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Current Issues in Childhood Apraxia of Speech

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Medical Management of Children with Childhood Apraxia of Speech

Presented By:

Amy Newmeyer, M.D.

Moderated By:

Amy Hansen, M.A.,CCC-SLP, Managing Editor, SpeechPathology.com

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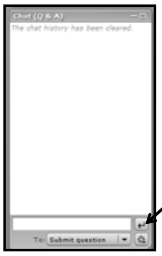

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Medical Management of Children with Childhood Apraxia of Speech

Amy Newmeyer, M.D.
Clinical Associate Professor of Pediatrics
Nationwide Children's Hospital

Overview

- Introduction
- Review of common medical complications seen in children with CAS
- Review reasons to consider referral of a child with CAS for neurological evaluation
- Review the medical testing protocol used to assess a child with CAS

Introduction

From a Neurological Perspective: What is Apraxia?

- Apraxia is defined as an abnormality in the planning and performing of movements
- Apraxia can affect any type of motor skill which requires sequencing of a series of movements
- Apraxia comes from the Greek word "a" meaning "to do without," and the Greek word "pratto" meaning "to do", which translates into "without being able to do."

What is Apraxia?

- Ideational Apraxia
  - Ideational apraxia is a disturbance of voluntary movement in which a person misuses objects due to difficulty identifying the concept or purpose behind an object
- Ideomotor Apraxia
  - Ideomotor apraxia is the inability to carry out a command to mimic limb or head movements performed or suggested by others

What is Apraxia?

- Buccofacial apraxia (facial or oral apraxia)
  - the inability to coordinate and carry out facial and lip movements such as whistling, winking, coughing etc on command.
- Verbal apraxia (childhood apraxia of speech)
  - A form of buccofacial apraxia which affects the ability to coordinate mouth movements to produce speech sounds accurately

ASHA 2007 Definition

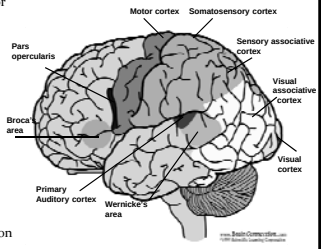
- Preferred terminology is now Childhood Apraxia of Speech (CAS)
- Key components of definition:
  - Neurological childhood speech sound disorder
  - Affects precision and consistency of movements that affect speech sound production and prosody
  - Occurs in the absence of other neuromuscular deficits (e.g. abnormal reflexes, abnormal tone)
  - For further information about the recent position statement visit www.asha.org

Etiology of CAS

- *Known neurological impairment, occurs with co-occurring genetic or neurological conditions*
- Idiopathic neurogenic speech sound disorder

Neurological Basis of CAS

- Components of the Audio-visual-motor Network
  - Posterior superior temporal cortex (involved in action control as interface between sensory and motor areas)
  - Frontal motor cortex (dorsolateral prefrontal, SMA, premotor and primary motor cortex) required to instigate movement
  - Pars opercularis (BA 44)
  - These areas are linked by direct connections, as well as circuits through the basal ganglia and cerebellum
    - Attentional and working and short-term memory processes also likely involved in completion of a motor action



Medical Complications in CAS

Medical Complications in CAS

Genetic Disorders

Genetic Disorders and CAS

- CAS has been identified in the following known genetic conditions:
  - Down Syndrome
  - Fragile X
  - Galactosemia
  - Joubert Syndrome
  - Velocardiofacial Syndrome

Down Syndrome

- Due to trisomy of chromosome 21
- Characterized by:
  - Intellectual Disability
  - Hypotonia
  - Childhood Apraxia of Speech

Other Syndromes

- Fragile X
  - Due to CGG repeat on X chromosome so males are most affected
  - Common Symptoms:
    - Intellectual Disability
    - Behavior Problems
    - Speech Disorder
  - Females are carriers and can have some of the phenotypic features of intellectual disability
- Galactosemia
  - Enzyme defect in breaking down Galactose
  - Common symptoms:
    - Speech Disorder
    - Ataxia
    - Diminished bone density
    - Cataracts

