

Differential Diagnosis in Young Children with Suspected CAS

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Speaker Disclosure

- Financial Disclosure: Cari Ebert owns Summit Speech Therapy, LLC/Cari Ebert Seminars and draws a salary. She receives royalties from product sales on her website. Ms. Ebert also receives compensation from CASANA for presenting this course.
- Nonfinancial Disclosure: Cari Ebert has a son with autism and apraxia and shares her personal experiences in her seminars.

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Learning Objectives

At the end of this course participants will be able to:

1. Summarize why CAS can be so difficult to diagnose in very young children and when it is clinically appropriate to make the formal diagnosis.
2. Compare and contrast the clinical symptoms of suspected CAS with Autism Spectrum Disorder, Dysarthria, Phonological Disorder and Expressive Language Delay.
3. Justify a working diagnosis of sCAS in very young, minimally verbal children by documenting specific signs and symptoms consistent with a motor planning disorder.

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Root Word: Praxis

- Praxis: Greek word which means "action" or "movement"
- Components of praxis include: imitation, initiation, grading of force, sequencing, timing, and motor planning
- Apraxia is a decrease in the ability to plan and perform purposeful actions/movements
- Apraxia can occur throughout the body

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3 Primary Types of Apraxia

- **Limb Apraxia:** Refers to the inability to make precise movements with the fingers, arms or legs on command (also called *dyspraxia*)
- **Non-Verbal Oral Apraxia:** Refers to the inability to perform oral/facial movements on command
- **Verbal Apraxia:** Refers to the inability to plan, coordinate & sequence sounds necessary for speech on command (CAS in children; AOS in adults)

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What ASHA Says...

ASHA states that the oral and limb apraxias should be of great interest to Speech-Language Pathologists

Why?

1. Because the presence of oral and/or limb apraxia in a child suspected to have CAS provides support for the diagnosis, especially in preverbal or minimally verbal children
2. Because the presence of limb apraxia will preclude the use of sign language

ASHA's Definition of CAS

"Childhood Apraxia of Speech (CAS) is a neurological childhood speech sound disorder in which the precision and consistency of movements underlying speech are impaired in the absence of neuromuscular deficits...the core impairment in planning results in errors in speech sound production and prosody."

ASHA Technical Report 2007

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Apraxia or Dyspraxia?

- Look at the prefixes in medical terminology:
 - "a" means absence or total loss
 - "dys" means partial loss
- In the past some SLPs used the term Dyspraxia to refer to a milder form of verbal apraxia
- But...Dyspraxia is a term used to refer to limb apraxia and it has its own ICD-10 code

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- A child may present with characteristics of just one type of apraxia, a combination of two types of apraxia, or a child may present with characteristics of all three types of apraxia (global apraxia)
- Apraxia may be the primary diagnosis or it may be a secondary diagnosis
- When verbal apraxia occurs in adults, it is acquired
- In children, the verbal apraxia can occur congenitally or be acquired anytime during the developmental period of speech acquisition

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- ### Etiologies of Childhood Apraxia of Speech
1. Neurologic Impairment – CAS can occur as a result of infection, illness, injury, trauma or stroke (MRI will be remarkable)
 2. Complex Neurodevelopmental Disorder – CAS can occur as a secondary diagnosis of other primary diagnoses including genetic, metabolic and/or mitochondrial disorders (e.g. Autism, Fragile X, Down Syndrome, Epilepsy)
 3. Idiopathic Neurogenic Speech Sound Disorder – CAS can occur as a disorder of unknown origin which means no observable neurological abnormalities or neurodevelopmental conditions are present

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- ### Genetics and CAS
- Lai, Fisher, Hurst, Vargha-Khadem & Monaco, 2001
- Primary findings related to genetics and CAS come from studies on a London family (1/2 had oral and verbal apraxia)
 - Findings indicate a mutation on the FOXP2 regulatory gene located on chromosome 7
 - There are likely subtypes of CAS as there are some children studied with CAS who do not have the FoxP2 genetic mutation

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Who Diagnoses CAS?

- CAS is a speech sound disorder, therefore, it is diagnosed by a speech-language pathologist
- There may be supporting documentation by a pediatrician or pediatric neurologist to support the neurologic or neurobehavioral component of CAS (remember the etiologies of CAS)
- The SLP should document the atypical speech development (motoric) and explain how this differs from a developmental language delay (linguistic)

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ICD-10 Codes

The ICD-10 went into effect on 10/1/15

- Code for Childhood Apraxia of Speech (CAS): R48.2
- Code for Phonological and Articulation Disorders: F80.0
- Code for Expressive Language Disorder: F80.1
- Code for Mixed Receptive-Expressive Language Disorder: F80.2
- Code for Dysarthria (in children/non post CVA): R47.1
- Code for Dyspraxia (limb apraxia, clumsy child syndrome, developmental coordination disorder): F82

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Should We Diagnose CAS In Children Under Age 3?

