

## Speech / Language Disorders in Children with PHACE Syndrome



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## Speech-Language Pathologist

Speech-language pathologists (SLPs) work to prevent, assess, diagnose, and treat speech, language, social communication, cognitive-communication, and feeding/swallowing disorders in children and adults.



## Speech vs Language

Language is made up of socially shared rules that include the following:

- What words mean (e.g., "star" can refer to a bright object in the night sky or a celebrity)
- How to make new words (e.g., friend, friendly, unfriendly)
- How to put words together (e.g., "Peg walked to the new store" rather than "Peg walk store new")
- What word combinations are best in what situations ("Would you mind moving your foot?" could quickly change to "Get off my foot, please!" if the first request did not produce results)

Language can be produced using verbal or nonverbal means of communication.



## Receptive and Expressive Language Disorders

Signs of a **receptive** language disorder:

- Difficulty understanding what words mean
- Difficulty following directions or answering questions
- Difficulty identifying objects and pictures
- Difficulty understanding abstract concepts

Signs of an **expressive** language disorder:

- Delayed production of first words
- Difficulty naming objects
- Delayed in putting words together into sentences
- Do not use correct pronouns, like "he" or "they"
- Inaccurate verb forms
- Poor conversational skills



## Speech vs Language

Speech is the verbal means of communicating.

Speech is produced through the complex interaction of multiple subsystems:

- Respiration
- Phonation
- Resonation
- Articulation
- Fluency / Prosody



## Articulation

How you adjust the shape and degree of *constriction* of the mouth, throat and nasal cavities as sound, air pressure and airflow pass through.

Speech sound production

"Pronunciation"



## Speech Sound Disorders

Phonological disorder  
Articulation disorder

Causes: abnormal learning, structural causes (e.g., cleft palate, dental/bite conditions, hearing loss, neurologic causes (e.g., paralysis)

Types of articulation errors:

- Substitutions
- Omissions
- Distortions
- Voicing errors



## Intelligibility

Degree of “understandability” of speech

Depends on the listener (familiar vs unfamiliar) and context

2 yrs old = 50% intelligible to strangers  
3 yrs old = 75% intelligible to strangers  
4 yrs old = 100% intelligible to strangers



## Treatment of Speech Sound Disorders

Speech therapy:

Demonstrating how to produce the sound correctly, learning to recognize which sounds are correct and incorrect, and practicing sounds in different words.

Should NOT include oral-motor strengthening exercises

Frequency and intensity of therapy depends on the age of the child, severity of disorder, and often, insurance limitations

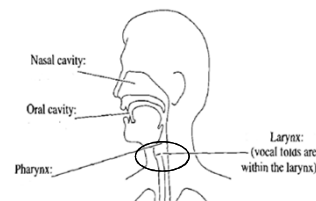


## Voice disorders

Dysphonia

Causes:

- Vocal nodules
- Vocal cord cysts
- Vocal cord paralysis
- Laryngeal webs
- Swelling/irritation (reflux)



## Resonance disorders

Too much or too little nasality in speech

Commonly causes of hypernasality (too much nasality):

- Cleft palate
- Submucous cleft palate
- Severe hearing loss
- Palate paralysis/weakness

Common causes of hyponasality (too little nasality):

- Severe cold/congestion
- Large adenoids/tonsils



## Fluency disorders (stuttering)

Approximately 5% of children exhibit atypical stuttering.

**Stuttering** = Repetitions of words or parts of words, as well as prolongations of speech sounds.

- Speech may become “blocked” → the mouth is positioned to say a sound, sometimes for several seconds, with little or no sound forthcoming. After some effort, the person may complete the word.
- Symptoms of **developmental stuttering** first appear between the ages of 2½ and 4 years. Most of the time the stuttering resolves within a few months.



