacologic) • Post-pubertal adolescent, or pre-pubertal child in selected cases	• Disturbed blood-brain barrier (eg, recent history of meningitis, recent neurosurgery) • Severe cardiovascular disease, stroke • Pregnancy, planned pregnancy or breast-feeding	• Pubertal status • Bone health, consider checking Vitamin D status • Linear growth • Weight/BMI • Infections • Pregnancy status

BMI = body-mass index.

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Szperka et al, Headache 2018

Case 1: Headache Prevention

- Cognitive Behavioral Therapy
  - In children and adolescents age 10-17, those who received CBT + amitriptyline vs Headache Education ("placebo") + amitriptyline had greater:
    - Reduction in headache frequency (SMD 0.48 [95% CI 0.14-0.82]
    - Likelihood of ≥50% reduction in headache frequency (RR 1.70 [95% CI 1.27-2.56]
    - Reduction in headache-related disability (SMD 0.43 [95% CI 0.09-0.77])

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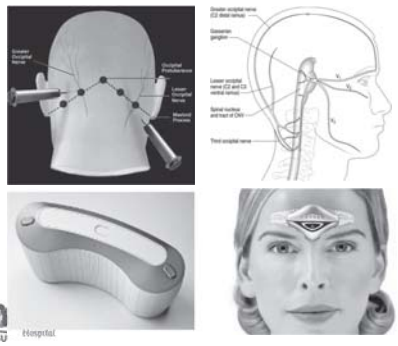
Powers et al, JAMA 2013

Case 1: Headache – Preventive Treatment



- Fourth-line:
  - Nerve blocks
  - Botox?
    - Insufficient evidence per practice parameter
  - Devices: TMS, Cefaly
  - Admission
    - DHE, thorazine, valproic acid

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Case 1: Headache

- Back to our case…
  - Acute plan:
    - Find a quiet place to rest
    - Mild/mod headache: Take naproxen 440 mg as needed up to 4 days/week
    - Mod/sev headache: Take sumatriptan 50 mg with naproxen 440 mg. Limit sumatriptan to 9 days per month.
  - Preventive plan:
    - Regular sleep, regular hydration, regular exercise, regular meals!
    - Take riboflavin 200 mg twice a day. This will take at least 8 weeks to see benefit.

Case 1: Headache – Take-Away Points

- "SSNOOPP" pneumonic for imaging indications
- ICHD-3: Excellent source for diagnostic criteria
- Migraine with aura: symptoms evolve/spread over ~5 min and last 5-60 minutes
- Acute treatment: NSAID +/- triptan (safe and approved in kids! With choice of formulation!)
- Preventive treatment: First, do no harm!
  - Emphasize on lifestyle and modification of risk factors
    - Think about CBT
  - New practice guideline from AAN/AHS in print
    - Amitriptyline, topiramate and propranolol may be considered

Case 2: Seizure

- Goals:
  - Identify features of spells concerning for seizure
  - Review differential for new onset seizures in childhood
  - Review general categorization of seizures
  - Outline steps of work up in a child with new concern for seizures
  - Discuss treatment indications and natural history

Case 2: Seizure

- CC: 4.5 yo boy with no significant PMH presents with three "spells" with alteration of consciousness over 10 days
- Description of spells:
  - Wakes from sleep and able to walk into mom's room
  - Behavioral arrest, unable to speak, appears "out of it", doesn't respond to mom's voice
  - On one occasion, made "gurgling sounds" in throat
  - No unusual movements of face or body, no LOC, no incontinence, no tongue biting, no post-ictal state
  - Duration: 45 seconds
- Any other symptoms?
  - Teacher has noticed some "staring spells" or "spacing out" episodes over the past 2-3 months
  - Has been more temperamental over the past 6 months (talking back, acting out)

Case 2: Seizure

- PMH: None
- Family history: First cousin with childhood epilepsy
- Medications: None
- Exposures: None
  - No recent illness
  - No known ingestions or possible ingestions
- Exam: Normal between attacks

Case 2: Seizure - Diagnosis

- Differential diagnosis:
  - Seizure
  - TIA
  - Parasomnia
  - Cardiogenic – arrhythmia, presyncope
  - Behavioral

Case 2: Seizure - Diagnosis

- What features are concerning for seizure?
  - Recurrent, stereotyped
  - Brief duration
  - Occurring out of sleep (or in sleep transition)
  - "Behavioral arrest"
- Commonly asked "seizure features"
  - Tongue biting (lateral)
    - 100% specificity, 30% sensitivity for seizure vs NES[1]
  - Urinary incontinence
    - 57% specificity, 38% sensitivity in differentiating syncope vs NES vs seizure[2]
  - Ictal eye closure
    - 80% specificity, 58% sensitive for PNES[3]

1Brigo et al Epilepsy Behav 2012; 2Brigo et al Seizure 2012; Brigo et al Seizure 2013

Case 2: Seizure - Diagnosis

- Differential etiologies for new-onset seizures in children
  - Structural lesions
  - Trauma
  - Vascular event (Ischemic or hemorrhagic stroke)
  - Infection (Meningitis, encephalitis)
  - Toxic (Ingestions, medication overdose)
  - Metabolic (Electrolyte disturbance, IEM)
  - Remote neurologic injury or abnormal brain development
  - Primary epilepsy

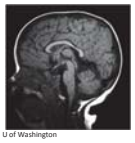
Case 2: Seizure - Etiology

- Types of seizures
  - Generalized seizures: Impaired awareness, bilateral motor symptoms
  - Focal ("partial") seizures: with our without impairment of awareness
    - Motor: may have spread ("Jacksonian march"), versive movement (head or eye deviation), vocalization or speech arrest (involvement of muscles of phonation)
    - Sensory: Paresthesias, distortion, olfactory or gustatory, auditory, visual
    - Autonomic: "Rising" sensation, sweating, pupil changes

Case 2: Seizure - Evaluation

- Laboratory evaluation and toxicology for patients seen in ED with first time seizure
- Head imaging
  - Emergent if concern for acute focal onset based on history, exam or EEG – rule out hemorrhage or ischemia
  - Outpatient MRI unless EEG confirms primary generalized epilepsy
- LP if febrile, concern for infection, not returning to baseline or <6 months of age[1]


Arimgsas.com.au

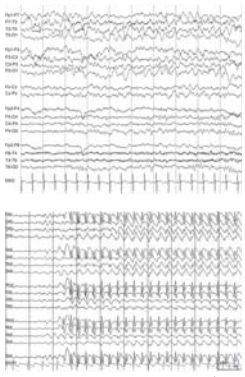

U of Washington


Aboutkidshealth.ca

American Academy of Neurology

Case 2: Seizure - Evaluation

- EEG recommended for all patients presenting with new-onset seizures
- When to get EEG?
  - Immediately post-ictal (<24 hours): can show generalized or focal slowing
  - If otherwise well and back to baseline, can be done as an outpatient
    - Consider more urgent EEG if not back to baseline or concern for subclinical seizure
- May provide insight into the etiology of seizure
  - Focal features OR characteristic findings of childhood epilepsy syndromes


Neupsykey.com

Case 2: Seizure - Prognosis

- Recurrence risk
  - All-comers: 42% recurrence
    - 88% of those in the first 2 years
  - Awake with normal EEG: 19%
  - Out of sleep with normal EEG: 37%
  - Out of sleep with abnormal EEG: 63%
  - >1 seizure in 24 hours: 41%

Shinnar Pediatrics 1996

Case 2: Seizure – Treatment

- "Seizure safety"
  - Caution around water, do not bathe or swim alone
  - No rock climbing or sky diving
  - Wear a helmet!
- Consider rescue medication if seizure was prolonged or child was endangered
  - First time seizure presenting in status has higher likelihood of recurring with status
    - Intranasal or buccal midazolam 0.2 mg/kg, max 10 mg
    - Rectal diazepam for younger children
  - Give instructions to call 9-1-1 with first administration

Case 2: Seizure – Treatment

- AAN guideline: Treatment with AED after first seizure may decrease risk of second seizure but dose not improve long-term prognosis[1]
- Recommend treatment after second afebrile seizure >24 hours apart
  - Focal seizures: Oxcarbazepine/carbamazepine, levetiracetam
  - Generalized seizures: Levetiracetam, topiramate, lamotrigine, valproic acid, zonisamide

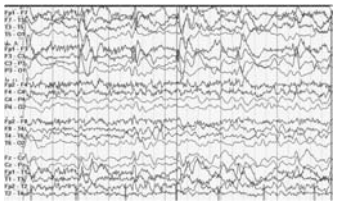
1Hirtz et al, American Academy of Neurology 2003, 2Specchio and Beghi CNS Drugs 2012

Case 2: Seizure - Treatment

- Duration of treatment: goal 2 years seizure-free[2]
  - 66-96% likelihood of seizure freedom at 1 year, 61-91% at 2 years
  - Higher risk of relapse: adolescent onset, underlying neurologic disorder, abnormal EEG

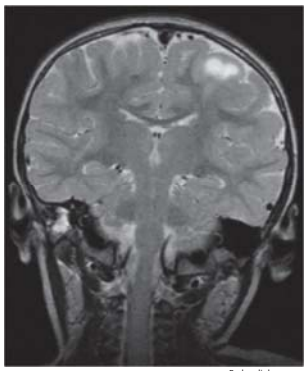
Case 2: Seizure

- Back to our patient…
  - Focal seizure by description
  - EEG showed significant left-sided epileptiform abnormalities and suggestion of underlying structural lesion



Case 2: Seizure

- Back to our patient…
  - MRI brain showed left parieto-occipital cortical dysplasia



Pedsradiology.com

Case 2: Seizure

- Back to our patient….
  - Started on levetiracetam on admission but switched to oxcarbazepine prior to discharge
  - Discussed future possibility of surgical intervention given focal cortical dysplasia

Case 2: Seizure – Take-Away Points

- Spell features concerning for seizure:
  - Stereotyped, behavioral arrest, occurring at sleep transition
  - Tongue biting>eye closure, incontinence to differentiate from NES and syncope
- Work-up of first-time seizure in Urgent Care/ED
  - Labs for all
  - Head imaging if acute focal onset or abnormal exam (otherwise outpatient)
  - LP if concern for infection or <12 months
- EEG indications: all patients with new seizures
  - Can be done outpatient unless not returning to baseline
  - Helps guide further work up
  - Helps predict recurrence risk
- Rescue medication for those presenting in status
- Initiation of AED after 2nd afebrile seizure

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Case 3: Stroke

- Goals:
  - Triage of acute onset of neurologic symptoms
  - Review basics of imaging techniques for stroke in children
  - Review of treatment protocol for acute stroke at OHSU
  - Recognize common presenting symptoms of stroke in children
  - Review risk factors for stroke in children
  - Review secondary work-up and stroke prevention in children

OHSU DOERNBECHER CHILDREN'S Hospital

Case 3: Stroke

- 8 year old previously healthy boy presents to the ED with acute onset of new headache, possible left facial droop and weakness 6 hours prior to arrival
- PMH: Unimmunized, limited primary care
- ROS: Fatigue, behavior changes and decreased PO for one week
- FH: No stroke, seizures, clotting or bleeding problems
- Exam: T 101F, VSS, follows commands on the R, R gaze preference, L facial droop, L upper and lower extremity weakness

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Case 3 - Stroke

- Differential
  - Stroke – ischemic or hemorrhagic
  - Seizure
  - Meningitis/encephalitis with focal infection
  - Migraine
  - Tumor or other lesion with acute change (hemorrhage)
- What next?

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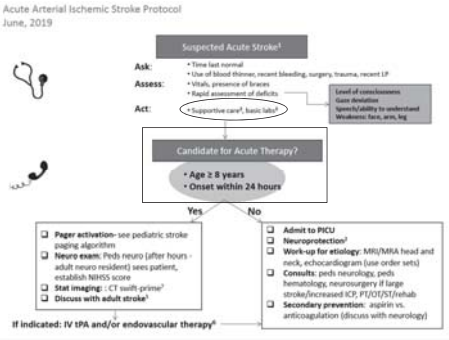
Case 3: Stroke



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Case 3: Stroke – Work-Up

- Labs: CBC, BMP, coags, type and screen, pregnancy test
- "Supportive care" while awaiting imaging
  - Bed rest with HOB flat
  - IV fluids
  - Neurochecks
  - Normothermia – avoid fever!!!
  - Normotension
    - Fluids for hypotension
    - Labetelol for hypertension
  - Consider AED if concern for seizure

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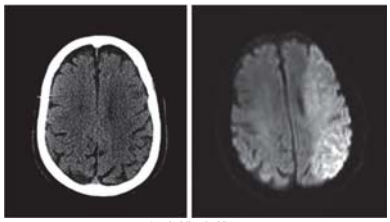
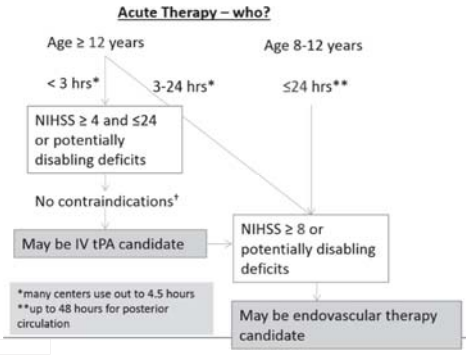
Case 3: Stroke – Work-UP

CT/CTA/CTP
- Very sensitive for blood
- Low sensitivity for acute ischemia
  - May show hypodensity after 6-12 hours
- Will show vessel occlusion
- Usually fastest to get!



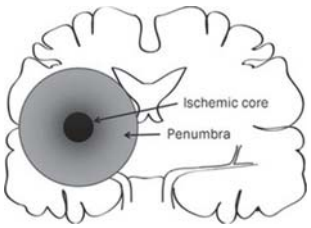
MRI/MRA
- Very sensitive for acute ischemia within minutes, up to 7-10 days
- Sensitive for blood
- Will show vessel occlusion

Case 3: Stroke – Acute Treatment



Case 3: Stroke – Acute Treatment

- Why the concern about timing?
  - Goal is to reperfuse the "penumbra" or "tissue at risk"
  - For tissue that is already infarcted, reperfusion increases risk
    - Hemorrhagic transformation
    - Reperfusion injury
    - Complications related to catheterization



Case 3: Stroke – Definitions and Epidemiology

- Stroke: "Acute onset neurological sign or symptom attributed to focal brain infarction or hemorrhage"
- 1-2 in 100000 children annually
  - Highest in children <5, boys>girls
  - "Neonatal" (>28 weeks gestation, <28 days postnatal) more common
- Etiology
  - Ischemic (~50%): Arterial ischemic stroke (AIS) or venous infarction due to cerebral sinovenous thrombosis (CSVT)
  - Hemorrhagic (~50%): intracerebral hemorrhage (ICH), intraventricular hemorraghe (IVH) or subarachnoid hemorrhage (SAH)

Ferriero et al, Stroke, 2019

Case 3: Stroke - Presentation

- Presenting symptoms:
  - Focal neurologic deficits
    - Hemiparesis and hemi-facial weakness (67-90%)
    - Speech disturbance (20-50%)
    - Vision disturbance (10-15%)
    - Ataxia (8-10%)
  - Altered mental status (17-38%)
  - Headaches (20-50%) – more common in children
  - Acute seizure (15-25%)

Ferriero et al, Stroke, 2019

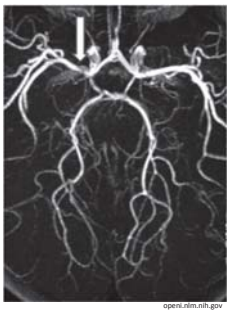
Case 3: Stroke - Etiology

- Cardiac (~30%)
  - Congenital heart disease
  - Endocarditis
  - Rheumatic heart disease
  - Arrythmias
- Vascular disease
  - Intracranial arteriopathy (~45%)
    - Focal Cerebral Arteriopathy (FCA)
    - Moyamoya
  - Extracranial arteriopathy (~7%)
    - Arterial dissection (esp posterior circulation)
- Hematologic
  - Sickle cell disease
  - Leukemia
  - Polycythemia
- Hypercoaguable state
  - Aquired: sepsis, nephrotic syndrome, liver failure, cancer, OCPs
  - Inherited: protein c/s deficiency, AT III deficiency, Factor V Leiden, MTHFR, prothrombin 20210
- Drugs
  - Cocaine
  - Chemotherapy (L-asp)
- Metabolic/Genetic
  - Homocystinuria
  - Fabry's disease
  - Fibromuscular dysplasia
  - Organic acidemias
  - Majewski's Osteopdysplastic Primordial Dwarfism, type II
  - Collagen vascular (e.g., Ehlers-Danlos)
  - SLE
- Neurocutaneous d/o's
  - Neurofibromatosis
  - Tuberous sclerosis
  - PHACE syndrome

Ferriero et al, Stroke, 2019

Case 3: Stroke – Focal Cerebral Arteriopathy

- "FCA": Unilateral stenosis and/or irregularity of the large intracranial arteries of the anterior circulation
  - Often involves junction of distal ICA and MCA/ACA
- Three types
  - Inflammatory
  - Dissection
  - Undetermined
- Has been associated with viral infections including HSV, VZV
- Course: Progression of symptoms over days-weeks, plateau over ~6 months, then subsequent improvement
  - BUT high 1-year recurrence rate (19-25%)

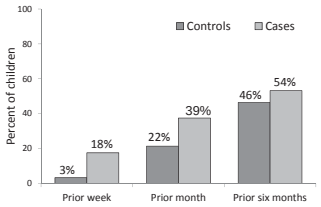


openi.nlm.nih.gov

Ferriero et al, Stroke, 2019

Case 3: Stroke - Etiology

- Infection as a risk factor for stroke
  - Large case-control international study of 355 children with AIS
  - 36% with definite arteriopathy, 10% with possible arteriopathy
  - Infection ≤1 week prior to stroke: 6.3-fold risk of AIS ($p<0.0001$; adjusted for age)
  - Unvaccinated: 7-fold risk of stroke ($p=0.0002$)



Fullerton et al *Neurology* 2015

Case 3: Stroke – Additional Evaluation

- Screen for common causes of stroke in children
  - Cardiac structure and function
  - Intracranial vessel imaging (including "vessel wall imaging" to look for inflammation of vessels if inflammatory FCA suspected)
  - Neck vessel imaging
  - Thrombophilia screening
  - Inflammatory markers
  - Screen for recent illness/infection
  - Lumbar puncture in the case of FCA
    - HSV PCR, VZV PCR and IgG/IgM

Ferriero et al, Stroke, 2019

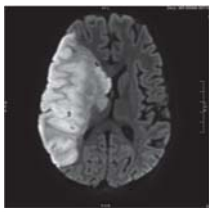
Case 3: Stroke – Secondary Stroke Prevention

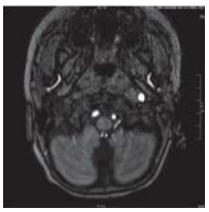
- High rate of recurrence
  - 10% for childhood ischemic stroke, 33% for arteriopathy
- No large studies to guide choice of antiplatelet vs anticoagulant therapy
  - For cardioembolic or thrombophlic stroke, consensus statement recommends anti-coagulation with LMWH or warfarin for 3-6 months
  - For all others, aspirin 3-5 mg/kg/d for ~2 years

Ferriero et al, Stroke, 2019; IPSS

Case 3: Stroke

- Back to our case…
  - MRI showed right MCA territory stroke with carotid occlusion





Case 3: Stroke

- Back to our case
  - RVP positive for parainfluenza 1 and 3
  - Not felt to be a candidate for acute intervention due to large territory of infarct and risk of reperfusion injury
  - Treated with aspirin and LMWH acutely, then long-term therapy with aspirin
  - Discharged home after inpatient rehab, ambulating independently

Case 3: Stroke – Take-Away Points

- Acute onset of focal neurologic symptoms is an emergency!
- CT is often faster, but MRI is more sensitive for ischemia
- Children > 8 years of age within 24 hours of onset of symptoms are candidates for acute intervention
  - tPA >12 years and <3 hours
  - Endovascular therapy >8 years and <24 hours
- Focal neurologic deficits are most common presenting symptoms
  - ***Headache*** and ***seizure*** also common in children
- Risk factors are more varied than in adults
  - Up to 45% of AIS in children are related to intracranial vasculopathy
  - Recent infection may be independent risk factor
- Long-term therapy typically includes aspirin for 2 years

OHSU DOERNBECHER CHILDREN'S Hospital

ON CONVERSION OR FUNCTIONAL NEUROLOGICAL DISORDERS

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Oregon Health & Science University
18 October 2019

Conflicts of Interest/Disclosure

I have no biomedical or financial conflicts of interest to disclose.

Who am I?

Craigan Usher

- Program Director, Child & Adolescent Psychiatry Training at OHSU
- Kienle Scholar for Medical Humanities through Penn State College of Medicine
- Assistant Editor—Book Forum, *Journal of the American Academy of Child & Adolescent Psychiatry*
- A few resource ideas:

LEARNING OBJECTIVES

By the end of this session, participants should be able to:

1) List three names that have been used to describe conversion phenomena
2) Name three stressors that are often "converted" in children/teens
3) Explain the psychoanalytic roots of the term conversion and what functional neuroimaging suggests are the underlying functional deficits that advance our understanding of conversion beyond the explanation offered by Freud
4) Discuss three ways to support youth with functional neurologic disorders

Why are we talking about?

DSM5 Criteria

A) One or more symptoms of altered voluntary motor or sensory function
B) Clinical findings provide evidence of incompatibility between the symptom and recognized neurological or medical conditions
C) This symptom or deficit is not better explained by another medical or mental disorder
D) The symptom or deficit causes clinically significant distress or impairment in social, occupational, or other important areas of functioning or warrants medical evaluation.

American Psychiatric Association. Diagnostic and statistical manual of mental disorders (DSM-5). American Psychiatric Pub; 2013 May 22.

Why are we talking about conversion disorder?

- Conversion disorder is a relatively common diagnosis in children and adults
- CD is rare before age 7 and common between ages 12-16
- Conversion disorders can lead to very significant distress, with patients, parents, school teachers, providers, and others often feeling confused, that there efforts are futile
- Up to 30% of new neurology outpatient visits may involve functional symptoms with 8% meeting criteria for conversion disorder
- 10-20% of patients with intractable epilepsy have non-electrical seizures (NES)
- The prognosis for adults is generally poor (50% improving), but many remain symptomatic
- The course of pediatric conversion is not well studied, but generally thought to improve more quickly

Hubschmid M, Aybek S, Maccaferri GE, Chocron O, Gholamrezaee MM, Rossetti AO, Vingerhoets F, Berney A. Efficacy of brief interdisciplinary psychotherapeutic intervention for motor conversion disorder and nonepileptic attacks. General hospital psychiatry. 2015 Sep 1;37(5):448-55.

Data on Prevalence & Characteristics

- Around 50% of children/teens have a "co-morbid" psychiatric disorder, the most common including anxiety and depression
- Often associated with/precipitated by stressors, including:
  - Family conflict
  - Bullying
  - Separation from a family member
  - Academic problems
- In 42 children at CHoP, in a 3yr period (02/2015 – 07/2018) they found:
  - Children with CD made up 10.7% of the CAP inpatient consults
  - Antecedent stressors (usually family structure, conflict) found in 95% of patients
  - A history of trauma found in only 14%
  - 25% demonstrated la belle indifference while 45% had moderate to severe distress
  - c/w other researchers, they found an even distribution of young men:young women at 13, but more females effected in later teens

Samuels A, Tuvia T, Patterson D, Briklin O, Shaffer S, Walker A. Characteristics of Conversion Disorder in an Urban Academic Children's Medical Center. Clinical pediatrics. 2019 Oct;58(11-12):1250-4.

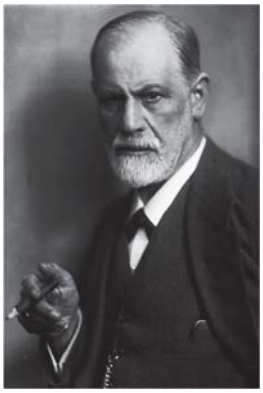
Vocabulary

"Hysteria" was first described by Egyptian and Greek philosophers and physicians and referred to a "wandering womb" etiological theory.

In the 19th Century, Jean-Marie Charcot noted that both men and women could suffer from "hysteria," but that male hysteria was due to trauma while female hysteria could be both traumatic *and* constitutional.

Charcot noted that many of his "hysteric" patient were more susceptible to hypnosis and that this may offer a cure.

A Clinical Lesson at the Salpêtrière by André Brouillet



Sigmund Freud's Original Conceptualization of "Konversion"

Originally wrote about conversion in "The Neuro-Psychoses of Defence" (1894) and in *Studies on Hysteria* (1895)

Conversion consists in a transposition of psychical conflict into, and it's attempted resolution through, somatic symptoms which may be either of a motor nature (e.g. paralyses) or of a sensory one (e.g. localised anasthesias or pains).

Essentially, Freud argued that "through bodily symptoms, repressed ideas 'join in the conversation'."

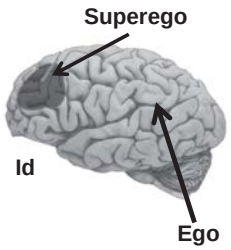
Pontalis JB, Laplanche J. The Language of Psycho-Analysis. New York: WW Norton; 1973. p90

Freud's Structural Model

Freud considered his models theoretical placeholders—until more sophisticated means of neural inquiry were available.

One can thus easily imagine Freud replacing his model with contemporary language, seeing:

1. The Id (*Das Es*) as the insistence of the Limbic System (amygdala, nucleus accumbens) pushing for pleasure or vengeance
2. The Ego (Ich) various regions of the posterior cortex responsible for how we represent the outside world
3. The Super Ego (*Das Über-Ich*) as the Prefrontal Cortex responsible for having a conversation restraint / top-down regulation

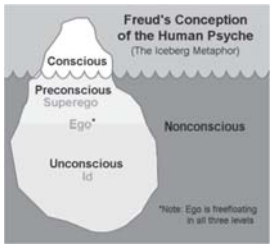


Freud's Topological Model

- Repression acts as a dam, actively keeping the individual keeping from conscious awareness painful thoughts, feelings, memories, and impulses.

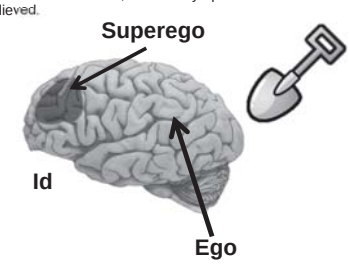
"If the perception of of reality entails unpleasure, that perception—that is, the truth—must be sacrificed."

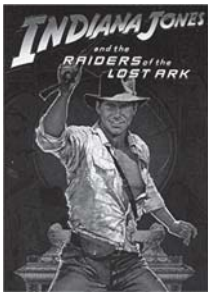
-Freud, SE XXIII, p 237



The Archaeological Model of Therapeutic Action

- If one could simply "dig" deeper, revealing to the patient what was being converted and hence kept from their awareness, then the symptoms could be relieved.



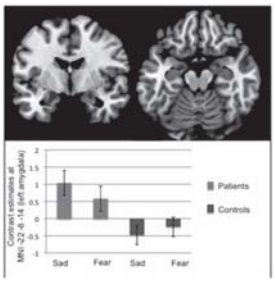


Classic Example: How this theory and "cure" are supposed to work

- A 12-year-old young man, Sam, discovers that his parents are not faithful to one another and are planning to separate. Sam cries and announces that he "can't stand this." When Sam wakes the next morning, his legs feel wobbly, his gait is unsteady, he has difficulty swallowing and he complains of nausea.
- That day, Sam sees his pediatrician for an urgent visit. She witnesses Sam's extremely abnormal gait that seems to change character. She finds that the patient's physical and neurological examinations are completely normal. Having read Freud and taken a clear history of the past 24 hours, Sam's doctor encourages him to recognize the link between his emotional pain, the traumatizing sudden rupture of expectation that he's gone through, his neurologic symptoms and things he's said to his parents ("you two make me sick" "I'm totally grossed out by you" "I can't stomach this" "I wont' stand for this" etc).
- With improved insight, the patient's symptoms resolve and he learns to cope with what he sees as his parents' betrayal.

PLEASE RAISE YOUR HAND IF YOU HAVE EVER HAD A REAL-LIFE CLINICAL EXPERIENCE THAT WORKED LIKE THIS.

CONVERSION DISORDER: NEUROIMAGING FINDINGS



Aybek S, Nicholson TR, O'Daly O, Zelaya F, Kanaan RA, David AS. Emotion-motion interactions in conversion disorder: an FMRI study. PLoS One. 2015 Apr 10;10(4):e0123273.

Roelofs JJ, Teodoro T, Edwards MJ. Neuroimaging in functional movement disorders. Current neurology and neuroscience reports. 2019 Mar 1;19(3):12.

Disrupted Emotional Processing

Traumatic experience may predispose individuals to having less activity in the supplementary motor area and greater activity in the R amygdala, insula, and bilateral posterior cingulate cortices.

Abnormal Sense of Agency

Agency is internal sense: "I made that happen." Neurologically, it appears to have an afterwardness.

Reduced activion in the temporo-parietal junction and reduced connectivity in sensorimotor cortex and cerebellum in pts w/ functional tremors.

Top Down Regulation Problems

One study found reduced activation of hand movement when observing hand movements (decreased mirroring) BUT NOT increased inhibitory control from frontal lobe.

TIPS FOR TALKING ABOUT CONVERSION DISORDER

Destigmatize & Legitimize

- This is a brain disorder. Period.
- What questions do you have about the nature of this problem?

Educate & Explore

- "Functional neurological disorders are common. They can be brought on by something painful in your life."
- "The amazing thing is, it come from your brain and your brain can be part of the solution."
- "But the part of your brain that CAN solve the problem, just doesn't know it yet. It needs training."
- Explore predisposing vulnerabilities, acute precipitants and perpetuating factors

TIPS FOR TALKING ABOUT CONVERSION DISORDER

Inquire about the details of a patient's (OT, PT, CBT, SLP) treatment

- Who are you meeting with?
- How often?
- What shared goals do you have?
- What therapeutic activities are you doing?
- Emphasize the importance of this work—call or email in front of your patient to collaborate.

Bring people up by their strengths, not their weaknesses

- Inquire about friends, hobbies, activities, things about which the patient is proud that do not relate to the functional deficit
- Demonstrate for parents how best to ignore/avoid reinforcing panic/concern about functional symptoms
- Highlight and reinforce engagement/patient's strengths and note that you would like to hear about an event or hobby, a favorite pet, book, movie, videogame, and perhaps for the patient to bring in a picture/sample at the next visit
- Encourage follow-up visits for progress NOT "if things are going well."

Adams C, Anderson J, Madva EN, LaFrance Jr WC, Perez DL. You've made the diagnosis of functional neurological disorder: now what? Pract Neurol 2018;18:323-330.

TIPS FOR TREATING CONVERSION DISORDER

Connect with School Personnel

- Create an assessment and safety plan
- Again, destigmatize and de-escalate sense of alarm that is often associated with PNES and other FNDs
- Clarify to whom they can reach out for support, when, and how

Document – Ideally in sharable EMR

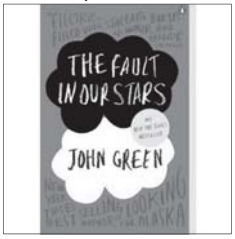
- Outline previous work-up and rationale behind diagnosis
- Delineate safety steps to take
- Note patient's strengths (the reader may not know that the Freudian archeological dig and reveal therapeutic approach is ineffective)
- Emphasize on-going outpatient treatment plan
- Clarify recommended treatment course that cautions against use of potentially habit-forming pharmacologic interventions

CONVERSION DISORDER: CASE EXAMPLES

A 12-year-old young man with urinary incontinence and LE weakness

A 16-year-old with NES, multiple ED and ICU treatments for acute episodes

A 13-year-old with headaches, weakness, speech articulation problems



QUESTIONS & YOUR CASES?

"Life can only be understood backwards; but it must be lived forwards."
- Soren Kierkegaard

LEARNING OBJECTIVES: REVISITED

So, today you learned that:

1) Conversion disorder and conversion phenomena have also been called:
   - hysteria
   - psychogenic disorders
   - non-organic syndromes
   - pseudoseizures
   - psychogenic non-epileptic seizures (PNES)
   - functional neurologic symptom disorder
   - functional neurologic disorder (FND)

2) Stressors that are often "converted" include:
   - family conflict
   - bullying
   - separation from a family member
   - academic problems

LEARNING OBJECTIVES: REVISITED

3) Sigmund Freud coined the term conversion disorder and he characterized this as a way that affects, ideas, and experiences that were actively being repressed by an dynamic unconscious force could "join the conversation" by being expressed neurologically. Functional neuroimaging has advanced this by discovering deficits in 1-emotional processing; 2-one's sense of agency; and 3-top down regulation/mirroring.

4) Some important ways of supporting children/teens and their families/friends/teachers:
- Combat stigma: these are real, treatable disorders
- Offer education and the neurological understanding of what maintains symptoms—it's skill opposed to will
- Refer and inquire about therapies including CBT, OT, PT; collaborate
- Bring people up by their strengths; reinforce these!
- Connect with schools
- Develop a treatment plan and place an alert/put this atop every note

CRAIGAN USHER, MD
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Tic Disorders and Tourette Syndrome

Evaluation, Diagnosis, and Treatments

October 18th, 2019 RP1

PRESENTED BY: Amelia B. Roth, MD

Slide 1

RP1 Enlarge font! Be proud!
Randall Phelps, 10/13/2019

Disclosures:

1. No financial disclosures
2. Clinical vignettes are used but patient information is protected
3. Off-label medication use is described, as is common in pediatrics

RP2

Vignette #1: RP3

- Smart, social 6 year old boy dx with ADHD at age 5 by PCP
- Continues to be disruptive, strong willed, anxious, and inflexible at home and school.
- Methylphenidate and Strattera have been tried, with mixed results…
- In the exam room he is fun and interactive, and frequently honks at me…
- On further questioning, he also has a history of repetitive throat clearing, grunting, crotch grabbing, and saying words over and over since toddlerhood

RP4

Slide 3

RP2 I prefer "vignette" to "Case". Case sounds cold and clinical. I think vignette sounds warmer.
Randall Phelps, 10/13/2019

RP3 Yeah, not so sure about this "case title". How about just giving him a name. Again, less objectifying.
Randall Phelps, 10/13/2019

RP4 I would leave this off for now. The implication here is that guanfacine is the treatment for TS, which, I don't think is the impression you want to leave. Yes, guanfacine can be a helpful and well-tolerated tool, but Rx shouldn't be emphasized as the primary cure for TS!
Randall Phelps, 10/13/2019

What is a tic?

- A fragment of normal behavior that occurs quickly and in isolation, but more repetitive and less variable
- Not voluntary and they are not involuntary, they are "*unvoluntary*"
- Can be easily described/reproduced by observers RP5
- Wax and wane, and can be suppressed at least temporarily
- Feels like an itch that has to be scratched or a sneeze that is hard to suppress RP6
- The tic itself is often not as much of a problem as the comorbidities…

Slide 4

RP5 No--I didn't coin this term. I heard it from Dr Sam Zinner of UW first, but I don't think he coined it either. I don't think you need to attribute the term.
Randall Phelps, 10/13/2019

RP6 subjectively, FEELS like an itch..
Randall Phelps, 10/13/2019

Tics versus Stereotypic Movements

- *Tics:* generally ego dystonic, most have a premonitory sensation and while they can be suppressed, tension exists when the tic is not released
- *Stereotypies:* ego syntonic, (though kids can become embarrassed by them), and suppression of the stereotypy does not cause as much tension
- Hand flapping, shuddering, complex hand movements, head nodding and banging, body rocking, sometimes accompanied by open mouth and staring, and sometimes vocalizations

Common Childhood Motor Tics

- Hard/frequent eye blinks, winks
- Eyes darting
- Facial grimaces, jaw movements
- Opening mouth
- Shoulder shrugging, neck stretching
- Torso shifting, jerking
- Hand to face/GU area/head/etc…
- Scrunching nose
- Copropraxia (rude gestures) and echopraxia (imitating gestures)
- Hopping, twirling, jumping
- Repetitive tensing of abdominal/limb muscles
- Truly dangerous tics are rare, but muscle soreness can occur, as opposed to stereotypes, which can include significant self-injurious behavior

RP9

Slide 6

RP9 contrast again with stereotypies. Stereotypies in contect of developmental disabilities, such as ASD or profound ID, can include significant self-injurious behavior
Randall Phelps, 10/13/2019

Common Childhood Phonic Tics

- Repetitive throat clearing
- Grunting, honking
- Meowing, hissing, barking
- Induced belching
- Making sounds with mouth
- Snorting, sniffing
- Gasping, sharp inhalations
- Short, sharp vocalizations: "oop" "eep"
- Rarely, coprolalia and echolalia, and palilalia (repeating own words)
- Hooting, shouting
- Words or phrases that are not part of a conversation (can be barked or grunted)

Premonitory Sensation

- Burning in the eye prior to a blink
- Tension in neck relieved with a stretch or jerk
- Feeling of tightness relieved with extension
- Kids get referred to PT's for "neck problems", and what is really occurring is a motor tic

"People believe that if you can shut off your Tourette's for a period of time, then you can always shut it off. I try to explain to people that if I spent my whole life trying to control my tics, that's all I would have time for." – Dash Mihok (actor)

RP7
RP8

Slide 9

RP7 I recommend consistency in font.
Randall Phelps, 10/13/2019

RP8 Great quote.
Randall Phelps, 10/13/2019

Types of Tic Disorders

- Transient: motor, phonic, or both for > 2 weeks and < 1 year
- Chronic Motor or Vocal Tic: Motor tic OR Vocal tic > 1 year
- Tourette Syndrome: At least 2 motor and at least one vocal tic > 1 year, (generally waxing and waning but mostly present)

Types of Tics

- Simple Tics: Sudden, brief, a limited number of muscle groups
- Complex Tics: coordinated between more than 1 muscle group (rolling eyes back while sniffing and shrugging shoulders)
- Complex Tic or OCD Ritual? Is a tic really a manifestation of OCD? On obsession followed by a compulsion?

Who gets tics?

- 1 out of 100 kids between 5 and 17 years of age has a tic disorder
- 1 out of 160 kids between 5 and 17 have Tourette Syndrome
- 3-4 boys diagnosed for every girl
- Tics tend to emerge around age 5/6, worsen around age 10/11, and improve by 18, then sometimes recur in middle age

Vignette #2: RP10

- 8 year old boy diagnosed with ADHD, ODD, and Social Anxiety at age 6 at the CDRC here for f/u
- Parents and Psychiatrist still think it's autism
- He has a 1:1 aid at school
- He is a perfectionist and easily escalates saying "I want to die", and now curses and hits walls
- He can be sweet, is eager to please, makes great eye contact, and is socially engaged. He hates that he curses and gets violent with objects…
- The only medicine tried so for was Risperidone
- I notice that older brother in room has a phonic tic…
- On further questioning, he makes a lot of random noises and m RP11 nents, and taps his forehead in a repetitive way...

Slide 13

RP10 again--I recommend "vignette" rather than "Case" and I would ditch the sub-titles.
Randall Phelps, 10/13/2019

RP11 where are you going with this vignette? Is there an epilogue? How do you address the diagnostic confusion (e.g. were they wanting ABA? DDS?) And what were the side effects of Risperidone?
Randall Phelps, 10/13/2019

Developmental Disability Services

- People seeking an autism diagnosis are sometimes seeking services…
- DDS offers respite care, personal support workers paid through the state, behavioral evaluations, and some money for the purchase of non-billable items (crash pads, sensory tools)
- Tourette Syndrome is now an eligibility for DDS, provided there is proof of global functional impairment, as are the diagnoses of an Autism Spectrum Disorder, Intellectual Disability, Global Developmental Delay, and FASD

Tourette Syndrome

- Most have normal IQ
- School performance often affected by OCD, anxiety, and ADHD
- Onset between ages of 2 and 15 years, the mean is around age 6 or 7 years
- Tics tends to be most severe in late childhood/early teen years
- Half of kids are tic free by age 18, though they can come back in middle adulthood
- Remember, mild cases are more common than severe cases!
- Only 15-20% have coprolalia or copropraxia

Vignette #3

RP12

- 14 year-old boy comes in with mom
- "Does he have autism or is he just a (a jerk)?", mom asks in front of son
- Difficulty making friends; annoyed with others easily
- Many annoying habits, including throat-clearing, coughing, making body function noises, bouncing, tapping, head-rolling, and fidgeting
- Teased about these behaviors and he would like to stop
- He has been diagnosed in the past with ADHD and treated with stimulant, which caused exacerbation of sounds/movements, weight-loss, and diminished energy. He has begun to hoard things and was dx with OCD.
- Aggression towards sister and cat had escalated and the family was beginning to consider residential treatment…
- A psychiatrist dx high functioning autism and prescribed an anti-psychotic medication, with some improvement in behavior, but also significant weight-gain and sedation

Slide 16

RP12 again: change to vignette, and use pseudonym instead of sub-title.
Randall Phelps, 10/13/2019

Vignette #3

RP13

- On Exam he is pleasant, cooperative, with typical social referencing and reciprocity, typical prosody of speech
- A few subtle tics seen in office, some fidgetiness
- ADOS—non-clinical
- Normal cognitive and language skills
- Now he is obese, secondary to atypical antipsychotic med
- He gained 30 lbs. in one year, and kept increasing doses
- He is now teased more for his weight than for his tics…

RP28

Slide 17

RP13 vignette, cont'd--data
Randall Phelps, 10/13/2019

RP28 Note that the obesity was the direct result of Risperidone, with 30 lbs weight gain in 1 year on it. Note that the Risperidone helped with tics initially, but that the benefits waned, necessitating increases in doses over the year. Note that child now says that he is teased more for being fat than he was ever teased for tics and that he would rather tic than have the extra 30#, and that he tics now anyway on Risperidone
Randall Phelps, 10/13/2019

Conclusions RP14

- Tourette syndrome, with secondary social impairments.
- The key is that the teen was very bothered by these habits.
- With new diagnosis, mom softened and was more receptive to him
- He was referred to counseling, and school accommodations where recommended, as well as sports/exercise
- On follow-up he was doing well, both academically and socially, and off of all medication

Slide 18

RP14 conclusion:
Randall Phelps, 10/13/2019

Comorbidities RP15

- ADHD
- Anxiety and OCD (20-40% have OCD, almost all have some elements of OCD)
- If you have OCD, you have a 20% risk of developing tics and 7% risk of TS
- Mood challenges
- "fiery temperaments"
- Social Development challenges
- Sleep challenges and parasomnias
- Comorbidities are often a bigger challenge than the tics!
- Target treatment to whatever causes the most interference with functioning

Slide 19

RP15 emphasize that the co-morbidies are OFTEN a bigger problem for folks than the tics themselves! Target treatment to whatever causes the most interference with function/participation!
Randall Phelps, 10/13/2019

Heritability

- Tourette Syndrome tends to be a highly penetrant dominant trait, males tend to have ADHD and tics, females tend to have OCD (externalization versus internalization)
- Stimulants provoke tics in predisposed kids, as can steroids, stress, illness, and lack of sleep

RP16

Slide 20

RP16 note that, in general, males externalize and females internalize, so this makes sense
Randall Phelps, 10/13/2019

Worsening Factors

- Sleep deprivation/Exhaustion
- Anxiety
- Excitement
- Anger
- Illnesses – virus, strep… RP17 RP20
- Pain, injury
- Being alone (feeling more comfortable) RP18
- Lack of exercise
- Feeling too hot or too cold
- Sensory irritants like tags, turtle necks, tight or itchy clothes

Slide 21

RP17 do you want to mention PANDAS here? That it seems that the issue is that infections generally increase tics and OCD sx, as well as other behavioral symptoms? Not necessarily immune-mediated--but that there is that hypothesis?
Randall Phelps, 10/13/2019

RP20 Ah--never mind. I see next slide. Good.
Randall Phelps, 10/13/2019

RP18 this could relate to feeling comfortable ticcing when alone
Randall Phelps, 10/13/2019

RP19

Do you Believe in Pandas?