Other Syndromes

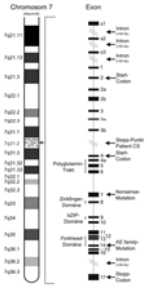
- Joubert Syndrome
  - Gene defect causes abnormal development of the cerebellum
  - Symptoms include:
    - Ataxia
    - Seizures
    - Speech Disorder
    - Intellectual Disability
- Velocardiofacial Syndrome
  - Due to deletion on chromosome 22q11.2
  - Symptoms include:
    - Cleft palate
    - Learning disability
    - Speech Disorder
    - Heart abnormalities

Other Less Common Genetic Mutations Associated with CAS

- FOXP2 mutations and deletions
- Chromosome 7 deletions and translocations
- Chromosome 4 deletions and translocations
- Rett Syndrome

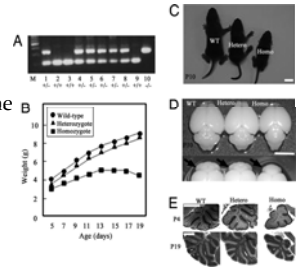
FOXP2 Mutation

- The FOXP2 mutation was first described in 1995 in a family in the UK
- FOXP2 is located on chromosome 7q31and is extensively expressed in the developing brain
- Appears to impact corticostriatal and corticocerebellar curcuits
- FOXP2 linked to gray matter abnormalities on brain MRI



FOXP2 Mutation

- Animal studies suggest that FOXP2 is involved in the development of circuits necessary for the motor planning of speech



Medical Complications in CAS

Other developmental disorders

Developmental Dyspraxia

- The Dyspraxia Foundation defines developmental dyspraxia as "an impairment or immaturity of the organization of movement. It is an immaturity in the way that the brain processes information, which results in messages not being properly or fully transmitted".
- "The term dyspraxia comes from the word praxis, which means 'doing, acting'. Dyspraxia affects the planning of what to do and how to do it. It is associated with problems of perception, language and thought".

Developmental Dyspraxia

- Also referred to as "developmental coordination disorder"
- Affects about 5 to 10% of school age children
- Can manifest as difficulties with handwriting and/or sports and recreational activities

Apraxia Clinic Data

- 181 patients were seen in the clinic between 7/03 and 6/07
  - 155 diagnosed with CAS in clinic
  - 43 females and 138 males
  - Race:
    - 2.8% Asian
    - 6.7% African-American
    - 87% Caucasian
    - 3.3% other
  - Age:
    - Avg. 55 months (21-171)

Findings from Apraxia Clinic

- Kaufman Speech Praxis Test
  - Oral Movements Standard Score 77 (14-110)
  - Simple Phonemic/Syllabic Level Standard Score 34 (0-106)
  - Complex Phonemic/Syllabic Level Standard Score 50 (6-84)
- Preschool Language Scale
  - Auditory Comprehension Standard Score 93 (50-125)
  - Expressive Communication Standard Score 77 (50-119)

Findings from Apraxia Clinic

- Peabody Developmental Motor Scales
  - Grasping Standard Score 7.1 (3-13)
  - Object Manipulation Standard Score 8.0 (4-12)
  - Visual Motor Integration Standard Score 8.1 (1-15)
- Sensory Profile also reviewed
  - Results indicated a difference from same age peers in a number of areas, including:
    - Fine Motor/Perceptual Skills
    - Behavioral Outcomes of Sensory Processing

Analysis of Preschool Children Ages 3 to 6 yrs

- Analysis done of 42 patients ages 3 to 6 year old (avg. age 44 months) who had both the KSPT and PDMS
- 8 females and 34 males studied
- In this analysis we controlled for presence of hypotonia and other developmental issues (cognitive disability, seizures, autism)
- A moderate correlation was found between a low score on the Oral Movement subtest of KSPT and a low score on PMDS visual motor integration subtest r= 0.46 (p =.0023); and for a low score on the PMDS object manipulation subtest r= 0.34 (p=0.039)
- No correlation of other components of the PDMS and simple phonemic/syllabic level subtest of the KSPT

