The Care of the Child with Down Syndrome: Otolaryngologic Considerations

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Disclosure Statement

• In the past 12 months, I have not had a significant financial interest or other relationship with the manufacturers of the products or provider of the services that will be discussed in my presentation.
DEVELOPMENTAL EXPECTATIONS IN EARLY COMMUNICATION SKILLS FOR CHILDREN WITH DS

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Developmental Communication Expectations

- For most individuals with DS, the ultimate communication system will be speech.
- The differences between communication, language and speech directly affect communication development and performance.
- Most toddlers will require a transitional communication system before they are ready to speak.
Communication

- Is Holistic
- May be unintentional or intentional/crying, smiling, pointing, soundmaking
- Communicative intent is learned when people in the environment react
- Can be powerful in affecting the environment
Language

• Is a shared code
• Is an arbitrary code which has rules that specify how to use the code
• Is used intentionally or purposefully
• Is learned through social interaction
• May include gestures, signs, pictures and/or speech
Speech

• Is verbal language
• Is overlaid on other systems in the body
• Involves muscle programming, movement and coordination
Birth to 1 Month

• **History**
  – Was hearing tested using Auditory Brain Stem Response (ABR) or Evoked Otoacoustic Emissions Testing?
  – Any difficulties with feeding?

• **Exam**
  – Weakness in lips?
  – Difficulty with coordination of suck-swallow-breathing synchrony?

• **Consults**
  – ENT
  – Audiology
  – Hearing evaluation
  – Feeding evaluation
Birth to 1 Month

Recommendations

• Feeding therapy as needed
• Information for family about early pre-speech vocalizations, and pre-language skills
• Information on how to encourage sound making and language development at home
• Refer to local parent support group
Early Communication Skills

- Communicative Intent
- Turn Taking
- Engaging
- Requesting
- Protesting
- Social Communication
Prerequisite Skills for Language

- Attentional Skills
- Sensory Processing
- Visual Skills
- Auditory Skills
- Tactile Skills
- Imitation Skills
- Motor Skills
- Cognitive Skills
Childhood: 1 to 12 months

• History
  – Parental concerns regarding hearing, vision, tactile skills, and feeding.
  – Parental report of sound making and responses to sound and words. Does the child respond to his name, environmental sounds? Does he appear to understand relationship between a word and its referent object? Will she look at the ball when you say “ball”? 
Exam (1-12 mo)

• Informal evaluation of pre-language skills such as reciprocal gaze, visual exploration, auditory localization, tactile sensitivities, tactile exploration including mouthing, cognitive skills such as object permanence, cause and effect, and means-end.

• Informal evaluation of pragmatics skills such as turn-taking, appropriate facial expressions, use of gestures, and social interaction.

• Informal evaluation of respiration, voice, and strength and range of motion of oral muscles, and oral structures for speech.
Consults/Recommendations

• Hearing testing every 6 months until normal bilateral ear specific test results. At that point, hearing tests should be done annually. (from AAP Health Care Guidelines).
• Speech-language pathology evaluation at 6-12 months to:
  • Evaluate oral motor skills
  • Evaluate pragmatics, language, and cognitive precursors for speech and language
  • Design and implement Total Communication program
  • SLP treatment and family education
Prerequisite Skills for Speech

- Respiratory Skills
- Feeding Skills
- Tactile Skills
- Imitation Skills
- Oral Motor Skills
- Sound Production Skills
Ready for Language

• Most children with DS have mastered the prerequisite skills for language and are ready to use language by 8-12 months of age.

• Children with DS speak their first word at an average age of 2-3 years.

• There will be a gap between readiness to use language and readiness to speak.
Recommendations

• Refer for early intervention program.
• Provide resources and information on early speech and language developmental milestones.
• Ongoing family involvement in speech-language pathology program is critical.
• Begin to facilitate AAC beginning at 8-12 months as appropriate.
Transitional Communication Systems

- Total communication/Sign Language
- Communication Boards
- Picture Exchange Communication (PECS)
- Communication Devices
- Apps
- Assistive Listening Devices
Childhood 1-3 Years

History

– According to parental report, is the child using speech? How many single words? Is the child using multi-word combinations? What length phrases?

– Does the child appear to have difficulty hearing? Understanding language? Following simple directions?

