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Communication Development and Characteristics

By Marjorie Goodban, Ph.D., CCC-SLP

This is the comprehensive outline of Dr. Goodban's presentation at the Arizona Connections meeting on June 24, 2000.

Development of Communication^[1]

- Overall development of communication skills is delayed for approximately 96% of the individuals with CdLS.
- Based on my 1993 study, 53% of the children who were four years or older combined 2 or more words into sentences; 33% had no words or only 1-2 words; 4% had language skills within normal to low-normal limits

Speech Development (Articulation)/Oral Motor Skills

- Many of the children exhibit errors in articulation (pronunciation).
 - Many consonants (e.g., "k, t, s") are omitted, distorted, or other consonants are substituted for them
 - Earlier developing consonants, such as "k" and "t" are frequently omitted in the initial and/or final positions in words, e.g., "ca_" for "cat"
 - "Front" sounds (speech sounds made in the front part of the mouth), such as "t" and "d," are often substituted for "back sounds," (sounds made in the back part of the mouth), e.g. "tat" for "cat," or more commonly, "ta_" for "cat, " which is a combination of omission and substitution
- Factors related to articulation errors
 - Oral motor apraxia (difficulty imitating or producing voluntary movements of the mouth, in the absence of paralysis or weakness); evidenced by the majority of the individuals with CdLS^[2]
 - Retro-micrognathia (the lower jaw is retruded and small)
 - Retrusion of the lower jaw makes the tongue movements necessary for pronunciation more difficult to make
 - It also may create misalignment of the teeth, which can affect pronunciation
 - Overall developmental delay
 - Hearing loss

Language Development

- By the age of four years, approximately half of the children are combining two or more words into sentences
- Asynchronous Development (uneven development of skills). Many aspects of the language development are marked by discrepancies between skills:
 - Receptive language, cognition, and expressive language



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- Receptive language skills (what the child understands) are typically much higher than the child's expressive language skills (what the child says)
- Receptive language skills appear to be commensurate with the child's cognitive developmental level (often synonymous with IQ or mental age)
- Typically, expressive language skills are much lower than the child's cognitive developmental level and the child's receptive language skills
- Nonverbal cognitive abilities (what the child is able to understand by watching) appear to be much higher than expressive language abilities
- Vocabulary skills (the number of words the child says) and utterance length (average or typical number of words in the child's sentences): Given the number of words the child can say, sentence length is shorter than that expected for the normally developing child
- Utterance length and syntactic skills (syntax or grammar): The child's ability to use standard grammatical forms is below that expected, given the typical number of words the child can use in a sentence; this includes the ability to transform words into the standard question form
- Recent research on children with specific genetic syndromes reveals performance not accounted for by their general cognitive abilities. For example, similar to my 1993 findings in CdLS, in the Down syndrome population, children evidence deficits in language production in comparison to language comprehension and nonverbal cognitive skills, as well as better development in vocabulary skills than in syntactic skills. A similar comparison is seen in the fragile X syndrome. In contrast, children with Turner syndrome appear to have better language communication skills than would be suggested by their general cognitive skills.

Vocal Quality and Vocalics (volume or loudness, pitch, intonation, phrasing)

- Most of the children exhibit an unusual vocal quality that is guttural and low in pitch
- Overall range of vocal inflection tends to be limited
- Although some children have a vocal quality that is more guttural and hoarse than other children, there does not seem to be a relationship between vocal quality and the development of expressive language abilities
- GERD (gastroesophageal reflux disease) and hearing loss may contribute to some of the vocal quality differences

Resonance (the vibration of sound)

- Hypernasality (excessive resonance or sound vibration in the nose or nasal cavity) may be present if the child has a cleft palate
- Hyponasality (not enough resonance or sound vibration in the nose or nasal cavity) may occur if the nasal cavity is unusually small
- Voice may sound muffled and not very loud as a result of deficiencies in oral resonance (not enough resonance or vibration of sound in the mouth or oral cavity); the retro-micrognathia may cause the tongue to be placed farther back in the mouth

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than usual, affecting resonance; also, the oral cavity will be slightly smaller as a result of the retruded lower jaw

Facial Expression/Gesture/Nonverbal Behavior

- Faces tend to lack the range of expression typical of normally developing children
- Gestures and nonverbal behaviors appear to be limited but may be commensurate with expressive language skills
- More information is needed in these areas

