DISCLOSURES

- Nothing relevant to disclose
INTRODUCTION

- Recently published research
- Ongoing research efforts
- Ongoing studies at CHOC
PRACTICAL QUESTIONS

• What is the best treatment for IS? Does it vary by etiology?
• How can we diagnose the underlying cause of IS?
• Which patient populations are high-risk for developing IS?
• Can we predict which children with IS will thrive and which will not?
FASCINATING QUESTIONS

• Why do these seizures only happen in infants?
• How can over 200 difference causes all result in the same EEG pattern and the same seizure type?
• Why does the whole brain often show hypsarrhythmia and then improve all at once, even with focal lesions?
• What causes hypsarrhythmia and spasms, biologically?
PREVIOUS RESEARCH

• Important factors for the best long-term outcomes
  • Short time from onset to treatment
    • Delaying diagnosis by even a few weeks can have developmental implications
  • Appropriate and efficient successful treatment
  • Underlying etiology - Better long-term outcome in patients with normal testing
CURRENT LARGE-SCALE RESEARCH

• Pediatric Epilepsy Research Consortium (PERC), part of American Epilepsy Society
  • National, multicenter epidemiological study (>400 patients to date)
  • Enrolling at time of diagnosis and following for a year
  • Trying to answer the big questions: Causes, treatment success rates, etc.

http://www.pediatricerc.com
PERC-RELATED PUBLICATIONS

• How Should Children With West Syndrome Be Efficiently and Accurately Investigated? Results from the National Infantile Spasms Consortium. Epilepsia 2015.

• 251 infants enrolled over 2 years
  • Causes found in 64% (161); 138 of which by Hx/Px/MRI, and 23 by lab testing
    • 14% genetic
    • 10% genetic-structural (i.e. tuberous sclerosis)
    • 11% structural-congenital
    • 22% structural-acquired
    • 5% metabolic
    • 2% infectious
  • Recommendation: Initial eval, MRI brain. If negative, then chromosomalCGH microarray followed by epilepsy gene panel, metabolic testing.
PERC-RELATED PUBLICATIONS

  • Responders: Clinical cessation of spasms and resolution of hypsarrhythmia for 3+ months
  • Standard treatments (46% response): ACTH, oral corticosteroids, vigabatrin
    • ACTH: 55% response
    • Oral steroids: 39% response
    • Vigabatrin: 36% response
  • Non-standard treatment response rate: 9%
RESEARCH AT CHOC

• 3 recent studies
  • PERC national study
  • Retrospective study – In collaboration with UC Irvine Biomedical Engineering
    • Will soon be expanded from 21 patients to 75+ patients in collaboration with UCLA
  • Prospective study – In collaboration with UC Irvine Biomedical Engineering
    • Recruitment ongoing
RETROSPECTIVE STUDY

- Overview
- Goals/hypotheses
- Findings
- Significance
Controls (n=21)

Pre-treatment spasms with hypsarrhythmia (n=21).
post-treatment, no hypsarrhythmia (n=17)

post-treatment, persistent hypsarrhythmia (n=4).
PROSPECTIVE STUDY

- Overview
- Goals/hypotheses
- Current progress
HIGH FREQUENCY ACTIVITY IN THE HUMAN BRAIN

In epilepsy, 80-250 Hz = “ripple”
NEW HFO DETECTOR BY LOPOUR LAB AT UCI

Background

HFOs
UC IRVINE RESEARCH COLLABORATORS

Beth Lopour, PhD
UCI Faculty

Rachel Smith
UCI Graduate Student

Amanda Sugijoto
UCI Undergraduate