SLPs should be cautious about giving a firm diagnosis of CAS prior to age 3 for two main reasons:

- First, we cannot formally diagnose CAS until the child is verbal - CAS is a speech disorder (motoric), not a language disorder (linguistic)
- Second, there is still a lot of brain development occurring prior to age 3

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Early Brain Development

Because most of brain development occurs prior to age 3, the earlier we treat these kids, the better their prognosis becomes.

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Early Brain Development

Babies are born with 100 billion neurons. Prior to age 3 the young brain must establish and reinforce connections between neurons. These connections are formed when impulses are sent and received between neurons. Axons send the messages and dendrites receive them. These connections form synapses. During the first 3 years of life the number of neurons stay the same but the number of synapses increases. (www.classbrain.com article 30)

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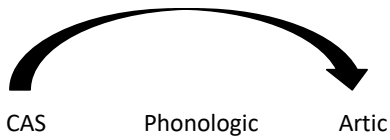
The Brain is Not Complete at Birth!

- Synapses = wiring
- Synaptic connections are created at a rapid rate through age three
- By age three, 85% of the core structures of the brain are formed
- While synapses are developing, the brain builds the potential to learn
- Repetition of appropriate interactions will help the brain reinforce existing synaptic connections & make new ones

(www.wccf.org, www.zerotothree.org, www.classbrain.com article 30)

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CAS is a Dynamic Speech Sound Disorder



Dynamic = Symptoms change over time

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CAS is a **Dynamic** Speech Disorder

“We need to remember that classifications or labels may change over time with neural maturation and appropriate treatment. For example, children with CAS often progress to the point at which speech characteristics are more appropriately labeled phonologic impairment or residual articulation errors.”

Strand & McCauley

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The Challenge of Diagnosing Very Young Children

“The complexity of diagnosis in young children under age 3 is that the child must be able to participate sufficiently in the assessment. Unless the child can attempt to imitate utterances that vary in length and phonetic complexity it is very difficult to make a definitive diagnosis.”

Dr. Strand

We do not yet have a blood test or brain scan that can lead to a clinical diagnosis of CAS. Therefore, SLPs must rely on signs and symptoms in very young, minimally verbal children.

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In order to administer a standardized apraxia test, we need 2 things:

1. A willing participant
2. A child with imitation skills

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Making a Diagnosis

- Once the child is verbal enough to participate in a formal evaluation using a standardized testing tool, it becomes easier to give an accurate diagnosis of CAS.
- Until that time, we report the characteristics, signs and symptoms of the motor planning difficulties and state that we are suspecting CAS as the cause for the lag in speech development.

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sCAS = Working Diagnosis

That's why we use the term *suspected* Childhood Apraxia of Speech (sCAS) when working with very young, minimally verbal children

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Misdiagnosing CAS

- Research has shown that many children with a diagnosis of CAS have been incorrectly diagnosed. (Davis, Jakielski, & Marquardt; ASHA 2007)
- Why is this occurring?
 - Diagnosing too young
 - Diagnosing speech disorder in children w/o speech
 - Professional other than SLP making the diagnosis
 - Lack of specific guidelines regarding when it's "ok" to make the diagnosis

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Why is it so Difficult to Diagnose Childhood Apraxia of Speech?

Question:

Why is it so difficult to correctly diagnose CAS and so easy to misdiagnose it?

Answer:

Because many of the characteristics overlap with other disorders; there is no blood test or genetic screening tool by which to make the diagnosis; CAS may be a secondary diagnosis instead of the primary diagnosis; and symptoms may change over time (Lewis et al. 2004)

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The Complexity of Speech (It's a miracle we speak at all!)

- CAS is a MOTOR disorder – and speech is the most finely tuned motor act we perform
- Speech = coordination of respiration, phonation, articulation and resonance
- Speech requires the coordination of more than 70 muscles and body parts
- "Given this complexity, even mild motor difficulties are enough to disrupt speech development."