Pragmatics (appropriate use of language in social settings; includes turn taking, interactions in conversation)

- Most of the children talk very little, even when they have highly developed vocabularies
- Some children exhibit difficulty in their relationships with peers
- The social behavior of some of the children with CdLS is similar to that of autism

Hearing

- Many children have conductive hearing losses (reduction in hearing sensitivity; otitis media; often caused by fluid in the middle ear cavity), often correctable with time, antibiotics and tubes)
- Hearing aids may be recommended, although parents often report their children seem to hear as well without the aids
- A number of children have sensorineural hearing losses (nerve damage), which are more serious; hearing aids are often recommended
- Unusual Results of Hearing Tests
 - Numerous reports from parents indicate that children are often diagnosed as deaf or profoundly hearing impaired immediately after birth, moderately hearing impaired at about 12 months, and mildly hearing impaired or with normal hearing at 18-24 months
 - I know of no other population where the hearing gets better instead of worse. These unusual findings may be related to audiologic testing procedures, the tiny structures of these infants, or some other unknown factor
 - Parents must be made aware that an initial diagnosis of deafness or severe hearing loss may be incorrect
 - It is important that parents talk to their infants as though they can hear
 - More information is needed about why the unusual hearing tests occur

Feeding Behavior

- Feeding problems seem to be very common
- Dr. Carrico will talk to you about this area.

Unusual Findings

- Unexpected Utterances
 - Clinical observations and reports from parents indicate that some children with CdLS clearly utter a word or phrase once or twice and then never use it again



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- This utterance is often many levels above the current level of performance, e.g., a child who is still struggling with simple, single-syllable words may unexpectedly produce a multi-syllabic utterance with perfect articulation
- This behavior is also observed in adults with apraxia and in children with autism

Prognostic Factors

- Children who do not talk at all or who are severely delayed in talking tend to have at least one of the following characteristics:
 - Birth weight under 5 lbs. (probably the least important factor)
 - Moderate-to-severe hearing impairment
 - Upper-limb malformations
 - Severe motor delay: sitting up after 25 months or walking after 35 months
 - Deficits in social relatedness: autistic-like behaviors, eye contact, overall ability to relate to people

OTHER COMMENTS

Communication Between Able-bodied Persons and Persons with Disabilities

- One of my other areas of specialization is Intercultural Communication
- The culture of the able-bodied is different from the culture of the disabled
- Because the able-bodied tend to feel uncomfortable and uncertain around disabled persons, they often react in predictable ways to the disabled and sometimes to the parents of the disabled
 - While the able-bodied may communicate verbal acceptance to the disabled, their nonverbal behavior may communicate rejection and avoidance
 - They may stand at a greater distance than usual
 - They may avoid eye contact
 - They may cut the conversation short
 - They may speak to the parent as though the disabled child is not present
 - Parents report friends and relatives may react differently to the birth of a disabled child
 - They may not send a birth present or a card for the disabled child
 - They may exclude the disabled child's name when sending greeting cards
 - They may not ask about the disabled child as they would the able-bodied child
 - The able-bodied often need to be educated on how to communicate with the disabled:
 - Treat persons with disabilities as persons first, recognizing that they are not dealing with a disabled person but with a person who has a disability



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- Don't avoid communication with persons who have disabilities simply because of discomfort or uncertainty
- Don't assume that persons with disabilities cannot speak for themselves or do things for themselves

TREATMENT

Groupings Based on Observed Characteristics^[3]

- Group I: "Talkers"
 - Approximately 3-4% of the population I have seen
 - Developmental milestones such as sitting and walking close to normal; may walk as late as 18 months; abilities in all areas are at low-normal to normal levels; no upper-limb malformations; normal ability to relate socially; minor hearing problems if any
 - Communication development is close to normal or maybe a little late; these children usually begin talking on their own but will probably benefit from speech therapy and other interventions
 - Educational placement may be in normal classroom with resource help for reading, math. etc.
 - Intervention will probably be necessary for some speech sounds, standard grammatical forms, increase in loudness, and optimal social interaction. Resource help may be necessary
- Group II: "Late Talkers"
 - Approximately 35-40% of the children I have seen
 - The children in this group will communicate verbally but these skills will develop later than for the Group I children and may not develop to the same extent; typically exhibit relatively good ability to imitate words they hear, although pronunciation may not be precise
 - The children in this group typically sit at or by 18 months and walk at or by 30 months; no upper-limb malformations; normal hearing, except for a possible mild conductive loss in one or both ears or a moderate conductive loss in one ear; moderate-to-good skills in social relatedness; birth weight at or greater than 5 pounds (although birth weight is probably not as important as the other factors)
 - First "real" words such as mama/dada may emerge as early as 12-18 months but often not until 24 -30 months; additional words will slowly emerge; two-word utterances at approximately 30-40 months; short sentences at approximately 40 months-six years; sentence length may never exceed five-seven words and will probably average three-five words per utterance; may be persistent problems with the formation of the standard question form; consistent omissions of substitutions of some consonants may persist; tendency to be very shy about talking except around family members
- Group III: "Limited" Talkers