- Tics and OCD tend to worsen with illness, and particularly with strep
- There is a theory that it's an immune mediated process, similar to Sydenham's Chorea

Slide 22

RP19 Randall Phelps, 10/13/2019

Alleviating Factors

- Sleep
- Calm
- Focusing on a task
- playing a musical instrument, (drums!)
- Vigorous exercise
- Regulating body temperature
- Staying healthy

RP21
RP22
RP24

Lifestyle and Behavioral Management

- **First:** optimize sleep! Decrease screen time!
- **Second:** optimize physical activity and outdoor time
- **Third:** get child into a physical or musical activity they enjoy like martial arts, running, swimming, ball sports, drumming, other musical instruments
- **Fourth:** Cognitive Behavioral Therapy (CBT) for anxiety/OCD and Comprehensive Behavioral Intervention for Tics (CBIT)
- Parents and teachers can redirect or distract when child is having tics, but should not keep asking child to stop, or make the child feel ashamed
- Celebrate neurodiversity in the home, school, and community

Slide 24

RP21 Great slide!
Randall Phelps, 10/13/2019

RP22 Add that working with schools is very important here--recommending 504 or IEP with scheduled sensory/tic breaks is important!
Randall Phelps, 10/13/2019

RP24 Ah, I see you got to this later, too--good!
Randall Phelps, 10/13/2019

Comprehensive Behavioral Intervention for Tics

- 1. Training the patient to be more self-aware of tics (but not more self-conscious)
- 2. Training the patient to do competing behaviors when they feel the urge to tic (slow breathing instead of throat clearing) → so, not suppressing the tic (which is exhausting), but practicing behaviors that are incompatible with ticcing until the urge goes away
- 3. making changes in daily routines that can be helpful in reducing tics (manage anxiety and stress)
- 4. Many people living with tics already use similar strategies they have discovered on their own

Medical Management: Optimize Sleep

- First, optimize sleep!
- Start with 0.25 mg Melatonin at bedtime if sleep onset is challenging, slowly increase as needed
- Next step would be Clonidine, start with 0.05 to 0.1 mg at bedtime
- Consider adding in long-acting Clonidine if waking up in night and ticcing
- If sleeping very well, AND still having problematic day time tics, consider day time medications as well, such as guanfacine

RP23

Slide 26

RP23 As you know, I agree. Clonidine helps with sleep and reduces tics, so it's a great choice. But, just to note: sleep specialists in attendance may object. One response to such an objection would be that not treating sleep ticcing can result in need for stronger Rx, with more side effects, so it is often in child's best interest to treat with Clonidine or Guanfacine...
Randall Phelps, 10/13/2019

Medical Management: Day Time

- Consider starting guanfacine, usually short acting
- For young kids, start with 0.25 mg BID, then can slowly increase as needed
- If starting long acting guanfacine, start at night if not already on clonidine, then move to AM once adjusted to soporific effects
- Once sleep is optimized, and day time tics are improved, consider addressing ADHD if needed with stimulants
- Consider managing anxiety/OCD with an SSRI if needed

Medical Management for ADHD in kids with tics

- Stimulants usually worsen tics, but occasionally can help
- Kids with Tourette Syndrome/Tics tend to do better with stimulants when used synergistically with alpha agonists
- Kids tends to do better with Dexmethylphenidate (Focalin) than Methylphenidate (Ritalin)
- Strattera can be helpful for some, though many report feeling unwell on this

Tips and Tricks in the Classroom

- Consider a 504 plan to allow for tic accommodations, or an IEP if significant ADHD also present interfering with learning
- Tic Breaks, or timing tics with other loud noises in the class (such as clapping or laughing)
- Sports water bottle at desk can help
- Chewing gum
- Fidgets in the hands or pockets like putty, pieces of felt
- Movement breaks
- Subtle hand signals between teacher and student to communicate needs
- Treat the underlying Anxiety, OCD, ADHD, Sleep Disorders

Resources for Families

- The Tourette Association of America, www.tourette.org, established in 1972
- Check out the video: "I have Tourette Syndrome but Tourette Syndrome Doesn't Have Me"
- If there are global adaptive impairments, kids can be RP26 eligible for Developmental Disability Services, and possibly SSI depending on family income

Slide 30

RP26 Yes. Highlight this earlier? in reference to a vignette? Perhaps one of the vignettes where the family insists on ASD--they want ASD so they can get DDS?
Randall Phelps, 10/13/2019

Adolescent suicide prevention:
Risk screening, assessment, and safety planning

Melissa Weddle, MD, MPH
Pediatric Review and Update
October 18, 2018

OHSU DOERNBECHER CHILDREN'S Hospital

Objectives

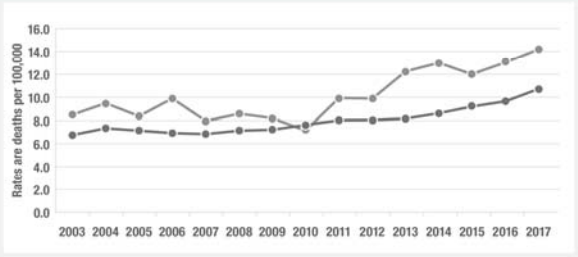
- Recognize adolescent suicide risk
- Identify strategies for screening of suicide risk
- Describe assessment and management of those at increased risk

PART 1

The Evidence
FOR SUICIDE RISK SCREENING

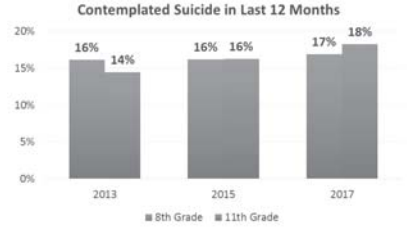
Youth Suicide in Oregon

Figure 1: Suicide rates among youth aged 10 to 24 years, U.S. and Oregon, 2003-2017



Source: CDC WISQARS and OPHAT

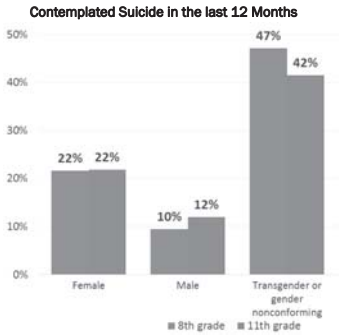
Youth Suicide in Oregon



PUBLIC HEALTH DIVISION
Adolescent and School Health

Source: 2013, 2015, 2017 Oregon Healthy Teens Survey

Youth Suicide in Oregon

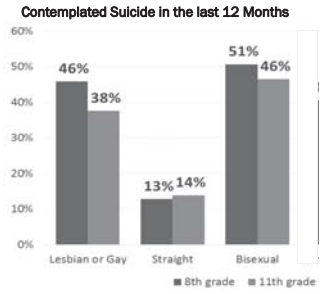


PUBLIC HEALTH DIVISION
Adolescent and School Health

Note: "Transgender or gender.." includes those who identified as transgender, gender fluid, genderqueer, gender nonconforming, intersex/intergender, multiple responses, and "not sure of gender"

Source: 2017 Oregon Healthy Teens Survey

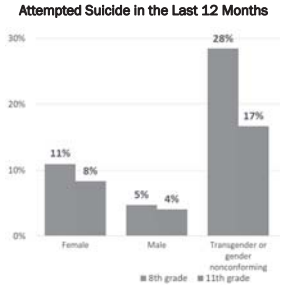
Youth Suicide in Oregon



PUBLIC HEALTH DIVISION
Adolescent and School Health

Source: 2017 Oregon Healthy Teens Survey

Youth Suicide in Oregon



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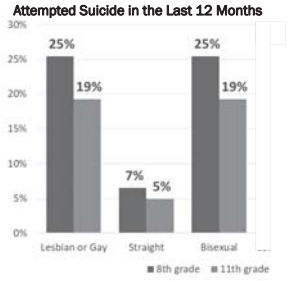
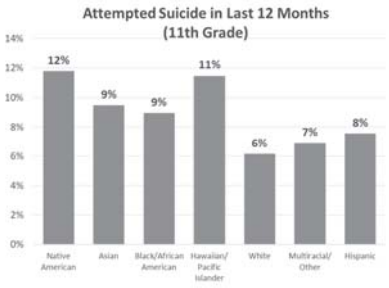


PUBLIC HEALTH DIVISION
Adolescent and School Health

Source: 2017 Oregon Healthy Teens Survey

Youth Suicide in Oregon

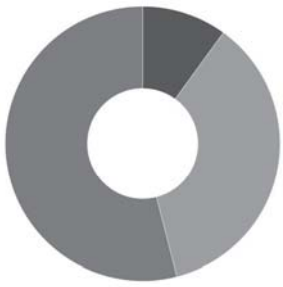


PUBLIC HEALTH DIVISION
Adolescent and School Health

Source: 2017 Oregon Healthy Teens Survey

Youth Suicide in Oregon

Suicide deaths by age, Oregon 2017



Source: Oregon Violent Death Reporting System

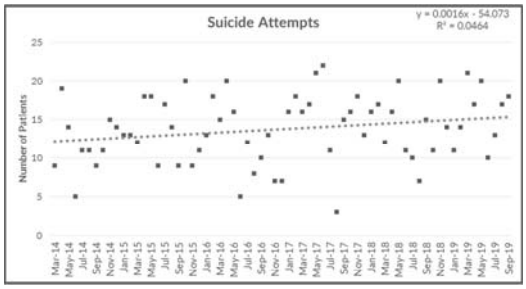
10-14 10% 15-19 36% 20-24 54%

Youth Suicide in Oregon

Suicide deaths by gender, Oregon 2017

Female 20% Male 80%

September 2019	2019 Year to Date
47 youth served	414 youth served
38% of cases were suicide attempts	34% of cases were suicide attempts
89% of suicide attempts were overdoses	89% of suicide attempts were overdoses
18 lockboxes provided to families	107 lockboxes provided to families
38% of referrals were from the ED	50% of the referrals were from the ED
34% of patients went to inpatient 9% went to subacute 2% went to residential 55% were discharged to outpatient	23% of cases went to inpatient 8% went to subacute 2% went to residential 68% were discharged to outpatient
70% of patients were female 28% of patients were male 0% of patients were trans: feminine 2% of patient were trans: masculine	62% of patients were female 35% of patients were male 0% patients were trans: feminine 3% of patient were trans: masculine



PART 2

Recommended
SCREENING & ASSESSMENT TOOLS

National Recommendations

American Academy of Pediatrics recommends that pediatricians ask questions about mood disorders, sexual orientation, suicidal thoughts, and other risk factors associated with suicide during routine health care visits

American Academy of Child and Adolescent Psychiatry recommends that physicians be aware of patients at high risk for suicide

American Medical Association Guidelines for Adolescent Preventive Services recommends that all adolescents be asked annually about behaviors or emotions that indicate risk for suicide

Why should Primary Care Practitioners Screen?

- Suicide is the #2 cause of death of 10 – 24 year olds
- 70% of adolescents seen by PCP annually
- Adolescents more comfortable with PCP
- Patients who died by suicide visited PCPs over 2 times as often as mental health clinicians

Barriers to PCP Screening & Assessment

Time **32.8%**

Adequate training **25.5%**

Adequate knowledge **32.9%**

Comfort discussing suicide **64.2%**

Why screen in the hospital or ED?

- 30% of adolescents have not been seen by a PCP in the past year
- PCP may not have screened or had adequate training

Minor Consent and Confidentiality

ORS 109.675 - a minor who is 14 years or older may access outpatient mental health, drug, or alcohol treatment without parental consent

ORS 109.860 - for mental health and chemical dependency services, the provider may disclose health information to a minor's parent or guardian if:

- It is clinically appropriate and in the minor's best interests
- The minor must be admitted to a detoxification program
- The **minor is at risk of committing suicide** and requires hospital admission.

Confidentiality Exceptions:

- Risk of harm to self or others
- Abuse

Risk Factors for Suicide

- Family history of suicide or child maltreatment
- Previous suicide attempt(s)
- History of trauma and/or personality or mood disorders
- History of alcohol and substance abuse

Risk Factors for Suicide

- Feelings of hopelessness
- Isolation
- Barriers to accessing mental health treatment
- Loss (relational, social, work, or financial)

Warning Signs

- Talking about wanting to die
- Talking about being a burden to others
- Increasing use of alcohol or drugs
- Acting anxious or agitated, behaving recklessly

Warning Signs

- Sleeping too little or too much
- Withdrawing from family or friends or feeling isolated
- Displaying extreme mood swings
- Saying good-bye to loved ones, giving belongings away

Protective Factors

- Family and community support (connectedness)
- Self-esteem and a sense of purpose and meaning
- Problem solving, conflict resolution, coping, and nonviolent communications skills
- Cultural or religious beliefs
- Effective clinical care

Components of Evaluation

- Screening
- Assessment
- Safety Plan
- Lethal Means Counseling
- Disposition

Suicide Risk Screening and Assessment Tools

Screening Tools

- PHQ-A (Patient Health Questionnaire for Adolescents)
- asQ (Ask Suicide-Screening Questions)
- C-SSRS (Columbia-Suicide Screening Rating Scale)

Assessment Tools

- asQ BSSA (Brief Suicide Screening Assessment)
- C-SSRS

Depression and Suicide Risk Screening


PHQ-9 modified for Adolescents (PHQ-A)

Name: _____ Clinician: _____ Date: _____

Instructions: How often have you been bothered by each of the following symptoms during the past two weeks? For each symptom put an "X" in the box beneath the answer that best describes how you have been feeling.

	(0) Not at all	(1) Several days	(2) More than half the days	(3) Nearly every day
1. Feeling down, depressed, irritable, or hopeless?				
2. Little interest or pleasure in doing things?				
3. Trouble falling asleep, staying asleep, or sleeping too much?				
4. Poor appetite, weight loss, or overeating?				
5. Feeling tired, or having little energy?				
6. Feeling bad about yourself – or feeling that you are a failure or that you have let yourself or your family down?				
7. Trouble concentrating on things like school work, reading, or watching TV?				
8. Moving or speaking so slowly that other people could have noticed? Or the opposite – being so fidgety or restless that you were moving around a lot more than usual?				
9. Thoughts that you would be better off dead, or of hurting yourself in some way?				

In the past year, have you felt depressed or sad most days, even if you felt okay sometimes? ☐Yes ☐No

If you are experiencing any of the problems on this form, how difficult have these problems made it for you to do your work, take care of things at home or get along with other people? ☐Not difficult at all ☐Somewhat difficult ☐Very difficult ☐Extremely difficult

Has there been a time in the past month when you have had serious thoughts about ending your life? ☐Yes ☐No

Have you EVER, in your WHOLE LIFE, tried to kill yourself or made a suicide attempt? ☐Yes ☐No

If you have had thoughts that you would be better off dead or of hurting yourself in some way, please discuss this with your Health Care Clinician, go to a hospital emergency room or call 911.

Office use only: Severity score: _____

Modified with permission from the PHQ (Spitzer, Williams & Kroenke, 1999) by J. Johnson (Johnson, 2002)

PHQ-9 Modified for Adolescents

PHQ-9 *plus* suicide questions

11-17 years old

The PHQ-A can be considered a suicide risk screening tool **ONLY** if suicide questions are included and everyone answers them (e.g. not only when PHQ-2 is positive)

Suicide Risk Screening - **asQ**


asQ Information Sheet

Screening Youth for Suicide Risk in Medical Settings

asQ Information Sheet

Developed for patients 10-24, for use in pediatric EDs, inpatient, and primary care settings

For use by non-psychiatric clinicians

12.1% of US adolescents experience suicide ideation, 4% develop a suicide plan, and 4.1% attempt suicide

Solely relying on depression screening through PHQ-9 missed up to 28% of participants at risk for suicide

Suicide Risk Screening - **asQ**


asQ Suicide Risk Screening Tool

Ask the patient:

1. In the past few weeks, have you wished you were dead? ☐Yes ☐No
2. In the past few weeks, have you felt that you or your family would be better off if you were dead? ☐Yes ☐No
3. In the past week, have you been having thoughts about killing yourself? ☐Yes ☐No
4. Have you ever tried to kill yourself? ☐Yes ☐No

If yes, how? _____

5. Are you having thoughts of killing yourself right now? ☐Yes ☐No

If yes, please describe: _____

If the patient answers Yes to any of the above, ask the following study question:

6. Are you having thoughts of killing yourself right now? ☐Yes ☐No

If yes, please describe: _____

Next steps:

Provide resources to all patients

asQ Suicide Risk Screening Tool

Available in **multiple languages**

Takes **1-2 minutes** to screen

100% Sensitivity in Primary Care

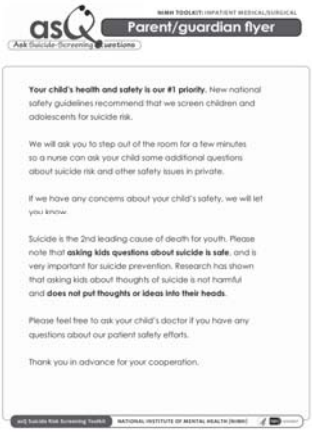
88% Specificity in Primary Care

Negative Screen: **"No"** on first 4 questions; end of screen

Positive screen: **"Yes"** to any of first 4 questions requires answer to question 5, **patients cannot leave until evaluated for safety**

Acute positive screen: **"Yes"** on question 5, patient requires STAT safety/full mental health evaluation

Non-acute positive screen: **"No"** on question 5, use asQ Brief Suicide Safety Assessment (BSSA) (~10-15 minutes)



asQ Parent/guardian flyer

Ask Suicide-Screening Questions

Your child's health and safety is our #1 priority. New national safety guidelines recommend that we screen children and adolescents for suicide risk.

We will ask you to step out of the room for a few minutes so a nurse can ask your child some additional questions about suicide risk and other safety issues in private.

If we have any concerns about your child's safety, we will let you know.

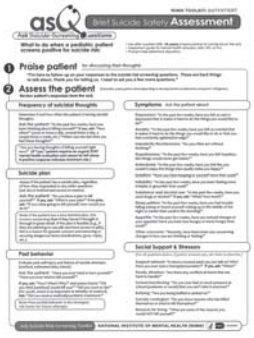
Suicide is the 2nd leading cause of death for youth. Please note that **asking kids questions about suicide is safe**, and is very important for suicide prevention. Research has shown that asking kids about thoughts of suicide is not harmful and **does not put thoughts or ideas into their heads**.

Please feel free to ask your child's doctor if you have any questions about our patient safety efforts.

Thank you in advance for your cooperation.

asQ Suicide Risk Screening Toolkit | NATIONAL INSTITUTE OF MENTAL HEALTH (NIMH)

Brief Suicide Safety Assessment



asQ Brief Suicide Safety Assessment

Ask Suicide-Screening Questions

asQ BSSA (Outpatient Version)

Developed for primary care

For use by non-psychiatric clinicians

Contains protocol and scripts for talking to pediatric patients and parents

Brief Suicide Safety Assessment

asQ Brief Suicide Safety Assessment

Ask Suicide-Screening Questions

3 Interview patient & parent/guardian together

4 Make a safety plan with the patient

5 Determine disposition

6 Provide resources to all patients

asQ BSSA (Outpatient Version)

Cues each step of process:

1. *Praise patient*
2. *Assess the patient*
3. *Interview patient & parent/guardian together*
4. *Make a safety plan with the patient*
5. *Determine disposition*
6. *Provide Resources to all patients*

BSSA Step 1: Praise Patient

NIMH TOOLKIT: OUTPATIENT

asQ Brief Suicide Safety Assessment

Ask Suicide-Screening Questions

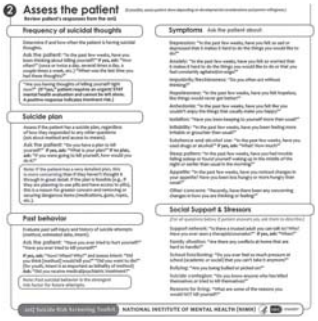
What to do when a pediatric patient screens positive for suicide risk:

- Use after a patient (10 - 24 years) screens positive for suicide risk on the asQ
- Assessment guide for mental health clinicians, MDs, NPs, or PAs
- Prompts help determine disposition

1 Praise patient *for discussing their thoughts*

"I'm here to follow up on your responses to the suicide risk screening questions. These are hard things to talk about. Thank you for telling us. I need to ask you a few more questions."

BSSA Step 2: Assess the Patient



2 Assess the patient

Review patient's responses from the asQ

Frequency of suicidal thoughts

Symptoms Ask the patient about:

Suicide plan

Past behavior

Social Support & Stressors

asQ BSSA (Outpatient Version)

Step 2: Assess the patient

Frequency of suicide thoughts

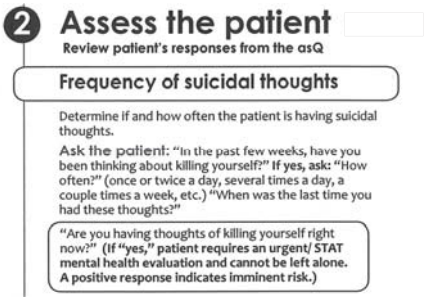
Suicide plan

Past behaviors

Symptoms

Social supports and stressors

BSSA Step 2a: Frequency of Suicidal Thoughts



2 Assess the patient

Review patient's responses from the asQ

Frequency of suicidal thoughts

Determine if and how often the patient is having suicidal thoughts.

Ask the patient: "In the past few weeks, have you been thinking about killing yourself?" **If yes, ask: "**How often?" (once or twice a day, several times a day, a couple times a week, etc.) "When was the last time you had these thoughts?"

"Are you having thoughts of killing yourself right now?" **(If "yes," patient requires an urgent/ STAT mental health evaluation and cannot be left alone. A positive response indicates imminent risk.)**

BSSA Step 2b: Suicide Plan

Suicide plan

Assess if the patient has a suicide plan, regardless of how they responded to any other questions (ask about method and access to means).

Ask the patient: "Do you have a plan to kill yourself?" **If yes, ask:** "What is your plan?" **If no plan, ask:** "If you were going to kill yourself, how would you do it?"

Note: If the patient has a very detailed plan, this is more concerning than if they haven't thought it through in great detail. If the plan is feasible (e.g., if they are planning to use pills and have access to pills), this is a reason for greater concern and removing or securing dangerous items (medications, guns, ropes, etc.).

BSSA Step 2c: Past Behavior

Past behavior

Evaluate past self-injury and history of suicide attempts (method, estimated date, intent).

Ask the patient: "Have you ever tried to hurt yourself?" "Have you ever tried to kill yourself?"

If yes, ask: "How? When? Why?" and assess intent: "Did you think [method] would kill you?" "Did you want to die?" (for youth, intent is as important as lethality of method) **Ask:** "Did you receive medical/psychiatric treatment?"

Note: Past suicidal behavior is the strongest risk factor for future attempts.

BSSA Step 2d: Symptoms

Symptoms Ask the patient about:

Depression: "In the past few weeks, have you felt so sad or depressed that it makes it hard to do the things you would like to do?"

Anxiety: "In the past few weeks, have you felt so worried that it makes it hard to do the things you would like to do or that you feel constantly agitated/on-edge?"

Impulsivity/Recklessness: "Do you often act without thinking?"

Hopelessness: "In the past few weeks, have you felt hopeless, like things would never get better?"

Anhedonia: "In the past few weeks, have you felt like you couldn't enjoy the things that usually make you happy?"

Isolation: "Have you been keeping to yourself more than usual?"

Irritability: "In the past few weeks, have you been feeling more irritable or grouchier than usual?"

Substance and alcohol use: "In the past few weeks, have you used drugs or alcohol?" **If yes, ask:** "What? How much?"

Sleep pattern: "In the past few weeks, have you had trouble falling asleep or found yourself waking up in the middle of the night or earlier than usual in the morning?"

Appetite: "In the past few weeks, have you noticed changes in your appetite? Have you been less hungry or more hungry than usual?"

Other concerns: "Recently, have there been any concerning changes in how you are thinking or feeling?"

BSSA Step 2e: Social Support & Stressors

Social Support & Stressors

(For all questions below, if patient answers yes, ask them to describe.)

Support network: "Is there a trusted adult you can talk to? Who? Have you ever seen a therapist/counselor?" **If yes, ask:** "When?"

Family situation: "Are there any conflicts at home that are hard to handle?"

School functioning: "Do you ever feel so much pressure at school (academic or social) that you can't take it anymore?"

Bullying: "Are you being bullied or picked on?"

Suicide contagion: "Do you know anyone who has killed themselves or tried to kill themselves?"

Reasons for living: "What are some of the reasons you would NOT kill yourself?"

BSSA Step 3: Interview Parent/Guardian Together

asQ Ask Suicide-Screening Questions

NIMH TOOLKIT: OUTPATIENT

Brief Suicide Safety Assessment

3 Interview patient & parent/guardian together

If patient is ≥ 18 years, ask patient's permission for parent/guardian to join.

Say to the parent: "After speaking with your child, I have some concerns about his/her safety. We are glad your child spoke up as this can be a difficult topic to talk about. We would now like to get your perspective."

- "Your child said... (reference positive responses on the asQ). Is this something he/she shared with you?"
- "Does your child have a history of suicidal thoughts or behavior that you're aware of?" **If yes, say:** "Please explain."
- "Does your child seem:
  - Sad or depressed?"
  - Anxious?"
  - Impulsive? Reckless?"
  - Hopeless?"
  - Irritable?"
  - Unable to enjoy the things that usually bring him/her pleasure?"
  - Withdrawn from friends or to be keeping to him/herself?"
- "Have you noticed changes in your child's:
  - Sleeping pattern?"
  - Appetite?"
- "Does your child use drugs or alcohol?"
- "Has anyone in your family/close friend network ever tried to kill themselves?"
- "How are potentially dangerous items stored in your home?" (e.g. guns, medications, poisons, etc.)
- "Does your child have a trusted adult they can talk to?" (Normalize that youth are often more comfortable talking to adults who are not their parents)
- "Are you comfortable keeping your child safe at home?"

At the end of the interview, ask the parent/guardian: "Is there anything you would like to tell me in private?"

BSSA Step 4: Make a Safety Plan with the Patient

4 Make a safety plan with the patient (Include the parent/guardian, if possible.)

Create a safety plan for managing potential future suicidal thoughts. A safety plan is different than making a "safety contract"; asking the patient to contract for safety is NOT effective and may be dangerous or give a false sense of security.

Say to patient: "Our first priority is keeping you safe. Let's work together to develop a safety plan for when you are having thoughts of suicide."

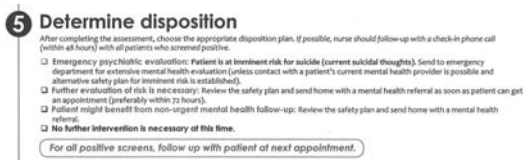
Examples: "I will tell my mom/coach/teacher." "I will call the hotline." "I will call _____."

Discuss coping strategies to manage stress (such as journal-writing, distraction, exercise, self-soothing techniques).

Discuss means restriction (securing or removing lethal means): "Research has shown that limiting access to dangerous objects saves lives. How will you secure or remove these potentially dangerous items (guns, medications, ropes, etc.)?"

Ask safety question: "Do you think you need help to keep yourself safe?" (A "no" response does not indicate that the patient is safe; but a "yes" is a reason to act immediately to ensure safety.)

BSSA Step 5: Determine Disposition


5 Determine disposition

After completing the assessment, choose the appropriate disposition plan. *If possible, nurse should follow-up with a check-in phone call* (within 48 hours) with all patients who screened positive.

- ❑ Emergency psychiatric evaluation: **Patient is at imminent risk for suicide (current suicidal thoughts).** Send to emergency department for extensive mental health evaluation (unless contact with a patient's current mental health provider is possible and alternative safety plan for imminent risk is established).
- ❑ Further evaluation of risk is necessary: Review the safety plan and send home with a mental health referral as soon as patient can get an appointment (preferably within 72 hours).
- ❑ Patient might benefit from non-urgent mental health follow-up: Review the safety plan and send home with a mental health referral.
- ❑ **No further intervention is necessary at this time.**

For all positive screens, follow up with patient at next appointment.

Outcomes based on assessment:

1. *Immediate referral to mental health provider*
2. *Safety planning with urgent referral to mental health provider within 72 hours*
3. *Safety planning with non-urgent referral to mental health provider*
4. *No further intervention needed at this time*

BSSA Step 6: Provide Resources to all Patients


6 Provide resources to all patients

- 24/7 National Suicide Prevention Lifeline 1-800-273-TALK (8255) En Español: 1-888-628-9454
- 24/7 Crisis Text Line: Text "HOME" to 741-741

asQ Suicide Risk Screening Toolkit | NATIONAL INSTITUTE OF MENTAL HEALTH (NIMH)

Oregon Resources:

Lines For Life - National Suicide Prevention Lifeline above re-directs here

YouthLine – a teen to teen crisis and help line; teens available to help daily from 4-10PM, off-hours call re-direct to Lines for Life

Call: 877-968-8491

Text: teen2teen to 839863

Chat: http://www.oregonyouthline.org

PART 3

Management, Referral, and Structured Follow-up

Safety Planning Template


Patient Safety Plan Template

Safety Plan Template (Brown and Stanley)

Free to use after registering on website

~20-30 minutes to complete with patient, collaborative process

Identifies

- Internal coping strategies
- Enhancing social support
- Professional Supports
- Emergency contacts

Safety Planning Intervention Example


SAFETY PLAN

Steps:

Step 1: Recognize warning signs

Step 2: Identify and employ internal coping strategies

Step 3: Use healthy social contacts as a means of distraction.

Step 4: Contact family and friends for help

Step 5: Contact MH professional or emergency services if needed

Step 6: Reduce access to lethal means

Lethal Means Statistics

What is it about guns?

- 85% lethality
- > 33% of households have guns
- Irreversible damage
- 85% come from the victim's home

Lethal Means: Special Issues Related to Suicidal Youth

Involve parents and guardians whenever possible. Ask questions about means restriction with parents privately.

Gently assume there may be guns in the home.

Example scripts:

"Let's talk about securing your guns so we can keep your child safe"

"Now might be a good time to give your guns to a friend or family member for safe-keeping"

Lethal Means: Special Issues Related to Suicidal Youth

It is important to remove and limit access to other lethal means:

- material that could be used for hanging
- medication lockbox

Means Safety Resources

Lockmed.com

Referrals

Local Mental Health Resources

Identify community mental health partners

OPAL-K

Can assist with diagnostic questions

Lines For Life

Can assist with identifying local community mental health providers and resources

PART 4

Implementation

Implementation

"It's not how are we going to do this, but how are we going to handle it if we lose one of our patients?"

~Ted Abernathy, MD
(Pilot Pediatrican for asQ Implementation)

Implementation

1. Education of staff about importance of screening
2. Identify a champion(s)
3. Provide information about confidentiality

Office Implementation

4. Establish flow of screening forms
   - When and where do patients receive screen?
   - Confidential space for patient to complete screen?
   - Who will review/score screen?
   - How is provider notified of results?
   - How are results documented in the chart?

Office Implementation

5. Can forms be embedded in EMR?
6. Establish tracking system to follow-up with patients

PART 5

Resources

OPAL-K
(Oregon Psychiatric Access Line about Kids)

Psychiatric phone consultation for medical practitioners who treat children and adolescents with mental health difficulties

9 am to 5 pm, Monday through Friday
855-966-7255 (toll-free) or 503-346-1000
(Portland metro)

Register online: www.ohsu.edu/opalk
Fax: 503-346-1389
Email: opalk@ohsu.edu

Other Resources/Toolkits

Resources for providers

- *OCCAP (Oregon Council of Child and Adolescent Psychiatry)*
- *Zero Suicide*
- *Suicide Prevention Resource Center (SPRC)*
- *Suicide Prevention in Primary Care Settings Toolkit (Deschutes County)*

Resources for youth

- *Lines For Life YouthLine*
- *Teens Finding Hope*
- *Trevor Project*
- *Youth ERA*

Resources for parents

- *Child Mind*
- *NAMI (National Alliance on Mental Illness) Toolkit*
- *OFSN (Oregon Families Support Network)*
- *Teens Finding Hope*

?

Thanks to Oregon Pediatric Society and the Adolescent Suicide Prevention Task Force members who generously provided their time and expertise

Barbara Long, MD, MPH
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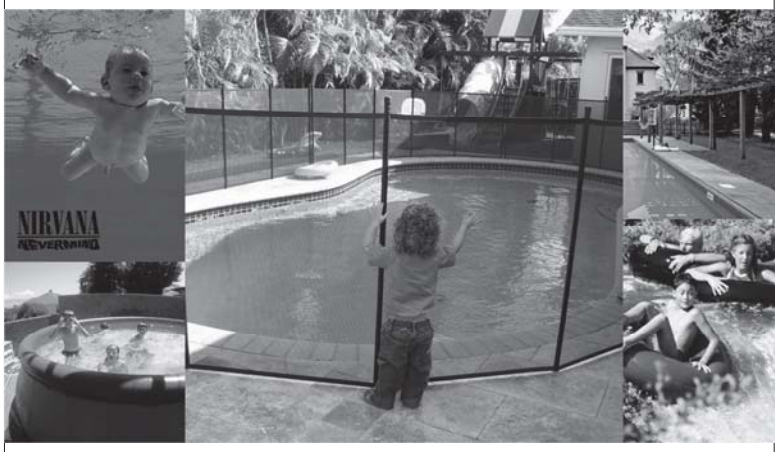
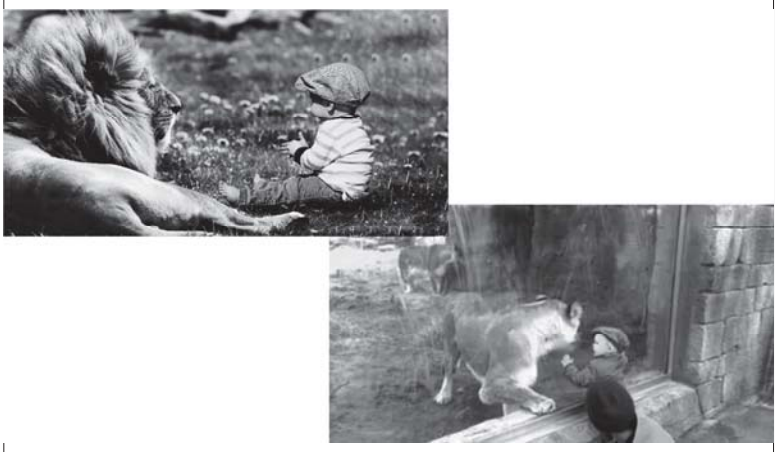
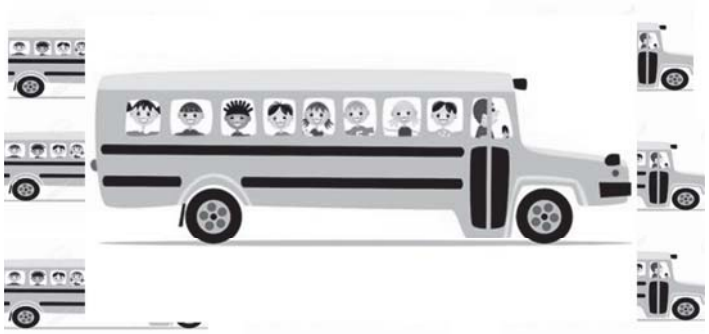
Kyle Johnson, MD
Rita Lahlou, MD
Stewart Newman, MD
Kristi Nix, MD
Teri Petterson, MD
Liz Stevenson, JD, MPH
Liz Thorne, MPH
Melissa Weddle, MD, MPH

What Every Pediatrician Needs to Know About Drowning

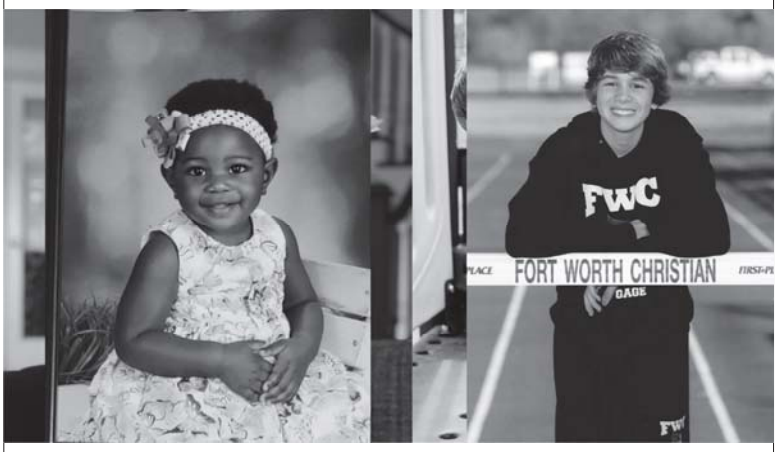
Benjamin Hoffman MD CPST-I FAAP
Professor of Pediatrics, Oregon Health and Science university
Chair, AAP Council on Injury Violence and Poison Prevention







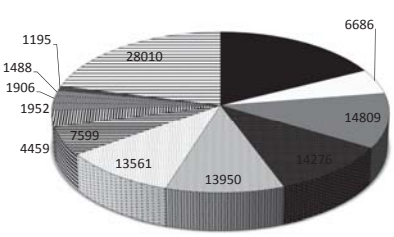
Drowning 1-18 years



Objectives

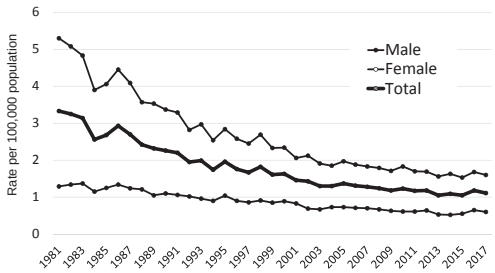
- By the end of this presentation, you should be able to:
  - Discuss the epidemiology of drowning for children and teens
  - Discuss the key points from the recently revised AAP policy statement on drowning prevention
  - List 5 key tips to help you decrease drowning risks for your patients and their families
  - Describe layers of protection in drowning prevention

Deaths 1-18 year 2007-2017



Rank	Age Groups <1	Age Groups 1-18
1	Congenital Anomalies 38,784	Unintentional Injury 44,613
2	Short Gestation 32,591	Suicide 14,278
3	SIDS 13,191	Homicide 13,930
4	Maternal Pregnancy Comp. 12,184	Malignant Neoplasms 13,561
5	Unintentional Injury 9,585	Congenital Anomalies 7,599
6	Placenta Cord Membranes 7,564	Heart Disease 4,459
7	Bacterial Sepsis 4,571	Influenza & Pneumonia 1,952
8	Respiratory Distress 3,903	Chronic Low Respiratory Disease 1,906
9	Circulatory System Disease 3,738	Cerebro-vascular 1,488
10	Neonatal Hemorrhage 3,360	Septicemia 1,195

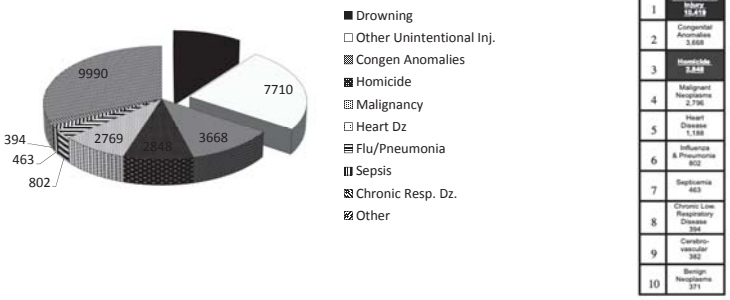
Unintentional Drowning Death Rate of US Infants and Children Ages 0-19 by Gender, 1981-2017



In 2017:
913 total unintentional drowning deaths

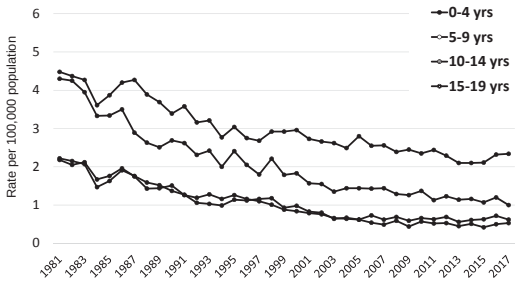
Source: AAP Analysis of CDC WISQARS fatal injury reports. February 2019 (https://webappa.cdc.gov/sasweb/ncipc/mortrate.html)

Deaths 1-4 year 2007-2017



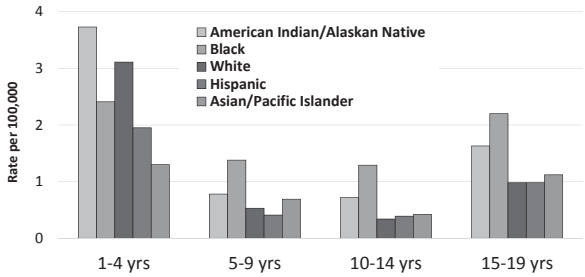
	Age Groups
Rank	1-4
1	Unintentional Injury 10,419
2	Congenital Anomalies 3,668
3	Homicide 2,848
4	Malignant Neoplasms 2,796
5	Heart Disease 1,188
6	Influenza & Pneumonia 802
7	Septicemia 463
8	Chronic Low Respiratory Disease 394
9	Cerebro-vascular 382
10	Benign Neoplasms 371

Unintentional Drowning Death Rate of US Infants and Children, by Age Group, 1981-2017



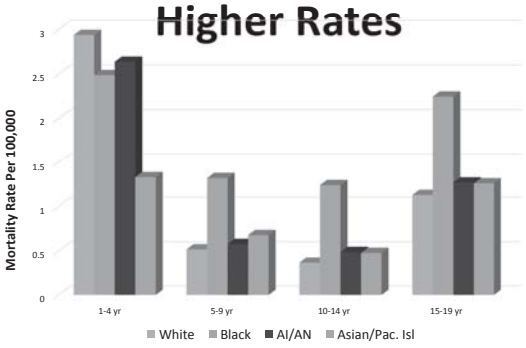
Source: AAP Analysis of CDC WISQARS fatal injury reports. February 2019
(https://webappa.cdc.gov/sasweb/ncipc/mortrate.html)

Unintentional Drowning Deaths (Rate per 100,000) among US Children (ages 1-19) by Race/Ethnicity by Age Group, 2008-2017 Average



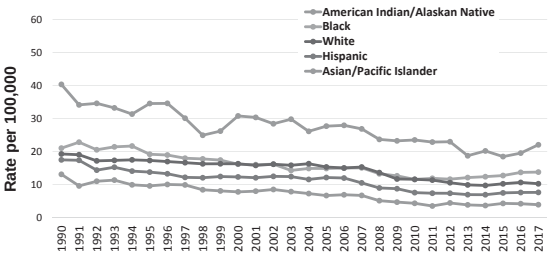
Source: AAP analysis of National Center for Injury Prevention and Control/CDC WISQARS™
(Web-based Injury Statistics Query and Reporting System), April 2019.
Note: American Indian/Alaskan Native, Black, White, and Asian/Pacific Islander refer to those who identify as non-Hispanic.