– If the child is not using speech, how is he communicating? Gestures? Grunts? Does he have a usable communication system? Is he frustrated?
1-3 Years Exam

- Interact with the child: Is the child socializing using gestures, facial expressions, and smiles?
- Check respiration, voice, oral-motor strength and range of motion.
- On observation, is the child using speech? How many single words? Is the child using multi-word combinations? What length phrases? Does the child appear to have difficulty hearing? Understanding language? Following simple directions? If he is not using speech, how is he communicating? Does he have a usable communication system? Does he appear frustrated?)
Consults

• The following evaluations should occur once yearly unless the child is in ongoing treatment. If the child is in treatment, data on progress can be ongoing.)
  – Hearing evaluation every 6 months until normal bilateral ear specific test results. At that point, hearing tests should be done annually. (from AAP Health Care Guidelines) Follow up referral to ENT for any difficulties and audiologist for assistive listening support.
  – Oral motor and oral sensory evaluation (pre-speech characteristics, posture, respiratory support for speech, oral sensory issues, tactile defensiveness, muscle strength and coordination, speech sound development)
Referrals

- Feeding evaluation (food texture progression)
- Pre-language evaluation
  - progress on pragmatics, language, and cognitive precursors
  - effectiveness of total communication
  - develop treatment plan with family participation in treatment
- Evaluation to explore reading instruction by age 3 years
Recommendations

- Development of Individualized Family Service Plan (IFSP) to include speech, language, oral motor, and feeding therapies, as appropriate
- By 1 year, use of Total Communication program, which may include sign language, communication board, PECS, I PAD or other touchscreen device, and/or synthesized speech (voice output) communication device to provide communication system until child starts to use speech
- Speech-language pathology treatment including oral-motor therapy, speech therapy, and language therapy
- By age 3, start to teach reading to support development of language
First Spoken Words

- Children with Down syndrome may begin to use speech to communicate between ages 2 and 4 years, but there is a wide range.
- They will usually understand far more than they can verbalize (receptive-expressive gap)
- Child’s intent to communicate and production of a variety of sounds should be noted.
Childhood 3-5 Years

History

• According to parental report, what is the size of the child’s vocabulary? How is the child communicating? Is he using multi-word combinations? Can he effectively make his needs known?

• Does the child appear to have difficulty hearing? Understanding language? Following directions? Ask for an example of an instruction that he can follow at home. Is he frustrated?

• Is the child speaking? Can he be understood?

• Is he in pre-school? How does he communicate in preschool?
Childhood 3-5 Years

Exam

• Interact with the child. Is he socializing using gestures, facial expressions, and smiles? Is he speaking? Are respiration, voice, oral motor strength, and range of motion adequate to support speech? What does he say to you? To family member present? Ask for an example of how the child would ask for a cookie, or would ask to go out to play. If he is not using speech, how is he communicating? Is the child using multi-word combinations?

• Does the child appear to have difficulty hearing? Understanding language? Following directions? Can he respond to yes/no or “wh-“ questions? (“What is your name? How old are you?”)
Childhood 3-5 Years: Consults

- (The following evaluations should occur once yearly unless the child is in ongoing treatment. If the child is in treatment, data on progress can be ongoing.)
- Hearing evaluation (at least once yearly)
- Speech evaluation
  - Oral motor/apraxia evaluation
  - Muscle strength and coordination
  - Difficulties in ease and clarity of speech production
Childhood 3-5 Years

Consults (continued)

• Language evaluation
  – receptive and expressive language
  – preschool language concepts
  – communication evaluation
  – referral to an augmentative communication team (AAC) when present communication system is not meeting the child’s needs
  – reading training
3-5 Years: Recommendations

- Development of Individualized Education Program (IEP) to include speech, language, and oral motor therapies as appropriate
- Provide resources for information on speech and language development, preschool concept development (e.g., colors, shapes), and literacy development
- Evaluate need for augmentative communication
- Services outside of school should be considered if school therapy is not provided, or is not meeting all communication needs.
- Treatment should include the family.
Speech

• Most children with DS will be speaking by age 5. If not, there is a need for a Speech Intelligibility Evaluation.

• All of the characteristics of speech intelligibility in children with Down syndrome occur in children with other conditions.

• It is the combination of anatomical, physiological, auditory, sensory processing and cognitive difficulties for each individual that necessitate individual treatment planning.
Speech Intelligibility Skills

• Speech intelligibility is important
• People underestimate a child’s skills when they don’t understand what the child is saying
• Familiar and unfamiliar listeners will have different ratings of intelligibility
• Factors affecting intelligibility can be identified and treated
WHAT AFFECTS SPEECH INTELLIGIBILITY?
EVALUATION & TREATMENT
Anatomical Factors

- A. Lips
- B. Tongue
- C. Teeth/Occlusion
- D. Hard Palate
- E. Soft Palate
- F. Upper Jaw (maxilla)
- G. Lower Jaw (mandible)
- H. Oropharynx
- I. Nasopharynx
- J. Tonsils/Adenoids
- K. Larynx
- L. Ears
Physiological Factors

• A. Lip Posture/Movement
• B. Tongue Posture/Movement
• C. Palatal Movement
• D. Intra-Oral Air Pressure
• E. Velopharyngeal Closure
• F. Jaw Movement/Stability
• G. Trunk Stability
• H. Vocal Vibration
• I. Breath Control/Support
• J. Other
  – Involuntary Movements
  – Drooling
  – Tooth grinding (Bruxism)
Neurofunctional Factors

- Neuromotor Component (Oral Motor)
- Childhood Apraxia of Speech (Motor Planning)
- Swallowing Pattern/-Feeding Pattern
- Hearing
Oral Motor Skills

• Oral motor skills refers to the movement of muscles of the face (e.g. lips and jaw) and oral area (e.g. tongue and soft palate), especially the movements related to speech. Oral motor skills also affect feeding, chewing, drinking and swallowing.