Medical Complications of CAS

Neurological Disorders

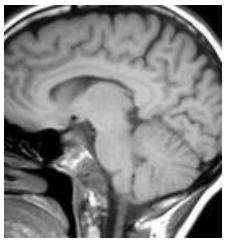
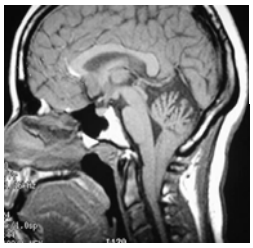
Brain Creatine Deficiency

- Brain creatine deficiency has been linked to intellectual disability and speech disorders
  - X-linked creatine transporter deficiency
    - Affects mainly males; female carriers may be symptomatic
  - Autosomal Recessive Liver enzyme defects

Chiari Malformations

- Downward displacement of the cerebellar tonsils through the foramen magnum, which may result in hydrocephalus and compression of the cerebellum
- Symptoms can include:
  - Headaches
  - Difficulty swallowing
  - Paralysis
  - Speech Disorder
- Surgical correction done if obstruction of flow of CSF and symptoms present

Chiari I malformation

Case Report

Case Report

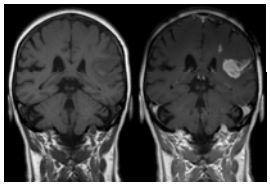
- 2 ½ year old male
- Presented for delayed speech development
- Frequent crying spells and feeding difficulties noted on review of history
- Clinical evaluation completed by developmental pediatrician, SLP and OT and subsequently diagnosed with CAS
- Brain MRI revealed Chiari I malformation

Case Report

- After decompression surgery completed, had improvement of speech
- At one year post-surgery, still receiving speech therapy
- Improvement in speech production and intelligibility

Stroke

- Also called Cerebrovascular Accident (CVA)
- Caused by acute loss of blood flow or oxygen to portion of the brain
- Can be perinatal (before or during birth) or postnatal causes
- May be related motor symptoms



Seizure Disorder

- Different Types of Seizures
  - Simple Partial Seizure
  - Complex Partial Seizure
  - Absence Seizure
  - Generalized Tonic-Clonic Seizure
  - Atonic Seizure
  - *Children with CAS may also have EEG abnormalities without seizure activity*

Seizure Disorder

- Landau Kleffner Syndrome
  - Abnormal EEG in deep stages of sleep causes sudden or gradual development of aphasia
  - Usually occurs between ages 5 and 7 years
  - Only identified in Stage 4 sleep on overnight EEG

EEG and Brain MRI Results

Role of EEG

Brain-based communication disorders
with EEG abnormalities:

- Autism/Autism spectrum disorders
- Landau-Kleffner Syndrome
- Benign Epilepsy of Childhood with Centro-Temporal Spikes
- Other epileptic syndromes

Aims of Study

Aim #1: To define the types of EEG abnormalities among subjects diagnosed with CAS.

Aim #2: To define the severity of the apraxia and describe other clinical findings among the study groups to more clearly define potential criteria for determining who should have an EEG study among children with CAS.

Methods: Criteria and Data

Inclusion Criteria
- Enrolled in IRB-approved research database
- Diagnosed with CAS

Exclusion Criteria
- Hearing loss

The database was reviewed for:
- Pertinent historical data
- Neurological exam findings
- EEGs, MRI, Speech/language testing

Methods: Language Testing

Language testing included:

- Kaufmann Speech Praxis Test, Part I: Oral Movements (KSPT)
- Receptive and Expressive language testing
  - Pre-School Language Scales—IV (PLS)
  - Clinical Evaluation of Language Fundamentals—IV (CELF)
  - Oral and Written Language Scales (OWLS)

Methods: EEGs

EEGs for this study:

- included short (routine, sleep-deprived) and prolonged (video, ambulatory) studies
- were standardized by one epileptologist
- were classified as
  - non-epileptiform
  - epileptiform—focality and morphology