## Risk factors for fluency disorders

- Males (4:1)
- family history of stuttering
- stuttering that has continued for 6 months or longer
- stuttering persisting past 4 years of age
- presence of other speech or language disorders
- strong fears or concerns about stuttering on the part of the child or the family



## Dysarthria

Collective name for a group of neurologic speech disorders resulting from abnormalities in the **strength, speed, range, steadiness, tone, or accuracy of movements** required for control of the respiratory, phonatory, resonatory, articulatory, and prosodic aspects of speech production. (Duffy, 2005)



## Typical characteristics of dysarthria

- Marked difficulties with speech and accuracy of articulatory movements (generalized imprecision)
- Reduced loudness
- Dysphonia
- Hypernasality
- Abnormal speaking rate
- Abnormal breath groups/rate
- Often have a history of feeding/swallowing problems, drooling, etc.



## Childhood apraxia of speech

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 (e.g. abnormal reflexes, abnormal tone).

The core impairment in planning and/or programming spatiotemporal parameters of movement sequences results in **errors in speech sound production and prosody** (ASHA, 2007).



## CAS signs and symptoms

The diagnosis of CAS is most confidently made after 3 years of age.

Other diagnostic features:

- Inconsistent speech sound errors
- Better receptive language (normal) than expressive
- Has difficulty imitating speech, but imitated speech is more clear than spontaneous speech
- May appear to be groping when attempting to produce sounds or to coordinate the lips, tongue, and jaw for purposeful movement
- Has more difficulty saying longer words or phrases clearly than shorter ones
- Is hard to understand, especially for an unfamiliar listener
- Sounds choppy, monotonous, or stresses the wrong syllable or word



## Risks for children with PHACE

1. Hearing
2. Speech / Language
3. Feeding / Swallowing




## Hearing loss in PHACE

Sensorineural hearing loss  
Conductive hearing loss

Hearing loss is often on same side as the hemangioma  
Higher risk for hearing loss if cutaneous hemangioma of the ear (and check for intracranial hemangiomas)

Newborn hearing screening  
Ongoing monitoring of hearing with re-testing as needed

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## Risk of speech-language disorders in PHACE

Risk factors:

- Posterior fossa malformations
- Lip/throat/airway hemangiomas
- Hearing loss
- Other developmental delays
- Prior stroke
- Multiple/prolonged hospitalizations
- History of cardiac surgery
  - Risk for vocal fold paralysis (breathy/weak voice)

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## Speech-language disorders in PHACE

Brosig et al. (2016):  
79% of children with PHACE had received or were currently in early intervention therapies


- 50% speech-language therapy
- 38% occupational therapy
- 67% physical therapy

Tangtiphaibootana et al. (2012):

- 40% speech-language therapy
- 46% occupational/physical therapy

Specific types of speech-language deficits in children with PHACE currently unknown

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## Feeding/swallowing disorders in PHACE

Failure to advance to solids  
Refusals/picky eater  
Discoordination of swallowing  
Penetration/aspiration (coughing/choking with feeds)

Swallowing problems may be secondary to hemangioma location (e.g., lip, oral cavity, airway)

Higher risk if history of cardiac surgery

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## When to request a referral to an SLP

Speech-language concerns:


- Reduced vocabulary for age; not combining words by age 2 years
- Intelligibility concerns: Child frustrated by not being understood
- All speech sounds not accurately produced by age 8 years
- Difficulty following verbal instructions

Known risk factors:

- Hearing concerns
- Posterior fossa malformations
- Lip/throat/airway hemangiomas
- Other developmental delays
- History of cardiac surgery
- Positive family history of speech-language disorder/delay

Consider full evaluation with SLP at 24 months of age (or sooner, if concerns)


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## Where to get SLP services

Pediatric hospitals: Birth to 18 years+  
County-based Early Intervention Program: 0-3 years  
School District: 3-18 years (sometimes 21)  
Private practices/clinics: Varies (suggest finding a pediatric-focused facility, if possible)

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American Speech-Language-Hearing  
Association (ASHA)

[www.asha.org](http://www.asha.org)



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