African American Kids Drown at Much Higher Rates



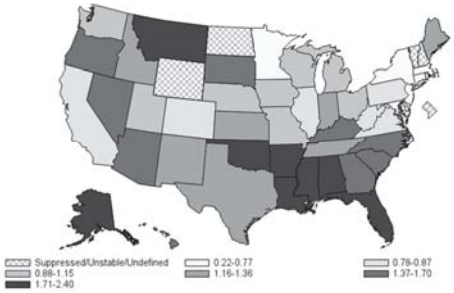
CDC WISQARS
2007-2017

Fatal Unintentional Injuries (rates per 100,000) among US Children (ages 0-19) by Race/Ethnicity, 1990-2017



Source: AAP analysis of National Center for Injury Prevention and Control/CDC WISQARS™
(Web-based Injury Statistics Query and Reporting System), March 2019.
Note: American Indian/Alaskan Native, Black, White, and Asian/Pacific Islander refer to those who identify as non-Hispanic.

Unintentional Drowning Death Rate (per 100,000 Population) of US Infants and Children Ages 0-19, by State, 2008-2014 Annualized Average



Source: NCHS Vital Statistics System for numbers of deaths; US Census Bureau for population estimates.
Retrieved from WISQARS Fata Injury Mapping. CDC

2002 - 2017, Arizona
Drowning Deaths and Rates per 100,000
All Races, Both Sexes, Ages 0 to 19
ICD-10 Codes: W65-W74,X71,X92,Y21

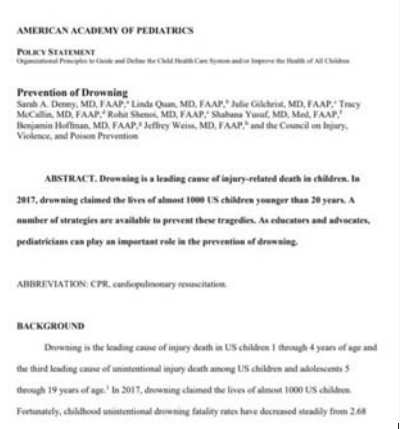
Age Group	Number of Deaths	Population***	Crude Rate
00-04	328	7,079,094	4.63
05-09	48	6,943,392	0.69
10-14	21	7,056,972	0.30
15-19	95	7,055,197	1.35
Total	492	28,134,655	1.75

2002 - 2017, Oregon
Drowning Deaths and Rates per 100,000
All Races, Both Sexes, Ages 0 to 19
ICD-10 Codes: W65-W74,X71,X92,Y21

Age Group	Number of Deaths	Population***	Crude Rate
00-04	86	3,706,162	2.32
05-09	23	3,766,908	0.61
10-14	39	3,887,091	1.00
15-19	114	4,002,331	2.85
Total	262	15,362,492	1.71

AAP Policy

"the AAP lays out strategies to protect children at each stage of their life. New parents are advised to be vigilant at bath time and to empty all buckets and wading pools immediately. All children should learn to swim, and children and teens should wear life jackets while near open bodies of water. Teens can learn CPR and other water safety skills."



AMERICAN ACADEMY OF PEDIATRICS

POLICY STATEMENT
Organizational Principles to Guide and Define the Child Health Care System and/or Improve the Health of All Children

Prevention of Drowning

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ABSTRACT. Drowning is a leading cause of injury-related death in children. In 2017, drowning claimed the lives of almost 1000 US children younger than 20 years. A number of strategies are available to prevent these tragedies. As educators and advocates, pediatricians can play an important role in the prevention of drowning.

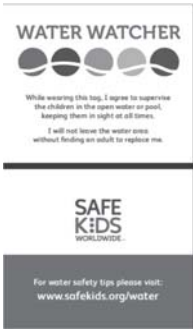
ABBREVIATION: CPR, cardiopulmonary resuscitation.

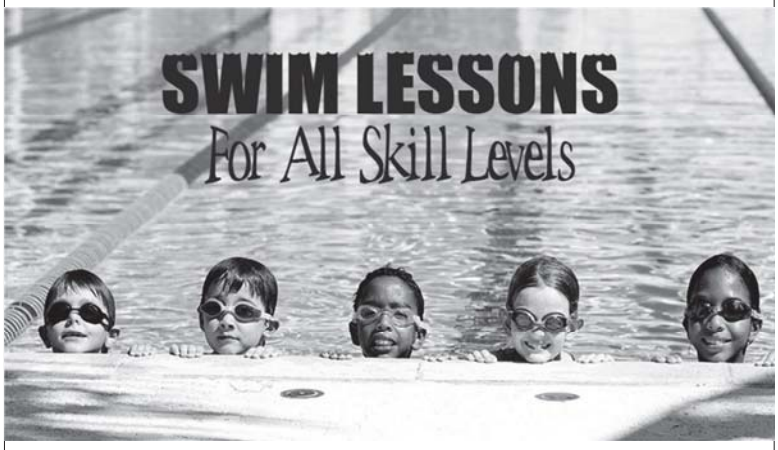
BACKGROUND

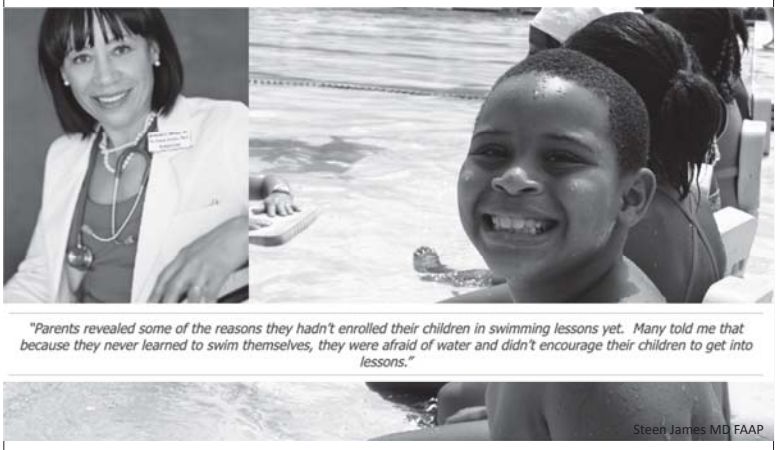
Drowning is the leading cause of injury death in US children 1 through 4 years of age and the third leading cause of unintentional injury death among US children and adolescents 5 through 19 years of age.[1] In 2017, drowning claimed the lives of almost 1000 US children. Fortunately, childhood unintentional drowning fatality rates have decreased steadily from 2.68

Barriers

Constant, Close and Capable Supervision







"Parents revealed some of the reasons they hadn't enrolled their children in swimming lessons yet. Many told me that because they never learned to swim themselves, they were afraid of water and didn't encourage their children to get into lessons."

Steen James MD FAAP

There is NO EVIDENCE that Infant Survival Swim Classes Work

Life Jackets

CPR

Aap.org/drowning

PSA with Nicole Hughes

https://www.youtube.com/watch?v=DQsro78hQC8

Questions?

Layers of Protection

- **Barriers**
- **Constant, Close, Capable Supervision**
- **Water Competence**
- **Life Jackets**
- **CPR**

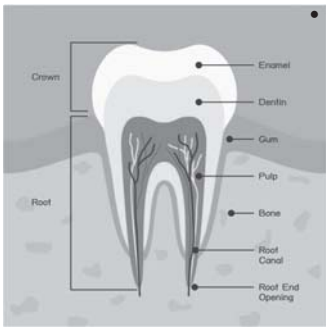
Pediatric Oral & Dental Care for the Primary Care Provider

Sarah Kate Lee, DDS

Outline

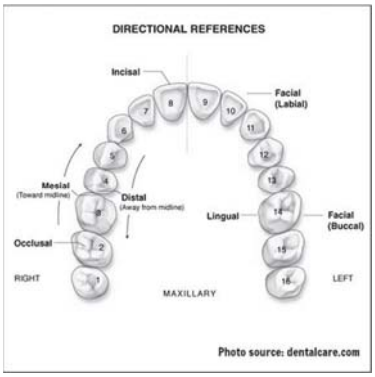
- Tooth Basics
- Caries
- Prevention
- Oral Pathology
- Dental Trauma
- Dental “Emergencies”

Basics



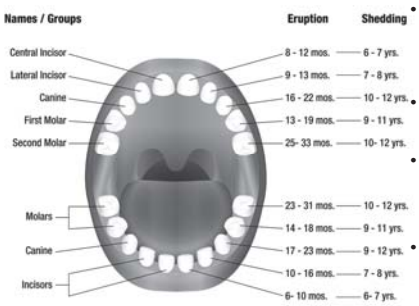
- Anatomy of a tooth
- Crown (portion seen in mouth) made of 3 layers
  - Enamel: outermost layer, white, strongest substance in the body, where cavities begin
  - Dentin: middle layer, yellowish, cavities progress much more quickly
  - Pulp: blood and nerve supply.
    - When cavities reach this far then endodontics (root canal therapy) or extraction indicated
    - Teeth either hurt a lot or not at all depending on health of pulp
- Root (portion in bone, surrounded by periodontal ligament)
  - Root canal is extension of the pulp down the root
  - Pulp is within the root and provides communication to the body via blood supply within

Basics



- Tooth Surfaces
  - Incisal (anterior) & Occlusal (posterior) – biting/chewing surfaces
  - Facial (anterior) & Buccal (posterior) – surface touching the lips & cheek
  - Lingual – surface touching the tongue
  - Palatal- surface towards the palate in upper arch
  - Proximal – surfaces that are next to each other
    - Mesial – surface facing towards the midline
    - Distal – surface facing away from the midline
- Cavities are most common –
  - Grooves on occlusal surfaces of posterior teeth
  - **On proximal surfaces where teeth touch (can only be cleaned w/ floss**)

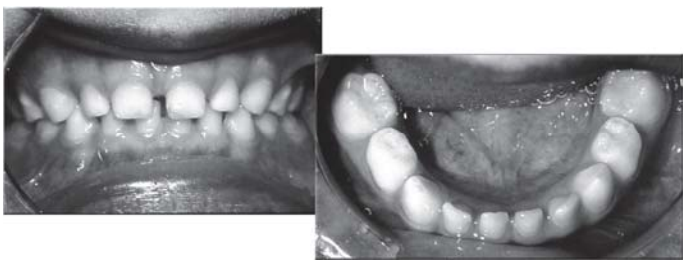
Primary Dentition – “Baby Teeth”



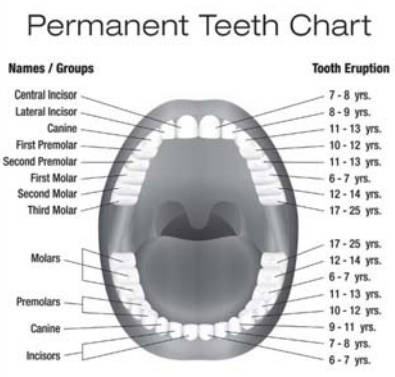
- Eruption of primary teeth begins around 6 months of age and continues until 30 months
- Order of eruption is also important
- First permanent tooth to erupt is the first molar
  - Erupts behind primary teeth around age of 6
- Primary teeth are replaced by permanent teeth

“Do baby teeth really matter?”

- Eating
- Preservation of facial form
- Preservation of arch length for permanent dentition
- Esthetics, social implications
- Healthy teeth aid in development



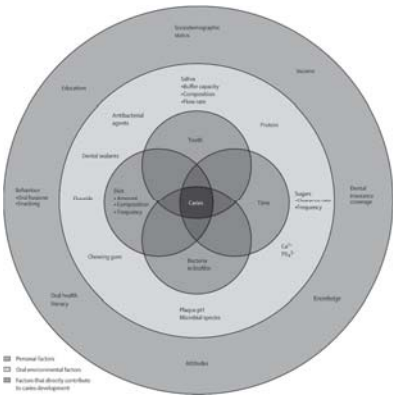
Permanent Dentition – “Adult teeth”



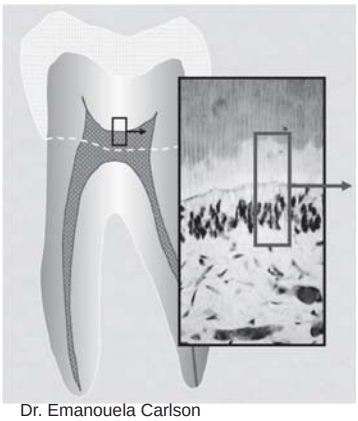
- Full permanent dentition around age 12
- Third molars (“wisdom teeth”) tend to cause symptoms in late teen years to early adulthood
- Order and timing are both important
- Mandible erupts prior to maxilla
- “Shark teeth” common

Caries Process

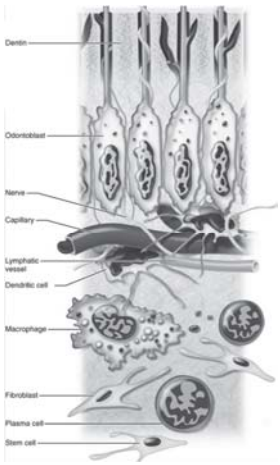
- Multifactorial disease that leads to the localized destruction of hard dental tissues
- Destruction of hard tissues by the weak acids produced by bacterial carbohydrate fermentation
  - Typically a slow process – remember the enamel is very strong!
- Left untreated, caries can lead to tooth pain, infection, and/or abscess
- **Most common chronic disease** of children aged 6 to 11 years and adolescents aged 12 to 19 years
  - Elementary school students miss an average of 2.3 days/yr for dental issues



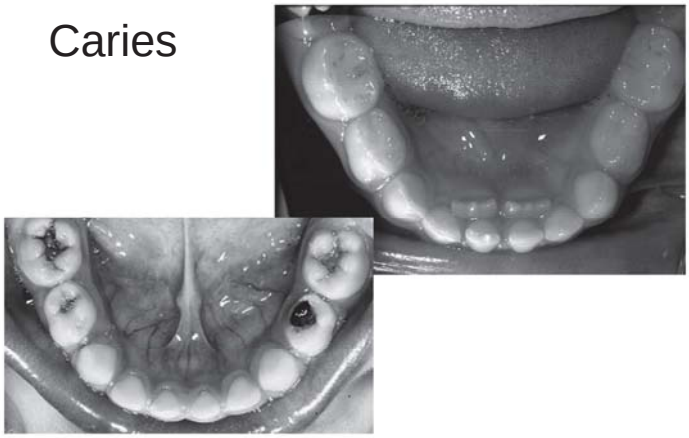
Caries Process



Dr. Emanouela Carlson

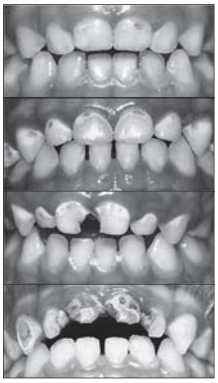


Caries



Early Childhood Caries-ECC

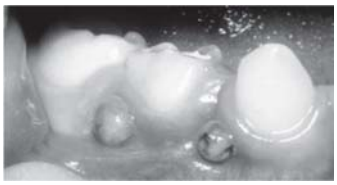
- Early childhood caries: presence of more than one decayed, missing (due to decay), or filled tooth surface in a child under 6 years old
- Severe ECC: any sign of smooth surface caries in a child under 3 years of age

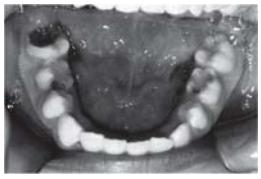


Caretaker education and establishing a dental home is vital!

Dentoalveolar Abscess & Infection

- Cause from caries, trauma, periodontal disease
- Systemic involvement (i.e. fever, facial swelling, asymmetry) warrant emergency attention
  - Concern for risk for endocarditis, brain abscess, Ludwig’s angina
- Tx. indicated is extraction or root canal therapy (only permanent teeth) – **urgent dental referral**
- Prescribe antibiotics
  - Amoxicillin and Clindamycin commonly





Caries Risk Factors

Factors	High Risk	Moderate Risk	Protective
Biological			
Mother/primary caregiver has active cavities	Yes		
Parent/caregiver has low socioeconomic status	Yes		
Child has >3 between meal sugar-containing snacks or beverages per day	Yes		
Child is put to bed with a bottle containing natural or added sugar	Yes		
Child has special health care needs		Yes	
Child is a recent immigrant		Yes	
Protective			
Child receives optimally-fluoridated drinking water or fluoride supplements			Yes
Child has teeth brushed daily with fluoridated toothpaste			Yes
Child receives topical fluoride from health professional			Yes
Child has dental home/regular dental care			Yes
Clinical Findings			
Child has white spot lesions or enamel defects	Yes		
Child has visible cavities or fillings	Yes		
Child has plaque on teeth		Yes	

Circling those conditions that apply to a specific patient helps the health care worker and parent understand the factors that contribute to or protect from caries. Risk assessment categorization of low, moderate, or high is based on preponderance of factors for the individual. However clinical judgment may justify the use of one factor (e.g., frequent exposure to sugar containing snacks or beverages, visible caries) in determining overall risk.

Overall assessment of the child's dental caries risk: High ☐ Moderate ☐ Low ☐

Prevention

- Caries is a 100% preventable disease!
- Diet
  - Avoid sticky, starchy, sweet
  - Duration and frequency matter
  - Juice, sports drinks, and soda are dangerous!
  - **Breast feeding vs bottle feeding**
- Establishing a dental home
  - First dental visit between eruption of first tooth & age 1

Prevention

- FLUORIDE
  - Converts hydroxyapetite to fluoroapetite
    - Fluoroapetite is 100 times less soluble!
  - Helps to slow demineralization
  - Promotes remineralization of tooth structure
  - Inhibits dental plaque bacteria metabolism
    - This reduces amount of acid produced
  - Public water fluoridation
    - Optimal level 0.7ppm F (mg/L)
  - Safety
    - Toxic dose 5 mg/kg (10 kg child= 1.8 oz of 1000ppm toothpaste (2 travel size tubes))
    - Lethal dose 32- 64 mg/kg

Prevention

- Fluoride Recommendations
  - Toothpaste
    - Brushing should be supervised until child has manual dexterity to tie their shoes or write their name in cursive
      - 2x/day for 2 minutes each time
    - "Smear" or "Grain of rice" – younger than 3 years old
    - "Pea-sized" – 3 to 6 years old

Figure. Comparison of a smear (left) with a pea-sized (right) amount of toothpaste.

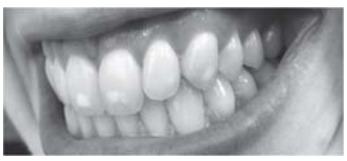
- Supplementation

Table. DIETARY FLUORIDE SUPPLEMENTATION SCHEDULE

Age	<0.3 ppm F	0.3 to 0.6 ppm F	>0.6 ppm F
Birth to 6 months	0	0	0
6 mo to 3 years	0.25 mg	0	0
3 to 6 years	0.50 mg	0.25 mg	0
6 to at least 16 years	1.00 mg	0.50 mg	0

Prevention

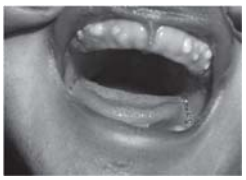
- Fluorosis
  - Occurs during tooth formation
  - White, opaque discoloration of enamel
    - Typically scattered around middle to incisal 1/3 of tooth
  - 84.5% of people unaffected in optimally fluoridated areas
    - Vast majority of cases are mild



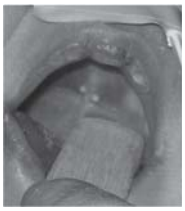
Oral Pathology

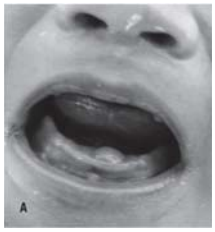
Neonatal White Spot Lesions

- Bohn's nodules
  - Mucus gland tissue present on maxillary alveolar ridge
- Epstein pearls
  - Trapped epithelial remnants on midpalatal raphe
  - Present in 80% of newborns
- Dental Lamina Cysts
  - Trapped epithelial remnants on alveolar ridge



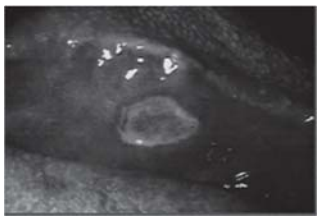
No treatment indicated.
Resolve spontaneously.

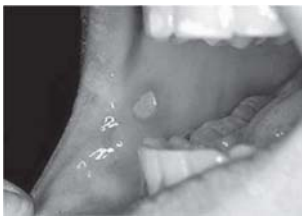




Aphthous Ulcer (canker sore)

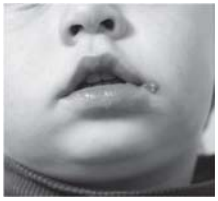
- Ulcerative appearance primarily and when matured
- Painful, self-limiting, no systemic manifestations
- Common locations: buccal mucosa, floor of mouth, oropharynx, vestibule, tongue
- One to few lesions present at a time typically
- Idiopathic, but can be associated with systemic disease: Bechet, Celiac, Crohn, Neutropenia, Immunodeficiency syndrome, GERD
- Treatment: Palliative, avoid trauma to area, topical steroid

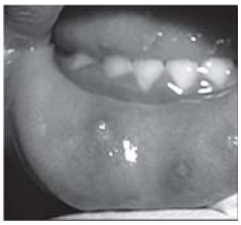




Herpetic Lesion

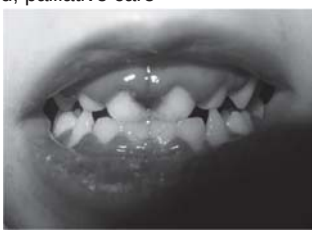
- Vesicle appearance primarily and ulcerative (shallow, punctate) when mature
- Common locations: attached gingiva, hard palate, vermillion border
- Few to several lesions present at a time typically
- Caused by HSV-1
- Treatment: palliative, systemic antiviral (valacyclovir) agents if within 72 hours





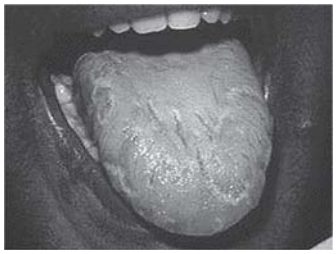
Primary herpetic gingivostomatitis

- Caused by HSV-1
- Most common under age 5
- Presents with fever, lymphadenopathy, headache, malaise, intense gingival erythema, painful oral vesicles throughout mouth
- Treatment: systemic acyclovir, valacyclovir may be warranted, palliative care



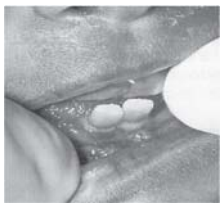
Geographic Tongue

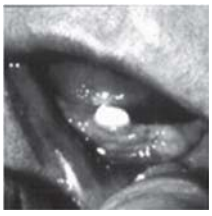
- Benign migratory glossitis
- Usually asymptomatic, but may have tingling or burning sensation
- May disappear and reoccur
- Tx: no treatment
  - If painful, can consider Candida infection



Natal Teeth

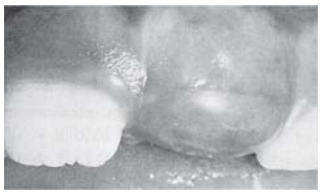
- Mineralized tooth-like structures present at birth or shortly thereafter
- 90% are the primary incisors
- Tx: Remove teeth if they are interfering with feeding or highly mobile and an aspiration risk

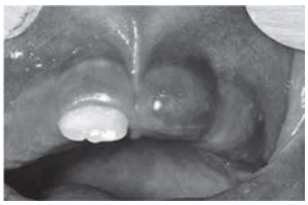




Eruption Cyst/Hematoma

- Red, purple gingival enlargement on the alveolar ridge
- Can occur in primary or permanent dentition
- Tx: None; resolve as tooth erupts
  - If symptomatic or causing delayed eruption, can make an incision



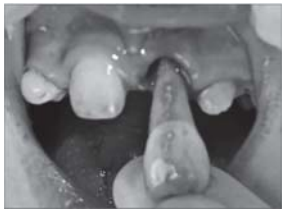


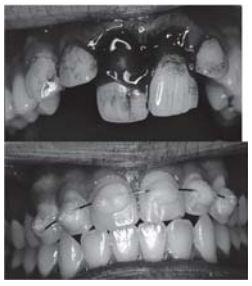
Dental Trauma

Avulsions

- **PERMANENT TEETH**:
  - Greatest chance of keeping tooth viable is **replanting** ASAP
    - Dry time of >60 minutes = no viable PDL cells
  - Only grab tooth by the crown (white part)
  - If dirty, rinse root with isotonic solution (Hank's Balanced Salt Solution), milk, cold running water
  - Reposition tooth in socket w/ firm finger pressure
    - If unable to – store tooth in milk, saline, or special storage media- NOT WATER!
  - Seek emergency dental treatment immediately
    - Tetanus status?

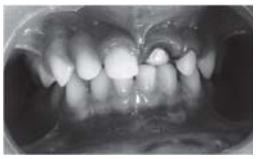
  NEVER replant a primary tooth

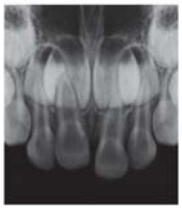




Intrusions

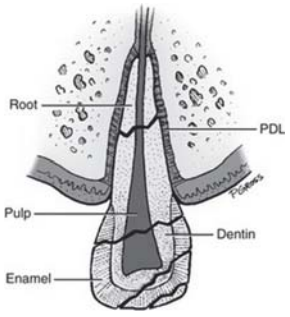
- Intrusions
  - Tooth pushed into the socket, which typically fractures as a result
  - **No immediate tx. needed**
    - **urgent referral to dentist for evaluation**
    - Pain management
  - Depending on extent on intrusion treatments include:
    - Waiting for spontaneous eruption or extraction (primary teeth)
    - Waiting for spontaneous eruption, orthodontic repositioning, or surgical repositioning (permanent teeth)

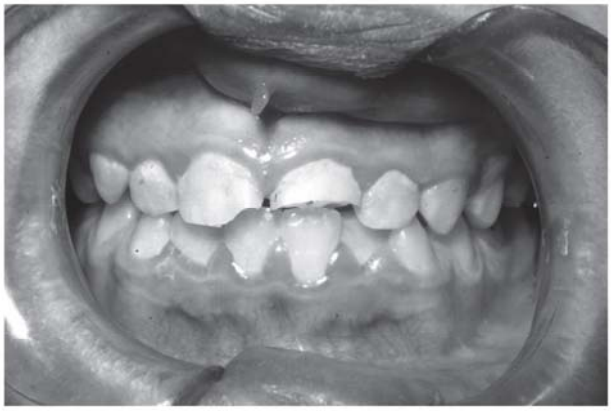


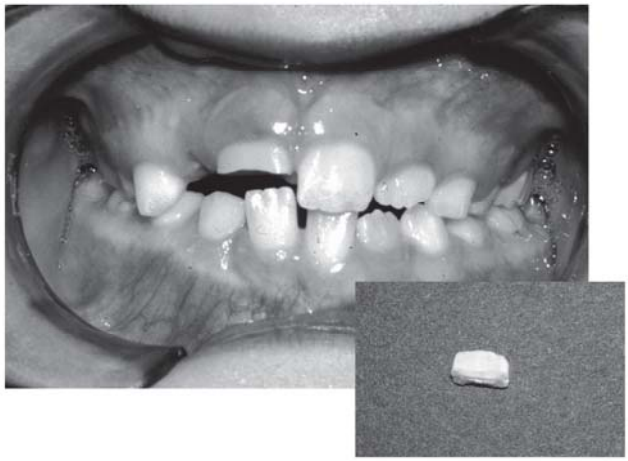


Dental Trauma

- Fractures
  - Tx. based on extent of fracture
    - Only enamel exposed: smooth sharp edges
    - Dentin exposed: seal w/ glass ionomer
    - Pulp exposed: pulp capping and restore or extraction (primary tooth), pulp capping or root canal therapy and restore or extraction (permanent tooth)
    - Root fracture: extraction likely
  - Pain Management (no antibiotics indicated)
  - **In ED: place dy-cal over pulp area, refer to see dentist ASAP**



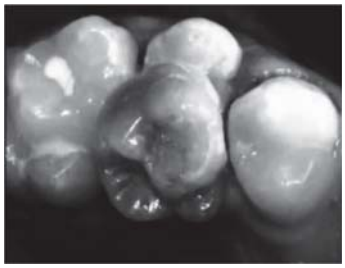


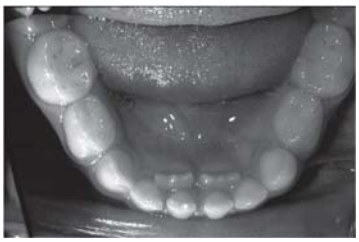


Dental “Emergencies”

Eruption

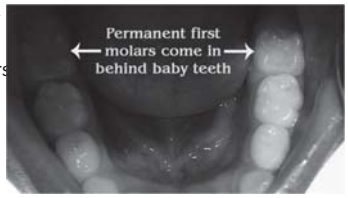
- There is tooth growing out the side of another tooth?!
  - Encourage child to wiggle out the tooth
  - If refuse and causing pain, dentist can extract
  - Concern for decreased oral hygiene in the area due to pain





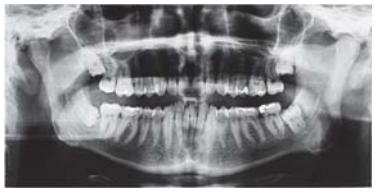
Teething Symptoms

- Pain during eruption
  - Cavities are a possible explanation, but pain in the back especially if it’s in multiple areas of the mouth may be related to eruption of first permanent molars
  - Teething
    - Occurs w/ eruption of primary dentition (btwn. 6-30 mos.) and permanent molars (6 & 12 yrs.)
    - Symptoms can include: drooling, rash (from drooling), pain
      - Teething does NOT cause fever!
    - Recommend cold washcloth, cool teething rings, ibuprofen or Tylenol



“Wisdom Teeth”

- Jaw pain posteriorly
  - Third molars or “Wisdom Teeth”
    - Can start erupting anywhere btwn. 15-21 years old
    - Most people don’t have space in their mouth for them (often impacted as a result)
    - **Pericoronitis** – gum inflammation around partially erupted tooth common
    - Proximity to Inferior Alveolar Nerve
    - Extraction recommended
      - Important consideration prior to chemo/radiation treatment (especially if IV bisphosphonates planned to avoid osteonecrosis of the jaw)



Special Thank You!

- Robert Steelman MD, DDS
- Ian Bell DDS

Top Endocrine Cases

Cheryl Hanna MD

Objectives

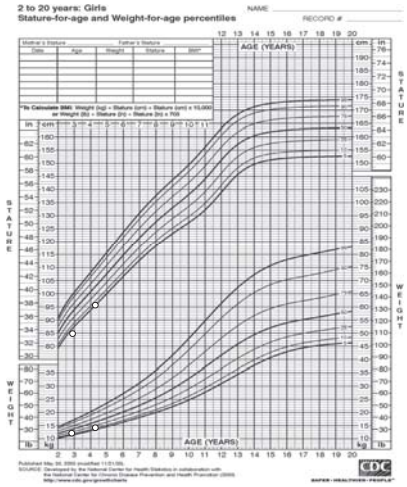
- Puberty early and late
- Growth throughout childhood
- Thyroid function : elevated of Free T4

Case 1

- EN is a 7y 4m girl referred for evaluation of early puberty
- Mother's observations
  - 6y 9m vaginal discharge, ? breast development
  - 7y papules on face, ↑hair in genital area → dermatology dx: acne
- Pediatrician evaluation
  - 17 OHP 244 ng/dl, Total Testosterone 34 ng/dl, normal thyroid function
  - Referral pediatric endocrinology

Pediatric Endocrinology visit

- PMHx
  - Born small; 5lb 3 oz, 18 ¾ inches at 38 weeks
  - Genetics evaluation at 2y 10m for mild developmental delay and short stature
  - Pediatrician w/u at 4y 3m for short stature
    - Bone age 3 proximally, 4y 2m distally
    - Genetics report
      - not Turners
      - complex chromosomal rearrangement of unknown significance
    - IGF 1 89 ng/ml (32-179), IGF BP3 2.9 mg/L (1.7-4.9)



Pediatric Endocrinology visit

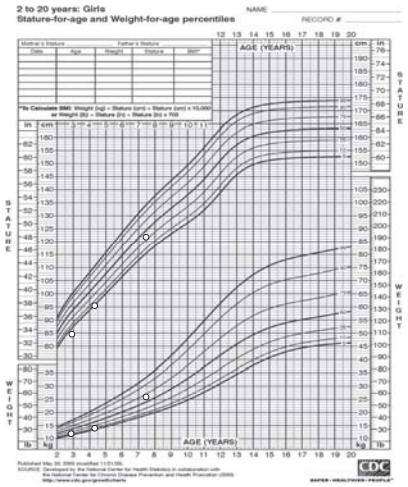
- PMHx
  - Born small; 5lb 3 oz, 18 ¾ inches at 38 weeks
  - Genetics evaluation at 2y 10m for mild developmental delay and short stature
  - Pediatrician w/u at 4y 3m for short stature
    - Exam- no puberty
    - Bone age 3 proximally, 4y 2m distally
    - Genetics report not Turners
    - IGF 1 89 ng/ml (32-179), IGF BP3 2.9 mg/L (1.7-4.9)
- FHx: Mother 63 in, Father 69.5 in

  Target height 63 3/4in

  Brother 50%
- Shx: Mother from Albania, shy but doing well in school

Pediatric Endocrinology visit

- Physical Exam at 7y 4 m
  - Ht 122 cm, weight 26kg
  - General not dysmorphic
  - Pubertal exam
    - Tanner III-IV breast
    - Tanner III pubic hair
  - Skin
    - Mild acne, increased hair on legs



Puberty Features
Breasts
Pubic hair
Accelerated growth

Differential Diagnosis

- 7 y 4m old girl with early puberty
  - Normal early puberty
  - Central precocious puberty
  - Mild congenital adrenal hyperplasia advancing bone age to the biologic time for puberty
  - Adrenal or ovarian tumor producing androgens and estrogens

ARTICLE

Recent Decline in Age at Breast Development: The Copenhagen Puberty Study

Lise Aksglaede, MD[a], Kaspar Sørensen, MD[a], Jørgen H. Petersen, PhD[a,b], Niels E. Skakkebæk, MD, DMSc[a], Anders Juul, MD, DMSc[a]

Pediatrics (2009) 123 e932

1991-1993
2095 girls 5.6-20years

2006-2008
1100 girls

Examined by palpation

TABLE 1 Estimated Mean Ages at Reaching Various Pubertal Stages in Girls Examined in 1991–1993 and 2006–2008

Stage	1991–1993			2006–2008		
	Mean	95% CI	95% PI	Mean	95% CI	95% PI
B2[a,b]	10.88	10.69–11.06	8.66–13.11	9.86	9.70–10.01	7.28–12.44
B3[a]	12.40	12.23–12.56	10.30–14.48	10.97	10.82–11.12	8.65–13.29
B4++	13.54	13.38–13.71	11.43–15.65	12.29	12.13–12.44	10.29–14.27
Menarche[c,d]	13.42	13.24–13.60	11.17–15.67	13.13	12.95–13.31	11.04–15.21
PH2[e,f]	11.29	11.13–11.46	9.28–13.30	11.09	10.95–11.23	8.94–13.24
PH3[a]	12.39	12.23–12.55	10.35–14.42	11.74	11.59–11.89	9.71–13.78
PH4++[a]	13.51	13.34–13.68	11.46–15.56	12.50	12.32–12.67	10.33–14.66

[a] $P < .0001$.
[b] $P < .0001$ after adjustment for BMI.
[c] $P = .021$.
[d] $P = .1272$ after adjustment for BMI.
[e] $P = .066$.
[f] $P = .10$ after adjustment for BMI.

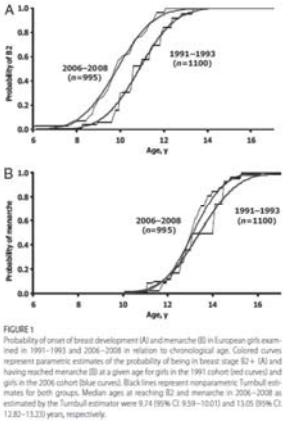


FIGURE 1
Probability of onset of breast development (A) and menarche (B) in European girls examined in 1991–1993 and 2006–2008 in relation to chronological age. Colored curves represent parametric estimates of the probability of being in breast stage B2+ (A) and having reached menarche (B) at a given age for girls in the 1991 cohort (red curves) and girls in the 2006 cohort (blue curves). Black lines represent nonparametric Turnbull estimates for both groups. Median ages at reaching B2 and menarche in 2006–2008 as estimated by the Turnbull estimator were 9.74 (95% CI: 9.59–10.01) and 13.05 (95% CI: 12.82–13.23) years, respectively.

The interval from B2 to menarche increased from 2.5 years to 3.3 years

Early puberty : when to evaluate

- EARLY
  - Boys younger than age 9
  - Girls with breast or pubic hair development before
    - age 7 (white)
    - age 6 (African American)
  - Older girls (6/7 to 8 years)
    - rapid progression of puberty
    - rapid bone age advancement
    - new CNS findings
    - emotional state adversely affected

Premature Adrenarche

Clinical signs of male androgen production (pubic hair, body odor, acne) without signs of true puberty (no enlargement of the penis, testis or breast development)

Idiopathic
Clinical features:
- tall for family
- mildly advanced BA

Associated with:
- Metabolic syndrome
- Obesity
- Insulin resistance
- FHx Type 2 diabetes
- SGA

Mild CAH

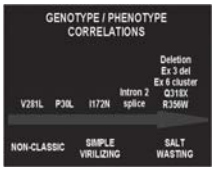
First sign of real puberty

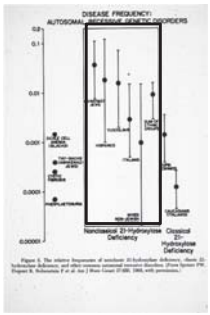
Exposure to topical testosterone

Adrenal tumor

Premature Adrenarche

Clinical signs of male androgen production (pubic hair, body odor, acne) without signs of true puberty (no enlargement of the penis, testis or breast development)

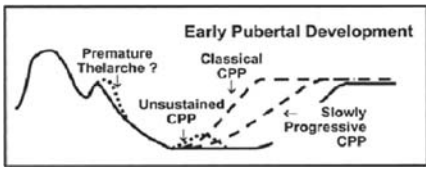




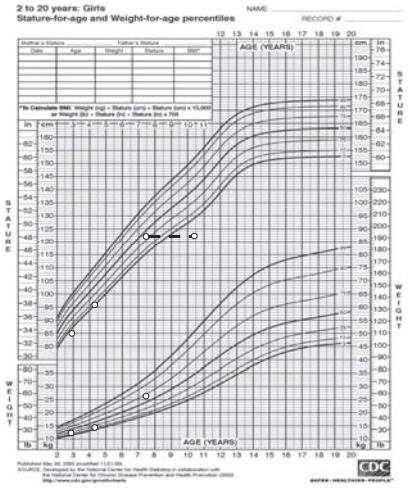
Mild CAH 21 hydroxylase deficiency
AM 17 OHP<100ng/dl rules it out
17 OHP >1000ng confirms dx

Evaluation

- Bone age: 10 proximally, 10 ½ distally
- Labs
  - LH 7.9 mIU/ml (prepubertal <0.3)
  - FSH 7.6 mIU/ml (prepubertal <4.2)
- Conclusion: she is in central puberty
- Potential explanations:



Could premature adrenarche have advanced BA and started normal puberty??

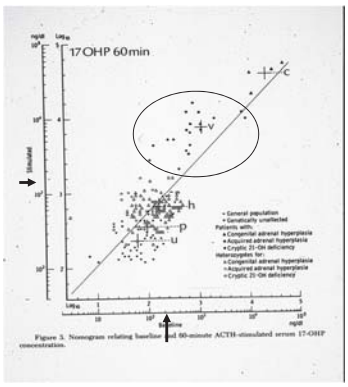


Puberty Features
Breasts
Pubic hair
Accelerated growth
Advanced BA

Evaluation

- Bone age: 10 proximally, 10 ½ distally
- Labs
  - LH 7.9 mIU/ml (prepubertal <0.3)
  - FSH 7.6 mIU/ml (prepubertal <4.2)

CAH work up
- AM 17 OHP 244 ng/dl
- 17 OH post ACTH 1233 ng/dl
- Cortisol post ACTH 26 mcg/dl



Treatment

- Treated central puberty
  - LHRH agonist
    - Estrogen is advancer of BA
    - More effective in younger girls
- Did not treat mild CAH
  - Treatment ↑ risk of adrenal insufficiency
  - Over treatment may stunt growth
  - Parents fearful of steroids

Case 2

- FL age 14y 8 m referred to pediatric endocrinology for short stature and delayed puberty
- PMHx
  - Birth history
    - 7lb 8 oz product of a term pregnancy
    - 2 days in NICU for meconium aspiration
    - No hypoglycemia or jaundice

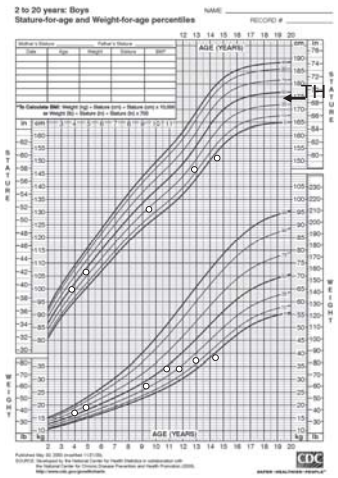
Significant past history

- 5 y crampy abd pain during/post eating
  - w/u age 10 negative H pylori, fecal calprotectin
  - w/u age 13 normal endoscopy
  - Miralax helps with constipation
- ADHD treatment started second grade
  - Gained 5 pounds in last several weeks off medication

Family History

- Mom 5'5"
  - menarche at 16-17
  - Irritable bowel syndrome
  - 2 maternal relatives with inflammatory bowel disease
- Dad 5'7"
  - Normal puberty
- Sister ADHD

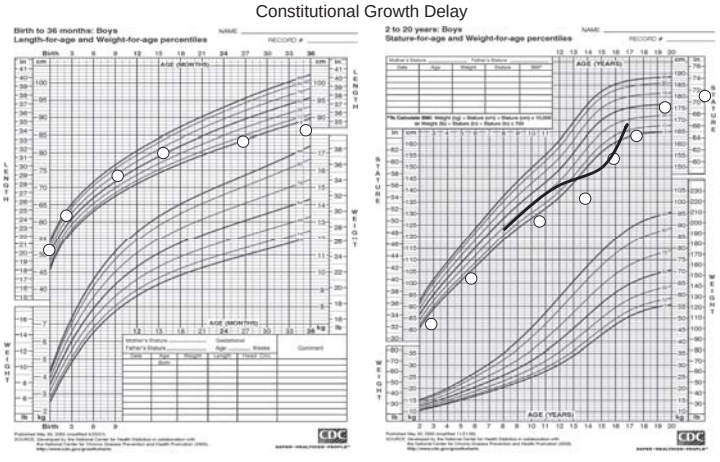
Exam:
Not dysmorphic
BP 107/65
GU tanner II
Testes 3-4 ml



Etiology of Delayed Puberty

Constitutional Growth Delay
Chronic illness
Endocrine disease which delays bone age
Failure of the hypothalamic pituitary gonadal axis

Constitutional Growth Delay



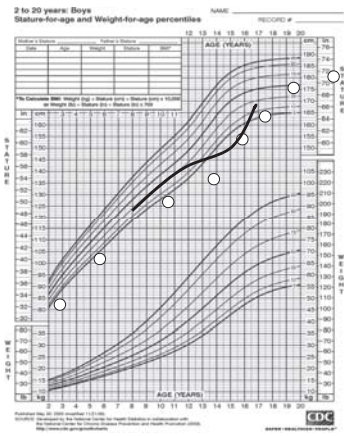
Constitutional Growth Delay

Positive Family History

Delayed BA

Delayed puberty

Chronic illness and hormone deficiency ruled out



OK135057

Growth Failure and Nutrition Considerations in Chronic Childhood Wasting Diseases

Nutrition in Clinical Practice
Volume 30 Number 2
April 2015 227–238

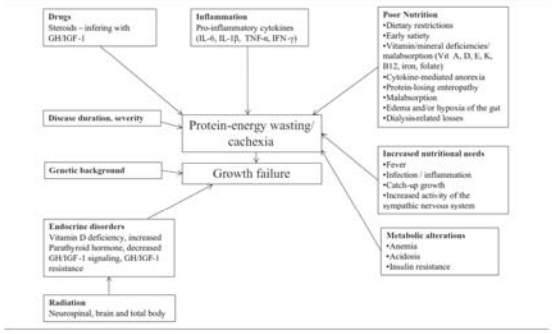
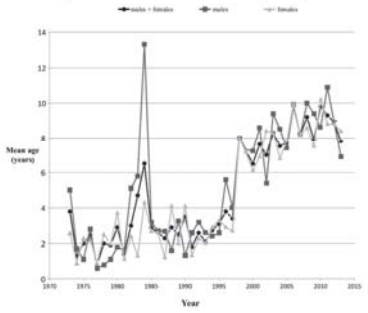


Figure 1. Contributing factors to growth failure in pediatric patients. GH, growth hormone; IFN-γ, interferon-γ; IGF-1, insulin-like growth factor-1; IL, interleukin; TNF-α, tumor necrosis factor-α.