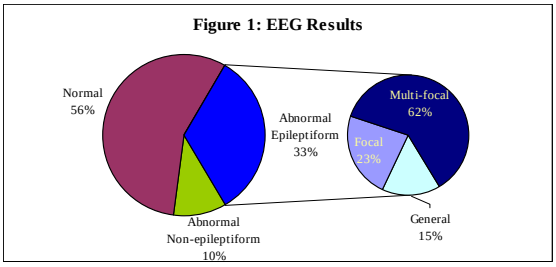
Results

Table 1: Demographics

	Abnormal EEG N = 17	Normal EEG N = 22	No EEG N = 117
Gender = Male	14 (82%)	18 (82%)	94 (80%)
Median age in Months (Range)	53 (33-148)	42 (26-114)	(21-171)
Race			
Caucasian	15 (88%)	19 (86%)	102 (87%)
African-American	1 (6%)	1 (5%)	5 (4%)
Other	1 (6%)	2 (9%)	10 (9%)

Results

Figure 1: EEG Results



Results-Neurological Examination

	Abnormal EEG N = 17	Normal EEG N = 22	No EEG N = 117
Hypotonia	6 (35%)	5 (23%)	26 (22%)
Drooling	7 (41%)	5 (23%)	13 (11%)
Abnormal reflexes	1 (6%)	2 (9%)	6 (5%)

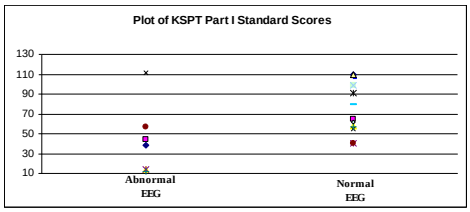
Results-MRI

	Normal EEG (N = 18)	Abnormal EEG (N = 17)	No EEG (N = 37)
Normal MRI	12	9	18
Abnormal MRI	6 (33%)	8 (47%)	19 (51%)
Congenital Malformation	2	4	10
Scar (gliosis, PVL)	2	3	3
Abnormal Myelination	1	2	5
Abnormal Cortex	1	0	1

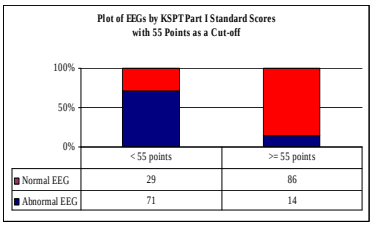
Results-Reason for Ordering EEG

Reason given	**Abnormal EEG**	**Normal EEG**
Stagnation	6	7
Staring spell	5	4
Clinical suspicion	5	6
Suspected convulsion	3	7
More than one indication	3	11
Perceived severity of apraxia	2	7
Regression	1	4
Abnormal sleep	0	1

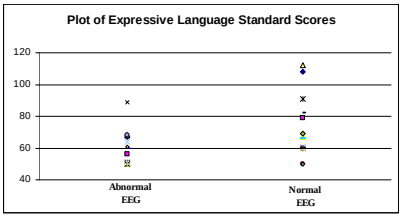
Results-EEG vs. KSPT Scores



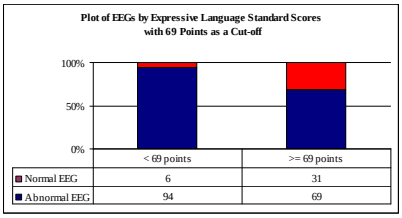
Results-EEG vs. KSPT Scores



Results-Expressive Language Scores



Results-Expressive Language Scores



	< 69 points	>= 69 points
Normal EEG	6	31
Abnormal EEG	94	69

Conclusions

- Right central area most frequently involved on EEG
  - Implicates motor and pre-motor cortex
- Abnormal EEG group had lower Expressive Language and KSPT Part I scores

Reasons to Consider Referral for Neurological Evaluation

When should a child with CAS be referred for Neurological Assessment?

- Signs of Seizures
  - Staring spells
  - Focal twitching of muscles
  - History of generalized tonic/clonic seizures
  - What about a febrile seizure?
- Loss or regression of speech and/or language skills

When should a child with CAS be referred for Neurological Assessment?