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- Approximately 20-25% of the children I have seen
- The children in this group will communicate verbally but these skills will not develop as early or to the same extent as found in the Group II children
- First real words may occur as late as four-seven years of age; additional words may develop as late as 10-11 years of age
- Developmental milestones, such as sitting and walking, are achieved at a later age than described for Group II children; typically no upper-limb malformations; skills in social relatedness may not be as good as for the Group II kiddies but would not be described as autistic-like
- Group IV: "Guarded Talkers"
 - Approximately 25-30% of the children I have seen
 - The children in this group may not develop verbal communication skills, or may never have more than one-three words
 - One or more of the following is almost always present: Upper-limb malformations; moderate to severe hearing loss; autism or autistic-like behaviors; very late attainment of developmental milestones, e.g., may not walk until after four-five years of age

Treatment Procedures

- Augmentative and Alternative Communication (AAC)
 - AAC is traditionally defined as strategies that include a communication board; Ameslan sign language; American Indian Hand Talk or Amer-Ind gestural code; Blissymbolics; Total Communication; Pantomime; a manual alphabet; eye-blinking encoding; or electronic communication aids. For children with severe upper-limb malformations there are gestural-assisted and neuro-assisted strategies available
 - Unfortunately, almost all augmentative communication strategies are difficult to learn for children who have difficulty understanding nonverbal or gestural communication. As we know, many children with CdLS have impairments involving vocal/verbal skills, hand skills and processing skills (sensory, e.g., hearing and tactile; cognition; attention)
 - Before considering the use of augmentative or alternative communication, a number of factors regarding the child need to be considered.
 - The level of cognition
 - Motor abilities
 - Receptive language abilities
 - The motivation to communicate
 - Before deciding on a program, the speech-language pathologist may want to consult as appropriate with professionals such as physical therapists, occupational therapist, physicians, psychologists, engineers, social workers, vocational counselors, wheel-chair seating specialists, nurses, and teachers



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- Sign language or a gesture system seems to be the easiest nonverbal system for many young children with CdLS. In general, gestures facilitate the learning of oral language
- Important to know that many children with disabilities are likely to become passive and give up on all communication attempts because the few attempts they do make are not noticed or are misunderstood
- These impairments disrupt the familiar communication routines found in most interactions between young children and their parents. Children with CdLS are less likely to initiate and to imitate behaviors
- It is critical that children learn that their behaviors are meaningful to at least one other person
- Because there are many children who will not be able to use the traditional AAC procedures, and because we do not want to lose time until a traditional procedure is established, I am introducing the definition of AAC as anything that helps children or adults communicate when traditional strategies are not sufficient to accomplish a communication goal (C.J. Cress, ASHA, 1999). In this definition, AAC refers to reacting to the child's behaviors, e.g., posture and body shifts; vocal behaviors.
- Example: With a very young child, watch what behaviors occur when the child wants to be picked up. If the shoulders are raised, touch the shoulders and say, "I see you are raising your shoulders. Do you want up?" Then pick up the child. Communication has occurred and it was successful. When the behavior becomes purposeful, it becomes augmentative and alternative communication.
- Useful for Groups I -IV
- Also see Buddy under the Case Studies section of this handout