The clinical presentation of coeliac disease in 1030 Swedish children: Changing features over the past four decades

Dimitrios Tapsas[a,*], Elisabet Hollén[b], Lars Stenhammar[a,c,d], Karin Fälth-Magnusson[a,e,f]

Digestive and Liver Disease 48 (2016) 16–22



[b] Pre-epidemic period: 1973–1983.
[c] Epidemic period: 1984–1996.
[d] Post-epidemic period: 1997–2013.

The clinical presentation of coeliac disease in 1030 Swedish children: Changing features over the past four decades

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Digestive and Liver Disease 48 (2016) 16–22

Table 2
Frequencies of presenting symptoms at different study periods and age groups at diagnosis.

	Gastrointestinal symptoms, *n* (%)	Extra-intestinal symptoms, *n* (%)	FTT and/or short stature, *n* (%)
Pre-epidemic[b] *n* = 98	72(82%)	38(39%)	69(70%)
Epidemic[c] *n* = 319	224(68%)	93(29%)	234(73%)
Post-epidemic[d] *n* = 613	436(71%)	118(19%)	158(26%)
p-Value[a]	0.82	<0.001	<0.001

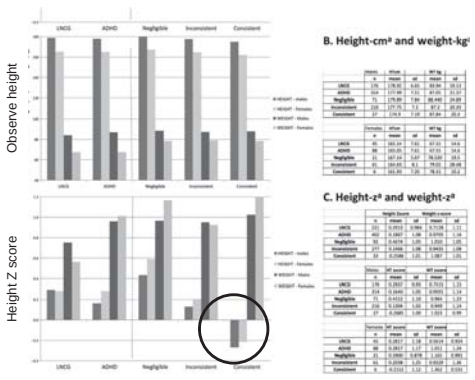
[b] Pre-epidemic period: 1973–1983.
[c] Epidemic period: 1984–1996.
[d] Post-epidemic period: 1997–2013.

Growth in Children on ADHD medications

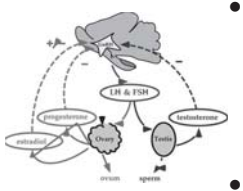
J Child Psychology Psychiatry:58;663 (2017)

- Observational long term follow up
  - 515 ADHD (age 7-10)
  - Treatment monitored to age 18
- 289 classmates without ADHD (LNGC)
- Height at age 25
- Conclusion: extended use of medication associated with suppression of adult height

J Child Psychology Psychiatry:58;663 (2017)



Failure of the Hypothalamic Pituitary Gonadal Axis

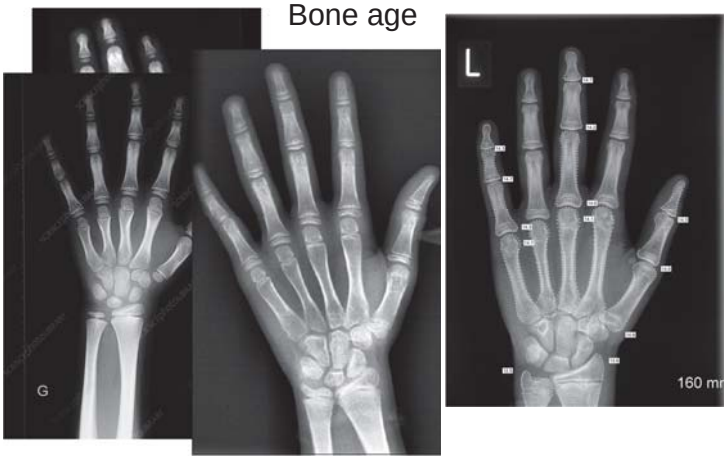


- **Hypothalamic Pituitary Dysfunction**
  - LH/FSH deficiency- isolated as in Kallmann's syndrome or as part of hypopituitarism
  - Hyperprolactinemia- prolactinoma or medication induced
  - Functional deficiency due to calorie insufficiency or excessive exercise
- **Gonadal Failure**
  - Females: Turner syndrome, oophoritis, galactosemia, chemotherapy, XX or XY gonadal dysgenesis
  - Males: vanishing testis syndrome, chemo or radiation

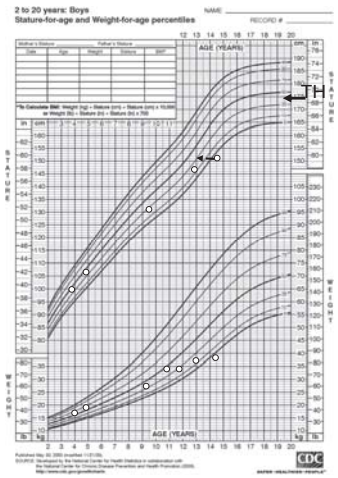
Laboratory evaluation

- Age 14
  - freeT4 1.36, TSH 1.66
  - Serum IgA 118, TTg 0
- Age 14 ½
  - BA 12 ½ to 13 ½
- Age 14y 8m
  - IGF 1 316 ng/dl (156-554)
  - LH 2.2 (prepubertal <0.3), FSH 3.8
  - Testosterone 67 ng/dl (Tanner II 18-150)

Bone age



Exam:
Not dysmorphic
BP 107/65
GU tanner II
Testes 3-4 ml



Conclusion

- Most likely constitutional growth delay
  - Bone age non specific test
  - At a bone age >12 ½ should be in puberty; exam and labs suggest he is
  - No idea about tempo, could be partial gonadotropin deficiency
- No lab evidence for growth hormone or thyroid hormone deficiency
- Are ADHD meds responsible? Does he have a hidden GI illness?
- Plan: observe progress in growth and puberty over next 6 months

Case 3

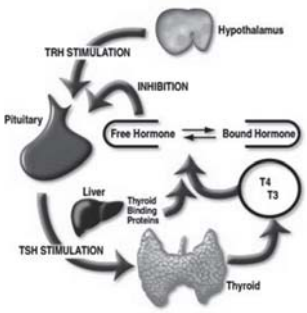
AV is a 15 year old girl referred for evaluation of abnormal thyroid function tests discovered in a work up for fatigue

Date	Free T4 (.58-1.64)	TSH (0.5-4.3)	
4/11/2019	4.4	0.58	
4/20/2019	5.71	0.94	

- Seen 4/26/2019
  - Fatigue since September 2018
    - Often naps after school
    - Always exhausted
  - Sleeps well at night time
  - Heavy periods since age 10 ½ started on ocp 4/11/2019
  - Often hot
  - Lightheaded when stands
  - No increased appetite, no racing heart beat
  - Biotin supplement for 1 year recommended by hair dresser
- Family HX negative for thyroid disease

- Exam: height 154.6 cm, weight 63.5 kg, BMI 26.6 BP 119/49
  - General- well appearing, no tremor, no sweaty hands
  - HEENT: no exophthalmos, no thyromegaly
  - Tanner V
  - Neuro: DTRs 2+

Interpreting Thyroid Function Tests



- Elevated free T4 and low TSH think hyperthyroidism
- Low free T4 and elevated TSH think primary hypothyroidism
- Low free T4 and normal TSH think hypopituitarism or non thyroidal illness

Elevated Free T4
(free T4 5.7, TSH 0.94)

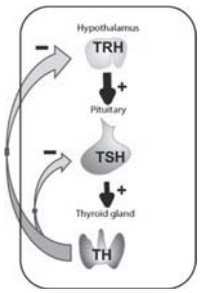
- TSH should be actually low in hyperthyroidism
- Could this be thyroid hormone resistance?
- Could this be an abnormality of thyroid binding showing up in the particular “direct” free T4 assay?
  - Birth control pills with estrogen raise TBG
- Could something be interfering with these assays?

Signs and Symptoms of Hyperthyroidism

- Goiter
- Prominent eyes
- ↑ HR
- Nervousness
- Sweating
- ↑ appetite
- Weight loss
- Deterioration in school
- Emotional disturbance
- Heat intolerance
- Fatigue/breathlessness
- Diarrhea

Graves’ Hyperthyroidism: Epidemiology

- Children 1:5,000
- Adults 1:500
- Peak age 11-15 years
- ♀:♂ **= 5:1**
- **Labs: ↑ Free T4, ↓ TSH**



Syndromes of reduced sensitivity to thyroid hormone: genetic defects in hormone receptors, cell transporters and deiodination

Best Practice & Research Clinical Endocrinology & Metabolism
Vol. 21, No. 2, pp. 277–305, 2007

- Thyroid hormone action defect
  - Thyroid hormone receptor beta gene defects
  - Increased T4
  - Normal TSH

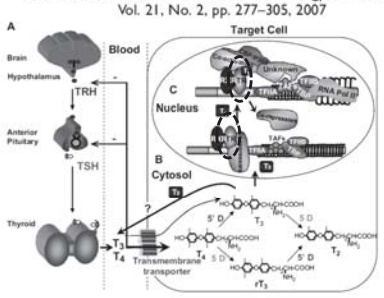


Figure 1. Regulation of thyroid hormone (TH) supply, metabolism, and genomic action. (A) Feedback control that regulates the amount of TH in blood. (B) Intracellular metabolism of TH, regulating TH bioactivity. (C) Genomic action of TH. For details see text. 5'D, deiodination by removal of an iodine from the 5' position in a iodothyronine; 5D, 5-deiodination; CBP/P300, cAMP-binding protein/general transcription adaptor; rT_3 reverse T_3; RXR, retinoid X receptor; T_2, diiodothyronine; T_3, triiodothyronine; T_4, thyroxine; TAF, TBP-associated factor; TBP, TATA-binding protein; TFIIA and TFIIB, transcription intermediary factor II, A and B; TR, TH receptor; TRH, TSH releasing hormone; TSH, thyroid stimulating hormone (thyrotropin).

Syndromes of reduced sensitivity to thyroid hormone: genetic defects in hormone receptors, cell transporters and deiodination

Best Practice & Research Clinical Endocrinology & Metabolism
Vol. 21, No. 2, pp. 277–305, 2007

Table 1. Clinical features: frequency of symptoms and signs (data derived from refs. 5, 16 and 50).

Symptom	Frequency (%)
Thyroid gland	
Goiter	66–95
Heart	
Tachycardia	33–75
Nervous system	
Emotional disturbances	60
Hyperkinetic behavior	33–68
Attention deficit hyperactivity disorder	40–60
Learning disability	30
Mental retardation (IQ < 70)	4–16
Hearing loss (sensorineural)	10–22
Growth and development	
Short stature (< 5%)	18–25
Delayed bone age > 2 SD	29–47
Low body mass index (in children)	33
Recurrent ear and throat infections	55

IQ, intellectual quotient; SD, standard deviation.

AV is a 15 year old girl referred for evaluation of abnormal thyroid function tests discovered in a work up for fatigue

Date	Free T4 (.58-1.64)	TSH (0.5-4.3)	
10/28/2016	1.37	0.92	
4/11/2019	4.4	0.58	
4/26/2019	5.71	0.94	

Drug Effects on the Thyroid

Henry B. Burch, M.D.

N ENGL J MED 381;8 NEJM.ORG AUGUST 22, 2019

- Interfere with endogenous thyroid function
- Interfere with thyroid hormone therapy
- Interfere with thyroid labs

Table 2. Drugs That Cause Spurious Thyroid Test Results in Euthyroid Persons.

Drug	Drug Class	Test Results			Condition Mimicked
		Thyrotropin	Free T_4	T_3	
Amiodarone	Class III antiarrhythmic agent	High end of normal range	High	Low end of normal range	Thyrotropin-secreting pituitary adenoma, thyroid hormone resistance
Biotin	Micronutrient	Low	High	High	Primary hyperthyroidism
Carbamazepine and oxcarbazepine	Antiepileptic agent	Normal	Low	Low end of normal range	Central hypothyroidism
Enoxaparin	Anticoagulant	Normal	High	High	Thyrotropin-secreting pituitary adenoma, thyroid hormone resistance
Heparin	Anticoagulant	Normal	High	High	Thyrotropin-secreting pituitary adenoma, thyroid hormone resistance
Phenytoin	Antiepileptic agent	Normal	Low	Low end of normal range	Central hypothyroidism
Salsalate	Nonsteroidal anti-inflammatory drug	Normal	Low end of normal range	Low end of normal range	Central hypothyroidism

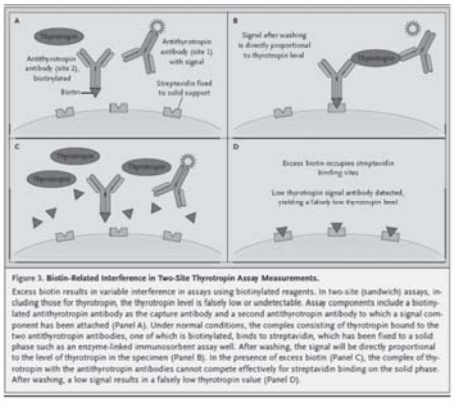


Figure 3. **Biotin-Related Interference in Two-Site Thyrotropin Assay Measurements.**
Excess biotin results in variable interference in assays using biotinylated reagents. In two-site (sandwich) assays, including those for thyrotropin, the thyrotropin level is falsely low or undetectable. Assay components include a biotinylated antithyrotropin antibody as the capture antibody and a second antithyrotropin antibody to which a signal component has been attached (Panel A). Under normal conditions, the complex consisting of thyrotropin bound to the two antithyrotropin antibodies, one of which is biotinylated, binds to streptavidin, which has been fixed to a solid phase such as an enzyme-linked immunosorbent assay well. After washing, the signal will be directly proportional to the level of thyrotropin in the specimen (Panel B). In the presence of excess biotin (Panel C), the complex of thyrotropin with the antithyrotropin antibodies cannot compete effectively for streptavidin binding on the solid phase. After washing, a low signal results in a falsely low thyrotropin value (Panel D).

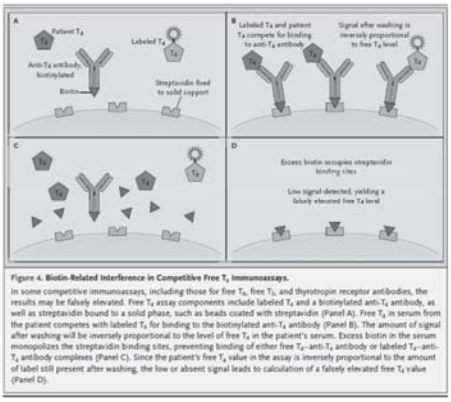


Figure 4. Biotin-Related Interference in Competitive Free T_4 Immunoassays.

In some competitive immunoassays, including those for free T_4, free T_3, and thyrotropin receptor antibodies, the results may be falsely elevated. Free T_4 assay components include labeled T_4 and a biotinylated anti-T_4 antibody, as well as streptavidin bound to a solid phase, such as beads coated with streptavidin (Panel A). Free T_4 in serum from the patient competes with labeled T_4 for binding to the biotinylated anti-T_4 antibody (Panel B). The amount of signal after washing will be inversely proportional to the level of free T_4 in the patient's serum. Excess biotin in the serum monopolizes the streptavidin binding sites, preventing binding of either free T_4– anti-T_4 antibody or labeled T_4– anti-T_4 antibody complexes (Panel C). Since the patient's free T_4 value in the assay is inversely proportional to the amount of label still present after washing, the low or absent signal leads to calculation of a falsely elevated free T_4 value (Panel D).

AV is a 15 year old girl referred for evaluation of abnormal thyroid function tests discovered in a work up for fatigue

Date	Free T4 (.58-1.64)	TSH (0.5-4.3)	Free T4 Equilibrium dialysis (0.8-1.7)
10/28/2016	1.37	0.92	
4/11/2019	4.4	0.58	
4/20/2019	5.71	0.94	
5/7/2019 off biotin	>6	0.76	

AV is a 15 year old girl referred for evaluation of abnormal thyroid function tests discovered in a work up for fatigue

Date	Free T4 (.58-1.64)	TSH (0.5-4.3)	Free T4 Equilibrium dialysis (0.8-1.7)
10/28/2016	1.37	0.92	
4/11/2019	4.4	0.58	
4/20/2019	5.71	0.94	
5/7/2019	>6	0.76	1.2

Free T4 by equilibrium dialysis

- Gold standard
- Helpful in patients on medications known to interfere with thyroid labs
- Helpful when things do not make sense

Peds ED Greatest Hits!
...well, actually, misses…
well, actually, *my* misses…

Beech Burns, MD,MCR
October 18th, 2019

Case #1

- CC: Abnormal anus
- HPI: 25 do boy with poor rectal tone, decreased PO intake. Adoptive mom noted rectal protrusion a few days PTP, feels it is getting worse. Constant trickle of stool, mustardy yellow, has not seen blood until today when she's noticed some small blood from area of mucosal breakdown on right perianal area
- No vomiting. Feeding reduced significantly in last 24h, usually eats 3oz at a time, now 1oz, still eating q3h. No fevers. No coughing. No significant nasal secretions.
- Normal MRI to evaluate sacral dimple 1 day PTP.

Case #1

- PMH:
  - C section. No complications. Immunizations utd. Full term
- FH:
  - Maternal asthma
- SH:
  - Lives with adoptive parents and adopted siblings
- ROS:
  - Otherwise negative

Case #1

- BP 83/61 T 36.7 HR 130 RR 48 SpO2 100%
- Gen: No distress. Interactive, sucking on pacifier
- Head: NCAT, AFSF, scattered petechiae around eyes bilaterally
- GU: normal
- Skin: Small ulcer with denuded head approximately 0.5cm diameter at right side immediately adjacent to anus. Abnormal appearing anus, no anal wink. Large low-lying sacral dimple

Case #1

What is this?

- Rectal prolapse?
  - Milk protein allergy
  - Neurogenic dysfunction related to tethered cord
  - Infection
- What about the facial petechiae?

What should we have done?

What we did

- CBC, BMP, Coags
  - Normal
- Neurosurgery consult for abnormal rectal tone
  - Imaging reassuring. Follow-up in clinic
- Pediatric Surgery consult
  - Close outpatient f/u, change formula to hydrolyzed, barrier ointment
- Evaluated by hospitalist at bedside
- Discharged

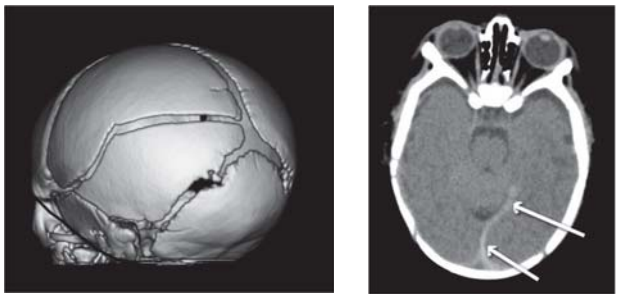
What happened?

- RTED 23 days later…
- "7 wo boy brought in for ALTE. Patient and Dad were home alone, patient was crying and inconsolable, Dad went to change his diaper when he felt that patient went limp and unresponsive for at least 30 minutes. Dad denies that he was choking or had repetitive shaking movements. Once Mom got back patient still was not acting normally. His eyes were pulled open and they were rolled upwards, Mom tried to open his mouth to check on his tongue (and make sure he wasn't choking on it) and felt that his jaw was clamped shut. 911 was called."

What happened?

- BP 123/72 T 35.9 HR 128 RR 48 SpO2 100%
- Well developed, active, strong cry
- Head: AFSF, some scalp tenderness on left side
- Skin: No bruising noted

Head CT



What happened next

- Neurosurgery, trauma consult
- Admitted to PICU
- SCAN team recommended CBC, CMP, urinalysis, UDS
- Social work consult
- DHS report made
- Child abuse investigation team from Portland Police Department came to Peds ED

What happened next

Intracranial injury:
Left parietal skull fracture, bilateral subdural hygromas, falcine subdural hematomas
Scattered subarachnoid blood
Concern for shear injury near corpus callosum

Skeletal injuries:
Bilateral wrist fractures
Subacute multi segmental bilateral rib fractures
Right proximal femur fracture

Ocular findings:
Bilateral diffuse scattered intra-retinal and pre-retinal hemorrhages
Diffuse roth spots scattered throughout periphery, both eyes

Abdominal injury:
Small liver lacerations x 2

What is a "sentinel injury"?

- A sentinel injury is a minor injury in a young child that is poorly explained and therefore concerning for physical abuse
- Abuse tends to get more severe over time
- Failure to recognize and take action when relatively minor, suspicious injuries occur may have devastating consequences for the infant and family.

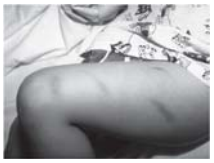
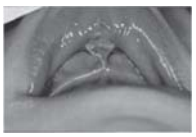
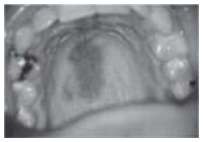
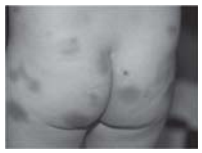
Sentinel Injuries

- About 30% of children with AHT and 20% of abusive fractures are initially missed
- In 2006 study, 30% of children who died of child abuse had documented health care visits for reasons other than routine well-child care in the year before their death
  - 19% of these children had visits 1 month before their death
- In 2013 study of 400 case controls, 27.5% of definitely-abused patients had a previous sentinel injury compared with 0% of non-abused children
  - In definitely-abused group, 42% of sentinel injuries were known by medical provider
- In 1999 study in JAMA, diagnosis of AHT more likely to be missed in intact, non-minority families

Possible Sentinel Injuries

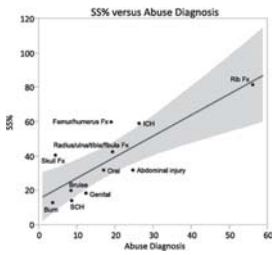
- Bruises in unusual locations
- Bruises in unusual patterns
- Burns
- Bite marks
- Intraoral injuries
- Fractures

Possible Sentinel Injuries

What are we likely to see?

- Highest Risk?
  - Fracture (rib)
- Most common?
  - 80% bruise
  - 11% intraoral injury
  - 7% fracture



Outcome

- 9 month well child check
- New adoptive family with 4 biological kids, 1 foster child, 1 exchange student, and 2 adopted children
- Normal growth and development

Key Takeaways (for me)

- Take a thorough history, scrutinize it
- Examine the patient closely
- Detection of sentinel injuries may save a child's life

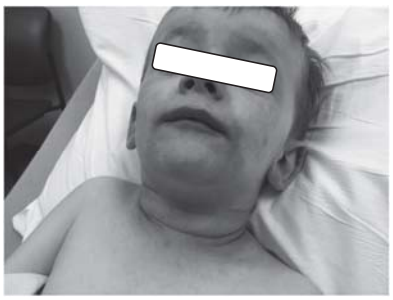
Case #2

- CC: Rash
- HPI: 3 yr old healthy boy with rash. Started 2 days ago as chapped lips. 1 day ago developed a neck rash + crustiness on left scalp. Parents also mention mild swelling on foreskin of penis (improving over past few weeks). Sore throat 1 day ago. No new exposures. No significant sun exposure
- PMH: None
- Relevant PSHx, Meds, Allergies, Social Hx:
  - No surgeries, daily medications, allergies, lives at home with parents, no sibs

Case #2

- HR: 106 BP: 122/82 RR: 22 Temp: 37.1 O2 Sat: 99% RA
- Gen: Well appearing, well developed, non-distressed
- HEENT: Small white patches on tonsils with mild erythema; no lesions on buccal mucosa; crusting in EACs. Ears are both erythematous but not tender. Perioral erythema. 3cm diameter crusted lesion on L parietal scalp w/o erythema
- GU: Penis normal, no swelling or erythema
- MSK: Patient holds arms flexed against body and resists attempts to move them upwards; erythema in AC fossae and the axillae bilaterally; no axillary LAD
- Skin: Blanching erythematous rash circumferentially around the neck, on the ears, and the perioral area

Case #2



What is this?

- Cellulitis
- Contact dermatitis
- Scarlet fever
- Id reaction to tinea infection
- Roseola
- Seborrhea
- Pharyngitis
- Stevens-Johnson Syndrome

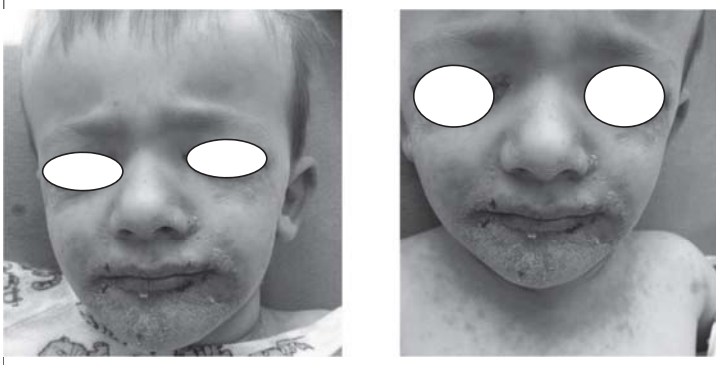
What should we have done?

What did we do?

- Rapid strep negative, culture negative (previous day)
- Received 400mg PO Tylenol + 9mg (0.6mg/kg) PO Decadron
- Observed with no change in appearance but improved pain. Discharged home with 14 day course of Griseofulvin. Return for high fever , n/v, decreased mental status

What happened?

- RTED 2 days later (day 4 of rash):
- Rash worse around lips and mouth (ulcerated). Now involving peri-orbital region. Extreme discomfort – refusing to open eyes or mouth with no oral intake in 1 day. Eyes not red but with yellow drainage. Sloughing rash in armpits
- Physical Exam:
- Vital Signs: HR: 107 BP: 107/62 RR: 22 Temp: 36.7 O2 Sat: 99% RA
- Skin: Skin is warm, CR < 2sec. Plaques with scale and crusting on neck, around mouth, behind ears. Crusting in external auditory canals with some draining. Lips are ulcerated and edematous. Desquamation present. Conj without injection but lids matted, yellow drainage bilaterally.
- GU: Inguinal folds with erythematous macules and papules



Return visit

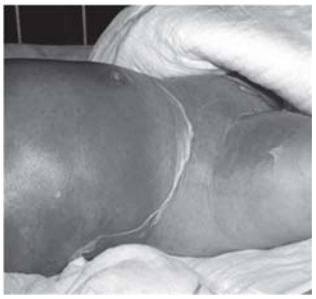
What happened next?

- CMP, CBC, lactate all reassuring
- CRP and ESR normal
- Rapid Strep: Negative
- Blood + Perioral Skin Cultures drawn
- Given IV fluids and Morphine
- Consulted Dermatology
- Admitted

Diagnosis?

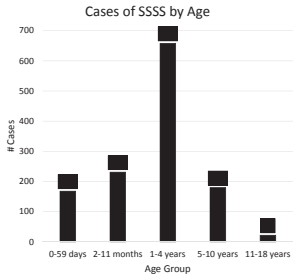
Staphylococcal Scalded Skin Syndrome!!

But isn't that for babies?

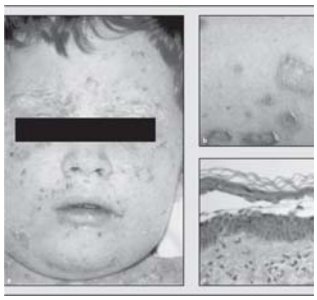


SSSS is all grown up…well, at least potty training

- 2018 study:
  - 1259 patients between 2011-2016
  - 84% ≤ 4 years old



But what does it look like in older kids?



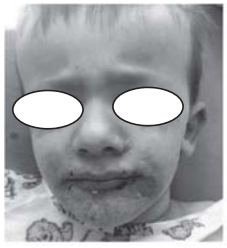
Patel, G.K. & Finlay, A.Y. Am J Clin Dermatol (2003) 4: 165. doi:10.2165/00128071-200304030-00003

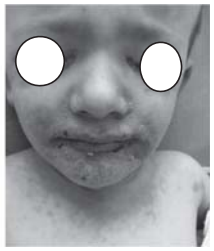
- "First signs are macular erythema and skin pain, initially accentuated in the skin folds, such as the neck, axillae, inguinal folds, and gluteal cleft"
  - 'Patient holds arms flexed against body and resists attempts to move them upwards; erythema in AC fossae and the axillae bilaterally'

What does it look like in older kids?

"Thick crusting and radial fissuring often develops around the mouth"

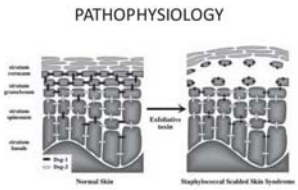
"The crusting, fissuring, and erythema can be striking and is classically referred to as SSSS 'sad face'"





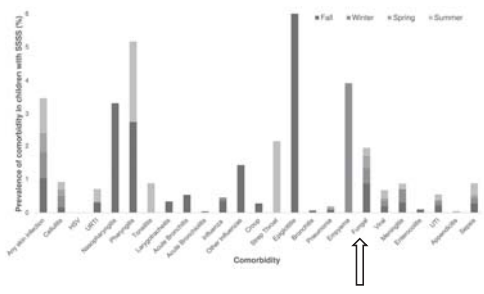
But why does it look different?

- "Protective antitoxin Abs in some children and most adults limits the lesions to a few localized blisters in milder forms, whereas lack of Abs in generalized SSSS allows hematogenous dissemination of ET to produce exfoliation that may cover the entire body surface." – *"Difficulties in diagnosis and management of the SSSS" in PIDJ 2000*



What about that fungal infection??

- Comorbidities for SSSS



Outcome

- ***Derm recs:***
  - IV Ancef (100mg/kg/day div q8) + IV Clindamycin (30mg/kg/day div q8)
  - Vaseline to affected skin areas
- ***Ophtho recs:***
  - No corneal/conjunctival involvement
  - Polytrim to lid margins TID
  - Warm or Cold Compress to break up eyelid crust
- **2 days after admission:**
  - Rash improving – Skin Cx: MSSA
  - Abx narrowed to IV Clindamycin
- **4 days after admission:**
  - Discharged on PO Keflex x 10 days
  - Polysporin BID (Eye lid) + Mupirocin/Vaseline TID
  - Ketoconazole shampoo daily x2 weeks then twice weekly as needed

Key Takeaways (for me)

- Staph scalded skin syndrome is not strictly a neonatal disease (children under 4 most common)
- The presentation may be more subtle…***skin pain*** is a key initial feature
- A kid who won't show you his armpits has SSSS until proven otherwise…or he's ticklish…

Case #3

- CC: Abdominal pain, hypoxia
- HPI: 5 yo boy transferred from OSH with concern for abdominal pain and hypoxia. N/V/D developed 5 days PTP. Tactile fever daily. Vomiting and diarrhea resolved, then congestion, cough developed. Today, family noted increased respiratory rate, working harder to breathe. Taken to OSH ED. There, febrile and hypoxic. Started on 3L NC. CMP with Na 128, K 3.2, Cl 88, AP 86, AST 58, ALT 48. CBC with WBC 26.8K, bands 49%

Case #3

- PMH:
  - Healthy, no hospitalizations, chronic med problems
- Relevant PSHx, Meds, Allergies, Social Hx:
  - Lives with parents
- Imm:
  - UTD per report

Case #3

Vital Signs: HR: 124 BP: 96/60 RR: 40 Temp: 38.4 O2 Sat: 88%
Constitutional: Listless young boy in moderate respiratory distress
CV: Tachycardia, no m/r/g
Resp: In respiratory distress. Decreased AM, very decreased on right. + retractions. No rales, rhonchi, wheeze
Abd: Soft, BS normal. No distension. Very TTP in RLQ, periumbilical area
Neuro: Normal
Skin: No rash, CR <3 sec

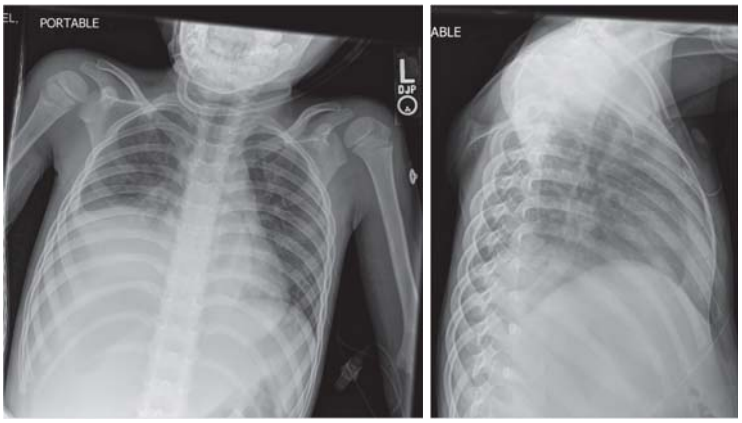
What is this? Why is this child hypoxic? This doesn't sound like appendicitis…

- Bacterial pneumonia
- Complicated PNA with effusion
- Atelectasis due to splinting from abdominal process

What should we have done?

What did we do?

- Increased oxygen to 4L
- Gave NS bolus
- Obtained a chest X-ray



Aha! Could this be pneumonia masquerading as appendicitis???

Let's look at the literature!

Basilar Pneumonia Simulating Acute Appendicitis in Children

Juda Z. Jona, MD, Robert P. Belin, MD

552 Arch Surg—Vol 111, May 1976

- N = 250 kids examined for acute abdomen between 1972-1975
- 12 cases of PNA (4.8%)
- All with pain severe enough to suggest acute appendicitis
- 3 had appendectomies (!)

- "(Our findings are) indeed a strong argument for obtaining chest roentgenograms on all children who have symptoms of an acute abdomen."

Pneumonia presenting with acute abdominal pain in children

D. RAVICHANDRAN* and D. M. BURGE
*Wessex Regional Centre for Paediatric Surgery and the *University Surgical Unit, Southampton General Hospital, Tremona Road, Southampton SO16 6YD, UK*
Correspondence to: Mr D. M. Burge

N = 1168
1986-1992, admitted with abdominal pain

 British Journal of Surgery 1996, 83, 1706–1708

Table 1 Patient details

No. of patients	19
Male	16
Age (years)	
3–5	9
6–12	10
Mean	6·5
Mean duration of symptoms	20·5 h (range 2 h to 4 days)
Symptoms of upper respiratory infection	16
Physical signs of lung infection	8
Respiratory rate on admission (per min)	
≥40	2
<40	17
General practitioner diagnosis	
Acute appendicitis	13
No firm diagnosis	6
Diagnosis of pneumonia	
On admission (<2 h)	8
Later than 2 h	11
Site of pneumonia	
Right lung	13 (upper lobe 2, middle lobe 6, lower lobe 5)
Left lung	6 (all lower lobe)
Mean (range) white blood cell count ($\times 10^9$/l) ($n = 15$)	19·3 (9·7–29·9)
Blood culture ($n = 14$)	
Positive	1 (*Staphylococcus aureus*)
Negative	13
Laparotomy performed	1

PEDIATRICS

Prevalence of pneumonia in children under 12 years of age who undergo abdominal radiography in the emergency department

Valérie Homier, MD;* Colette Bellavance, MD;*† Marianne Xhignesse, MD‡

- Retrospective study.
- N = 1613 pts under 12 who got KUB and CXR
- 30 cases of pneumonia (1.89%)
- All but 2 had fever, cough, or URI symptoms

Back to our case…

- Cough? ☑
- Fever? ☑
- URI symptoms? ☑

Nice try, kid! You have to wake up pretty early in the morning to fool me…

MDM

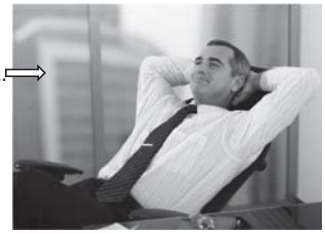
- 5 yo boy who presents with nausea, vomiting, and diarrhea, followed by development of fever and cough found on exam to have decreased breath sounds on the right and hypoxia concerning for bacterial pneumonia. Differential diagnosis also includes viral pneumonia, pleural effusion, atelectasis with abdominal pathology including appendicitis. Patient has marked leukocytosis with bandemia concerning for bacterial infection.
- CXR obtained, which revealed right lower lobe and right middle lobe pneumonia.
- Oxygen saturations were 88% on 3 L on arrival. Increased 4 L with increase in oxygen saturations to 98%. With oxygen requirement, tachypnea, signs of dehydration, patient warrants admission for further care. We gave ampicillin IV ×1 dose. We also gave a normal saline bolus. Given mild hyponatremia, hypokalemia, and hypochloremia, electrolytes should be followed on admission

Timeline of Care

• 0023	T 38.4 HR 124 BP 96/60 RR 40 SpO2 88% 3L
• 0047	CXR ordered
• 0107	Ampicillin, NS bolus ordered
• 0130	HR 115 BP 89/58 RR 42 SpO2 98% on 4L
• 0238	T 37.1 HR 112, patient reports belly feels better. Decreased WOB. Awaiting admission.
• 0306	Admitted to ward

Another job well done…

- Me at the nurse's station… ⟹

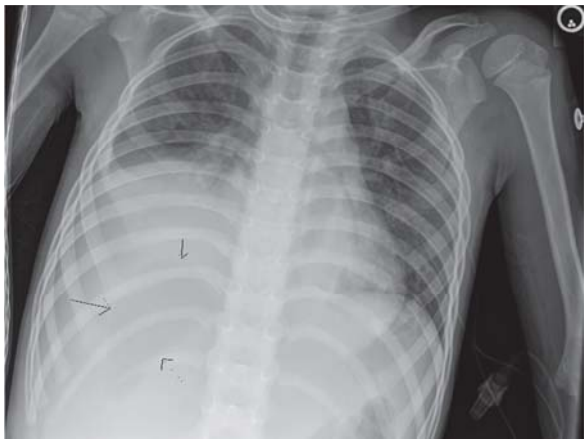


Following admission

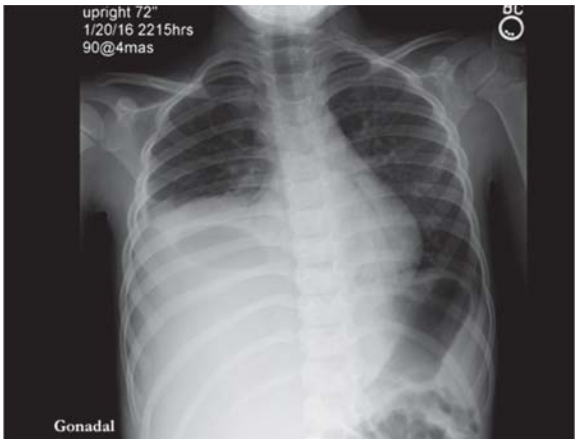
• 0837	Radiology calls inpatient team after reading the film…

Impression: Extensive right lower lobe consolidation is noted with adjacent pleural effusion. Left lower lobe atelectasis also present. Bilateral airway thickening and low lung volumes. Ovoid loculated gas projects over the liver not definitively within bowel. If there is concern for bowel perforation or abscess, consider a left lateral decubitus view.

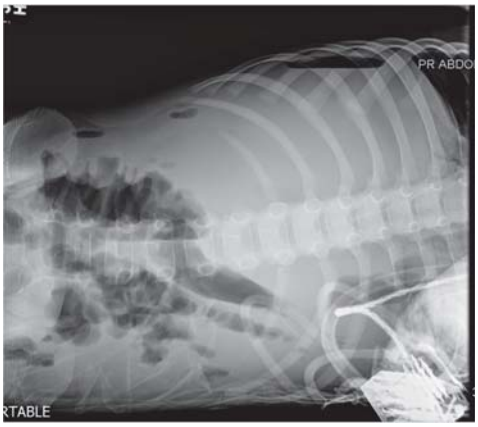
A Closer Look…



Outside Film



LL Decub Film



Radiology Read:

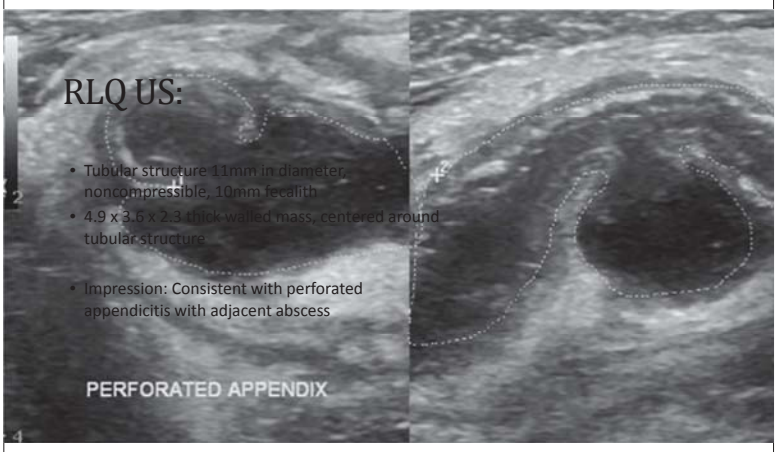
- IMPRESSION
  Pneumoperitoneum, gaseous bowel distention and multiple air-fluid levels.
- Findings are concerning for perforated appendicitis given history of fevers and right lower quadrant pain.