• Low muscle tone, weak muscles, and difficulty with coordination of movements are common
Oral Motor Skills

- Muscle tone
- Muscle strength
- Range of motion (distance)
- Speed
- Coordination
- Dissociation (ability to move structures such as tongue and lips independent of one another)
Childhood Apraxia of Speech (CAS)

• CAS is a motor speech disorder where children have difficulty planning, coordinating, producing and sequencing the sounds for speech.

• CAS interferes with the child’s ability to say sounds and to combine them into syllables, words, phrases and conversations.
CAS Developmental Speech History

- When you review a case history form for the SLP, the red flags for CAS are:
  - Children with CAS are quiet babies who use little cooing and babbling
  - They may have difficulties feeding
  - Children with CAS are late talkers. Speech develops at a much slower rate in children with CAS and many do not begin to speak until after age 5
  - Children with CAS usually develop gesture systems to communicate their needs.
  - In early words, they often omit the first sound in the word (e.g. *up* for *cup*)
  - Words may be said correctly, and then *disappear*
CAS Evaluation

- The child has difficulty producing both consonant and vowel sounds. Child may omit or distort vowel sounds.
- Inconsistent production of speech sounds. Sometimes your child can say a sound or word clearly, but not at other times.
- Struggle or groping when speaking. Your child tries so hard, but cannot say words clearly.
CAS Evaluation

• As a word or sentence length increases, the child has more difficulty (key, monkey, monkey bars and light, lightning, lightning bug)
• Difficulty combining and sequencing phonemes (efelant for elephant)
• Child omits sounds and syllables in speech, but also child may add sounds and syllables to words (hamburgurgger)
• Automatic speech is easier. Child may be able to count or say the alphabet clearly, or say “I don’t know”. 
CAS Evaluation

- Difficulty saying unfamiliar words. Familiar words and scripts are easier.
- Child has difficulty imitating words.
- Child may move their tongue or lips into place (silent posturing) but can’t make the sound right.
- Child has difficulty with rhythm, stress, and timing in speech. He may speak very fast, or accelerate as he speaks.
Formal CAS Testing

The formal tests most frequently given to diagnose CAS are:

• *The Kaufman Speech Praxis Test for Children* (KSPT)
• *The Apraxia Profile*
• *Verbal Motor Production Assessment for Children* (VMPAC).
Therapy Approaches for CAS

- Oral motor approaches
- Phonemic and articulation approaches
- Visual cueing approaches
- Multimodal or Total Communication approaches
- Prosodic approaches
- Shaping approaches
Perceptual Speech Symptoms

- Sound Errors
  - Articulation
  - Phonological Processes
- Voice
  - Volume
  - Pitch
  - Voice Quality
  - Resonance (Oral/Nasal Balance)
- Rate
- Fluency Pattern-Stuttering or Cluttering
- Prosody
Pragmatic Language Factors

• A. Social Language Skills
• B. Conversational Skills
• C. Narrative Discourse Skills
• D. Other Language Factors
Nonverbal Factors

• A. Eye contact
• B. Gestures
• C. Facial expressions
• D. Proxemics
Language Message Factors

• A. Greetings
• B. Routine/Automatic Verbalizations
• C. Longer Verbalizations
• D. Complex Messages
• E. Other
External/Environmental Factors

• A. Visual
• B. Auditory
• C. Listener Variables
• D. Other
SPEECH INTELLIGIBILITY TREATMENT-AAC

• Assistive Technology needs
  – AAC for classroom use/general use
  – www.ataccess.org; www.resna.org
  – Assistive listening devices

• Supports and modifications needed
  – “Native language”
  – Test and response, e.g. word processed responses or use of AAC
SPEECH INTELLIGIBILITY TREATMENT

• Team Approach:
  – SLP
  – Pediatrician
  – Otolaryngologist (ENT)
  – Audiologist
  – Neurologist
  – Psychologist
  – Occupational Therapist (OT)
  – Sensory Processing SPD/ Sensory Integration Specialist
  – Feeding specialist
Resources

- For more information, consult the following:
- [www.ndsccenter.org](http://www.ndsccenter.org) (Resource guides in English and Spanish)
- [www.ndss.org](http://www.ndss.org)
- [www.apraxia-kids.org](http://www.apraxia-kids.org)
- [www.asha.org](http://www.asha.org)
- [www.disabilitycompass.org](http://www.disabilitycompass.org) (volume 5 has 3 issues on speech intelligibility from *Disability Solutions* can be downloaded)