- Lack of progress with intensive therapy
- Family history of known genetic conditions
- Strong family history of speech disorders
- Suspicion of other neurodevelopmental disorders, such as intellectual disability and autism

Neurodevelopmental Testing Protocol

Neurodevelopmental Evaluation of a Child with CAS

- Past Medical History
  - Brain injury
  - seizures
  - PE tube placement
  - hearing loss
- Developmental History
  - autism spectrum disorder
  - Intellectual disability
  - language disorder
  - Gross motor delay/fine motor delay
  - Sensory processing problems

Neurodevelopmental Evaluation of a Child with CAS

- Family History
  - Language disorder
  - Apraxia or another speech disorder
  - Autism spectrum disorder
  - Learning disabilities
- Physical Examination
  - Thorough oromotor examination
  - Neurological examination including reflexes, tone
  - Observation of communication, any unusual behaviors

Neurodevelopmental Evaluation of a Child with CAS

- Medical Testing to Consider
  - Genetic testing
  - Brain MRI to rule out brain malformation, evidence of stroke or other brain injury
  - Brain MRS to rule out creatine deficiency
  - EEG
    - Particularly if there is a history of language loss or variability in acquisition of language/speech skills

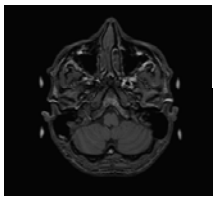
Genetic Testing

- Karyotype
- Microarray
- Fragile X

- FOXP2 not a routine test at this time, although Microarray may identify deletions or duplications on chromosome 7 in FOXP2 region

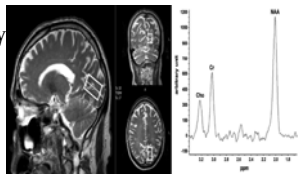
Brain MRI

- Brain MRI
  - Can help to rule out structural disorder, stroke, or other injury to developing brain
- T1 and T2 weighted images are obtained



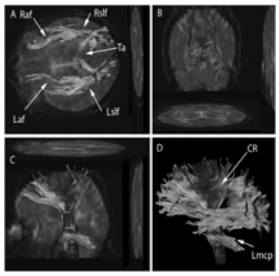
Brain MRS

- Clinical test for chemical levels in the brain
- Brain creatine deficiency has been linked to speech disorders and intellectual disability



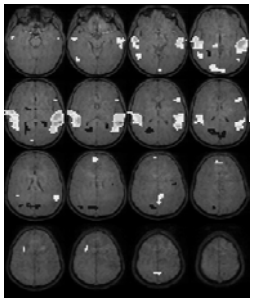
Brain DWI and DTI

- Diffusion weighted MRI (DWI) sensitive to acute stroke
- Diffusion tensor MRI (DTI) allows evaluation of white matter tracts in the brain
- Available clinically but for limited uses

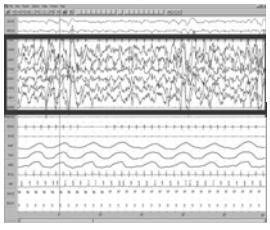


Functional MRI

- Only available for research purposes
- Allows analysis of increased blood oxygen utilization in areas of the brain during a cognitive or motor task



EEG



Questions?

Conference Schedule

Monday: ***Childhood Apraxia of Speech: An Overview and Assessment Considerations*** **- Rebecca McCauley, Ph.D., CCC-SLP**

Tuesday: ***Medical Management of Children with Childhood Apraxia of Speech*** **- Amy Newmeyer, M.D.**

Wednesday: ***Principles for Childhood Apraxia of Speech Across Childhood*** **– Shelley L. Velleman, Ph.D., CCC-SLP**

Thursday: ***Genetic and Neurological Correlates of Childhood Apraxia of Speech*** **– Barbara A. Lewis, Ph.D., CCC-SLP**

Friday: ***Current Issues in CAS: Round-Table Discussion*** **– Rebecca McCauley, Ph.D., CCC-SLP, Amy Newmeyer, M.D., Shelley Velleman, Ph.D., CCC-SLP, Barbara Lewis, Ph.D., CCC-SLP**