Typical Therapy Procedures for Groups II and III

- Assumes the necessary prognostic factors for success with verbal language: physical ability to produce speech sounds; sufficient level of cognition; adequate vision; sufficient attention span; ability to imitate sounds and words; adequate hearing; prepubescent; lack of autistic-like behaviors; and absence of upper-limb malformations.
- Therapy moves from gross motor imitation, to vocal imitation, to sound and word imitation.
- The earlier sessions are marked by playful activities involving oral-motor play and use of noise-making objects. Sessions are designed to provide maximum visual and auditory stimulation and to increase attending behavior.
- A very successful stimulation approach for eliciting vocal imitation consisted of saying "ba" or some other sound, word or phrase into a plastic ring and encouraging the child to imitate this utterance, while holding a ring near his/her mouth.
- All sessions must include therapy specifically designed for oral-motor apraxia; this therapy approach consists of:



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- Prolonged and highly inflected simple words and utterances (like motherese); numerous repetitions of these utterances, accompanied by holding the stimulus item or fingers near the parent's/therapist's mouth; slightly louder than normal loudness
- This exaggerated inflection is probably more successful because it intensifies auditory and visual stimulation; allows for greater auditory processing time; is closer in form to the motherese appropriate for children at a younger language age; and is "musical" and more likely to evoke a response
- Speaking this way can be remarkably difficult for some people to learn; I know because of the time it takes for my student therapists to learn it!
- Use of gestures, movements, tapping with blocks, or clapping to mark the rhythm of each syllable or word
- Reinforcement of successive approximations by the child.
- For the most part, therapy should follow the lead of the child. Be prepared with many interesting toys/items so that the child maintains interest and is frequently surprised. These items also serve as motivators
- Activities are designed to approximate those of normal daily routines, such as pretend grocery shopping, meal time, playing with doll houses, using a vacuum cleaner
- Try to find something the child will work hard to obtain. Food, toys, or fascinating items often work
- Tactile (touch) stimulation is often avoided because of the disturbing effect this has on many children with CdLS
- The use of sign language or gestures frequently facilitates the development of oral language. Most of us gestured before we talked, and many of us still rely on gestures to communicate or facilitate our recall of words
- For the child who has developed some expressive vocabulary, drill and repetition are useful in maintain and developing longer utterances. The "Wh-" question form often requires much practice.

Therapy Procedures for Groups III and IV

- Communication Board: If your child is completely nonverbal and unable to communicate wants and needs, this procedure is useful in helping your child communicate. At the same time, you will be teaching your child the basic nature of communication.
 - Provide two-to-four actual objects, imitation objects, or large, very clear pictures or photographs of objects, per context. By context I mean, meal time, play time, selection of toys, etc.
 - For example, at meal or snack time, you may have a communication board with four different pictures of food, or these pictures may be on the refrigerator door. When your child indicates a desire to eat, pair the touching of the picture (first by the parent as an example; later by the child) with the presentation of that particular food or drink. This provides a method of



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communication for the nonverbal child who has no other method of communication.

- There are a number of variations on the above approach. One is to have laminated pictures of objects on a large, circular key ring. This method is very portable and can be used by individuals other than the parents.
- Picture Exchange Communication System (PECS): This system is often used in schools for children in Group IV. PECS is usually established by a speech-language pathologist and used by the therapist as well as by the teacher and/or an aide. The basic idea of PECS is that the child learns to exchange a picture, and gradually a more sophisticated form of communication, for the desired outcome.
- Therapy for Children with Autism Spectrum Disorders: Some children exhibit behaviors that are very similar to those seen in children who are diagnosed with autism. According to reports from parents, such children have qualified for treatment programs for children with autism.

General Intervention Recommendations

- All children should receive a communication assessment as early as possible. Pre-verbal and verbal assessment can be obtained from interviews of caregivers, formal test administration, informal observations, and medical and educational reports.
- Communication intervention should be initiated as early as possible. Above all, the parents should talk to their child as though they expect a response and continue to expect a verbal response for as long as appropriate.
- Hearing ability is a critical factor in the development of speech and language.
 - Early and frequent tests are necessary, particularly with the child who has a suspected hearing loss.
 - Babies with CdLS have very tiny structures and testing may be difficult. It is advisable to consult an audiologist and/or otolaryngologist who is familiar with CdLS or who is experienced in working with infants.
 - Tubes may be useful for middle-ear drainage but again it is necessary to consult with a physician experienced in working with small infants.
- If a hearing loss is suspected, headsets and hearing aids may be prescribed for infants and children. Even a mild hearing loss can result in a speech and language delay.
 - Smaller aids are available so it is not necessary or advisable to use an adult-sized aid.
 - If the child will not leave on the aid, an audiologist or behavioral therapist may be helpful.
 - Appropriate audiological management should include selection and fitting of suitable amplification for all listening environments.
 - While the child's personal hearing aid may be sufficient some of the time, the use of FM amplification may be necessary in the school setting.
- Cleft lip and palate should be closed as early as possible. This improves the ability to eat, enhances the normal speaking process, and reduces the likelihood of ear infections leading to hearing loss. Evaluation and treatment for cleft lip and/or palate, insufficient velopharyngeal closure, and submucous cleft requires a team

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approach, usually comprised of at a speech-language pathologist, surgeon and dentist.