What happened next?

- Surgery consult
- Started Zosyn
- RLQ US

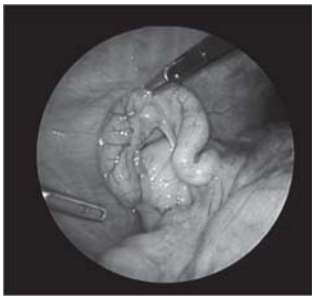
RLQ US:

- Tubular structure 11mm in diameter, noncompressible, 10mm fecalith
- 4.9 x 3.6 x 2.3 thick walled mass, centered around tubular structure
- Impression: Consistent with perforated appendicitis with adjacent abscess

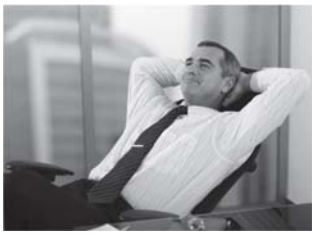


What happened next?

- Laparascopic appendectomy
- Omentum adherent to right abdominal wall. Large abscess in right pericolic gutter. Second larger abscess found in the suprahepatic space. JP drain left in place.



What about the pneumonia? What am I supposed to tell this guy?



Ah…the old reactive-pleural-effusion-secondary-to-abdominal-abscess trick…

Q: How common is this?

- Empyema and lung abscess as complication of a perforated appendicitis in a pregnant woman. *Int J Surg Case Rep.*, 2012; 3(12)
- Right postoperative pleural effusion following laparascopic appendectomies: a case series. *Ann R Coll Surg Engl.,* 2010; 92(5)
  - 3 consecutive cases, all with ruptured appendix
- Empyema. A rare presentation of perforated appendicitis. *JAMA,* 1978; 240(23)
  - 2 cases – 50 yo woman, 5 yo boy

A: Not very…

Outcome

- POD #0: PICU for monitoring
- POD #7: Recurrent fevers. CT abdomen showed 2 recurrent abscesses. IR drained and percutaneous drain left in place.
- POD #15: Discharged with IR drain in place with plan for clinic f/u. Discharged on Augmentin.

Key Takeaways (for me)

1. Review all available diagnostics
2. Phone your (radiology) friends
3. Think horses…then zebras… then think about some animal you've never heard of…
4. Practice humility in all things

Questions?

THANKS FOR YOUR ATTENTION!

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Challenging Pediatric Otolaryngology Cases

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Multi-disciplinary clinics within pediatric otolaryngology

- 1. Craniofacial clinic (Cleft lip and palate, craniofacial abnormalities)
- 2. Aero-digestive clinic (Pediatric ENT, Pulmonary, Speech and Feeding, GI)
- 3. Vascular anomalies (Pediatric ENT, Dermatology, Interventional Radiology)
- 4. Voice clinic (Pediatric ENT, Voice therapy)
- 5. Hearing loss clinic (Pediatric ENT, Speech, and Audiology)
- 6. Thyroid clinic (Pediatric ENT, Endocrinology, Radiology)

Case 1. – Vascular anomalies

- 3 year old girl with a known lymphatic malformation on her neck comes in to see you in clinic with increased pain and swelling on neck. It has doubled in size and she also has a URI symptoms (fever, cold).
- No difficulty breathing. Otherwise stable.

How do you treat her?

A. No treatment, it will get better on alone
B. Order an MRI or CT on patient
C. Refer to ENT immediately
D. Tx with antibiotics
E. Treat with antibiotics and steroids

Lymphatic malformations

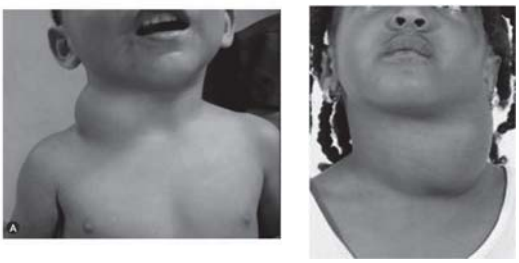


Figure 1: http://www.sickkids.ca/PlasticSurgery/What-we-do/Vascular-Anomalies-Clinic/Vascular-Malformations/Lympathic%20Malformations/index.html

Lymphatic malformations-

- Lymphatic malformations are collections of dilated lymphatic channels which can vary widely in terms of their size and age of presentation
- Can get infected very easily, especially with onset of sickness
- Short term treatment is to treat infections with both antibiotics and steroids (usually 2 weeks abx, 5 days steroids at 2 mg/kg/day)
- Later treatments can include sclerotherapy, surgery, and new treatments such as sirolimus

Case 2 – Hoarseness

- 2 year old ex 28w preemie with a history of cardiac surgery(PDA ligation) comes in with a chronic history of hoarseness and voice straining during a well child check up. Mom complains that she still tends to cough and choke with liquids.

What is the most likely cause of her hoarseness?

1. Vocal cord nodules
2. Vocal cord paralysis
3. GERD
4. Laryngeal cleft

Vocal cord paralysis

- 1. Most commonly associated with cardiac surgery, prolonged intubation, thyroidectomy, and TEF repair.
- 2. Weak cry in infants
- 3. Difficulty feeding
- 4. Breathy soft voice
- 5. Tx include, voice therapy, **surgery**

Vocal cord paralysis

Vocal cord nodules

- 1. Most common cause of chronic hoarseness in school age children
- 2. Boys> Girls
- 3. Located at the junction of the anterior 1/3 and posterior 2/3 of vocal cords
- 4. Develop from repeated trauma to vocal cords
- 5. Voice therapy most indicated treatment- rarely surgery

Vocal cord nodules

Case 3

-15 month old comes in with choking with liquids and recurrent pneumonia.
-Also with chronic cough and not gaining weight.
-Mom said symptoms are worse when lying down.
- Has tried a trial of omeprazole with no help.
- No stridor. No history of intubation.
-prior MBBS at 6 months of age showed aspiration

What is the best type of test to order at this time?

1. Repeat swallow study (MBBS)

2. FEES (flexible fiberoptic laryngoscopy and function endoscopic evaluation) – better than MBBS in visualizing laryngeal function

3. GERD Testing –pH probe

4. Chest- xray (normal)

MBBS (Modified Barium swallow study)

- Inconsistent micro aspiration with the thin and nectar thick liquids. Resolved with nectar Plus thick liquids and pureed to dry soluble solid foods.

What is the child's diagnosis

- 1. Laryngomalacia
- 2. GERD-
- 3. Laryngeal cleft
- 4. Vocal cord paralysis

Type I Laryngeal cleft

- Feeding issues
- Failure to thrive
- Recurrent pulmonary issues (aspiration)
- Hoarseness
- 75% will have aspiration on MBBS

Laryngeal cleft

Type I –extends to level of vocal cords
Type II – extends below vocal cords into cricoid cartilage
Type III –extends to trachea/esophagus
Type IV – extends to level of trachea/esophagus

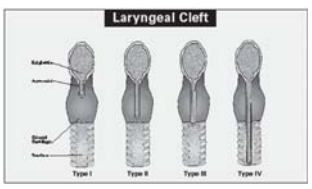


Figure 2: http://www.laryngeal-cleft.com/What-is-laryngeal-cleft

Treatment of laryngeal cleft

- 1. Direct laryngoscopy to view airway
- 2. If cleft is present (type I, II), endoscopic repair considered
- 3. Surgical repair outcomes favorable for cessation of aspiration
- 4. Repeat swallow study in 3 months

Case 4

- 3 month female former preemie 32 week old presents with loud stridor and follow up from ED.
- Was diagnosed with croup and RSV, but unresponsive to treatment in ED. Mom said noisy breathing is worse when feeding. Seems to be getting louder. No history of intubation.
- Gaining good weight
- No wheezing
- Chest x-ray is normal
- Swallow study normal

What is the cause of her stridor?

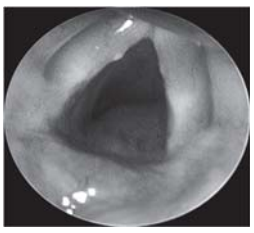
1. Laryngomalacia
2. Foreign body
3. Airway hemangioma
4. Vascular ring

Case 4

- **ENT consulted to do beside scope – No evidence of laryngomalacia**
- **Taken back for airway evaluation (MDL, Bronch)**

Airway hemangioma

Figure 3
https://www.sciencedirect.com/science/article/pii/S0030666508000844



Presentation of airway hemangioma

- More common in preemie, Caucasian, F > M
- Stridor occurring around 6-8 weeks of age- worse with feeding
- Other hemangiomas in “beard” distribution

Airway hemangioma

- Treated with propranolol 2mg/kg/day divided tid till 6 months of age, then can go to bid dosing- must give with feeding
- Stay on this dose till one year of age –PCP can adjust
- Symptoms resolve (stridor) in one to two weeks- no need for f/u airway exam.

Case 5 – Voice clinic

- 14 year old presents with shortness of breath with activity that began 2 years ago.
- Worse with activity. She is a competitive soccer player and began experiencing symptoms when starting more competitive play
- She is a straight A student, highly motivated
- Keeps her from performing sport, has tried albuterol inhaler given by PCP for exercise induced asthma, but not helping

Exercise induced laryngeal obstruction

- Usually in Adolescent females involved with competitive sports
- Anxiety and high stress commonly noted
- Majority treated with albuterol inhaler although PFT were normal.

What we do in Voice Clinic…

- Seen by Voice/Speech Therapist first who does flexible laryngoscope (may re-create symptoms by running up and down stairs)
- ENT evaluates scope with Speech (looks for an airway abnormalities that may be causing stridor- webbing, nodules, stenosis, etc..
- Come up with a treatment plan for the patient and family

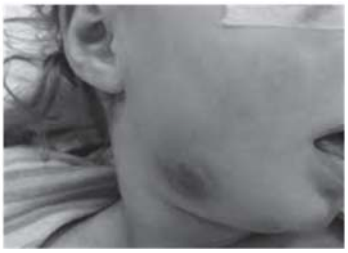
Vocal cord dysfunction

- Misdiagnosed as Asthma
- Triggered by exercise, stress
- Co occurs with asthma – 50%
- Responds very well to voice therapy (preventative and interruption technique)
- Sensation of throat tightness, sudden onset, trouble breathing in, and stridor on inhalation
- Non-responsive to inhalers

Case 6

- 2 year old girl, enlarging neck mass for 3 weeks
- Non-tender
- No associated symptoms
- Healthy child
- NO cats, one dog

- PE: Afebrile,
- Left submandibular mass -2.5 X 2 cm, starting to turn purple and has had some drainage



What is this neck mass?

Labs: Bartonella,

PPD

Atypical mycobacterial lymphadenitis

- 2-5 years of age; rare > 12 years
- Otherwise healthy
- Fish, turtles, birds
- Painless mass – overtime skin changes
- Submandibular
- Usually unilateral
- > 3cm in 80%
- Onset over weeks
- 35-40% suppurate

Treatment of NTM (non-tuberculosis mycobacteria)

- Usually no need for imaging
- Surgical excision better than FNA
- Surgical excision has a 96% cure
- About 67 % respond to antibiotics (clarithromycin + rifampin(lots of choices)
- (takes 12 weeks to respond to abx and need monitoring)
- Could go away without any treatment in 12 months (observation)

Case 7

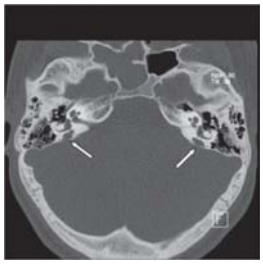
- 8 year old boy comes in for a well child check with a known left sided SNHL(sensorineural hearing loss). He has recently moved and tells you he feels like his hearing has changed.

After reviewing his records, You find the reason for his hearing loss is an enlarged vestibular aqueduct found on MRI.

Enlarged vestibular aqueduct (EVA)

- 40 % of kids with EVA with experience progression of hearing loss over time, on one side or both
- Head trauma may cause symptoms to worsen-controversial
- Accounts for about 23% of unilateral hearing loss
- Can have issues with balance and dizziness

Enlarged Vestibular Aqueduct



Unilateral hearing loss

- 59% of children with unilateral hearing loss can have academic or behavioral problems- speech, etc
- Preferential seating
- FM system
- Keeping other ear healthy- monitoring for ear infections
- HA use (binaural hearing)
- Regular audiograms
- Some may be candidates for cochlear implants

Immunizations for cochlear implants – CDC recommendations

- Infants below 2- Prevnar 13 routine
- Children (between 2nd and 6th birthday) – two doses of Prevnar 13 if they have not gotten PCV 7 or 13 previously. If they finished PCV 7- one dose of PCV 13
- Between ages 6 -18 – single dose of Prevnar 13 regardless of whether they received PCV7 or PPSV23.
- In addition all children age 2 years and older who have completed the Prevnar series should receive one dose Pnenumovax 23 (PPSV 23). Wait at least 2 months after last dose of Prevnar to receive Pneuomovax23.

Case 8

- 2 year old with a history of recurrent ear infections. She had tubes placed 6 months ago by ENT. She presents to your office with a draining ears. You start on her on ear drops and drainage goes away. She is back in your office again a month later with draining ears.

What to do?

- 1. Put her on an oral antibiotics
- 2. Get an ear culture and start antibiotic ear drops
- 3. Refer back to ENT
- 4. Do not treat

Clinical practice guidelines- American academy of otolaryngology

- Topical antibiotic eardrops only, without oral antibiotics, for children with uncomplicated acute tympanostomy tube otorrhea

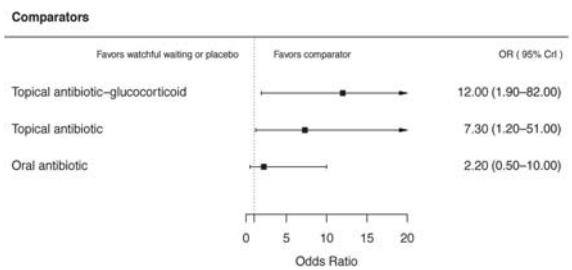


Figure 6: https://pediatrics.aappublications.org/content/139/6/e20170667

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Thank You

Top Cases in Pediatric Infectious Diseases
(and lessons learned)

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Disclosures

I have no financial relationships or Conflicts of Interest (COIs) to disclose

Objectives

At the conclusion of this session, participants will be able to:

1. Describe an approach to the differential according to presenting signs and symptoms for common pediatric complaints
2. Discuss the diagnostics of challenging cases in infectious diseases
3. Describe emerging infectious diseases affecting patients in the Pacific Northwest

It is really hard to be a PCP

Disclaimer: these are weird cases!

Imagine you had these patients – would you do anything differently?

What tools/resources/tests are available?

RESPECT

Presenting Symptom: Headache

14 year old male with several weeks of worsening headache.

HPI:

- Several weeks of headaches; Cold around Christmas.
- PCP visit– social issues highlighted (mental health, drug use).
- "Headache is **located across his forehead and is described as throbbing**. He rates the **pain at 9/10 currently, but 10/10 at night**. The intensity of the pain waxes and wanes. He reports associated **photophobia and hyperacusis**. He also reports mild dizziness when walking. He denies any fevers. No nausea or vomiting.
- **"He is very argumentative during our visit saying that he is just going to sleep and not go to school and he is going to eat whatever the hell he wants."**
- **ED: Non contrast Head CT normal; labs normal**
- **Initial diagnoses: Acute non-intractable headache vs. Tension type**

Pertinent History

- **PMH**: **Type 1 DM** (poorly controlled A1c 10.6); depression; obesity; immunized;
- **FMH**: Crohn's disease, stroke (father), Substance abuse (mother); Autoimmune (diabetes, thyroid) – maternal side
- **ROS**: No focal neuro deficits noted;
  - No fevers/vomiting/rashes/cough

1 day prior to arrival

- **ED:** "Headache is **throbbing, anterior, and equal bilaterally**. Pain has been temporarily **alleviated with use of ibuprofen and tylenol** at home. Today he reports his pain is **uncontrolled** despite use of ibuprofen around 1:30. The patient reports associated nausea with one episode of **vomiting**"
- Concern about exposure to antidepressants at the patient's mother's house. DHS is involved.
- No recreational or other substance exposure has been confirmed.

Dx: Acute non-intractable headache, unspecified headache type

Challenging case

Worsening headache

Vomiting / Nausea

Photophobia or other signs

Could this be a migraine?
CT is negative
Neuro visit is months away
Social overlay

Increasing number of ED visits → things not getting better

Day of admission (January)

- Parents report: child was **asleep most of day** and then woke up for dinner
- Began having tingling in hands and fingers
- Rapid neurologic change: incomprehensible speech and not following commands appropriately. Didn't know where he was
- **To ED** – concern for ingestion
- Exam: dilated pupils, combative, afebrile
- CTA: No official report - verbal was that there were **no concerns for stroke**.
- Intubated for LP: **CSF - R 2750 W: 680 L: 71 M:9 N:20; Glc: 108 Protein 145; Meningoencephalitis panel negative.**
- Transfer to DCH PICU for altered mental status

Differential?

Meningitis

Encephalitis

Sphenoid sinusitis

Brain abscess

Other brain lesion


IDSA GUIDELINES

The Management of Encephalitis: Clinical Practice Guidelines by the Infectious Diseases Society of America

IDSA GUIDELINES

Practice Guidelines for the Management of Bacterial Meningitis

News

West Nile virus cases reported in Deschutes, E. Oregon

By: AP
Posted: Sep 18, 2019 08:57 AM PDT
Updated: Sep 18, 2019 08:57 AM PDT

Tunkel AR et al. Practice guidelines for the management of bacterial **meningitis**. Clin Infect Dis. **2004** Nov 1;39(9):1267-84.
Tunkel AR. **The management of encephalitis: clinical practice guidelines by the Infectious Diseases Society of America.** Clin Infect Dis. 2008 Aug 1;47(3):303-27.

Exposures and Social History

- **Lives:** Recently in Southern Oregon: lived near woods with known ticks. Moved back up with father and stepmother in Eugene (town).
- **Recreation:** Camping in southern Oregon.
- **Food:** No raw or uncooked meats; no hunting; likes to garden, particularly, tomatoes.
- **Animals:** 2 cats, multiple dogs.
- **MRSA/TB/HSV:** Stepmother + HSV cold sores, none in past 2 months. No foreign travel.
- **SDRR:** Not sexually active, smoked marijuana, none in past month; Mom and her boyfriend smoke a lot.
- **Mental Health:** Recently removed from mother's home due to maternal substance abuse (alcoholism and MJ); sister attempted suicide with Benadryl overdose

Exam following transfer

T 39.7 C P 65 R 20 BP 110/58 SpO2 100%

Intubated, heavily sedated

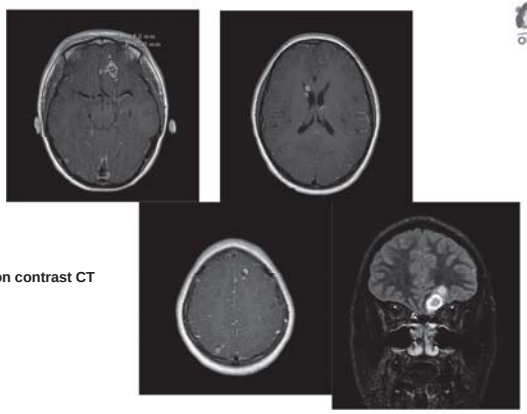
Neuro/Psych:
- Becomes **agitated and combative** with exam, not oriented
- When disturbed very agitated, opens eyes, no redirectable, beats hand on bed, not cooperative
- Normal reflexes

Exam otherwise normal

Outside Non contrast CTA: No extra-axial fluid collections. No parenchymal or subarachnoid hemorrhage. No midline shift or mass effect. **There is an area of nonspecific hypoattenuation within the inferior left frontal lobe. This measures 4.5 x 2.1 cm in size.**

Additional small focus of hypoattenuation is seen within the anterior left frontal lobe on axial image 20. A **small focus of hypoattenuation** is seen in the right parietal cortical region on axial image 26.

No morphological abnormalities of the ventricles. The sellar and pineal regions are unremarkable. No abnormalities of the basal cisterns.



What the non contrast CT missed

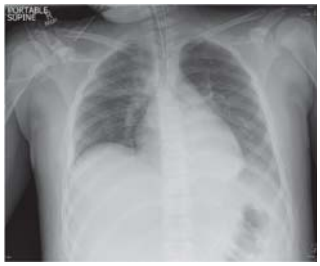
Brain MRI

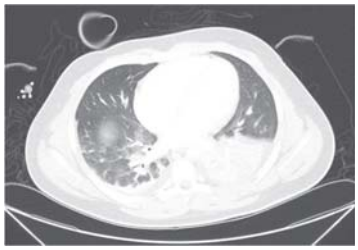
Multifocal rim-enhancing circular lesions. Largest at the left inferior frontal lobe. Mostly at grey-white matter junction

Left lateral ventricular intraventricular cyst

Mild diffuse leptomeningeal enhancement

Chest imaging: LLL PNA





Differential

Infectious
- Neurocystercercosis
- Toxoplasmosis
- Fungal/mold
- TB
- Echinococcosis
- Bacterial abscesses

Neoplastic
- Lymphoma
- Metastatic lesions from unknown primary site

Autoimmune

What antimicrobials would you start?

Vancomycin, ceftriaxone, and liposomal amphotericin B

Other considerations:

Acyclovir; doxycycline

RIPE or empiric treatment for toxo or neurcystercercosis?

steroids?

Additional studies

- LP: Opening pressure: **52 cm** WBC 594 (47% PMN), RBC 4, P79, G: 21
- **Meningoencephalitis panel: negative**
- EVD was placed for high ICP
- Right frontal endoscopic approach for biopsy of right frontal ventricular lesion
  - **Findings: Small pink exophytic mass; Not a cyst/larva/worm;** Touch prep revealed **abnormal cells**

Microbiology

- **Broad Range PCR**: Amplified something with fungal primers (ultimately unable to further identify)
- **Day 4: CSF CrAg 1:10 Serum CrAg 1:2560**
- **Path:** Yeast forms identified in specimen B, most compatible with Cryptococcus neoformans
- **Fungal CSF culture:** *Cryptococcus gatti* Molecular subtype: VGIIa
- – took several weeks to get this;

Hospital course

Amphotericin B (5 mg/k/day) and 5-FC

Full neurologic recovery by day 7 of amphotericin
- EVD removed at this time

Complicated by acute kidney injury: Serum creatinine: 0.56 →1.63

Remained inpatient for entire **6-week course of AmphoB**

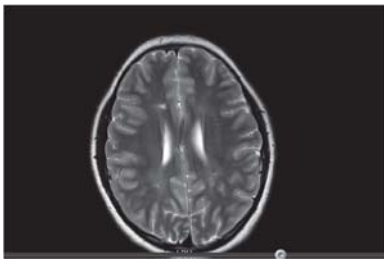
Discharged on **fluconazole**; has chronic kidney disease (Stage 3)

Outpatient Course

Has been on fluconazole for over 1.5 years.

MR this summer: There is continued regression of multiple contrast enhancing foci involving the supratentorial brain.

Plan: **continue until completely resolved**



Imaging considerations

- CT: In patients with neurologic symptoms such as moderate or severe impairment of consciousness or neurologic deficits (not including cranial nerve abnormalities), performing CT before lumbar puncture is recommended.

  → consider contrast CT to rule out abscess

- MR: essential in detecting complication of meningitis such as venous thrombosis, small vessel infarct/ischemia, cerebritis, ventriculitis, subdural/epidural empyema, and vasculitis, and to discern the etiology and route of spread of infectious meningitis.
- Advanced MRI techniques such as magnetic transfer sequence, diffusion-weighted imaging (DWI), diffusion tensor imaging (DTI), MR spectroscopy, and perfusion imaging have significantly contributed to the evaluation of meningitis complications

Topics in Magnetic Resonance Imaging. 23(5):315–325, OCTOBER 2014

Despite fancy diagnostics, there are still challenges

Date	WBC	RBC	Glucose	Protein	M/E	Fungal culture	CSF CrAg	Serum CrAg
1/6	680	2750	108	145	Negative			
1/8	594	4	21	79	Negative	C gattii		
1/9	20	1333					1:10	
1/13								1:2056
1/29	36	2	89	104		Negative		
2/20	20	1340	109	58			1:40	

Remember all the various diagnostic tools available to us: serologies, culture, broad range PCR, pathology!

Meningoencephalitis Panel

1. Make sure you know which organism are on your institutional panel
2. Know what is NOT on the panel (GAS, Staph aureus, other gram negatives)
3. Know when the ME panel is most helpful
- Pretreated Bacterial Meningitis
- Entero/parechovirus
- When it is not helpful: HHV6
4. Know there are concerns regarding false negative and false positive results, particularly with HSV.
- Obtain alternative testing

Virus	Bacteria	Other
CMV	E. Coli	Cryptococcus
VZV	H. Influenzae	
Enterovirus	Listeria m.	
HSV 1/2	Group B Strep	
HHV 6	S. pneumoniae	
Parechovirus		

Bard JD and Alby K. Point-Counterpoint: Meningitis/Encephalitis Syndromic Testing in the Clinical Laboratory. J Clin Microbiol. 2018 Apr; 56(4): e00018-18.
Radmard S et al. Clinical Utilization of the FilmArray Meningitis/Encephalitis (ME) Multiplex Polymerase Chain Reaction (PCR) Assay. Front Neurol. 2019; 10: 281.
Leber et al. **Multicenter Evaluation of BioFire FilmArray Meningitis/Encephalitis Panel for Detection of Bacteria, Viruses, and Yeast in Cerebrospinal Fluid Specimens.** J Clin Microbiol. 2016 Sep;54(9):2251-61.

New e attii

- First identified in
- Travel history to:
- Disease in healt romised.
- 65 cases occurre ortable in Oregon since 2011.
- Association: dec Northwest (deBess 2014). S nette Valley.
- Concern for expa te rain forest zone


HEALTH

Killer fungus rode into Oregon on ships, a tsunami and a flood, researchers say

Updated Oct 08, 2019; Posted Oct 08, 2019

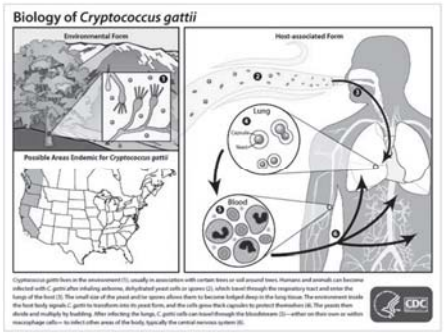
19 1.2k shares

By Fedor Zarkhin | The Oregonian/OregonLive

DeBess E et al. Isolation of Cryptococcus gattii from Oreg
BMC Microbiol. **2014** Dec 21;14:323.
Smith RM et al. Treatment and outcomes among patients with Cryptococcus gattii meningitis in the United States Pacific Northwest. PLoS One. **2014** Feb 19;9(2):e89075.
https://www.oregon.gov/oha/PH/DISEASESCONDITIONS/COMMUNICABLEDISEASE/DISEASESURVEILLANCEDATA/ANNUALREPORTS/Documents/2017/2017-Cryptococcus.pdf

Incidence of cryptococcosis by county of residence: Oregon, 2013–2017

https://www.oregon.gov/oha/PH/DISEASESCONDITIONS/COMMUNICABLEDISEASE/DISEASESURVEILLANCEDATA/ANNUALREPORTS/Documents/2017/2017-Cryptococcus.pdf



https://www.cdc.gov/fungal/diseases/cryptococcosis-gattii/index.html

- **Acquisition:** inhalation of spores from the environment. No zoonotic transmission.
- **Incubation:** 2 to 13 months, with a median of 6–7 months
- **Clinical presentation: cryptococcomas in the** lung and brain (often large, multifocal lesions)

Isn't this a weird presentation?

Why did this boy get so sick?

Sentinel Immunodeficiency

Newly recognized immune deficits in otherwise healthy persons

- **Auto-antibodies against GM-CSF detected in serum**
  - Also causes acquired Pulmonary alveolar proteinosis (PAP), leading to a build-up of surfactant in the alveoli and inhibiting gas exchange
- Ig Deficiencies **(CVID)**
- HIV or Idiopathic lymphopenia

Rosen LB et al. **Anti-GM-CSF autoantibodies in patients with cryptococcal meningitis.** J Immunol. 2013 Apr 15;190(8):3959-66.
Marr KA. Cryptococcus gattii infection in healthy hosts: a sentinel for subclinical immunodeficiency? Clin Infect Dis. 2012 Jan 1;54(1):153-4.

Pearls

Non contrast CT may miss early lesions.

→ **If infection concern, need contrast (or diffusion weighted) to exclude infection**

Meningo-encephalitis panel may not always be helpful (concerns about false positive/false negatives)

→**If index of suspicion is high, confirm with other tests**

Severe cryptococcal disease (namely C gattii) can occur in previously healthy hosts here in the Pacific Northwest

→ **It's here! Be on the look out**

If a weird infection presents and is severe, think about possible immunodeficiency.

→ **We now have a pediatric immunologist!**

Presenting Symptom: Swollen glands

5 year old with a lump on R neck in May accompanied by 1 day fever (102.5 F)

HPI:

- Previously healthy; **had a cold prior to this.**
- ER: Temp 37.5, red pharynx, diagnosed Strep without a rapid strep and gave amoxicillin.
- Saw PCP for follow-up: Didn't think this was strep. Large lymph node at R neck. Not fluctuant or tender. Ordered labs – including cat scratch and presumptively started him on azithromycin and clindamycin.
- Over 1 month: Node continued to enlarge. Referred to **local ENT** (adult). CT scan.
- Admitted for IV antibiotics and surgery with partial IandD. Discharged on Augmentin. No growth.
- Referred to ID for persistent draining wound.

Pertinent History

ROS: 1 day of fever; +night sweats; decreased activity/fatigue

- Pertinent negative: **no blue, pink or purple toned skin;** no weight loss, no belly pain, no other lymph nodes

PMH: Hydrocele and hernia repair x2; Lymphangioma removal right scalp

FMH: Mom- Celiac disease; MGGF-TB in the 1950s

Exposure History

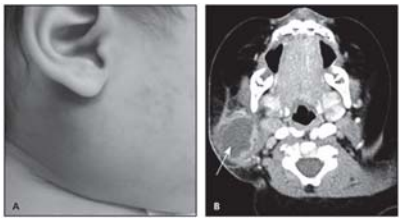
- Attends preschool and otherwise home
- TB/MRSA/HSV: No homeless shelter/ jail exposures. No one with cold sores.
- Food/Water: Well water for bathing and teeth brushing
- Recreation: No hot tub/warm springs; lots of gardening; no hanging plants; no composting on site; Organic soil delivered 2 years ago. Tonka trucks in dirt; has eaten dirt per mom.
- Animals: 2 dogs, 1 fish, 15 chickens, 5 ducks; Scratched by cat 6 months ago.

Common causes of lymphadenopathy

Bacterial Adenitis

Fever
Edema
Erythema
Tenderness
WBC

Remember the pharyngeal spaces



https://www.aafp.org/afp/2014/0301/p353.html

Mycobacteria

Scrofula: TB vs. NTM

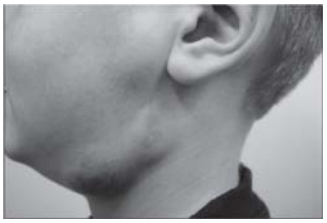
Sx: Develops over weeks to months; Tender and rubbery, Discolored skin over the node. Cervical>>Axillary>Groin

Dx: clinical; biopsy shows necrotizing granulomas; culture or PCR +

Tx: Excisional biopsy preferred; If involves facial nerve, may require abx (azithromycin, rifampin +/- ethambutol).

Cx: If Iand D is done→may lead to sinus tract and cutaneous drainage for up to 12 months

HELPFUL CLUES: AGE, LOCATION, APPEARANCE



https://cmr.asm.org/content/24/4/701

Cat Scratch: Bartonella henselae

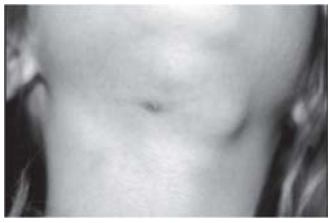
SX: A small papule may develop at the site of inoculation; can take 2+ weeks to develop adenopathy

Dx: Serology, Blood or PCR

Tx: Azithromycin, Bactrim

Cx: Retinitis, osteomyelitis, hepatosplenic lesions, endocarditis.

Consider Tularemia with an eschar



https://www.dermatologyadvisor.com/dermatology/cat-scratch-disease-bartonella-infection/article/691691/

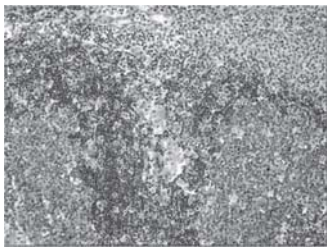
Toxoplasmosis

Sx: malaise, fever, sore throat, and myalgia.

Dx: Serologic testing; Tissue PCR

Tx: Self limited; pyrimethamine/sulfadiazine + leucovorin rescue

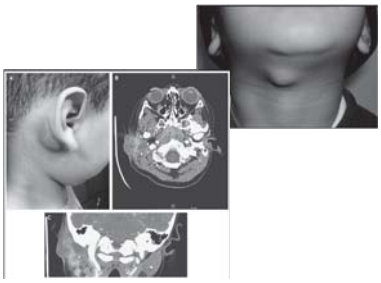
Cx: Retinitis, myocarditis and pneumonitis.



https://www.pathologyoutlines.com/topic/lymphnodetoxoplasma.html

Congenital Conditions Mimicking Adenopathy

- Thyroglossal duct cyst (see picture)
- Dermoid cysts or tumors
- Branchial cleft (see picture)
- Lympho-vascular malformations
- Hemangioma
- Ectopic thymus
- Epidermoid cyst
- Cystic Hygroma



http://fortworthent.net/ear-nose-throat/thyroglossal-duct-cyst-removal/ https://www.nejm.org/doi/full/10.1056/NEJMcm1503044

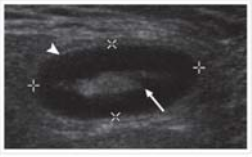
Imaging Considerations

Chest x-ray → prolonged fevers, constitutional symptoms; concern for TB, cancer, etc.

Ultrasonography → defining the presence and extent of an abscess; Liver/spleen/Masses

CT → congenital / structural concerns; pre-op

MR → not used often – helpful if want to avoid radiation but may need sedation



American Journal of Roentgenology. 2012;199:1105-1113. 10.2214/AJR.12.8629. Imaging of Cervical Lymphadenopathy in Children and Young Adults. https://www.ajronline.org/doi/10.2214/AJR.12.8629

BACK TO OUR CASE

Microbiology

- 6/1: WBC 18.8 HCT 32.6 Plt 390 77N ESR 83
- Bartonella IgG = <1:64 (neg); IgM= <1:16 (neg)
- TST: negative / Quantiferon Gold: negative
- Wound culture from OR on 6/13: negative for bacteria.
- No AFB or fungal cultures were done.
- Path report: **Necrotizing granulomatous lymphadenitis.** GMS and AFB stains for mycobacteria and fungus were negative.
- **ID Eval: Large infectious serological panel was negative (toxoplasmosis, fungal, tularemia).**

Differential

- Broad Range Bacterial PCR: Legionella longbeachae

Mycobacterium species

Unusual fungus

Nocardia

Other (who knows?)

Broad Range PCR: no nontuberculous mycobacteria detected; no TB detected

Final Pathologic Diagnosis:
Right neck lymph node, biopsy (SP-16-17290, 6/13/16):
- Necrotizing granulomatous lymphadenitis with neutrophils, see comment

Comment: We appreciate the opportunity to review this case and agree with the original diagnosis. special stains for mycobacteria and fungi (provided AFB and GMS) are negative for organisms. This histologic pattern is nonspecific and seen in cat scratch disease, mycobacterial and fungal infections. Culture and clinical correlation is required for a definitive diagnosis.

Course

- Underwent a more extensive excision and debridement with Peds ENT.
- Confirmed the Broad range PCR result with a second sample
- Completed 21 days of azithromycin with clinical resolution.

Em ...is

- At least 60 (
- Most disea
- Longbeach
- First isolat
- Highly rec
- Bacteria is
- **Diagnosis** (paired); PC

Hot tub display at North Carolina fair kills 2 after infecting 134 with Legionnaires' disease

Mike Wehner @MikeWehner

Share Tweet

Whitley H, Bentham R. *Legionella longbeachae* CID Volume 38, Issue 10, 15 May 2004, Pages

ew Zealand. EID Jul;23(7):1148-1154.

Legionellosis

- **Transmission:** No Person-to-person; Inhalation and ingestion are possible modes
- **Clinical:** early symptoms include fever, chills, headache, shortness of breath, sometimes dry cough, and muscle aches and pain. Pontiac fever (without pneumonia)

  Other: Osteomyelitis; Cutaneous (**non healing wound**); Adenopathy
- **Risk factors:** Exposure to compost or potting mix. Gardening behaviors, including having unwashed hands near the face after exposure to or tipping and troweling compost or potting mix.

McClelland M. **Pneumonia and Osteomyelitis Due to *Legionella longbeachae* in a Woman with Systemic Lupus Erythematosus**

Red Flags

- **Non healing wound**
- **Night sweats** or weight loss
- Lack of infectious symptoms in the ear, nose, and throat regions
- Unexplained fevers > 1 week
- Lymph nodes > 2 cm in size; does NOT Wax / Wane
- Supraclavicular or axillary lymph nodes
- Hard, rubbery consistency; fixed/matted
- Abnormal CXR
- Hepatosplenomegaly
- Abnormal labs (CRP, ESR, WBC, etc.)

Pearls

Avoid I and D unless bacterial abscess

- →Poor wound healing may signify continued infection
- → Excisional biopsy is best.

You may not be able to identify the underlying etiology in every patient.

- →Newer molecular diagnostic studies (Broad Range PCR) may be helpful, particularly with more unusual presentations.

Know the red flags

- → If it waxes/wanes – you are generally ok, but close follow-up can identify early lesions

Presenting Symptom: Difficulty Seeing

6 year old girl with new onset vision loss

HPI

- Used father's reading glasses when looking at books.
- Over the next few months noted she was holding objects close to face; unable to read large projection screen at church;
- Denies eye pain or headaches or preceding illnesses
- **Visited the optometrist where glasses were recommended.**
- Incidental screening exam abnormal
  - Strabismus
  - Fundus exam: disc edema

Referral to Casey Eye

Pertinent History

ROS negative: NO fever, headache, neurologic or constitutional symptoms; +vision loss

PMH: normal pregnancy, labor, delivery, infancy
- Hx of urticaria lasting 12 months between ages 3 and 4 years; symptomatic treatment; resolved spontaneously
- No medications
- Unimmunized

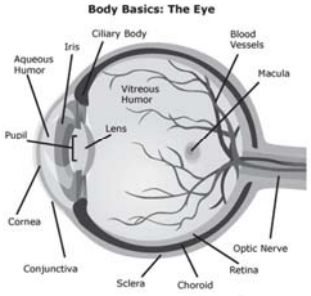
Social History and Exposures: youngest of 11 children; lives on the northern Oregon coast; No travel; 3 cats, dogs and chinchilla; no sick contacts

FMH: brother with myopia and astigmatism

A quick review of the eye exam

1. Visual acuity
2. Pupils (with afferent check – swinging light)
3. Extraocular motility and alignment (both and monocular)
4. Intraocular pressure
5. Visual fields
6. External exam
7. **Slit lamp:** Lids/lashes/lacrimal system:; Conjunctiva/sclera; Cornea; Anterior chamber; Iris; Lens, Anterior vitreous
8. **Funduscopic:** Optic nerve, macula, vessels, periphery

Eye Anatomy



https://kidshealth.org/en/parents/eyes.html

Eye Concerns

	Keratitis	Scleritis	Uveitis	Retinitis/optic neuropathies
Mechanism	inflammation and ulceration of the cornea	Inflammation of sclera	Inflammation of iris/ciliary body, choroid	Optic nerve lesions Macular lesions
Etiology	HSV, bacteria/fungi	RA, Crohns	Syphilis, TB, toxoplasmosis, histo; autoimmune	Infection, autoimmune, ischemic, neoplasms
Symptoms	Pain, decreased acuity, irritation, tearing, photophobia, mild conjunctivitis	Dull pain, intense redness, loss of vision	Pain, photophobia, blurred vision, redness, pupillary constriction	Pain is variable Vision loss

Initial Evaluation at Casey Eye Institute

Acuity
- Right 20/60
- **Left 20/200**

Slit Lamp- normal

Slit Lamp Exam

	Right	Left
Lids/Lashes	Normal	Normal
Conjunctiva/Sclera	White and quiet	White and quiet
Cornea	All layers clear	All layers clear
Anterior Chamber	Deep and quiet	Deep and quiet
Iris	Normal	Normal
Lens	Clear	Clear
Vitreous	Normal	Normal

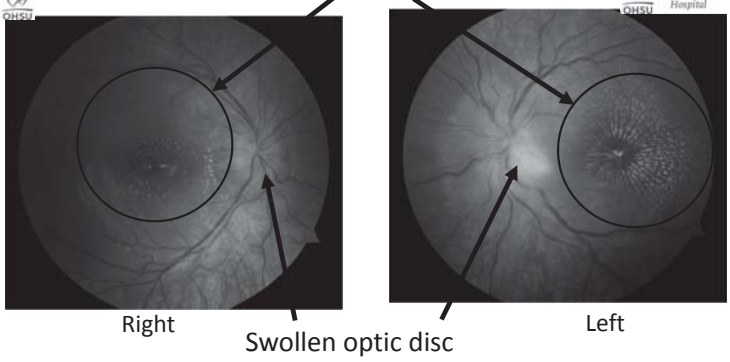
Refraction
- Sphere
  - Right +1.25
  - Left +0.75

Pressure: normal

Motor
- **Intermittent exotropia**
- **Deprivation amblyopia**

Fundus Exam

	Right	Left
Disc	Disc fullness with exudative material temporal to disc	Disc edema especially infero-nasally
Macula	Faint macular scar seen nasally	Prominent macular star
Vessels	Normal	Normal
Periphery	Normal	Normal



Macular Star and Strabismus

- **Macular star** formation is caused by the deposition of lipid exudates along the outer plexiform layer of the macula.
  - Vision loss due to maculopathy NOT optic nerve issue
- **Strabismus:**
  - Failure of eyes to look in the same direction at the same time
  - Weakness of muscles of one eye: (superior oblique, interior oblique, lateral)
  - Childhood: associated with amblyopia (decreased vision in one eye)
  - Types
    1. Esotropia: convergent - cross eye of one eye
    2. **Exotropia**: divergent - one eye turns outward

Differential Diagnosis: neuroretinitis

Inflammatory: MS, sarcoid, Behcet, Sjogren, Lupus, Guillain-Barre, Wegner's, IBD

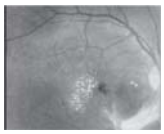
- Post infectious: measles, mumps, varicella, influenza, EBV

Infections:

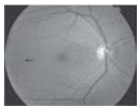
- Complication of meningitis or encephalitis either as a direct effect of the infectious organism or from a secondary vasculitis

- **Acute viral infections (CMV), Toxoplasmosis, Syphilis**, **Tuberculosis, Cat Scratch Disease**, **West Nile, Cryptococcus, Ebola, Zika, Lyme, RMSF**

Infectious Neuroretinitis

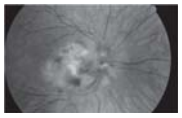


Toxoplasma retinitis

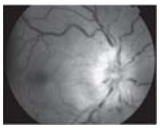


Syphilis retinitis

CMV retinitis



Lyme retinitis



Balansudaram et al. Outbreak of Acquired Ocular Toxoplasmosis Involving 248 Patients. Arch Ophthalmol. 2010 Jan;128(1):28-32.