Jennejahn & Turner

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Demographics/Prevalence

- 86% of kids with CAS have at least 1 family member with speech-language disorders & 59% of kids with CAS have at least 1 affected parent – which means there is a strong heritability factor (Velleman, 2006)
- Prevalence: estimated to be about 5% of children with speech sound disorders present with CAS (Delaney & Kent, 2004; apraxia-kids.org; Strand, 2010)
- ASHA reports that *“as with several other complex neurobehavioral disorders (e.g. autism, ADHD), the prevalence of CAS has reportedly increased substantially during the past decade.”*

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Key Diagnostic Features of CAS

- Atypical development (carefully scrutinize the birth to 15 month period of development)
- Strong desire to talk
- Effort associated with talking
- Difficulty sequencing sounds & syllables
- Inconsistent speech sound errors
- Vowel errors
- Prosodic abnormalities
- History of “pop-out” words
- Use of “go-to” sound/word

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Rome's
“go-to”
sound
Age 31
months



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Rome's "go-to" sound



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Differential Diagnosis

Differential diagnosis is the process of "ruling out" some disorders to ensure proper treatment

Ongoing diagnostic therapy is a crucial component of the therapeutic process

We must be skilled at diagnostic therapy in order to make a differential diagnosis by identifying specific characteristics to confirm or rule in or rule out our working diagnosis of suspected CAS

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Suspected CAS or Autism?

Some young children with Childhood Apraxia of Speech (CAS) may be mis-diagnosed as having Autism Spectrum Disorder (ASD) because there are 4 primary overlapping symptoms that commonly occur in both disorders including:

1. Child is minimally verbal
2. Child has social deficits
3. Child has poor eye contact
4. Child has sensory issues

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Suspected CAS	Autism Spectrum Disorder
<ul style="list-style-type: none"> • Receptive language stronger than expressive • Communicates wants and needs effectively – just not verbally • Strong desire to interact & communicate w/ others • Exhibits typical play time interests (limb apraxia may interfere with execution of play skills) 	<ul style="list-style-type: none"> • Impaired receptive language skills • Does not communicate effectively either verbally or non-verbally • Limited desire to interact and communicate with other people • Exhibits atypical, absent or aberrant play skills

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Play Skills are Critical for Making a Differential Diagnosis

- Play is a reflection of development - Children with limb apraxia/dyspraxia may have overly simplistic play schemes that are not consistent with their cognitive abilities; they may play in a repetitive manner and prefer to line up toys
- Children with limb apraxia/dyspraxia may not wave or point in a timely manner
- These characteristics are often considered consistent with a diagnosis of autism spectrum disorder
- ASD and CAS are not treated the same way...*won't vs. can't; non-compliance vs. motor planning difficulties*

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Suspected CAS	Autism Spectrum Disorder
<ul style="list-style-type: none"> • Strong social referencing/averts eye gaze when pressured to talk • Limited speech production attempts because child anticipates failure based on past talking experiences and is NOT a communication risk-taker • May have sensory issues 	<ul style="list-style-type: none"> • Lacks social referencing/overall poor eye contact • Limited speech production because child lacks symbolism/doesn't understand that words have power or inappropriate speech production due to echolalia and scripting • Likely has sensory issues

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It is important to remember that while kids with autism have sensory dysfunction, not all kids with sensory dysfunction have autism!

BEWARE!

CAS + SPD can mask as ASD

Differential diagnosis is critical!

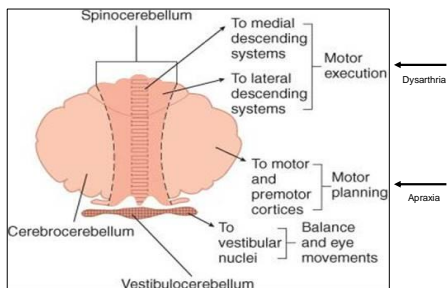
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Differential Diagnosis

Suspected CAS vs. Dysarthria

- "Dysarthria manifests as disrupted or distorted oral communication due to paralysis, weakness, abnormal tone, or incoordination of the muscles used in speech. Symptoms may include slurred speech, weak or imprecise articulatory contacts, weak respiratory support, low volume, incoordination of the respiratory stream, and hypernasality." Strand & McCauley
- CAS and Dysarthria are both motor speech disorders, but they occur at different levels of the motor cortex

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Reflex and Voluntary Control of Posture & Movement - Ganong's Review of Medical Physiology, 24th Edition

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Suspected CAS	Dysarthria
<ul style="list-style-type: none"> • Difficulty planning the movements necessary for speech – lack of consonants and vowels • Difficulty with motor PLANNING • Not associated with weakness • Receptive language better than expressive 	<ul style="list-style-type: none"> • Difficulty in the actual production of speech – distortion of consonants and vowels • Difficulty with motor EXECUTION • Characterized by weakness • No significant difference between receptive & expressive language skills

