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Verbal dyspraxia in children with galactosemia

The term praxis (Greek: $\pi\rho\acute{\alpha}\xi\iota\varsigma$) describes a learned ability to plan and direct a temporal series of coordinated movements toward achieving a result. It is a skilled act that is strictly human and cortically directed. The motor acts requested by the physician, when performing a neurological examination, for example, are praxic in nature and are really quite simple when compared with the complicated requirements of articulating even relatively simple words. For instance, if we consider only the movements of the lips and the tongue in the production of the word "speech" we can see how complicated the process is. For the sound "s" the tongue must be grooved slightly so that the air stream moves smoothly forward to create a friction sound against the front teeth. The "p" sound occurs instantaneously following "s" when the lips come together and then pop open slightly to produce a mild "plosive" sound. The "ee" sound is difficult to describe, but the arch of the tongue must be perfectly positioned to get that precise vowel and not another. And finally, the "ch", a "plosive-sibilant" sound is produced when the tip of the tongue makes quick contact with the alveolar ridge and is quickly retracted.

Fortunately, we do not have to think about "how" we perform these articulatory miracles. For most of us articulating the sounds that make up words is produced automatically and effortlessly through sophisticated neurophysiological function and coordination.

This is not the case, however, with children who have dyspraxia of speech. In this disorder there is a sensory motor disturbance of articulation characterized by impaired capacity to program the positioning of speech musculature and the muscle movements for the volitional production of speech sounds [1, 3, 4]. Usually, there is also

some inability to relate the sequence of movements or motions to each other, commonly called "motor planning".

It is this disorder, dyspraxia of speech, or verbal dyspraxia as it will be referred to in this paper, that occurs frequently in children with treated galactosemia. We reported the speech characteristics of 24 patients treated for galactosemia [2]. Fifty-four percent had the speech disorder, verbal dyspraxia, and indicate the association of a specific and unusual speech defect with a specific and rare metabolic disorder.

It is the purpose of this presentation to:

1. Describe in more detail the characteristics of the disorder of verbal dyspraxia.
2. Offer suggestions relative to early diagnosis and treatment.

Characteristics of verbal dyspraxia

It should be mentioned first of all, that there still is great uncertainty as to what causes children without galactosemia to have verbal dyspraxia. It is known, however that many children in the public schools, who have severe articulation disorders meet the criteria for the diagnosis of dyspraxia [5]. It is not even certain what the specific relationship is in galactosemic children. We can only speculate that dyspraxic children, with or without galactosemia, appear to possess an impairment of sensory processing and in particular proprioceptive input with an ensuing failure to program, organize and carry out movements necessary for expressive speech.

1. High probability indicators are the following:
 - a. presence of vowel and diphthong errors
 - b. errors increase on longer responses; isolated sound patterns may be adequate, but polysyllabic words and phrases cause an increase in errors
 - c. indications of presence of an oral dyspraxia

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- d. groping trial and error behavior is very common; sound prolongations, repetitions, circumlocutions and silent posturing of the articulators typify imitative utterances.
 - e. sound and syllable transpositions of re-orderings are very common; "bakskek" for "basket;" "mukis" for "music"; "plakskik" for "plastic," and "patching" for "package".
2. Related probability indicators are the following:
 - a. poor diadochokinesia
 - b. deficient auditory memory span
 - c. in repeating short word lists, there is often evidence of re-ordering, substitutions of words, and poor memory
 - d. poor imitative responses and ability
 - e. errors are highly inconsistent and dependent on phoneme environment
 3. Additional descriptive characteristics are the following:
 - a. absence of specific paralysis or weakness of speech mechanism
 - b. significant delay in speech and language development; delayed or altered babbling development; history of late onset – or – apparent normal onset but with long periods of little or no progress
 - c. normal or disproportionately higher receptive language in comparison with expressive language
 - d. slow or limited progress in conventional speech intervention programs
 - e. increased likelihood of other school problems – reading, spelling, written language

Discussion and treatment considerations

Verbal dyspraxia is an unusual and characteristic speech disorder that appears with some regularity in children

with classical galactosemia. Early diagnosis is important because the disorder is recalcitrant to standard types of speech intervention and requires special methods of treatment. These methods may include:

1. Insuring acquisition and development of basic skills necessary to underpin articulation such as breathing, phonating, and resonating.
2. Raising awareness of oral sensation, position and relationship of approximating oral areas, attributes of movement and self-monitoring.
3. Aiming of reliable and consistent sound production with the help of visual, proprioceptive, and tactile feedback.
4. Establishing a key word vocabulary by teaching a few words or phrases that are necessary in the persons's everyday life.
5. Improving vowel and diphthong errors because speech intelligibility improves with good production of these sounds.
6. Emphasizing movement sequences; the use of nonsense syllables is helpful.
7. Teaching rhythm and syllable use.
8. Using visual, visual-auditory, and kinesthetic feedback procedures rather than auditory discrimination. Not only does sight help in the placement and production of visible consonants at the syllable level but in proper order at the polysyllable level.

Finally, it is important for the families to be aware of the high incidence of the impairment and to be given information that will assist them in providing suitable and consistent speech and language stimulation in the early years. If symptoms of the disorder do appear, then the families must also be strong advocates for their children to insure that regular and appropriate speech intervention programs are provided.

References

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