Arq. Bras. Oftalmol. vol.77 no.5 São Paulo Sept./Oct. 2014
Mora D. Int J Med Sci 2009; 6(3):124-125.

Lab Evaluation

CBC and CMP were normal

Sarcoid: ACE normal

Syphilis: NR

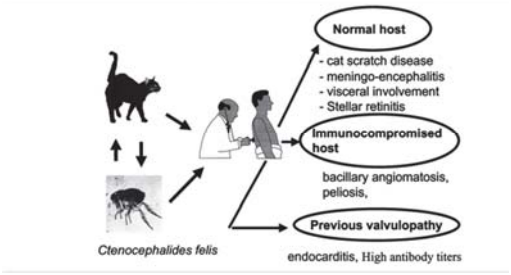
Tuberculosis: Quantiferon - NR

B. henselae: IgG 1:1024, IgM 1:16

Emerging Infection: Bartonellosis

- Bartonella henselae: Facultative, intracellular gram negative rod; fastidious
- NOT Bartonella Quintana (trench fever) or Bartonella bacilliformis (Carrion's disease)
- Incidence highest among in the southern United States (6.4 cases/100,000 population) and among children 5–9 years of age (9.4 cases/100,000 population).
- 12,000 outpatients are given a CSD diagnosis and 500 inpatients are hospitalized for CSD.
- Normal flora in kittens; maintained through contact/fleas; transmission from cat bite, scratch, lick

Nelson Cat-Scratch Disease in the United States, 2005–2013. EID Volume 22, Number 10—October 2016

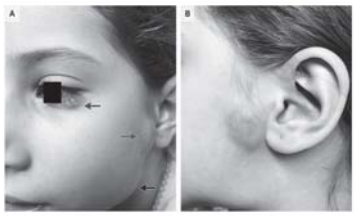


https://cvi.asm.org/content/9/1/8/figures-only

Rash
Hepatosplenic dissemination;
Osteomyelitis
Encephalopathy
Endocarditis
Eye Disease

Cat Scratch Ocular Disease

- Ocular involvement of cat-scratch disease occurs in 5–10% of cases, and is the most common non-lymphatic organ involvement.
- Parinaud's oculoglandular syndrome: occurs in 5% of cases.
- Neuroretinitis is seen in 1-2%



Ghazi A case of cat-scratch disease with unusual ophthalmic manifestations. Middle East Afr J Ophthalmol. 2012 Apr-Jun;19(2):243-6. Arango-Ferreira. Parinaud's Oculoglandular Syndrome in Cat Scratch Disease N Engl J Med 2018; 37

Treatment

Unclear benefit in healthy hosts, but lesions may resolve faster

Many agents potentially active: Macrolides, tetracyclines, aminoglycosides, TMP-SMX

Retinitis – visual prognosis is usually excellent

- Doxycycline plus Rifampin or Fluoroquinolone based on case series
  - 2-4-6 weeks
  - +/- steroids

Eye Exams

	Initial	+10	+30	**+42 (stop therapy)**	**+360**
Acuity (R)	20/60	20/40	R: 20/25	**20/30**	**20/20**
Acuity (L)	20/200	20/70	L: 20/70	**20/80**	**20/50**
Macula (R)	faint macular scar seen nasally	trace macular star	trace macular star	**trace macular star**	**Normal**
Macula (L)	prominent macular star	Trace exudate, macular star		**mild exudate/macular star**	**Small hypopigmented scarring on inferior fovea**
Disc (R)	Disc fullness with exudative material temporal to disc	Mild edema	mild disc edema	**mild disc edema**	**Normal**
Disc (L)	disc edema especially inferonasally	Disc edema especially inferolaterally		**Disc edema especially inferonasally**	**Inferior temporal gliosis**

Pearls

- Acute vision changes should prompt referral to an eye specialist

  → Dilated exam and slit lamp key

- Common infections can have unusual presentations

  → the most common cause of neuroretinitis is cat scratch disease.

- While CSD is usually self-limited, use of doxycycline or fluroquinolones may be needed for disseminated disease

  → could consider other regimens: azithromycin or trim-sulfamethoxazole

Summary

1. Described a differential according to presenting signs and symptoms for headache, lymphadenopathy, and vision loss
2. There are many diagnostic tools available in infectious diseases, including newer molecular tests
3. Cryptococcal disease, Legionellosis, and Cat Scratch disease are rare but emerging infections affecting patients in the Pacific Northwest

Questions?

Thank You

Can a Screening EKG Save A Pediatric Athlete's Life

Brendan Kelly, MD
Pediatric Cardiology
Oregon Health & Science University
NW Permanente Physician

Conflict of Interest Statement

- I have no financial disclosure
- This presentation does not contain trade names
- This presentation does not contain advertising.

BUT I AM A CARDIOLOGIST!

Hayward Demison

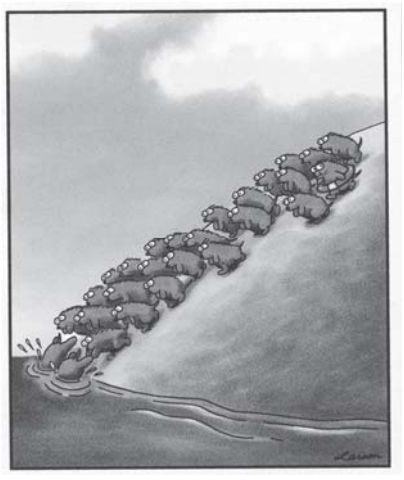
- Portland Central Catholic High School Star athlete
- Cardiac arrest during a football game in 2010 after scoring a touchdown
- Successful resuscitation by members of the audience.
- Echo showed: Anomalous left coronary artery
- Probable cause of event: VF due to acute ischemia.
- Recovered after surgery for coronary artery re-implantation

Hank Gathers '90

"Pistol" Pete Maravich '88

Marc Vivien Foe '03

Reggie Lewis '93

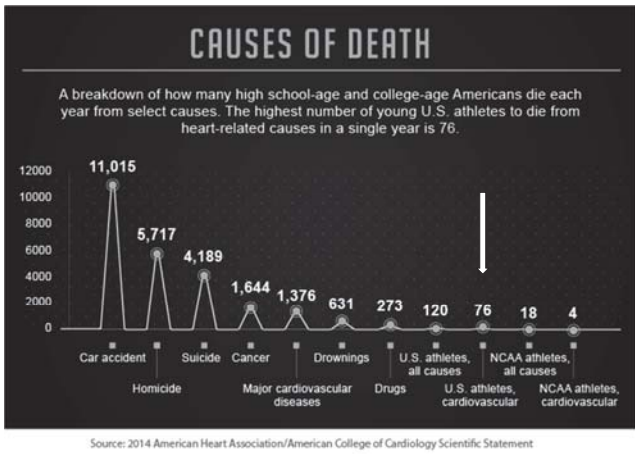


Sudden Death in Children

- 1.3 per 100,000 (1-22yrs)
  - Minnesota, Driscoll et al 1985
- 3.3 per 100,000 (1-20yrs)
  - Northern England, Wren et al 2000
- 2.7 per 100,000 (1-18yrs)
  - Taiwan, Wu et al 2009

How about Portland Oregon?

- Chugh et al, Oregon Sudden Unexpected Death Study. Heart Rhythm 2009.
- 7.5 per 100,000 (0-17yrs)
- 1.9 per 100,000 (1-17yrs)
- 3.0 per 100,000 (1-4yrs)
- 2.4 per 100,000 (5-9yrs)
- 1.7 per 100,000 (10-14yrs)



Harmon et al 2015

- 514 deaths in NCAA athletes 2003-2013
- Accidents responsible for 6.1/100,000 pty
- Sudden cardiac death in 79 athletes
  - 1.9 deaths/100,000 pty
  - Male vs female NCAA athletes 2.6 vs 0.8/100,000 pty
  - Black vs white NCAA athletes 4.7 vs 1.5 /100,000 pty
  - Male NCAA div I basketball player 19.2/100,000 pty
- 25% were autopsy negative SCD
- Different risk for different populations

Deaths During Sports are RARE

- But…..
  - Highly visible deaths, prime of life
  - Kids being active like doctors prescribe
- "No child should die that way."
- "Can you tell me my child won't die?"
- "I have good insurance, I want all the tests."
- Community screening programs
  - EKG only
  - EKG and echo
- What about the non-athletes?

What are the causes of the problem?

Maron et al 2007

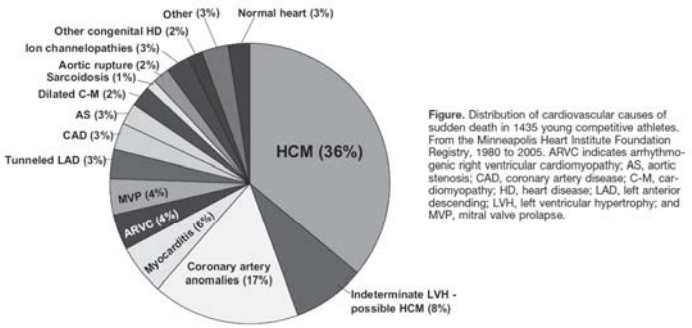


Figure. Distribution of cardiovascular causes of sudden death in 1435 young competitive athletes. From the Minneapolis Heart Institute Foundation Registry, 1980 to 2005. ARVC indicates arrhythmogenic right ventricular cardiomyopathy; AS, aortic stenosis; CAD, coronary artery disease; C-M, cardiomyopathy; HD, heart disease; LAD, left anterior descending; LVH, left ventricular hypertrophy; and MVP, mitral valve prolapse.

Disease	ECG abnormal?	Echo/MRI abnormal?	Inherited?
HCM	Y*	Y	Y
Abnormal Coronary	N	Y*	N
Long QT	Y	N	Y
ARVC	Y*	?	Y
CPVT	?	N	Y
Brugada syndrome	Y	N	Y
WPW	Y	N	N
CHD	Y*	Y	N*
Myocarditis	NA	NA	N
Commotio cordis	NA	NA	N

Screening In The US:

Pre-Participation Evaluation (PPE)
for the 12-25 year old athletes

Prevention of SCD

- Primary Prevention
  - History
  - Family History
  - Physical examination
  - ECG
  - Echocardiography
  - Stress test
- Secondary Prevention
  - CPR + AED Programs

Currently a part of pre-participation screening

Pre-participation Evaluation (PPE)

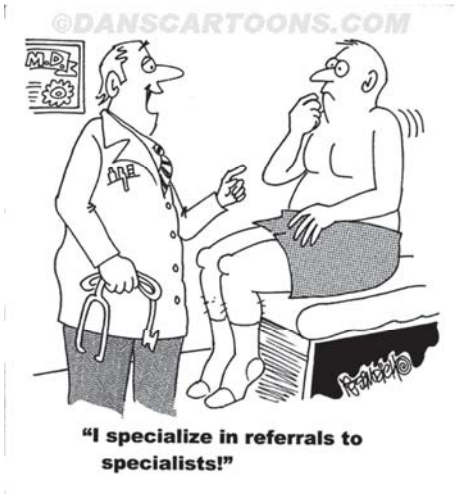
- Current AHA recommendation
- 14 point History & Physical components
- Most states including Oregon have adopted this document

AHA Statement Recommendations. Maron 1996, Revised 2007

TABLE 1 The 14-Element AHA Recommendations for Preparticipation Cardiovascular Screening of Competitive Athletes

Medical history*
Personal history
1. Chest pain/discomfort/tightness/pressure related to exertion
2. Unexplained syncope/near-syncope†
3. Excessive and unexplained dyspnea/fatigue or palpitations, associated with exercise
4. Prior recognition of a heart murmur
5. Elevated systemic blood pressure
6. Prior restriction from participation in sports
7. Prior testing for the heart, ordered by a physician
Family history
8. Premature death (sudden and unexpected, or otherwise) before 50 y of age attributable to heart disease in ≥1 relative
9. Disability from heart disease in close relative <50 y of age
10. Hypertrophic or dilated cardiomyopathy, long-QT syndrome, or other ion channelopathies, Marfan syndrome, or clinically significant arrhythmias; specific knowledge of genetic cardiac conditions in family members
Physical examination
11. Heart murmur‡
12. Femoral pulses to exclude aortic coarctation
13. Physical stigmata of Marfan syndrome
14. Brachial artery blood pressure (sitting position)§

AHA indicates American Heart Association. *Parental verification is recommended for high school and middle school athletes. †Judged not to be of neurocardiogenic (vasovagal) origin; of particular concern when occurring during or after physical exertion. ‡Refers to heart murmurs judged likely to be organic and unlikely to be innocent; auscultation should be performed with the patient in both the supine and standing positions (or with Valsalva maneuver), specifically to identify murmurs of dynamic left ventricular outflow tract obstruction. §Preferably taken in both arms. Modified with permission from Maron et al. (3).



Before You Refer

- Think about asthma
- Rule out anemia
- Vasovagal episodes are common
- We all have symptoms with exertion
- In my practice:
  - Close family member means 1st degree relative
- Innocent murmurs are common

What about other developed countries?

Italian Experience

- Screening program for athletes introduced 1982.
- 12-35 year old athletes screened per Italian law
  - History, physical exam, 12 lead EKG
- Corrado et al in 2006
  - 1979-2004 55 SCD in 50 males and 5 females
  - SCD age mean 23.3yrs, median 23yrs
  - 90% white population
  - Decrease in SCD per 100,000 person years with screening
    - 4.19 (1.94-7.59) to 0.87 (0.46-1.28)
  - Greatest decline in death from cardiomyopathies (especially arrhythmogenic right ventricular cardiomyopathy)
    - 36% to 17%
- 7-9% false positive rate of screening

Corrado et al JAMA 2006. Veneto region of Italy

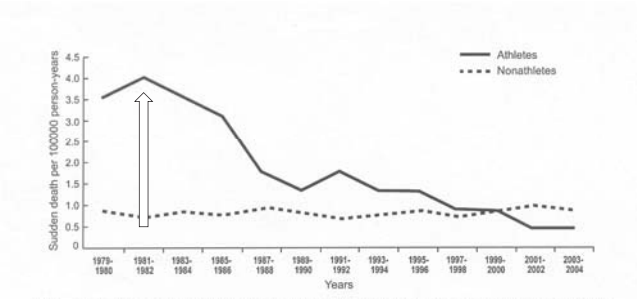


Fig. 2. Annual incidence rates of SCD per 100,000 persons, among screened competitive athletes and unscreened non-athletes 12 to 35 years of age in the Veneto Region of Italy, from 1979 to 2004. During the study period (the nationwide preparticipation screening program was launched in 1982), the annual incidence of SCD declined by 89% in screened athletes (P for trend <0.001). In contrast, the incidence rate of SCD did not demonstrate consistent changes over time in unscreened nonathletes. (*Modified from* Corrado D, Basso C, Pavei A, et al. Trends in sudden cardiovascular death in young competitive athletes after implementation of a preparticipation screening program. JAMA 2006;296:1593–601; with permission.)

But….

- Risk of SCD goes up with age
  - 0.13/100,000 in 12-19yo
  - 1.45/100,000 in 20-24yo
- Risk of SCD is higher in males vs females
  - 0.75/100,000 vs 0.13/100,000
- SCD risk in US 12-25yo <1/100,000
- SCD risk in Italy 12-35yo 3/100,000
  - 82% were males
- In US 1/3 SCD due to hypertrophic cardiomyopathy
- In Italy 1/4 SCD due to ARVC

Italian approach

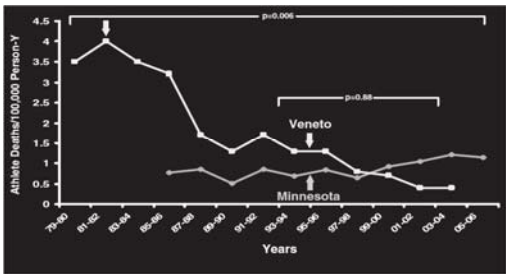
- Specialized sports medicine physicians
- Has been adopted with modifications by:
  - ESC, IOC, FIFA, many US professional sports teams
- Universities
  - Harvard, UW, Stanford, UVA, U Wisconsin, Georgetown
- US military for aviators

Comparison of U.S. and Italian Experiences With Sudden Cardiac Deaths in Young Competitive Athletes and Implications for Preparticipation Screening Strategies

Barry J. Maron, MD[a,*], Tammy S. Haas, RN[a], Joseph J. Doerer, BS[a], Paul D. Thompson, MD[b], and James S. Hodges, PhD[c]

(Am J Cardiol 2009;104:276–280)

- Used insurance claims to assign cause of death
- 2.28M person-y versus 2.93 person-y



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Pre-Participation Athletic Screening

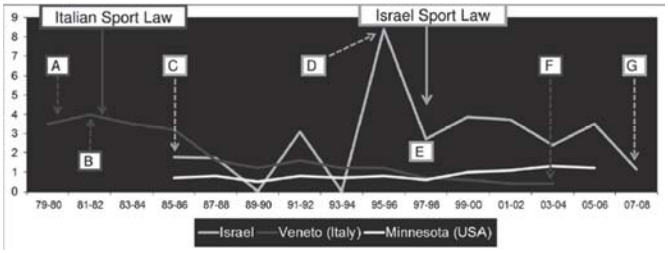
Mandatory Electrocardiographic Screening of Athletes to Reduce Their Risk for Sudden Death

Proven Fact or Wishful Thinking?

Arie Steinvil, MD,* Tamar Chundadze, MD,* David Zeltser, MD,* Ori Rogowski, MD,* Amir Halkin, MD,† Yair Galily, PhD,‡ Haim Perluk, MD,§ Sami Viskin, MD†
Tel-Aviv, Israel

- Review of 24 newspaper reported sudden death events during sports in competitive athletes from 1985-2009 in Israel
- 1997 mandated screening instituted
  - H&P, resting ECG, exercise test screening by certified physicians
  - 12-44 years old
- No decrease in event rate with ECG
  - 2.54 events per 100,000 athlete-years prior to 1997
  - 2.66 events per 100,000 athlete-years after 1997

Steinvil et al 2011



Not All ECGs Are Typical!

- HCM: 10% normal; sub-clinical/pre-clinical
- WPW: can be intermittent, or subtle
- LQT: can be tough & subtle.
  - Can even be normal.
- Brugada: often normal.
  - May need provocative testing (fever, IV Procainamide).
- ARVC: subtle repolarization abnormalities
- CPVT: usually normal
  - PVCs or VT with exercise; usually suspected when story suggests LQT but ECG is "normal".

Accuracy of Interpretation of Preparticipation Screening Electrocardiograms

Allison C. Hill, MD, Christina Y. Miyake, MD, MS, Stafford Grady, MD, and Anne M. Dubin, MD

(J Pediatr 2011;159:783-8)

- 53/212 pediatric cardiologists who returned a survey
- 8 normal EKGs
- 10 abnormals (LQT, WPW, HCM, PHTN, myocarditis)

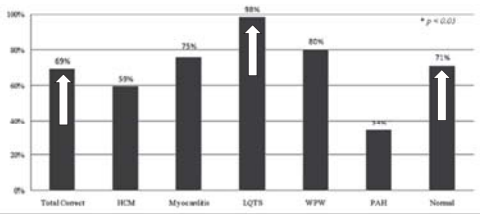


Figure 1. Percentage of correct ECG interpretations: respondents' scores of accurate findings in ECG interpretation broken down by underlying disease.

Let's Talk About the False Positive ECGs

Author	Population	Positive ECG
Fuller 1997	5615 HS athletes	2.6%
Pelliccia 2006	32652 athletes Italy	9%
Pelliccia 2007	4450 athletes Italy	12%
Magalski 2008	964 college athletes	10%
Wilson 2008	2720 HS athletes UK	4%
Bessem 2009	428 athletes NL	6%
Baggish 2010	510 college athletes	16%
Weiner 2011	510 college athletes	10%
Vetter 2011	400 children 5-19	8%
Chandra 2014	7764 non athletes UK	22%
Chandra 2014	4081 athletes UK	33%

So Change EKG Reading Criteria for Athletes?

European Society of Cardiology 2010
Seattle Criteria 2013
Refined Criteria 2014
International 2017

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Pediatric Cardiology

Prevalence of Electrocardiographic Anomalies in Young Individuals

Relevance to a Nationwide Cardiac Screening Program

Navin Chandra, BSc (Hons), MBBS,*† Rachel Bastiaenen, MA, MBBS,* Michael Papadakis, MBBS,*† Vasileios F. Panoulas, MD, PhD,† Saqib Ghani, MBBS,* Jennifer Duschl, MBBS,† David Foldes, MBBS,† Hariharan Raju, MBBS,* Rebecca Osborne, MSc,*† Sanjay Sharma, BSc (Hons), MBChB*†

London, United Kingdom

Objectives This study sought to investigate the prevalence of potentially abnormal electrocardiographic (ECG) patterns in young individuals to assess the implications for a nationwide screening program for conditions causing sudden cardiac death (SCD).

Background The Italian experience suggests that pre-participation screening with ECG reduces the incidence of SCD in athletes. However, the majority of SCDs occur in nonathletes. In the United Kingdom, screening for cardiac disorders is confined to symptomatic individuals or those with a family history of inherited cardiac conditions or premature cardiac death.

Methods Between 2008 and 2012, 7,764 nonathletes ages 14 to 35 years underwent ECG screening. Electrocardiograms were analyzed for group 1 (training-related) and group 2 (potentially pathological) patterns presented in the 2010 European Society of Cardiology position paper, which advocates further evaluation for individuals with group 2 ECG patterns. Results were compared with 4,081 athletes.

Results Group 1 patterns occurred in 49.1% of nonathletes and 87.4% of athletes ($p < 0.001$). Group 2 patterns occurred in 21.8% of nonathletes and 33% of athletes ($p < 0.001$). In nonathletes, QTc interval abnormalities comprised the majority (52%) of group 2 changes, whereas T-wave inversions constituted 11%. Male sex and African/Afro-Caribbean ethnicity demonstrated the strongest association with group 2 ECG patterns.

Conclusions The study demonstrates that 1 in 5 young people have group 2 ECG patterns. The low incidence of SCD in young people suggests that in most instances such patterns are non-specific. These findings have significant implications on the feasibility and cost-effectiveness of nationwide screening programs for cardiovascular disease in young nonathletes and athletes alike, on the basis of current guidelines. (J Am Coll Cardiol 2014;63:2028–34)

Chandra et al. JACC 2014

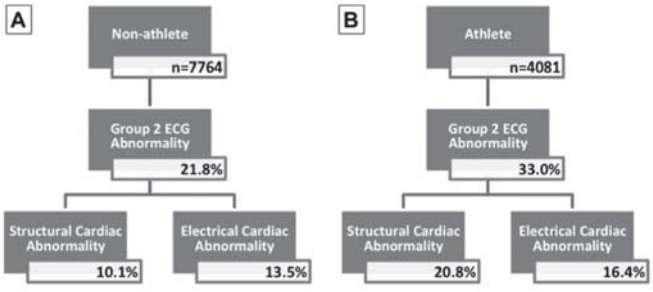
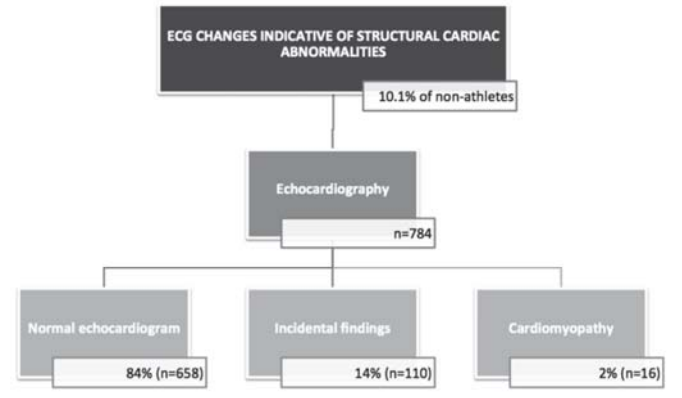


Figure 3 Flow Chart Represents the Proportion of Nonathletes and Athletes With Group 2 ECG Patterns That Would Require Further Evaluation

ECG changes suggestive of cardiomyopathy or structural cardiac abnormalities included T-wave inversion, ST-segment depression, pathological Q-waves, left atrial and right atrial enlargement, left-axis deviation and right-axis deviation, right ventricular hypertrophy, left bundle-branch block, and right bundle-branch block. ECG changes suggestive of ion channelopathy or electrical cardiac abnormalities included ventricular pre-excitation, long- and short-QTc interval, and Brugada-like early repolarization.

Chandra et al. JACC 2014



Sharma et al: International recommendations for electrocardiographic interpretation in athletes. European Heart Journal 2018.
Asymptomatic Athletes 12-35 year old

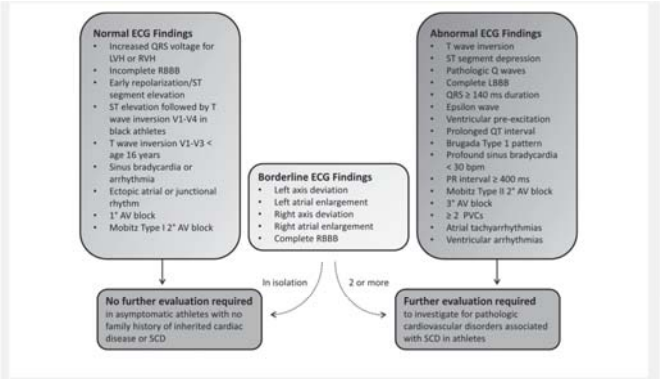


Figure 1 International consensus standards for electrocardiographic interpretation in athletes. AV, atrioventriular block; LBBB, left bundle branch block; LVH, left ventricular hypertrophy; RBBB, right bundle branch block; RVH, right ventricular hypertrophy; PVC, premature ventricular contraction; SCD, sudden cardiac death.

Abnormal ECG findings in athletes

These ECG findings are unrelated to regular training or expected physiologic adaptation to exercise, may suggest the presence of pathologic cardiovascular disease, and require further diagnostic investigation.

ECG abnormality	Definition
T wave inversion	≥ 1 mm in depth in two or more contiguous leads; excludes leads aVR, III, and V1
• Anterior	• V2–V4 – excludes: black athletes with J-point elevation and convex ST segment elevation followed by TWI in V2–V4; athletes < age 16 with TWI in V1-V3; and biphasic T waves in only V3
• Lateral	• I and AVL, V5 and/or V6 (only one lead of TWI required in V5 or V6)
• Inferolateral	• II and aVF, V5-V6, I and AVL
• Inferior	• II and aVF
ST segment depression	≥ 0.5 mm in depth in two or more contiguous leads
Pathologic Q waves	Q/R ratio ≥ 0.25 or ≥ 40 ms in duration in two or more leads (excluding III and aVR)
Complete left bundle branch block	QRS ≥ 120 ms, predominantly negative QRS complex in lead V1 (QS or rS), and upright notched or slurred R wave in leads I and V6
Profound nonspecific intra-ventricular conduction delay	Any QRS duration ≥ 140 ms
Epsilon wave	Distinct low amplitude signal (small positive deflection or notch) between the end of the QRS complex and onset of the T wave in leads V1-V3
Ventricular pre-excitation	PR interval < 120 ms with a delta wave (slurred upstroke in the QRS complex) and wide QRS (≥ 120 ms)
Prolonged QT interval*	QTc ≥ 470 ms (male) QTc ≥ 480 ms (female) QTc ≥ 500 ms (marked QT prolongation)
Brugada Type 1 pattern	Coved pattern: initial ST elevation ≥ 2 mm (high take-off) with downsloping ST segment elevation followed by a negative symmetric T wave in ≥ 1 leads in V1–V3
Profound sinus bradycardia	< 30 bpm or sinus pauses ≥ 3 sec
Profound 1° atrioventricular block	≥ 400 ms
Mobitz Type II 2° atrioventricular block	Intermittently non-conducted P waves with a fixed PR interval
3° atrioventricular block	Complete heart block
Atrial tachyarrhythmias	Supraventricular tachycardia, atrial fibrillation, atrial flutter
PVC	≥ 2 PVCs per 10 s tracing
Ventricular arrhythmias	Couplets, triplets, and non-sustained ventricular tachycardia

Sharma et al: International recommendations for electrocardiographic interpretation in athletes. European Heart Journal 2018.

Malhotra et al BMJ 2019

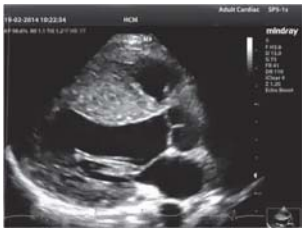
Accuracy of the 2017 international recommendations for clinicians who interpret adolescent athletes' ECGs: a cohort study of 11 168 British white and black soccer players

- 11,168 soccer players between 1996-2016
- Health questionnaire, EKG, echocardiogram.
- 95% male, 91% white
- Compared ESC 2010, Seattle 2013, Refined 2014, and International 2017
- All four criteria identified 36 of 42 athletes with serious cardiac conditions (86%)

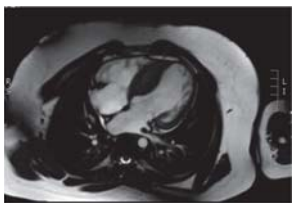
Malhotra et al BMJ 2019

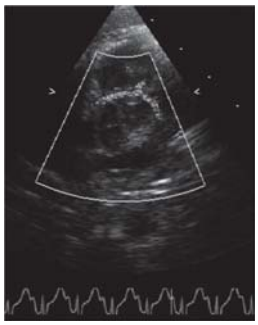
- International criteria was the best
  - Specificity of 98%
  - Sensitivity 86%
  - PPV 17%
- History
  - Specificity 96%, Sensitivity 7%, PPV 2.8%
- Physical
  - Specificity 98%, Sensitivity 5%, PPV 1.9%

But there is always echo and MRI right?



HCM

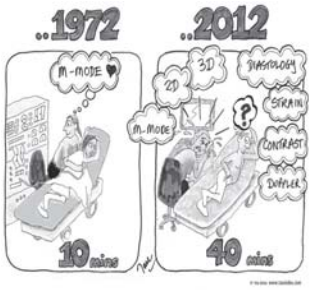




Anomalous Left coronary

Screening echo

- Can detect obvious cardiomyopathy
- Imaging coronary arteries takes skills
  - Often need cardiac CT in suspicious cases



Talk to me about the $$MONEY$$

A life – how much is it worth?

- Priceless….in theory.
- Screening societal threshold
  - $50,000-$100,000/life year
- Is this money better spent in other areas of health care?

ARTICLE | Annals of Internal Medicine

Cost-Effectiveness of Preparticipation Screening for Prevention of Sudden Cardiac Death in Young Athletes

Matthew T. Wheeler, MD, PhD; Paul A. Heidenreich, MD, MS; Victor F. Froelicher, MD; Mark A. Hlatky, MD; and Euan A. Ashley, MB ChB, DPhil

Ann Intern Med. 2010;152(5):276-286.

- Addition of EKG to H&P
  - $42,900/life year
    - Assumes <9% abnormal EKGs
    - Assumes EKG cost of $5
    - Assumes secondary testing of $330
    - Assumes risk reduction of 84%
      - 50% reduction leads to $63,600/life year

Leslie et al. Circulation 2012

- Simulation models incorporating prevalence, sensitivity, specificity
  - HCM, LQTS, WPW
  - 2 EKG screening populations ADHD and athletics
- Treatment algorithms generated and analyzed
- Screening at age 8: $91,000/life year
- Screening at age 14: $204,000/life year

Maron 2007	$3.4 million per life saved
Fuller 2000	$44,000 per life year saved
Wheeler 2010	$46,000 per life year saved
Malhotra 2011	$69,000 per diagnosis
Leslie 2012	$91,000 per life year saved
Halkin 2012	$10-14 million per life saved
Dhutia 2016	$36,000 per diagnosis

USA

- ~10 million competitive athletes.
- 60 million people aged 12-25.
  - Can screening ethically be restricted to "athletes"?
- Current cost estimate:
  - $2.5-3.5 billion per year.
- Not enough cardiology providers to read all the EKGs
- Insurance company payment is an issue

Cost

- *"A billion here and a billion there and pretty soon you are talking real money!"*
  Senator Everett Dirksen

Evidence pro ECG screening

- ECG abnormal in almost all HCM patients with hypertrophy.
- Can detect LQT, WPW, Brugada, CPVT etc
- A high cut off QTc value > 460 in boys and 480 in girls can pick up clear LQT.
- ECG is "cheap" and easy to do.
- Current PPE is less cost effective screening than ECG.
- False positives can be reduced with a clear & modified EKG reading protocol

Evidence con ECG screening

- Italian study has not been replicated even in Italy.
- USA study (Maron) and Israel Study did not support ECG screening.
- False positives: Almost 30% in athletes?
  - Mild LVH, mild RVH, borderline QTc.
- False negatives: will occur regardless of technique
  - Coronary artery abnormalities are hard to detect.

ACC AHA guidelines

- AHA/ACC panel does not support mandatory national ECG screening.
- They cite:
  - Low prevalence
  - Low risk in those with conditions associated with SD
  - Large population size
  - Imperfections of ECG
- Do support local efforts in small cohorts with close physician involvement.

So, where are we?

- Debate at every meeting. Both sides have good points but seem to selectively choose data.
- Data being collected. Child Safety Research Consortium; mainly pediatric EP and cardiology (PACES), lay advocates and FDA. Working to set common standards on data collection and reporting.

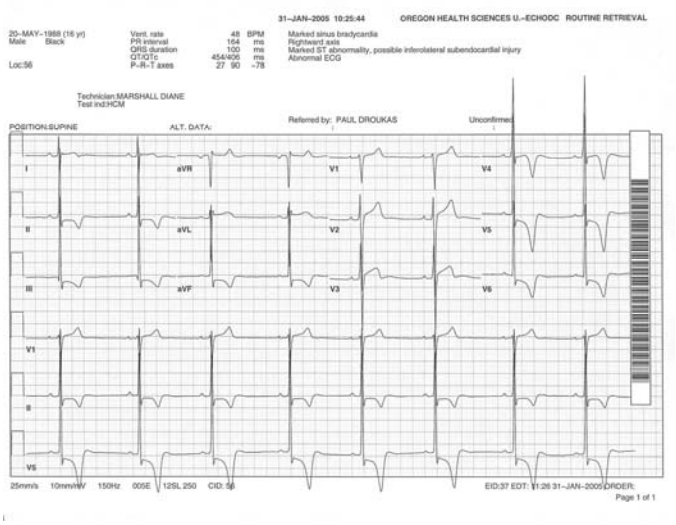
What can we do?

- Screen patients with symptoms
  - Syncope or near-syncope: ECG.
  - Syncope or chest pain during exercise: ECG + cardiology referral (will likely need echo)
- Screen patients with positive family history.
  - Cascade screening.
- Widespread availability of AED.
  - ~$53,000/QALY
- Widespread CPR & AED training.

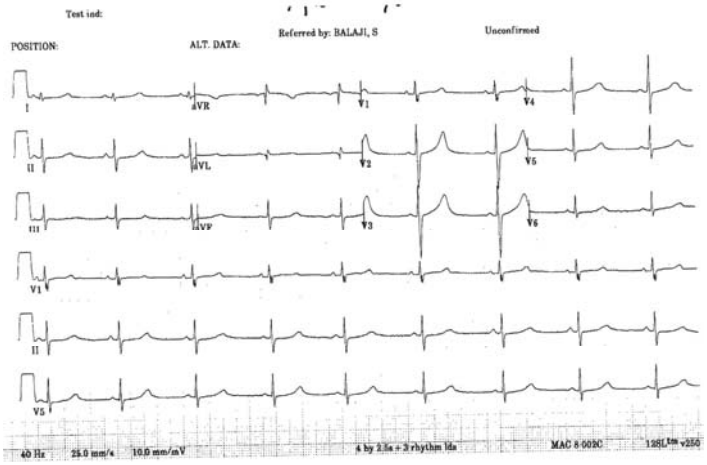
The logic of Cascade screening

Disease	ECG abnormal?	Echo/MRI abnormal?	Inherited?
HCM	Y*	Y	Y
WPW	Y	N	N
Long QT	Y	N	Y
ARVC	Y*	?	Y
CPVT	?	N	Y
Brugada syndrome	Y	N	Y
Abnormal Coronary	N	Y*	N
CHD	Y*	Y	N*
Myocarditis	NA	NA	N
Commotio cordis	NA	NA	N

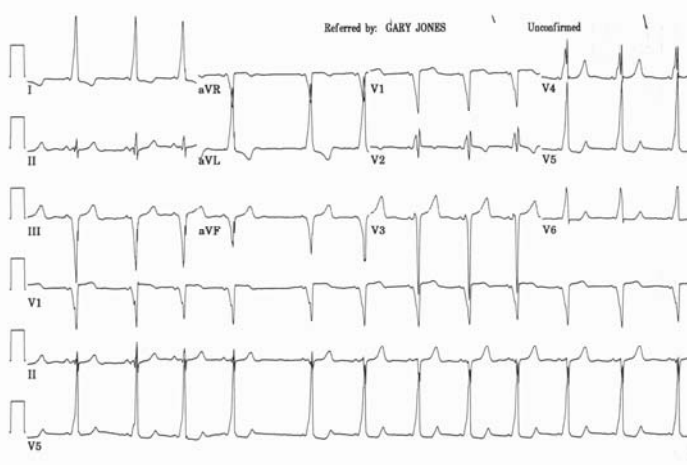
Abnormal EKG Examples



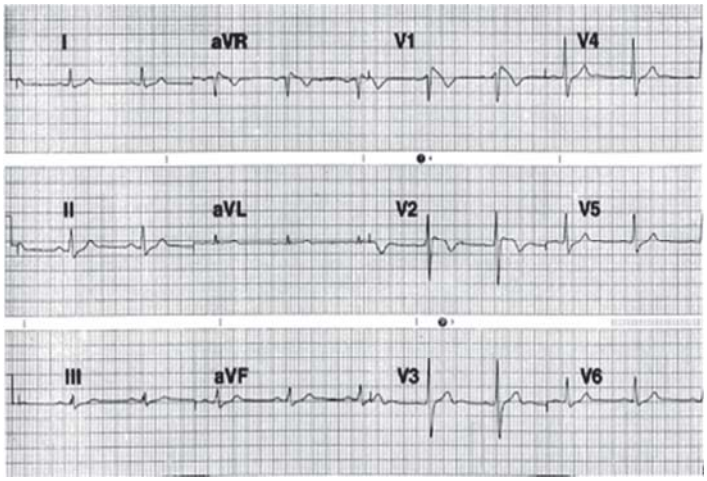
Long QT: QT = 600 ms; with sinus bradycardia



WPW



Brugada ECG



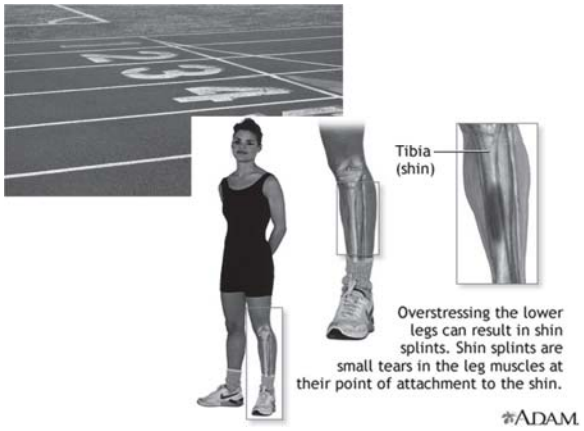
Final Thoughts and Questions?

Prevent an Eating Disorder- Save an Athlete

Dr. Melissa Novak D.O.
Primary Care Sports Medicine
Oregon Health Sciences University

- NO disclosures

20 Year Old Collegiate Track Athlete



2014 Female Athlete Triad Coalition Consensus Statement

nsensus statement

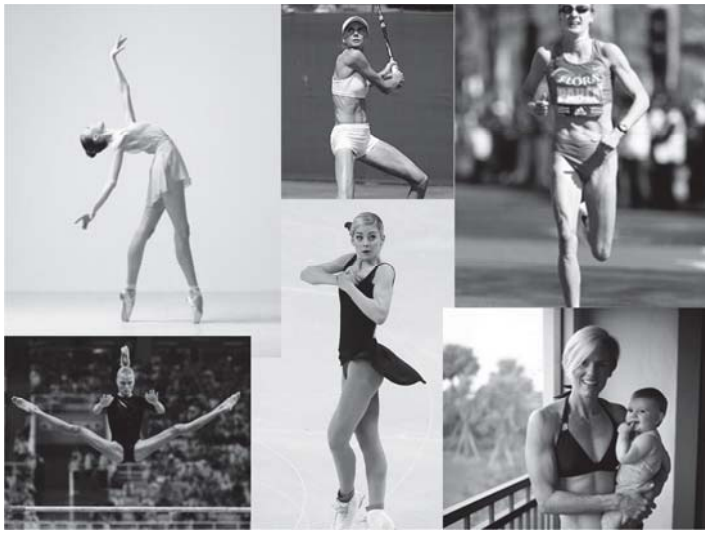
Risk Factors	Magnitude of Risk		
	Low Risk = 0 points each	derate Risk = 1 point each	High Risk = 2 points each
Low EA with or without DE/ED	☐ No dietary restriction	Some dietary restriction‡; urrent/past history of DE;	☐ Meets DSM-V criteria for ED*
Low BMI	BMI ≥ 18.5 **or** ≥ 90% EW** or weight stable	☐ BMI 17.5 < 18.5 **or** < 90% EW **or** 5 to < 10% weight loss/month	☐ BMI ≤17.5 **or** < 85% EW **or** ≥ 10% weight loss/month
Delayed Menarche	☐ Menarche < 15 years	Menarche 15 to < 16 years	☐ Menarche ≥16 years
Oligomenorrhea and/or Amenorrhea	☐ > 9 menses in 12 months*	} 6-9 menses in 12 months*	☐ < 6 menses in 12 months*
Low BMD	☐ Z-score ≥ -1.0	☐ Z-score -1.0*** < - 2.0	Z-score ≤ -2.0
Stress Reaction/Fracture	☐ None	1	☐ ≥ 2; ≥ 1 high risk or of trabecular bone sites†
Cumulative Risk (total each column, then add for total score)	____ points +	____ points +	____ points = Total Score

Consensus statement

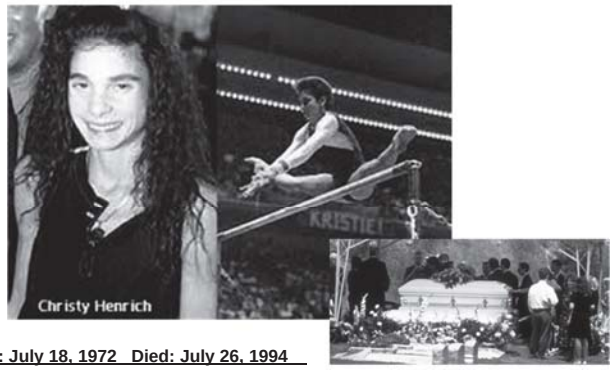
	Cumulative Risk Score*	Low Risk	Moderate Risk	High Risk
Full Clearance	0 – 1 point	☐		
Provisional/Limited Clearance	2 – 5 points		☐ Provisional Clearance ☐ Limited Clearance	
Restricted from Training and Competition	≥ 6 points			Restricted from ...ining/ Competition-Provisional ☐ Disqualified

What we are going to talk about

- Define Female Athlete Triad Syndrome
- Explain How ***YOU*** can Prevent and Screen in the during routine well child checks
- Explore Diagnosis and Return to Play Guidelines



Age 22, Multi-organ Failure, 60lbs
Christy Henrich



Born: July 18, 1972 Died: July 26, 1994

TO THIN TO TRAIN??