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Suspected CAS	Dysarthria
<ul style="list-style-type: none"> • No difficulty with involuntary motor control for eating (unless there is also oral apraxia) • Inconsistent speech errors • Prosody is disrupted – rate, rhythm, inflection patterns & stress impaired – better control of pitch and loudness • Voice quality is intact 	<ul style="list-style-type: none"> • Difficulty with involuntary motor control for eating due to muscle weakness and incoordination • Articulation is imprecise, but errors are consistent • Monotone voice common; difficulty controlling pitch and loudness levels • Voice quality may be impaired depending on type of dysarthria

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Differential Diagnosis
Suspected CAS vs. Dysarthria

Kids with CAS don't have strength issues, they have movement issues. "We don't need *strong* articulators, we need *agile* articulators." Dr. Lof 2007

Speech production requires rapid & accurate alternating movements of the articulators (i.e. speed & agility)

Diadochokinetic Rate (measures how accurately person can produce a series of rapid alternating sounds)

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Differential Diagnosis

Suspected CAS vs. Dysarthria

- Both CAS and Dysarthria will result in poor speech intelligibility - determining the etiology of the unintelligible speech will guide our treatment methods
- Weakness problems (Dysarthria) vs. Praxis problems (CAS)

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Differential Diagnosis

Suspected CAS vs. Phonological Disorder

- Phonology is the sound system of the language
- Children with a phonological disorder haven't learned the rules for how sounds fit together to make words
- A phonological disorder involves patterns of sound errors
- Children with a phonological disorder produce consistent sound errors and can imitate correctly when provided with auditory and visual cues

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Differential Diagnosis

Suspected CAS vs. Phonological Disorder

- Common phonological processes young children use: final consonant deletion, cluster reduction, gliding, fronting, stopping, deaffrication, assimilation
- Atypical phonological processes include backing and initial consonant deletion (Pam Marshall said that backing and initial consonant deletion are indicative of a more severe and a more persistent speech sound disorder)

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Suspected CAS	Phonological Disorder
<ul style="list-style-type: none"> • Motorically based • Vowel errors • Inconsistent errors • Effortful speech • Errors increase as length/complexity of utterance increases • Impaired prosody • Success with speech is situationally dependent - "on demand" vs. "automatic" 	<ul style="list-style-type: none"> • Linguistically based • Vowels typically intact • Consistent error patterns • Speech is not effortful • Errors consistent regardless of utterance length • Prosody is intact • No difference in how easily speech is produced based on the situation

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Differential Diagnosis

Suspected CAS vs. Language Delay

- While children develop skills at different rates, the most important factor is that the milestones are achieved in a typical or sequential manner.
- Milestones may be achieved late, but if they are acquired in the correct developmental sequence, the child is likely exhibiting a delay. If the developmental sequence is out of order/atypical then the child is more likely exhibiting a disorder.
 - Delay: child follows a typical path of development, it just takes longer
 - Disorder: child acquires milestones out of sequence/ scattered skills/lacks the foundation skills on which to build

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Suspected CAS	Language Delay
<ul style="list-style-type: none"> • Slow, inconsistent progress • Noticeable difficulty with vowel sounds • Limited babbling history • Restricted sound inventory • Disruption in the normal sequence of development- "atypical development" • Impaired prosody • Plays silently (even during high energy play) 	<ul style="list-style-type: none"> • More rapid, consistent progress • Vowels intact • Typical babbling history • Wider variety of speech sounds in repertoire • Speaks like a child who is chronologically younger - "delayed or late talker" • Prosody is intact • Is noisy during play

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Differential Diagnosis

When assessing young, minimally verbal children, SLPs will be differentially diagnosing between the following:

- Language Delay (late talker)
- Suspected CAS (motor planning/praxis problems)
- Dysarthria (motor execution/weakness)
- Phonological Disorder (patterns of sound errors)
- Autism Spectrum Disorder (communicative intent)

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When Families Ask About Prognosis

- We need to be honest and tell families that progress is often slow - that learning to talk is a marathon not a sprint. There is no "fast fix."
- Remember, that slow progress adversely affects parents' confidence in the therapy process.
- Duration of therapy for a toddler with suspected apraxia is likely to be 3+ years.
- Child with CAS is at risk for reading, spelling & writing difficulties as oral language problems often precede written language problems.

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What Affects Prognosis for Becoming Verbal?

- Severity
- Cognitive skills
- Child's personality/temperament
- Age at which therapy was initiated
- Co-existing conditions
- Motivation
- Appropriateness of the therapy
- Family involvement
- Etiology

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