Landau-Kleffner syndrome: An exploration of parent experience of the diagnostic process

Presented by Daunette Lemard-Reid, PhD, MSPH, MPAS

Landau-Kleffner syndrome

- Rare childhood disorder
  - Seizures
  - Loss of response to spoken words
- Often misdiagnosed
- Sleep EEG important

Research Problem

- LKS misdiagnosis
  - Restricts children's potential
  - Lowers quality of life
  - Preventable
  - Underrecognized

Research methods

- Medical model
- Semistructured interviews
- Online support group

•7 women

•6 boys, 1 girl

•5 rural, 2 suburban, 1 urban

Results

Parents reported their experience included

- Frustration
- Poor communication
- Multiple barriers
- Becoming advocates

Misdiagnosis was prevalent

- Childhood seizure disorders
- Developmental delay
- PDD-nos
- Autism
- Mental retardation

Sleep EEG was required for LKS diagnosis

- 6 children diagnosed by EEG
- 1 child diagnosed by return of language with seizure control
- 5 lay persons were first to suspect diagnosis
- Diagnosis usually made by neurologist

Treatment for LKS included

- •Antiepileptics
- •Steroids
- –Best response
- •IVIG
- •Speech therapy

Lack of awareness greatest barrier to LKS diagnosis

- Prevented effective communication between parents and providers
- Caused doctors to
  - Miss the diagnosis
  - Fail to recognize symptoms even when children presented with classic LKS signs
  - Ignore inconsistent symptoms
  - Blame parents for the child's illness behaviors
  - Forced parents into advocacy role

Socioeconomic status contributed to misdiagnosis

- Geographic barriers
  - Distance to appointments
  - Appointment delays
  - Lack of skilled resources

- Financial barriers
    - Out of pocket expenses

Contribution to the literature

- First to describe LKS from parent point of view
- Support findings of failure to refer for EEG in absence of overt seizures (Sarco & Takeoka, 2009)
- Support conclusions regarding the importance of early LKS diagnosis and treatment of LKS (Thomsen (2007); Garcia-Penas (2010); Fandino, Connolly, Usher, Palm, & Kozak, 2011)

Summary

LKS is characterized by seizures in the speech center and loss of response to spoken words. It can be misdiagnosed as autism, benign childhood seizure disorders, or mental retardation. Early diagnosis using a sleep EEG and neurodevelopmental screening is vital to promoting early treatment allowing addition of steroid to seizure medications and promoting functional language development after seizure recovery. The difficulty distinguishing LKS from its confounding syndromes is overshadowed by limited provider awareness of its presence or the overlapping presentation with other developmental syndromes. This lack of awareness can prevent or delay LKS diagnosis in the presence of aggressive parent advocacy and long established EEG criteria for diagnosis. Public health professionals must act to improve LKS awareness among pediatric medical providers because of the far-reaching personal and societal consequences of prolonged epileptiform activity.