- For children with esophageal reflux, the parents are advised to seek early treatment or surgery to help reduce pain and discomfort, thus improving behavior. This treatment may also avoid irritation of the oral-pharyngeal-laryngeal areas as well as the eustachian tube and help promote better vocal quality and hearing.
- The procedures of self-talk (describing to the child what you are doing); parallel talk (describing to the child what the child is doing); and expectant waiting (using an expectant manner while waiting for the child to respond) are recommended for the parents and the therapists. Gestures and sign language are also encouraged as methods to facilitate and motivate oral communication. All babies gesture before they use words and everyone uses gestures to facilitate communication.
- Children with CdLS from bilingual families seem to do equally well in both languages (or not do equally well in both languages as the case may be). I do not recommend that intervention be limited to only one language.
- If the child has a gastric tube, feeding therapy may be indicated so the oral mechanism functions as normally as possible. However, it is not necessary for the child to have eaten normally in order for talking to occur.
- Work with the child and other therapists as necessary to reduce deficits in social relatedness. For the older child who has autistic-like behaviors and constantly plays with or holds the same toy, strive to have that object be an age appropriate object
- Also see my Pre-speech and Speech Intervention Procedures handout.

CASE STUDIES

Buddy The following case study illustrates a child who benefited from non-vocal strategies. The description of Buddy by Buzolich (1987) demonstrates the successful use of Blissymbols with a 12-year-old ambulatory nonverbal boy with Cornelia de Lange Syndrome. Buddy received his first non-oral communication evaluation at the age of 12 years upon admittance to a private school for severely behaviorally and educationally handicapped children. At the same time, he was placed in a group home due to the death of his foster parent. He was described as having no symbolic communication system on admittance to this private school and it was determined he had a severe oral-motor, verbal, and motor apraxia. In addition he had an attention-deficit disorder, and he exhibited both impulsive and compulsive behaviors.

Buddy's eye gaze, facial expressions, gestures, and vocalizations were his primary means of communication. He reportedly had mastered 60 signs although they were idiosyncratic versions of standard signs. Expressive verbal language was below the one-year level and receptive language functioning was at approximately the four-year level. His strength was in his visual modality; he could readily recognize letters of the alphabet and some common words. After 8 months of training he was easily trained to express himself with 100 Blissymbols and was able to combine these symbols into more complex expressions. After outgrowing his communication book of symbols, Buddy was trained to use the Wolf Voice-

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Output Communication Device (cited in Buzolich, 1987) and achieved an improved level of functional communication. Through an appropriate assessment, educational and therapeutic program, Buddy was able to learn and compensate for his handicapping condition.

Sammy The following case study illustrates the use of facilitative communication.

According to the parent, Sammy, a six-year-old girl with CdLS, has benefited greatly from the use of facilitative communication (Ask the Doctor, 1993). Both the therapist and the parent believe this approach has enhanced this child's ability to communicate.

Facilitative communication is a procedure involving a therapist or a "facilitator" who assists an individual with physical and communication disabilities to point to desired objects, pictures, printed letters and words, or to a keyboard. The scientific validity and reliability of this treatment technique has not been established (American Speech-Language-Hearing Association, 1995). As such, this procedure should not be used without the informed consent of the individual and family. The critics of this technique claim the facilitator and not the child is the one communicating. Proponents claim there are instances in which they had no prior knowledge of the information that ultimately unfolded.

^[1] Goodban, M. T. (1993) Survey of speech and language skills with prognostic indicators in 116 patients with CdLS. *Am J Med Genet* 47:1059-2063.

^[2] Nine doctors present research update at Arizona convention. (1994) *Reaching Out: The Newsletter of the CdLS Foundation*. XIII: 4, p. 11.

^[3] ©M. M. Goodban, June, 2000.