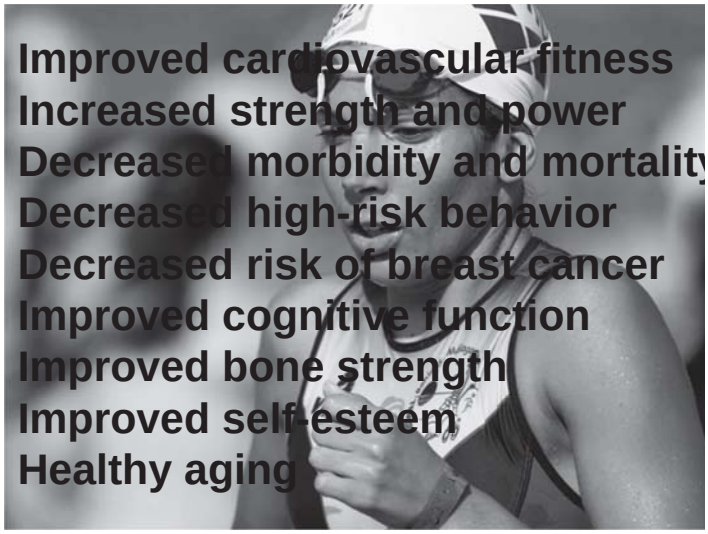
TO THIN TO TRAIN?

Meet Sarah.

- "I realized that as I worked harder and lost some weight, my times were improving,"
- "So I figured that if a little weight loss was good, a lot would be even better."

Simple Logic:

- Sarah's downward spiral into the depths of anorexia is perhaps most disturbing for its simple logic:
- If a few pounds were good for performance, a lot of pounds would be amazing…



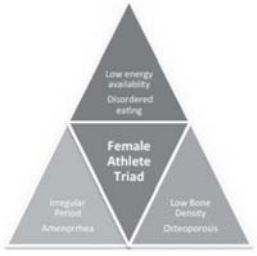
Unrealistic standards of appearance and performance

If a little weight loss is good, More is Better

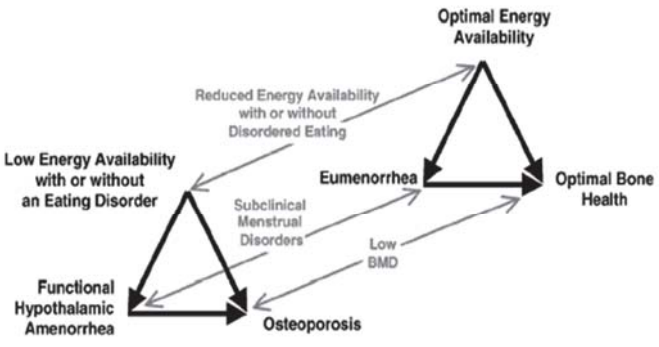
"Smarten up"

- "Even though your score is suppose to be based on your routine, you must know that you are giving the judge lots of signals…approach the apparatus with your head high, clothes tidy, hair in place. You will be "saying" to the judge you have trained well…Judges will see you in a positive light. They may even be tempted to run out on the floor and pinch your cheek because you are killing them with "cute". Judges love "cute" so work it babe!"

Female Athlete Triad- Defined in 1992



The Female Athlete Prism-The Spectrum of the Female Athlete Triad



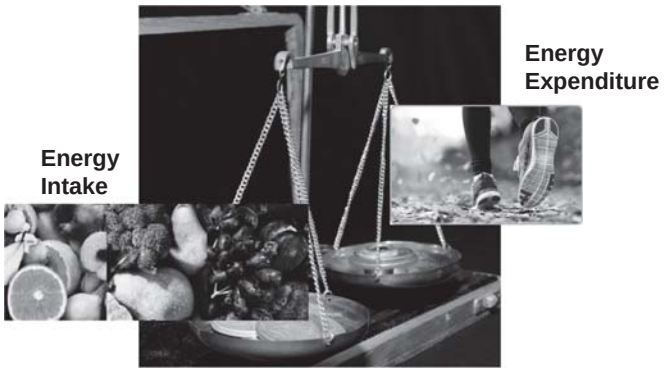
Screening Recommendations

- Female Athlete Triad Coalition recommends screening once a year with self reported questionnaire
- If there is any one symptom of the triad further investigation should be initiated

Female Triad Coalition Questions??

- Have you ever had a menstrual period?
- How old were you when you had your first menstrual period?
- *When was your most recent menstrual period?
- How many periods have you had in the last 12 months?
- *Are you presently taking any female hormones (estrogen, progesterone, birth control pills)?
- Do you worry about your weight?
- Are you trying to or has any one recommended that you gain or lose weight?
- Are you on a special diet or do you avoid certain types of foods or food groups?
- Have you ever had an eating disorder?
- Have you ever had a stress fracture?
- Have you ever been told you have low bone density (osteopenia or osteoporosis?)

Low Energy Availability

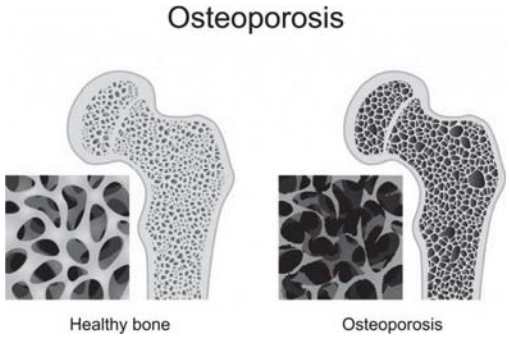


Energy Balance

How Can You Assess Low Energy Availability

- Energy availability calculator on Female Athlete Coalition Website
  - **http://www.femaleathletetriad.org/calculators/**
- Nutrition assessment with sports dietician
- Energy expenditure apps

Consequences of Low Energy Availability



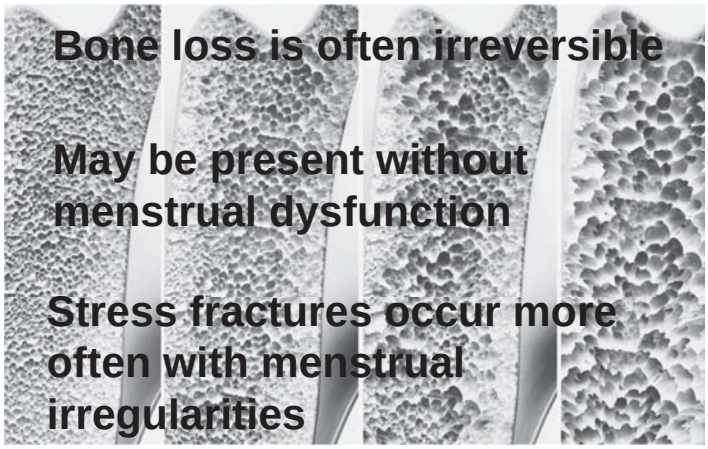
How Athlete's Reduce Energy-disordered eating

- Abnormal eating behaviors
  - Fasting
  - Binge-eating
  - Purging
  - Diet pills
  - Laxatives
  - Diuretics
  - Enemas
- Eating disorders/mental health disorder
  - Anorexia/Bulimia

Menstrual Dysfunction

- Amenorrhea: primary or secondary
  - Primary: delay of menarche
  - Secondary: cessation after regular menstrual cycles have been established
- Underlying factor is inadequate energy availability
- Amenorrheic women are infertile due to absence of ovulation, *BUT* they may ovulate before menses is restored = unintended pregnancy!

Osteopenia/Osteoporosis



Health Consequences

- Psychological Health
  - Low self esteem, depression, anxiety
  - 5.4% athletes with eating disorders reported suicide attempts
- Medical Complications
  - Cardiovascular, endocrine, reproductive, skeletal GI, renal and central nervous systems

Sarah: "I felt alone…"

- For most health issues, off to the PCP…
- "When I went to see my PCP, it was not helpful"
  - "I was told I should gain weight to reach 120 pounds"
  - "That's more than I ever weighed before I even began running"

Well Meaning Useless Advice… "I FELT ALONE"

- Disconnect between a PCPs advice and the goals of an athlete
  - No constructive path for an athlete to follow
  - Yes, she needed to add some pounds back on, but she wasn't willing to give up her athletic dreams to do so

"I felt alone"

Prevention/Early Detection

- Education!!
  - Athletes, parents, coaches, athletic trainers, judges, administrators
- Pre-participation Physical
- Presentation with any associated clinic syndrome
- Rule changes
  - Discourage unhealthy weight loss practices

Identify Athletes at Greatest Risk

- Restrict dietary energy intake
- Exercise for prolonged periods
- Vegetarian
- Limit the foods they will eat
- Early start of sport-specific training and dieting, injury and sudden increase in training volume

Identify Athletes Most at Risk for Stress Fracture

- Low BMD
- Menstrual disturbance
- Late menarche
- Dietary insufficiency
- Genetic predisposition
- Biomechanical abnormalities
- Training errors
- Bone geometry

Nonpharmacologic Treatment

- Main goal of treating the triad is increasing energy availability
- Goals: Improved bone health and menstrual function
- Multidisciplinary team is key
- Time course is different for each athlete

Recovery

- Recovery of Bone Mineral Density
  - Process: YEARS
- Recovery of Menstrual Cycle
  - Process: MONTHS
- Recovery of Energy Status
  - Process: DAYS TO WEEKS

Treatment

- Recommend increasing dietary energy intake and decrease exercise energy expenditure or both
- Individual treatment plans: diet quality, timing, incorporation of energy dense foods, adjustments for training
- Increase energy intake gradually 20-30% over baseline needs
- Weight gain of approx 0.5 kg every 7-10d
- Regular monitoring with sports dietitian

Treatment

- Weight gain to achieve a BMI of >18.5
- Return of body weight associated with normal menses
- Reversal of recent weight loss

Calcium and Vitamin D

- 9-18 years
  - Vitamin D: RDA 600 units
  - Calcium: RDA 1300mg
- 19-50 years
  - Vitamin D: RDA 600 units
  - Calcium: RDA 1000mg

Pharmacological Therapy

- Lack of evidence based studies to recommend pharmacological therapy
- Would only be considered in athlete if lacking response to non-pharmacologic management with low BMD + clinical significant fracture history
- In general we do NOT treat with oral contraceptives as they mask the menstrual problems and do not increase bone density

Triad Clearance

- Conundrum: many athletes cleared without proper management and assessment
- Return to Play:
  - Athletes often return after triad associated injures or illness without adequate management or follow up

Evidence Based risk factors associated with Poor outcomes

- Low energy availability with or without disordered eating/eating disorder
- Low BMI
- Delayed menarche
- Oligo/amenorrhea
- Low BMD
- Stress reaction/fracture history
- Leanness sport

Female Athlete Triad Cumulative Risk Assessment

Risk Factors	Magnitude of Risk		
	Low Risk = 0 points each	Moderate Risk = 1 point each	High Risk = 2 points each
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Low BMI	☐ BMI ≥ 18.5 **or** ≥ 90% EW** or weight stable	☐ BMI 17.5 < 18.5 **or** < 90% EW **or** 5 to < 10% weight loss/month	☐ BMI ≤17.5 **or** < 85% EW **or** ≥ 10% weight loss/month
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Stress Reaction/Fracture	☐ None	☐ 1	☐ ≥ 2; ≥ 1 high risk or of trabecular bone sites†
Cumulative Risk (total each column, then add for total score)	____ points +	____ points +	____ points = ____Total Score

Athlete Participation in Sport

- Athlete must agree:
  - To comply with all treatment strategies
  - To be closely monitored by health-care professionals
  - Place a precedence on treatment over training and competition
  - Modify type, duration, and intensity of training and competition
- Often useful to have a written contract with the agreements

Return to Play- Complex Equation

- Willingness of athlete to comply with goals
- Sport-specific training demands
- Is the sport an increased risk of medical and/or psychological risk to the athlete
  - Yes: consider limiting or withholding training/competition
  - Withholding training/competition can be motivating

Clearance…

- Need to respect the athletes privacy, very sensitive issue
- However communication with coaching staff extremely important
  - Coaches may be a part of the solution
- If disqualified specific steps need to be outlined for the athlete
  - Who should they meet with
  - What are the consequences
  - Timeframe for return to training and competition

Questions before I summarize?

Female Athlete Triad- Summary

- Spectrum of health and disease based on energy availability
  - Disordered Eating
  - Menstrual Dysfunction
  - Bone Mineral Density
- Identification of those at risk
- Treatment team is multi-disciplinary

Sarah's parting words-

- **"Your body can't run on nothing. Eventually, you will crash and burn. If a friend or coach says something, be open to considering what they're telling you. The sooner you get help, the easier it will be to get your life back."**

Thank you!

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2014 Female Athlete Triad Coalition Consensus Statement on Treatment and Return to Play of the Female Athlete Triad: 1st International Conference Held in San Francisco, CA, May 2012, and 2nd International Conference Held in Indianapolis, IN, May 2013

Primary Authors: De Souza MJ, Nattiv A, Joy E, Misra M, Williams NI, Mallinson RJ, Gibbs JC, Olmsted M, Goolsby M, Matheson G

Expert Panel Members: Barrack M, Burke L, Drinkwater B, Lebrun C, Loucks AB, Mountjoy M, Nichols J, Sundgot-Borgen J

Endorsed by the American College of Sports Medicine, the American Medical Society for Sports Medicine and the Female Athlete Triad Coalition

Published in: British Journal of Sports Medicine, Vol 48, Feb 2014

Clinical Journal of Sport Medicine, Vol 24 (2), March 2014


Cases of Horses and Zebras of Pediatric Sports Injuries

Ryan Petering MD, CAQSM
OHSU Sports & Family Medicine
OHSU Sports Medicine Fellowship Director

Disclosures

- Author of chapter in one of text will recommend
- Otherwise – none.

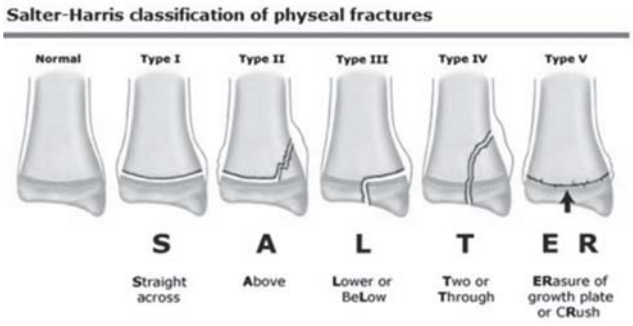
Agenda

- Case based approach to common and uncommon Pediatric Sports injuries

RESOURCES

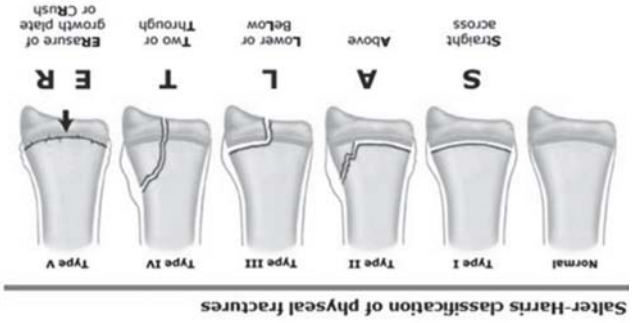
- Orthobullets
- AFP (American Family Physician)
- Fracture Management Primary Care

Salter Harris Classification



UpToDate: General principles of fracture management: Fracture patterns and description in children

Salter Harris Classification



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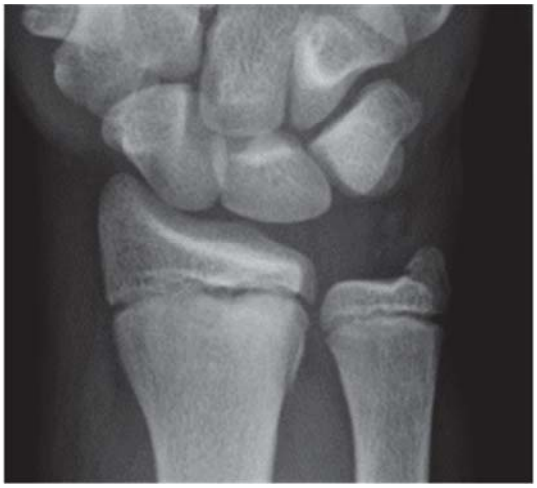
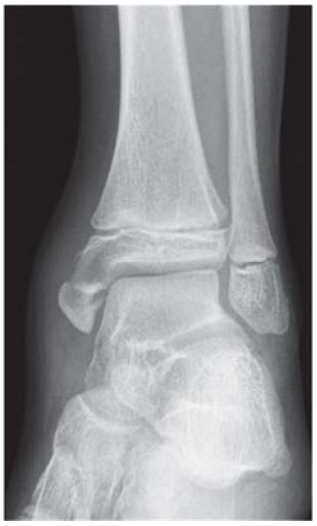
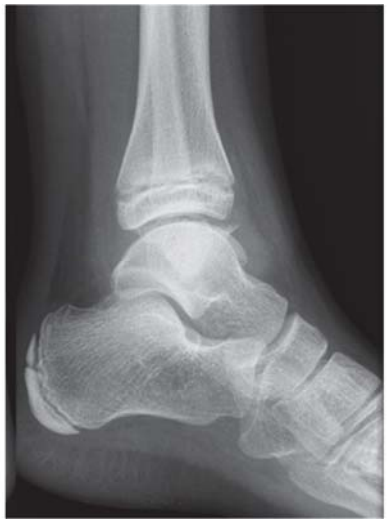
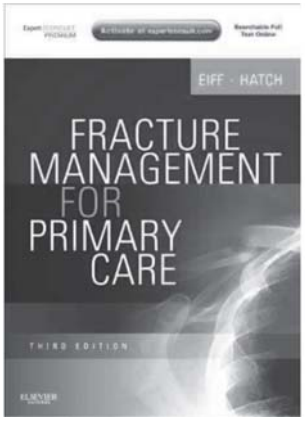
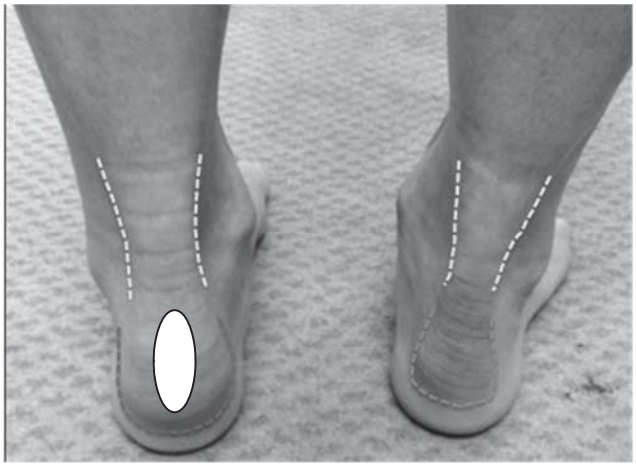


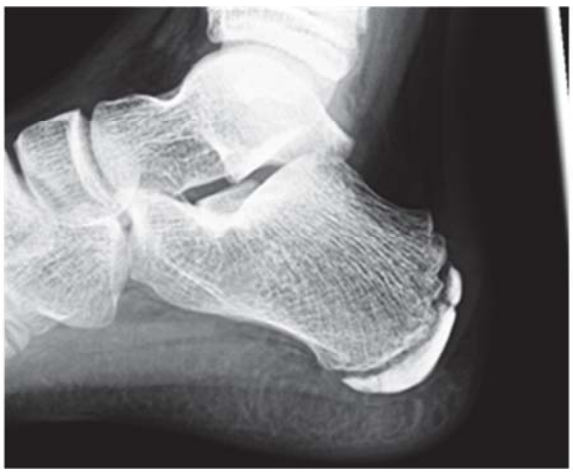






10 yo male soccer player with 2-3 weeks of heel pain. No injury.



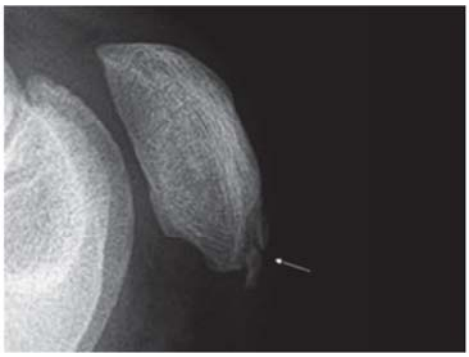


Severs Disease

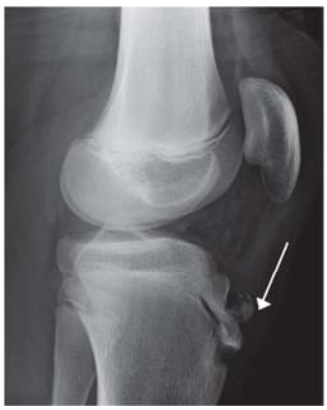
- Apophysitis of the Achilles tendon at calcaneus insertion
- 8-11 years old/boys > girls
- Primarily relative rest treatment
- Heel raise insert
- Does NOT require strict rest or sports avoidance.
- Does NOT mandate xray (preferred to avoid)
- Questionable if PT helpful when compared to wait and see, heel raise (n = 101)
  - J Pediatr Orthop. 2016 Mar;36(2):152-7. Treatment of Calcaneal Apophysitis: Wait and See Versus Orthotic Device Versus Physical Therapy: A Pragmatic Therapeutic Randomized Clinical Trial.

13yo male basketball player with recurrent/chronic knee pain – anterior – with no trigger - worse with activity.

Sindig Larsen Johannson



Osgood Schlatter



Osgood Schlater/Sindig Johanson Larsen Disease

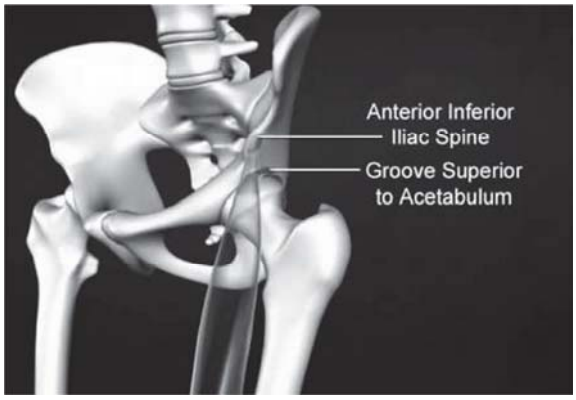
- Apophysitis of the patellar tendon at tibia (OS) or the patella (SJL)
- Primarily relative rest treatment
- Chopat strap
- Does NOT require strict rest or sports avoidance.
- **Does NOT mandate xray** (preferred to avoid)

Pediatrics. 2011 Nov;128(5):e1121-8. Epub 2011 Oct 3. Hyperosmolar dextrose injection for recalcitrant Osgood-Schlatter disease.

- N = 65
- Compared with usual care at 3 months, unaltered sport was more common in both dextrose-treated (21 of 21 vs 13 of 22; P = .001) and lidocaine-treated (20 of 22 vs 13 of 22; P = .034) knees, and asymptomatic sport was more frequent in dextrose-treated knees than either lidocaine-treated (14 of 21 vs 5 of 22; P = .006) or usual-care-treated (14 of 21 vs 3 of 22; P < .001) knees.
- At 1 year, asymptomatic sport was more common in dextrose-treated knees than knees treated with only lidocaine (32 of 38 vs 6 of 13; P = .024) or only usual care (32 of 38 vs 2 of 14; P < .0001).

14yo sprinter, during competition, sudden onset of right thigh/anterior hip pain.

Rectus Femoris Avulsion

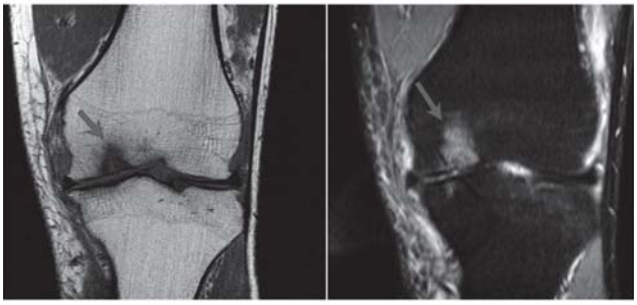


Rectus Femoris Avulsion

- Sudden onset anterior hip pain
- Sprinter/soccer/explosive sports player
- Often initially unable to walk
- **Xrays ARE needed** – to assess if bony avulsion
- Ultrasound or MRI needed often to quantify if tear in tendon or just avulsion
- Nonoperative management – weight bearing as tolerated
- Large bone avulsion, tendon retraction – indication to refer Ortho.
- Typically 6-12 week full return to sport

15yo active male with recurrent, chronic left knee pain and effusion – no focal event/injury – and an exam normal except for effusion

Osteochondral Lesion



Osteochondral Dissecans (OCD)

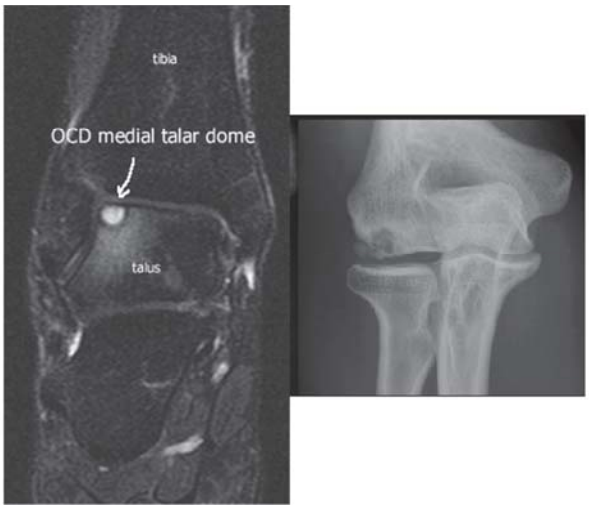
Most common location:
- Femoral condyles
- Capitellum humerus @ elbow
- Talar dome

Epidemiology
- Most common age group: – Adolescence

Presentation:
- Joint pain
- SWELLING = EFFUSION
- Limited ROM
- Mechanical symptoms

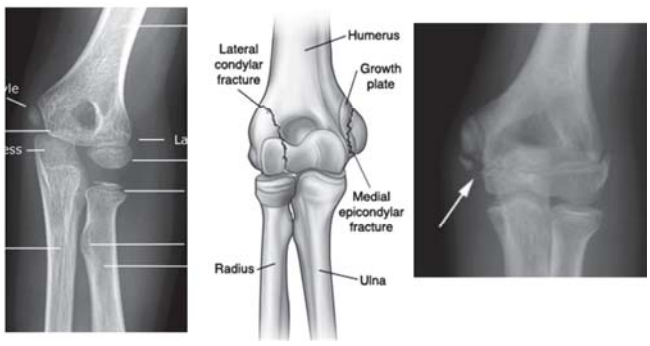
Diagnosis:
- **Xray: Tunnel view of the knee (4th view)**
  - Consider getting bilateral films
- MRI: Confirm/Staging

Management
- Activity restriction/reduction
- Non weight bearing if severe pain
- Bracing
- Surgical options



15yo baseball pitcher with medial elbow pain x 2 weeks.

Throwers Elbow



Little Leaguers Elbow

- Risk factors
  - Greater than 80 pitches per game
  - More than 8 months of competitive pitching per year
  - Fastball speed > 85mph
  - Continued pitching despite arm fatigue/pain
  - Participating in showcases/tournament

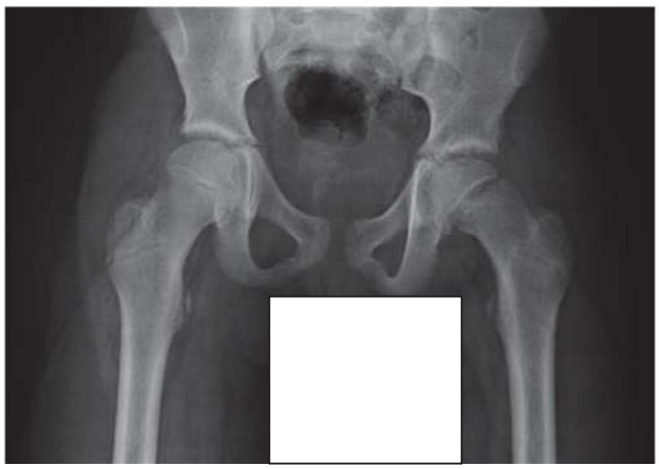
AGE	DAILY MAX (PITCHES IN GAME)	REQUIRED REST (PITCHES)				
		0 Days	1 Days	2 Days	3 Days	4 Days
7-8	50	1-20	21-35	36-50	N/A	N/A
9-10	75	1-20	21-35	36-50	51-65	66+
11-12	85	1-20	21-35	36-50	51-65	66+
13-14	95	1-20	21-35	36-50	51-65	66+
15-16	95	1-30	31-45	46-60	61-75	76+
17-18	105	1-30	31-45	46-60	61-75	76+
19-22	120	1-30	31-45	46-60	61-75	76+

http://m.mlb.com/pitchsmart/pitching-guidelines

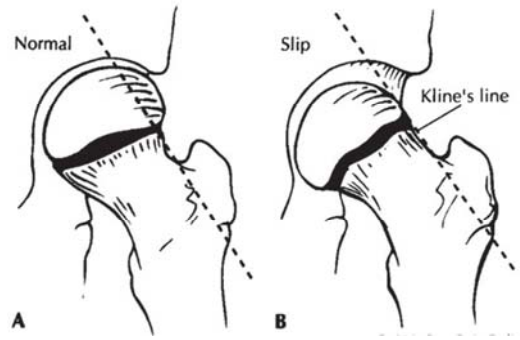
Little Leaguers Elbow

- Treatment
  - Nonoperative most common
  - Surgical consideration if bony fragment (debate as to size of fragment needed for surgery)
  - Pitch count adherence
  - Gradual return to baseball – with delayed return to pitching

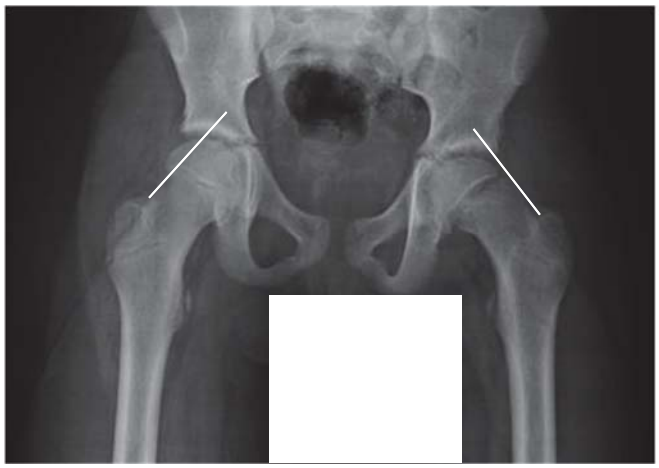
11yo obese male football player – new/acute onset left hip pain with gradual worsening over 2-3 days – no trigger/focal event



SCFE (Slipped Cap Femoral Epiphysis)



Pediatric Rheumatology 7(1):10 · June 2009

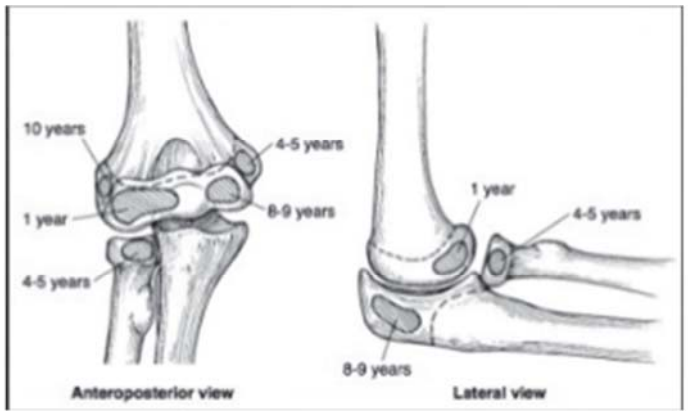


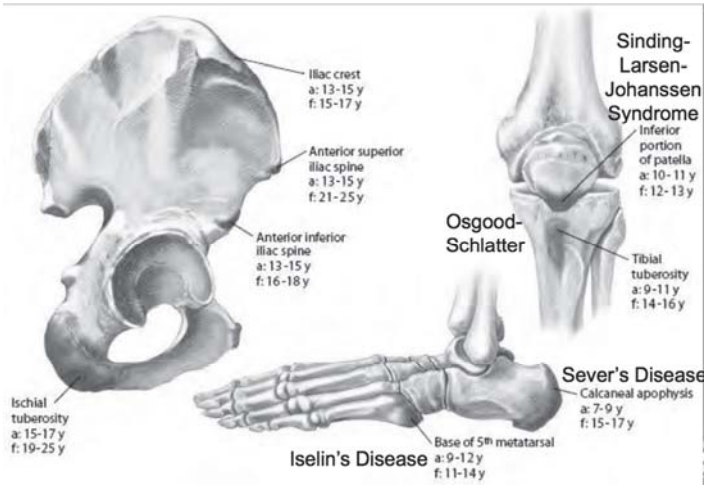
SCFE (Slipped Cap Femoral Epiphysis)

- Greatest risk factor?
- Most common age onset?
- Presenting sign?
- Management?

SCFE (Slipped Cap Femoral Epiphysis)

- Greatest risk factor?
  - Obese
  - Male
  - African American/Islanders
- Most common age onset?
  - 13yo boys/12yo female
- Presenting sign?
  - Groin/hip pain most common
  - Knee pain not rare
- Management?
  - Surgery
  - Crutches/emergent referral



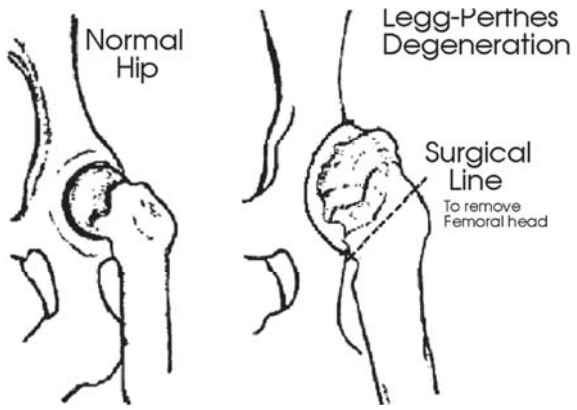


5yo male soccer play with limp, thigh pain and knee pain x 4 months with no trigger/causative event. Normal knee and thigh exam.

Legg Calve Perthes

- Idiopathic avascular necrosis of proximal femoral epiphysis
- M>F (5:1)
- 4-8yo (5yo most common)
- Bilateral ~ 10-15%
- Risk factors
  - positive family history
  - low birth weight
  - abnormal birth presentation
  - second hand smoke
  - Asian, Inuit, and Central European decent

Legg Calve Perthes



Legg Calve Perthes

- Nonoperative
  - Activity modification
  - Maintain motion
  - No role for bracing/casting/splinting
- Operative
  - Typically > 8yo

15yo female soccer player with sudden onset knee pain while cutting/pivoting

ACL tear

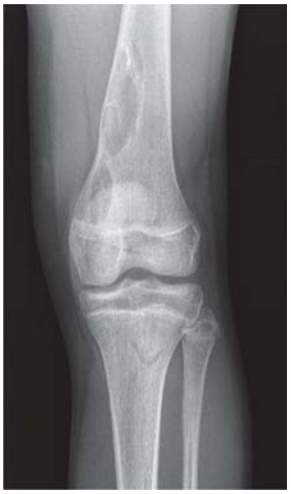
- F > M (~ 5:1)
- Common noncontact/pivoting
- Effusion can occur in < 1 hour
- Pain vs instability
- Surgery or no surgery?
- Timeframe for surgery?
- PEP program

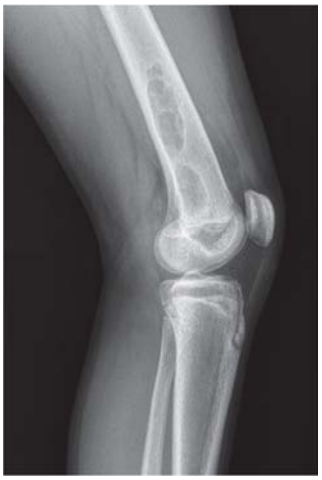
http://smsmf.org/files/PEP_Program_04122011.pdf

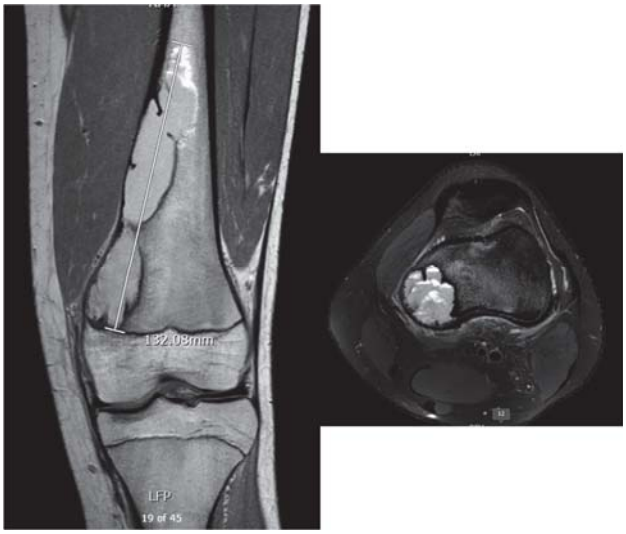
Anterior Drawer

Lachman

15yo female ballerina with acute worsening of chronic knee pain – medially knee





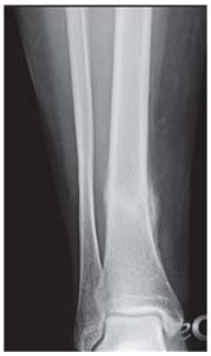


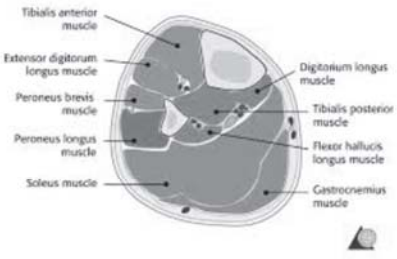
Aneurysmal Bone Cyst (ABC)

- Benign & nonneoplastic bone lesion
- 75% < 20yo @ diagnosis
- Spine (25%), long bones (25%)
- Pain and swelling
- May present as pathologic fracture
- Missed often on plain films
- Treatment – usually surgical
  - Curettage +/- bone grafting
  - Cements, other adjuvants

15yo baseball player with shin pain – anterior – x 4 weeks, now preventing running.

Stress Fracture vs Shin Split





Risk Category Stress Fracture

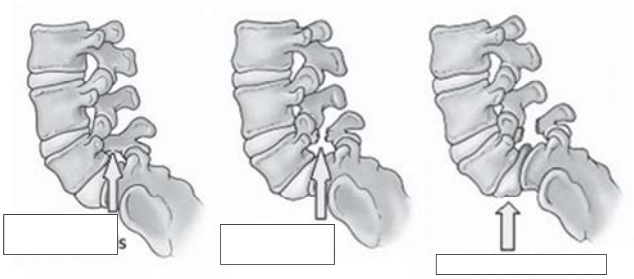
High-risk fracture sites	Low-risk fracture sites
Femoral neck	Femoral diaphysis
Patella	Medial tibia
Anterior diaphysis of tibia	First to fourth metatarsals
Medial malleolus	Fibula
Talus	Calcaneus
Tarsal navicular	Pelvic skeleton
Fifth metatarsal	
Sesamoids of the great toe	

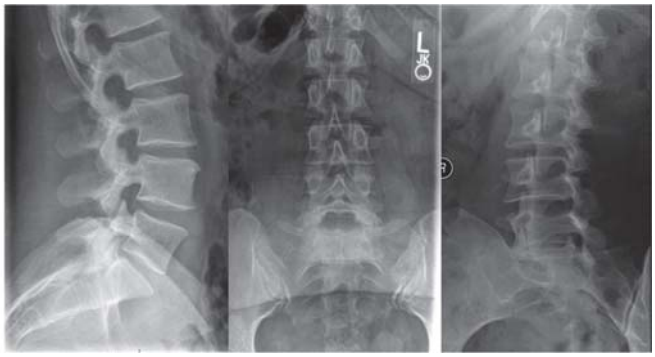
Shin Splint vs Stress Fracture

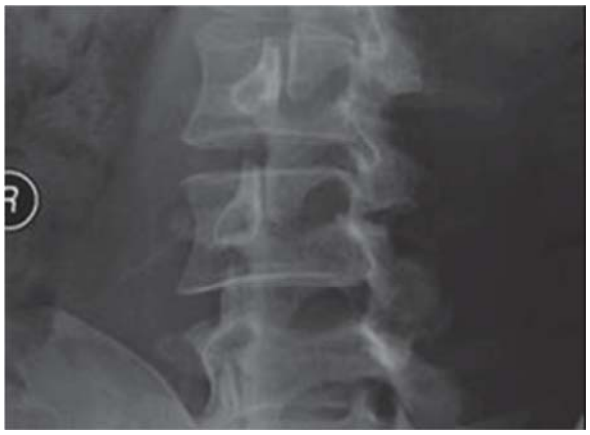
Shin splint (Medial Tibial Stress Syndrome)	Stress Fracture
Diffuse pain location anterior tibia; arch collapse/medial knee deviation/hip drop	**Focal** pain location; arch collapse/medial knee deviation/hip drop
May improve with running (initially)	Worse with running
Associated with activity – does not have to be intense activity – or with increase; more common in novice exercisers	Typically associated with dramatic (relative) increase in volume of exercise
Xray negative	Xray negative (unless 3-4 weeks of symptoms – may have callus); MRI or bone scan typically needed
Activity reduction is key pain management	Typically non weight bearing and consider boot/cast initially
Shoe changes, inserts, arch support and lower leg strengthening exercises	Gradual ramp up of activity – medial tibial stress reaction – typically 6-8 weeks out of running (best case)

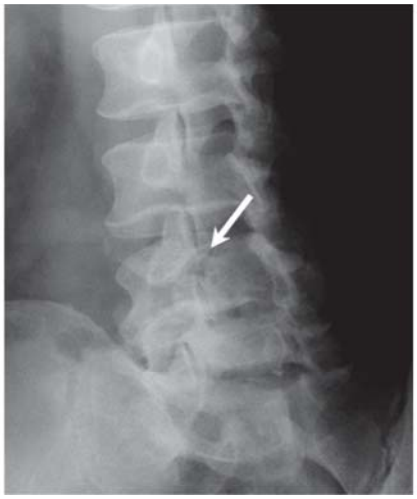
13yo female gymnast with 3-4 months of low back pain

Spondylolysis vs Spondylolisthesis









Spondylolysis vs Spondylolisthesis

- Stress reaction/fracture pars interarticularis = spondylolysis
- Anterior motion of lumbar vertabrae – spondylolisthesis
- Grading based on how much anterior motion (law of 25%)
- Adolescents with recurrent hyperextension of back
- Typically many months duration when diagnosed – misdiagnosed as low back strain.
- **Xray – need oblique views bilaterally vs MRI/CT**
- Non operative if less than 50% anterior motion
- Back bracing controversial/unclear benefit if less than 25-50%
- Typically 90 days of noncompetition if spondy

R. Grazina et al. / Physical Therapy in Sport 37 (2019) 34e43

Spondylolysis vs Spondylolisthesis

- Return to play at any level was approx. 90% return to the pre-injury sports activity level
- The mean time to return to sports was 4+ months.
- Approx 90% return with nonsurgical/conservative management
- Surgically managed patients had 6+ months to return to sports
- Approx 80% return with surgical management.

R. Grazina et al. / Physical Therapy in Sport 37 (2019) 34e43

16yo football kicker – collision on field – likely LOC for approx. 10sec – assessed on sideline – and SCAT 5 assessment performed

Concussion

- Recognize, Remove, Recover, Return to Learn/Play
- Physical activity recommendations
- Sub-symptom threshold
- Return to play
- Avoid ED, avoid imaging, avoid predetermined time off
- Focus on common primary care topics – poor sleep, anxiety and other psychosocial issues

Summary

- Respect the physis!
- Remember these common diagnosis – approach patient with goal to make sure your patient does not have one of these!
- When there is trauma – think broken bone
- Use references

Thank you

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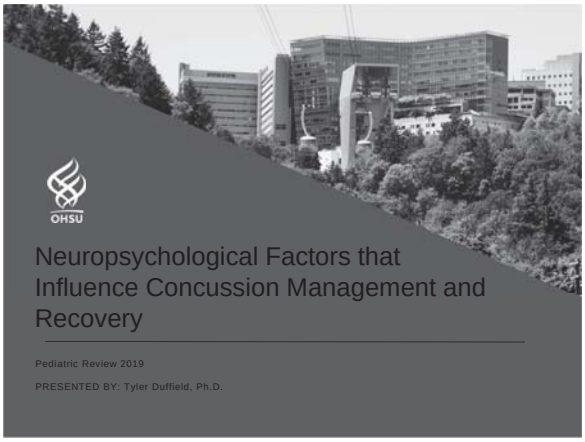
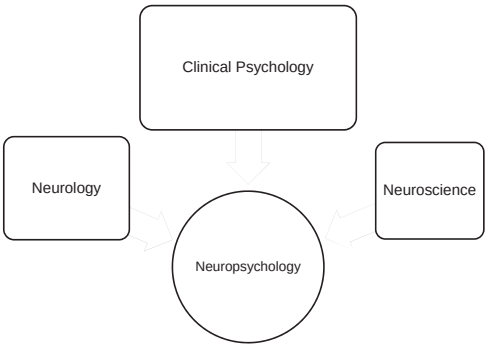


Neuropsychological Factors that Influence Concussion Management and Recovery

Pediatric Review 2019

PRESENTED BY: Tyler Duffield, Ph.D.

What is neuropsychology?



Neuropsychology Application

- Distinguish injury from non-injury factors:
  - Neurologic vs.
  - Psychiatric vs.
  - Neurodevelopmental vs.
  - Psychosocial/Family factors
  - Or more often the case, a combination of these factors

Epidemiology of Concussion

- CDC: approximately 1.7 million Americans sustain annual traumatic brain injury (TBI)
  - approximately 70% (i.e., 1.2 million) considered mild (mTBI)
- Several groups of authors have noted that the actual number of TBIs annually is likely much higher, as many go undiagnosed, unreported, and thus uncounted.
- Estimated total expenditures exceeding $21.5 billion per annum for mTBI alone

Characterizing TBI

Table 1 Classification of traumatic brain injury

Mild ("concussion")	Moderate	Severe
Normal imaging	Normal or abnormal imaging	Abnormal imaging
Loss of consciousness (LOC): 0–30 min	LOC > 30 min < 24 h	LOC > 24 h
Alteration of consciousness (AOC): up to 24 h	AOC > 24 h	
Post-traumatic amnesia (PTA): 0–1 day	PTA > 1 and < 7 days	PTA > 7 days


NEUROSURGERY

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Article Contents

Abstract

METHODS

RESULTS

DISCUSSION

CONCLUSION

REFERENCES

Concussion Guidelines Step 2: Evidence for Subtype Classification

Angela Lumba-Brown, MD, Masaru Teramoto, PhD, MPH, PStat, O Josh Bloom, MD, MPH, David Brody, MD, PhD, James Chesnutt, MD, James R Clugston, MD, MS, Michael Collins, PhD, Gerard Gioia, PhD, Anthony Kontos, PhD, Avtar Lal, PhD ... Show more

Neurosurgery, nyz332, https://doi.org/10.1093/neuros/nyz332

Published: 21 August 2019 **Article history**

- 5 subtypes:
  - Cognitive
  - Ocular-motor
  - Headache/migraine
  - Vestibular
  - Anxiety/mood
- Also considered sleep disturbance and cervical strain as associated conditions

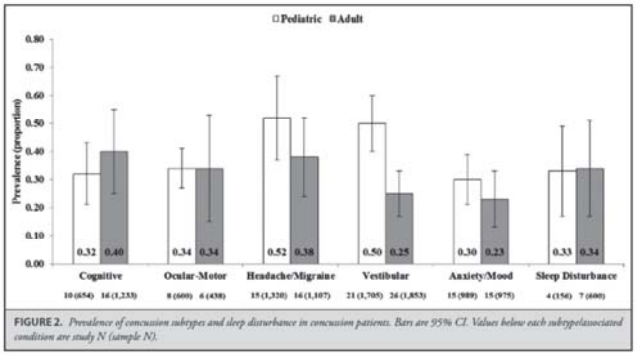


FIGURE 2. *Prevalence of concussion subtypes and sleep disturbance in concussion patients. Bars are 95% CI. Values below each subtype/associated condition are study N (sample N).*

Oregon Legislation

- Max's Law (2010) applies only to Oregon School Districts.
- Jenna's Law (2014) extends the intent of Max's Law to Oregon youth sports and referee organizations.
- Both Max's and Jenna's Laws require school and non-school youth athletic programs to:
  - Create policies and procedures.
  - Provide training.
  - Track training.
  - Ensure that staff practice good concussion management.
  - Restrict play when a concussion is suspected.
  - Provide educational materials/programs.

Oregon Legislation

- Senate Bill 1547 (2018) takes effect in 7/2020
  - Allow a larger range of medical professions to make medical clearance decisions if they undergo an education module
  - Previously allowed:
    - Physicians, nurse practitioners, physician assistants and (neuro)psychologists
  - Now also allowed:
    - Chiropractors, naturopaths, physical therapists and occupational therapists
    - However, not athletic trainers!

Research

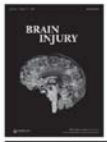
Original Investigation

Point of Health Care Entry for Youth With Concussion Within a Large Pediatric Care Network

Kristy B. Arbogast, PhD; Allison E. Curry, PhD; Melissa R. Pfeiffer, MPH; Mark R. Zonfrillo, MD, MSCE; Juliet Haarbauer-Krupa, PhD; Matthew J. Breiding, PhD; Victor G. Coronado, MD, MPH; Christina L. Master, MD

Why a Neuropsychologist in Primary Care?

- 82% (n = 6624) first visit within primary care
- 5% (n = 418) within specialty care (e.g., neurology)
- 12% (n = 947) within the ED
  - Age: Significantly higher rate of <4 y/o
  - Race/Ethnicity: 42% AA patients compared to 5% white patients
  - Payor:
    - 37% children insured by Medicaid
    - 24% self-pay
    - 7% private insurance



Brain Injury

ISSN: 0269-9052 (Print) 1362-301X (Online) Journal homepage: https://www.tandfonline.com/loi/ibij20

Products and activities associated with non-fatal traumatic brain injuries in children and adolescents – United States 2010-2013

Bina Ali, Bruce A. Lawrence, Ted Miller & Jennifer Allison

- TBI rates averaged 1,237 per 100,000 population
  - 1,457 for males and 1,006 for females
- Majority of TBI cases (92%) were treated in an ED and released
- Most TBIs are unintentional (93.7%), but small subset due to assault (6.1%)
- Nearly three-fourths (72.2%) associated with consumer product
- Product-related TBIs were more frequent among:
  - <1 year (90.6%)
  - 1–4 years (81.4%)
  - 5–9 years (71.9%)
  - 10–14 years (75.1%)
  - 15–19 years (54.8%)

Table 2. Leading product groups associated with non-fatal TBI in children and adolescents by age group, 2010–2013.

	Age Group					
Product Group	<1 (n = 380,842)	1–4 (n = 1,085,680)	5–9 (n = 682,826)	10–14 (n = 834,565)	15–19 (n = 1,107,463)	Total (n = 4,091,376)
Sports & recreation	0.8%	7.8%	31.8%	53.9%	38.3%	28.8%
Home furnishings & fixtures	42.7%	32.0%	14.6%	5.3%	4.5%	17.2%
Home structures & construction materials	23.6%	28.6%	17.9%	10.6%	8.1%	17.1%
Child nursery equipment	18.8%	3.3%	0.2%	0.0%	0.0%	2.7%
Toys	1.0%	2.7%	3.0%	2.8%	1.9%	2.4%
Personal use items	1.6%	3.5%	1.2%	0.3%	0.3%	1.4%
Home electronics & hobby	0.8%	1.2%	0.8%	0.4%	0.4%	0.7%
Other product groups	1.2%	2.1%	2.4%	1.6%	1.4%	1.8%
No product involved	9.4%	18.6%	28.1%	24.9%	45.2%	27.8%

Table 3. Top ten leading products contributing to non-fatal TBI in children and adolescents by age group, 2010–2013.

Rank	<1 Year (n = 380,842)	1–4 Years (n = 1,085,680)	5–9 Years (n = 682,826)	10–14 Years (n = 834,565)	15–19 Years (n = 1,107,463)	Total (n = 4,091,376)
1	Beds 25.4%	Stairs 9.9%	Floors 6.1%	Football 13.7%	Football 8.8%	Floors 6.4%
2	Floors 14.1%	Beds 9.7%	Bicycles 5.2%	Basketball 6.4%	Basketball 4.8%	Beds 6.1%
3	Sofas/couches 7.4%	Floors 9.7%	Beds 4.1%	Bicycles 5.4%	Soccer 3.8%	Football 5.7%
4	Stairs 6.2%	Tables 5.2%	Stairs 3.3%	Soccer 4.8%	Bicycles 3.0%	Stairs 4.6%
5	Car seats 4.6%	Chairs 5.2%	Football 3.0%	Baseball/softball 4.4%	Floors 2.7%	Bicycles 3.1%
6	Tables 3.1%	Sofas/couches 4.2%	Ceilings and walls 2.8%	Floors 3.6%	Baseball/softball 2.4%	Basketball 2.9%
7	Baby strollers 2.7%	Ceilings and walls 3.4%	Monkey bars/playground gyms 2.6%	Ceilings and walls 2.3%	Stairs 1.9%	Ceilings and walls 2.3%
8	Chairs 2.5%	Grocery/shopping carts 3.1%	Baseball/softball 2.4%	Stairs 1.9%	Skateboards 1.5%	Chairs 2.3%
9	High chairs 2.2%	Doors 1.6%	Tables 2.0%	Skateboards 1.6%	Ceilings and walls 1.4%	Soccer 2.2%
10	Baby changing tables 2.1%	Cabinets/shelves 1.6%	Swings 1.9%	Cheerleading 1.2%	Wrestling 1.3%	Tables 2.2%

Big Take Aways

- <10 y/o, beds leading cause of TBI
  - Consistent with prior findings
- Placing infants on beds/furniture and fall/roll off
- Bunk beds are especially risky – danger of top bunk
- <1 y/o, car seats problematic, particularly when used as carrier inappropriately
  - e.g., placing on countertop and falling/knocked off

Big Take Aways

- 5-9 y/o, bicycle crashes often contribute to TBI
- 10–19 y/o sustain TBIs from contact sports, primarily football
- TBIs associated with floors and stairs are common in children and adolescents of all ages (account for approximately 11%)
  - Structural designs, such as uneven flooring and prefabricated stairs
  - Hard or non-resilient surfaces, such as asphalt and concrete, are associated with skull and upper extremity fractures

Prevention Strategies in and Around the Home

- Removing tripping hazards such as area rugs
- Improving lighting
- Avoiding hard surface playgrounds
- Increasing use of home safety devices
  - Stair gates and guard rails that are easily grasped and no sharp edges
- Avoid use of prefabricated stairs
  - Create tripping hazard when the builder raises/lowers the top-step riser to adjust the stairway height to match the actual height rise between floors
- Caregiver education and home safety visits
- Enforcement of game and playground safety rules, consistent and proper use of safety gear, notably helmets, adult supervision, and education of youth athletes, parents, and coaches

Published in final edited form as:
J Pediatr. 2019 July ; 210: 13–19.e2. doi:10.1016/j.jpeds.2019.04.001.

Risk of Repeat Concussion Among Patients Diagnosed at a Pediatric Care Network

Allison E. Curry, PhD, MPH[1,2], Kristy B. Arbogast, PhD[1,2], Kristina B. Metzger, PhD, MPH[1], Ronni S. Kessler, MEd[1], Matthew J. Breiding, PhD[3], Juliet Haarbauer-Krupa, PhD[3], Lara DePadilla, PhD[3], Arlene Greenspan, DrPH[3], Christina L. Master, MD[1,4]

- 16% history of concussion at index concussion
  - 22% of those repeat within 2 years vs 15% w/o history of concussion
- 8.4% (n = 45) repeat concussion within 1 year
- 16.2% (n = 87) repeat concussion within 2 years
  - including 3.4% (n = 18) with 2 additional concussions
- Median (IQR) time to repeat concussion was 11.8 (5.8–17.8) months
- Risk among 12-to 15-y/o was 1.85 times that of 9-to 11-y/o
- Risk was 1.5 times higher ≥1 pre-existing co-occurring condition
  - Migraine/headache (28.6%)
  - Anxiety (25.0%)

- Clinical course and symptom burden risk of repeat injury:
  - 2 times greater for one month vs. one week of symptoms
  - 2.5 times greater for ≥11 symptoms vs. 0–2 symptoms
  - Highly correlated, constructed multivariate models
    - Predicted risk increased
    - Presence of co-occurring condition non-significant
- The 2-year risk of a repeat concussion did not vary by:
  - Sex
  - Insurance payor
  - Mechanism of injury

Concussion Severity/Grading & Return to Play (RTP)

- 14 guidelines identified by Collins et al. (1999)
- 3 emerged as the most widely used:
  - The Cantu Grading Scales
  - The Colorado Medical Society Guidelines (CMS)
  - The American Academy of Neurology guidelines (AAN)
- All use mild, moderate, severe ratings
- Generally based upon symptom duration, post-traumatic amnesia (PTA), and loss of consciousness (LOC)
- "An examination of the grading systems reveals little agreement in grading concussion severity."
  - Echemendia, Giza, and Kutcher (2015)

Defining Concussion…

Review

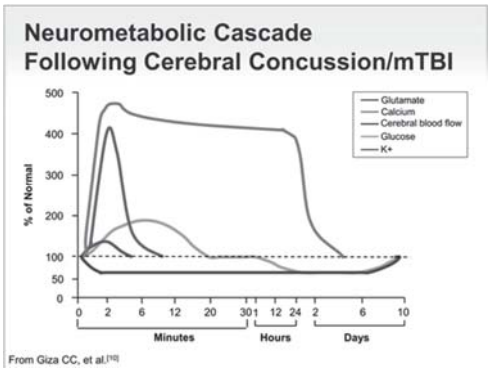
What is the definition of sports-related concussion: a systematic review

Paul McCrory,[1] Nina Feddermann-Demont,[2,3] Jiří Dvořák,[3,4] J David Cassidy,[5,6,7] Andrew McIntosh,[8,9] Pieter E Vos,[10] Ruben J Echemendia,[11,12] Willem Meeuwisse,[13] Alexander A Tarnutzer[2,3]

Defining Concussion…

- 1601 articles screened, 36 studies included
- 14 reported on criteria for SRC definitions
- 22 on biomechanical aspects of concussion
- 6 different operational definitions

- **Summary/Conclusions:** SRC is a TBI that is defined as a complex pathophysiological process affecting the brain, induced by biomechanical forces with several common features that help define its nature.



Clin Sports Med. 2011 January ; 30(1): 19–vii. doi:10.1016/j.csm.2010.08.009.

Biomechanics of Concussion

David F. Meaney, PhD[a,*] **and Douglas H. Smith, MD**[b]
[a]Department of Bioengineering, University of Pennsylvania, 240 Skirkanich Hall, 210 South 33rd Street, Philadelphia, PA 19104-6392, USA
[b]Department of Neurosurgery, University of Pennsylvania, 105D Hayden Hall, 240 South 33rd Street, Philadelphia, PA 19104-6392, USA

- Direct or Impulsive forces
- Linear and rotational forces
- 70 – 100 g of force

- Hitting your head does <u>not</u> equate concussion
- Linear/sequential recovery process
- Physiologic recovery continues after resolution of clinical symptoms

Symptoms of concussion usually fall into four categories:

Thinking/ Remembering	Physical	Emotional/ Mood	Sleep
Difficulty thinking clearly	Headache	Irritability	Sleeping more than usual
	Fuzzy or blurry vision		
Feeling slowed down	Nausea or vomiting (early on)	Sadness	Sleep less than usual
	Dizziness		
Difficulty concentrating	Sensitivity to noise or light	More emotional	Trouble falling asleep
	Balance problems		
Difficulty remembering new information	Feeling tired, having no energy	Nervousness or anxiety	

https://www.cdc.gov/traumaticbraininjury/symptoms.html

Non-specific Symptoms

- Symptoms of concussion have large overlap with:
  - Sickness (e.g., cold)
  - Poor sleep
  - Stress
  - Anxiety
  - Depression

General Symptom Resolution Trajectory

- Resolution of clinical symptoms from self-report and objective testing typically 1-2 weeks with age moderation
- Physiologic recovery as demonstrated by MRS, fMRI, qEEG, etc. is variable and outlasts clinical recovery, but latter recovery is 45 days to 3 months typically.
  - Kamins et al., 2017
- As a provider:
  - Linear/sequential recovery process, symptoms do not wax and wane
    - Consideration of premorbid/concomitant factors for prolonged recovery
    - Exception is symptom exacerbation with physical exertion in acute recovery period
  - Symptom report in acute recovery period is most reliable

Neuropsychology
2005, Vol. 19, No. 3, 283–296

Neurocognitive Outcomes and Recovery After Pediatric TBI: Meta-Analytic Review of the Literature

Talin Babikian and Robert Asarnow
David Geffen School of Medicine at UCLA

Figure 2. Summary diagram of trends in neurocognitive outcomes and recovery over time.

Neuropsychology
2014, Vol. 28, No. 3, 321–336

The Neuropsychological Outcomes of Concussion: A Systematic Review of Meta-Analyses on the Cognitive Sequelae of Mild Traumatic Brain Injury

Justin E. Karr, Corson N. Areshenkoff, and Mauricio A. Garcia-Barrera
University of Victoria

- Single mTBI vs. multiple mTBI very small differences (d = .06)
  - Limited to trivial cumulative impact
- Executive functions most susceptible to multiple mTBI
  - White matter maturation occurs last in frontal lobes
- Yet to identify threshold (e.g., 5th concussion) that predicts longstanding neuropsychological impairment

- The long term cumulative effects of concussion regarding cognition is a contentious research topic:
  - Some reviews find negligible impairments or inconclusive findings
    - Karr, Areshenkoff, & Garcia-Barrera, 2014;
    - Solomon, Ott, & Lovell, 2011;
    - Yumul & McKinlay, 2016
  - While others show long-term cognitive effects from repeated concussion primarily related to elite athlete status
    - Manley et al., 2017
    - Vos, Nieuwenhuijsen, & Sluiter, 2018

Concussions vs. Repetitive Sub-Concussive Impacts

- High contact athletes (football) perform worse than low contact athletes (basketball, baseball, soccer, wrestling, volleyball, paddling, and cheerleading) on ImPACT testing.
  - Tsushima et al. (2016)
- High contact (lineman) youth football players perform worse than low contact (receivers and defensive backs) players on ImPACT testing.
  - Tsushima et al. (2017)

Concussions vs. Repetitive Sub-Concussive Impacts

- Exposure to <u>contact</u> football before or after age 12
  - >2 times increased odds for problems with behavioral regulation (e.g., easily angered), apathy, and executive function (e.g., organizing/planning)
  - >3 times increased odds for depression
    - Alosco et al., 2017

1-time NP Consultation as PCS Intervention

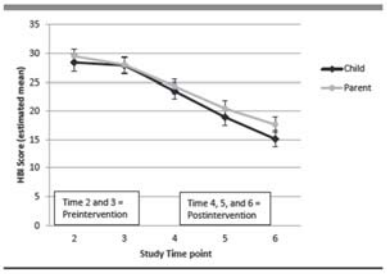
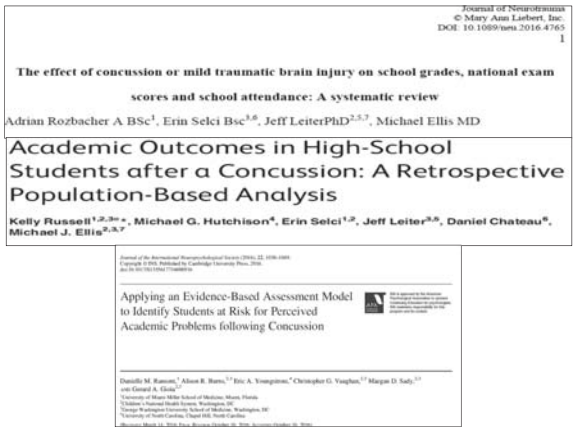


Figure 2. Child and parent estimated postconcussive symptom mean scores across time for the HBI.

Kirkwood et al., 2016



The effect of concussion or mild traumatic brain injury on school grades, national exam scores and school attendance: A systematic review

Adrian Rozbacher A BSc[1], Erin Selci Bsc[3,6], Jeff LeiterPhD[2,5,7], Michael Ellis MD

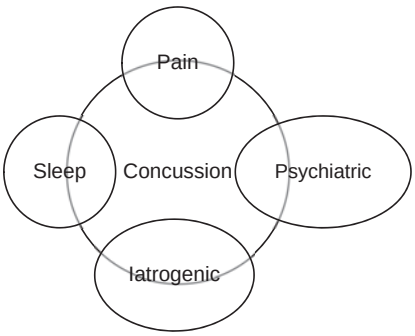
Academic Outcomes in High-School Students after a Concussion: A Retrospective Population-Based Analysis

Kelly Russell[1,2,3]*, Michael G. Hutchison[4], Erin Selci[1,2], Jeff Leiter[3,5], Daniel Chateau[5], Michael J. Ellis[2,3,7]

Applying an Evidence-Based Assessment Model to Identify Students at Risk for Perceived Academic Problems following Concussion

- Minimal impact on school grades, national exam scores, and graduation rates at a group level.
- PCS symptoms and self-reported executive dysfunction more predictive of poor school performance than cognitive testing.
- Concussion team at school still very important for reintegration into school following rest.
- How much does missed school matter?

Adapted from Iverson et al. (2008)



Review

OPEN ACCESS

Predictors of clinical recovery from concussion: a systematic review

Grant L Iverson,[1,2] Andrew J Gardner,[3] Douglas P Terry,[1,2] Jennie L Ponsford,[4] Allen K Sills,[5] Donna K Broshek,[6] Gary S Solomon[7]

Best Predictors of Outcome in Concussion

- **Age**: mixed findings
- **Sex**: mixed findings
- **Prior Concussions**: mixed findings
- **Migraine**: mixed findings
- **ADHD, LD, etc.:** minimal support
- **LOC**: minimal support
- **PTA**: minimal support
- **Headache (post-injury)**: worse outcomes
- **Total symptom report**: strong evidence of worse outcomes
- **Mental health history**: strong evidence of worse outcomes

Cerebrum, December 2016

Finding the Hurt in Pain

By Irene Tracey, Ph.D.

- Co-morbid problems like depression, anxiety, and sleeplessness are inherent in chronic pain.
- The brain responds to 'painful' or nociceptive events in a host of brain regions/ circuits in a flexibly accessible manner:
  - Sensory
  - Discriminatory
  - Emotional/affective
  - Cognitive/decision making
  - Brainstem modulatory
  - Motor

- **People have higher ratings of pain when sad**
  - Higher activations in emotional regulatory circuitry (e.g., orbitofrontal cortex)

➔

  - higher pain processing activation (e.g. amygdala, insula, inferior frontal gyrus, anterior cingulate).

- **People have higher ratings of pain when anxious**
  - hippocampus/entorhinal complex with interactions to the anterior insula and mid anterior cingulate

➔

  - higher pain processing activation (e.g. amygdala, insula, inferior frontal gyrus, anterior cingulate).

Depression, anxiety, and threat act as a physiological amplifier for pain.

- **Descending Pain Modulatory System (DPMS)**
  - Inhibitory and facilitatory modulatory action largely based upon expectation
  - Healthy controls given intravenous painkiller during brain-imaging study while given painful stimuli throughout.
    - Hidden injection
    - Positive expectation
    - Negative expectation

Original Investigation

Factors Associated With Concussion-like Symptom Reporting in High School Athletes

Grant L. Iverson, PhD; Noah D. Silverberg, PhD; Rebekah Mannix, MD, MPH; Bruce A. Maxwell, PhD; Joseph E. Atkins, PhD; Ross Zafonte, DO; Paul D. Berkner, DO

IMPORTANCE Every state in the United States has passed legislation for sport-related concussion, making this health issue important for physicians and other health care professionals. Safely returning athletes to sport after concussion relies on accurately determining when their symptoms resolve.

OBJECTIVE To evaluate baseline concussion-like symptom reporting in uninjured adolescent student athletes.

DESIGN, SETTING, AND PARTICIPANTS In this cross-sectional, observational study, we studied 31 958 high school athletes from Maine with no concussion in the past 6 months who completed a preseason baseline testing program between 2009 and 2013.

RESULTS Symptom reporting was more common in girls than boys. Most students with preexisting conditions reported one or more symptoms (60%-82% of boys and 73%-97% of girls). Nineteen percent of boys and 28% of girls reported having a symptom burden resembling an International Classification of Diseases, 10th Revision (ICD-10) diagnosis of postconcussional syndrome (PCS). Students with preexisting conditions were even more likely to endorse a symptom burden that resembled PCS (21%-47% for boys and 33%-72% for girls). Prior treatment of a psychiatric condition was the strongest independent predictor for symptom reporting in boys, followed by a history of migraines. For girls, the strongest independent predictors were prior treatment of a psychiatric condition or substance abuse and attention-deficit/hyperactivity disorder. The weakest independent predictor of symptoms for both sexes was history of prior concussions.

CONCLUSIONS AND RELEVANCE In the absence of a recent concussion, symptom reporting is related to sex and preexisting conditions. Consideration of sex and preexisting health conditions can help prevent misinterpretation of symptoms in student athletes who sustain a concussion.

JAMA Pediatr. doi:10.1001/jamapediatrics.2015.2374
Published online October 12, 2015.

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Corresponding Author. Grant L. Iverson, PhD, Center for Health and Rehabilitation, Department of Physical Medicine and Rehabilitation, Harvard Medical School, Flyfish 10th St, Charlestown Navy Yard, Charlestown, MA 02129 (giverson@mgh.harvard.edu).

The Database

- 32,855 student athletes from the state of Maine
- Age: 13-18
- No athlete reported sustaining a concussion in the past 6 months.

Iverson et al., 2015

ICD-10 Criteria for Postconcussional Syndrome

- Must endorse symptoms in at least 3 domains
  - Physical
  - Emotional
  - Cognitive
  - Insomnia
  - Other domains not considered: Excessive worry over symptoms and intolerance for alcohol.

Iverson et al., 2015

What percentage of boys and girls meet ICD-10 Criteria for a Mild Post-Concussional Syndrome During Baseline Preseason Testing?

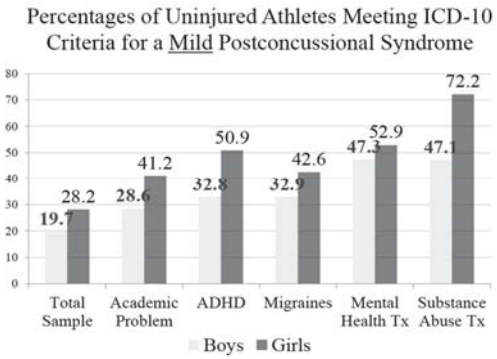
- Boys = 19.7%
- Girls = 28.2%

Iverson et al., 2015

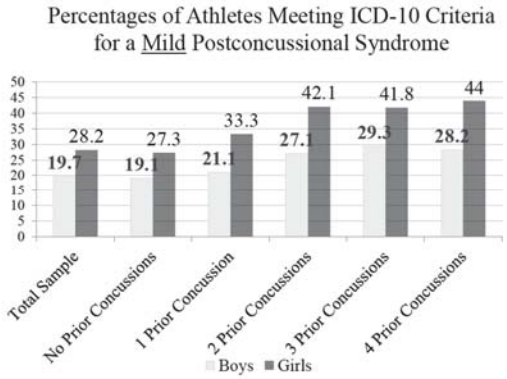
What percentage of boys and girls meet ICD-10 Criteria for a Moderate-Severe Post-Concussional Syndrome During Baseline Preseason Testing?

- Boys = 4.4%
- Girls = 7.2%

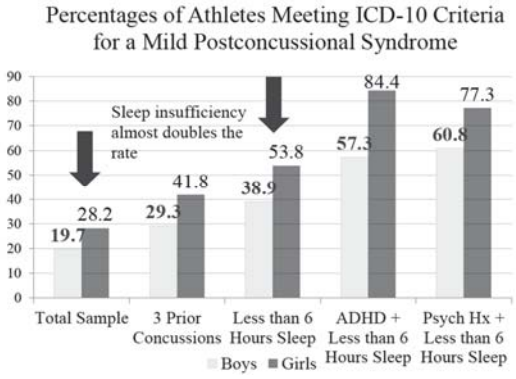
Iverson et al., 2015



Iverson et al., 2015



Iverson et al., 2015



Iverson et al., 2015

Article

Prolonged Activity Restriction After Concussion: Are We Worsening Outcomes?

Marc DiFazio, MD[1], Noah D. Silverberg, PhD[2,3], Michael W. Kirkwood, PhD[3,4], Raquel Bernier, MD[5], and Grant L. Iverson, PhD[6,7,8]

Clinical Pediatrics
2016, Vol. 55(5) 443–451

DOI: 10.1177/0009922815588914

cpj.sagepub.com

- The most methodologically rigorous studies to date have not demonstrated benefit of an initial period of 5 to 6 days of complete rest over an earlier return to activity.
- Authors could not find studies suggesting that thinking, reading, or studying cause neurometabolic demands, or changes in the brain that could be harmful.

Harmful Effects?

- Nocebo effect
  - Remember the DPMS
  - Priming effects
- Activity Restriction Model of Depression
- Physical Deconditioning

- **Conclusion**: Gradual/graded return to normal life activities following 2-3 days in most cases.

- Similarly, a more recent systematic review concluded 24-48 hours of cognitive and physical rest is a appropriate for most patients.
  - Schneider et al., 2017

Additional Psychological Factors Related to Recovery

- **Coping Style/Illness Perception**
  - Anderson & Fitzgerald, 2018
- **Good Old Days Bias**
  - The tendency to underestimate pre-injury problems and overestimate pre-injury health.
    - Brooks et al., 2014
- **Cogniphobia**
  - Avoidance of mental exertion out of a fear of developing or exacerbating a headache.
    - Silverberg, Iverson, & Panenka, 2017
- **Diagnosis Threat**
  - Form of stereotype threat - reduced cognitive/academic performance due to beliefs or reminders following a neurologic injury.
    - Fresson, Dardenne, & Meulemans 2018

Review

Traumatic Brain Injury, Sleep Disorders, and Psychiatric Disorders: An Underrecognized Relationship

Anne M. Morse [1,*] and David R. Garner [2]

1 Janet Weis Children's Hospital, Department of Pediatric Neurology and Sleep Medicine, Geisinger Medical Center, MC 14-12, 100 N Academy Blvd, Danville, PA 17822, USA
2 Department of Pediatrics, Geisinger Medical Center, Danville, PA 17822, USA; Drgarner@geisinger.edu
* Correspondence: amorse@geisinger.edu

Received: 28 December 2017; Accepted: 5 February 2018; Published: 15 February 2018

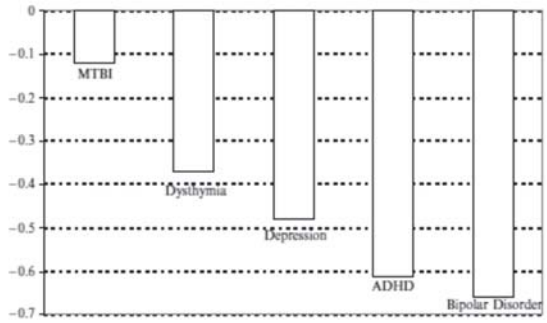


Fig. 22.3 Effects of MTBIs and various psychiatric conditions on neuropsychological functioning. MTBI (Binder et al. 1997), 11 studies, $n=314$ MTBI, $n=308$ controls; Dysthymia, Depression, and Bipolar Disorder (Christensen et al. 1997), 3 comparisons for dysthymia, 97 comparisons for depression, and 15 comparisons for bipolar disorder; ADHD (Frazier et al. 2004), based on Full Scale IQ, 123 studies

Sleep and Mental Health – Blake et al., 2017

- 30-40% of US youth experience inadequate sleep
- 30% have a sleep disorder
  - Insomnia
  - Delayed Sleep Phase Disorder
- Pervasive in psychiatric disorders
  - Share highest % of connected symptoms within all symptoms of DSM-IV

- May precipitate and maintain psychiatric conditions

  – ↓ Sleep → ↑ Anxiety & Depression

  MORE THAN

  – ↑ Anxiety & Depression → ↓ Sleep

Sleep and General Health

- Sleep deprivation increases risk of:
  - Illness susceptibility (4x increase of cold less than 6 hours)
  - Orthopedic injuries
  - <8 hours 2x increase in concussion rates in youth
  - Lifestyle disease (e.g., diabetes, obesity, heart disease)
  - Dementias
    - 60% of Alzheimer's patients have sleep disorder that preceded diagnosis by several years
  - Mortality
    - Decades decrease in life expectancy with chronic sleep deprivation

Sleep and TBI risk

- Sleep deprivation hinders:
  - Reaction time
  - Judgment
  - Balance
  - Coordination
  - Proprioception
  - General cognition (learning, memory, problem solving, etc.)

Sleep Disturbance Following Concussion

- 30-70% report sleep difficulties 1-3 weeks post-injury
  - Hypersomnia is common
- Following acute phase of recovery
  - 30% report insomnia
  - Approximately 40% can have circadian rhythm shift (delayed)
  - 40-70% report fatigue
  - 30% report sleep apnea
- The pattern and time frame of sleep disturbance may vary substantially among patients who have sustained a concussion.
  - Mosti, Spiers, & Kloss, 2016

Sleep and Concussion

- Subjective sleep complaints are 3x more likely to develop concomitant headaches in the first 6 weeks following an MTBI.
  - Also more likely to have depressive symptoms and irritability.
    - Chaput et al., 2009
- Sleep disturbance in the acute TBI period was associated with increased symptoms of depression, anxiety and apathy (mild TBI group only) 12 months post-injury.
    - Rao et al., 2014
- In fact, sleep disturbance, even in the acute post-TBI period, predicted the development of anxiety and depression in the chronic period for all severities of TBI.
    - Morse & Garner, 2018

Sleep and Concussion

- Switching between sleep and wake is complex:
  - Ventrolateral preoptic nucleus
    - γ-aminobutyric acid and Galanin producing neurons that, when stimulated, are responsible for normal sleep
  - Posterior lateral hypothalamus
    - Produces orexin
  - Tuberomammillary nucleus
    - Releases histamine
  - Dorsal Raphe Nucleus
    - Produces Serotonin
  - Locus Coeruleus
    - Produces Noradrenaline
- Similar "switches" regulate the transitions between NREM and REM sleep

Sleep & Concussion

- Exact mechanisms by which concussion affects sleep are not yet fully understood.
- Disturbances in orexin, serotonin, histamine, and noradrenaline have all been proposed as potential mechanisms for concussion-induced sleep dysregulation.
- In addition, neuro-inflammation and disturbances in the newly described glymphatic pathway could also play a role in the concussion-sleep disturbance relationship.

Consultation & Management Model

Brain Inj. 2015 ; 29(2): 195–206. doi:10.3109/02699052.2014.965210.

Multimodal Evaluation and Management of Children with Concussion: Using our heads and available evidence

Gerard A. Gioia, Ph.D
Division of Pediatric Neuropsychology, Children's National Health System, Departments of Pediatrics and Psychiatry & Behavioural Medicine, George Washington University School of Medicine

Generally

- Set positive and realistic expectation!
  - Expectancy effect
  - Importance of early education – well validated intervention
  - Null effects for cognitive rehabilitation per 2 systematic reviews and empirical support for vision therapy is tenuous
- Resume normal activities as soon as reasonably possible, including light exercise!
- Reinforce progress!
  - Prolonged symptom pacing recommendations = iatrogenic

JAMA | Original Investigation

Association Between Early Participation in Physical Activity Following Acute Concussion and Persistent Postconcussive Symptoms in Children and Adolescents

Anne M. Grool, MD, PhD; Mary Aglipay, MSc; Franco Momoli, PhD; William P. Meehan III, MD; Stephen B. Freedman, MDCM, MSc; Keith Owen Yeates, PhD; Jocelyn Gravel, MD; Isabelle Gagnon, PhD; Kathy Boutis, MD; Willem Mouwitsse, MD, PhD; Nick Barrowman, PhD; Andrée-Anne Ledoux, PhD; Martin H. Osmond, MDCM; Roger Zemek, MD; for the Pediatric Emergency Research Canada (PERC) Concussion Team

- Prospective, multicenter cohort study (9 EDs)
  - 5-18 cohort (average was 12)
  - 2413 participants (40% female)
- Physical activity participation and PCS severity were rated using standardized questionnaires in the ED and at days 7 and 28 post-injury.
- Physical activity within 7 days of acute injury compared with no physical activity was associated with reduced risk of PCS at one month.

JAMA Pediatrics | Original Investigation

Early Subthreshold Aerobic Exercise for Sport-Related Concussion A Randomized Clinical Trial

John J. Leddy, MD; Mohammad N. Haider, MD; Michael J. Ellis, MD; Rebekah Mannix, MD; Scott R. Darling, MD; Michael S. Freitas, MD; Heidi N. Suffoletto, MD; Jeff Leiter, PhD; Dean M. Cordingley, MSc; Barry Willer, PhD

- N = 103
  - (aerobic exercise: n = 52; 24 female [46%]; stretching, n = 51; 24 female [47%])
- Exercise group seen a mean (SD) of 4.9 (2.2) days after SRC
- Stretching group seen a mean (SD) of 4.8 (2.4) days after SRC
- No differences in age, sex, previous concussions, time from injury, initial symptom severity score, or initial exercise treadmill test and physical examination results.
- Exercise recovered in a median of 13 (IQR = 10-18.5) days
- Stretching recovered in a median of 17 (IQR = 13-23) days
  - (P = .009 by Mann-Whitney test)
- Nonsignificant lower incidence of delayed recovery in the aerobic exercise group (2 participants [4%] in the aerobic group vs 7 [14%] in the placebo group; P = .08).

Neuropsychology Service in Family Med/Sports Med

- **Evaluation**:
  - Half day and full day evaluations
    - Concussion/mTBI
    - Neurodevelopmental disabilities
    - General neurological conditions

OHSU Concussion Program

- **Concussion Treatment Clinic**:
  - The concussion follow-up clinic: 3-6 sessions
  - Partnership of ATC, NP, Sports MD
    - ATC: treadmill test, sensory/motor intervention
    - NP: sleep protocol, behavioral activation, exposure

	5/3/2017	5/9/2017	5/16/2017	6/6/2017
PHQ-15 (Somatization Symptoms)	17 (Severe)	13 (Moderate)	8 (Mild)	3 (No clinical concern)
GAD-7 (Anxiety Symptoms)	13 (Moderate)	10 (Moderate)	9 (Mild)	2 (No clinical concern)
PHQ-9 (Depressive Symptoms)	21 (Severe)	20 (Severe)	11 (Moderate)	2 (No clinical concern)
PSQI (Sleep Quality)	9 (>poor quality)	-	-	6 (>5 poor quality)

	6/12/2017	6/19/2017	7/11/2017
PHQ-15 (Somatization Symptoms)	11(Moderate)	10 (Moderate)	7 (Mild)
GAD-7 (Anxiety Symptoms)	8 (Mild)	6 (Mild)	2 (No clinical concern)
PHQ-9 (Depressive Symptoms)	9 (Mild)	4 (No clinical concern)	2 (No clinical concern)
PSQI (Sleep Quality)	8 (>5 poor quality)	-	6 (>5 poor quality

Practical take homes

- Pre-injury mental health and sleep quality will predict outcomes
- High acute symptom burden (particularly headache), onset of sleep dysregulation and/or activity withdrawal will prolong recovery
  - Dr. Herring's perspective on disability
- Early exercise and sleep intervention will likely improve clinical outcome
- Returning to normal daily activities (physical, recreational, social) as soon as possible (2-3 days), often gradually/incrementally, will likely improve clinical outcome

Practical take homes

- Linear/sequential recovery process, symptoms do not wax and wane
  - Consideration of premorbid/concomitant factors for prolonged recovery
  - Exception is symptom exacerbation with physical exertion in acute recovery period
- Symptom report in acute recovery period is most reliable
- Consider the person who sustained the concussion, not just persistent symptoms through the medical lens.
  - The more distal from injury, consider referring to a mental health therapist rather than a rehabilitation therapist.