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The child with developmental disorders of speech and language

EPNS Training Course

Budapest

10-11 March 2016

Prof. dr. Philippe F. Paquier
Université Libre de Bruxelles (ULB)
Vrije Universiteit Brussel (VUB)
Universiteit Antwerpen (UA)

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Speech *vs*. Language

Language

- A socially agreed on, rule-governed system of symbols that is used to represent ideas about the world (Sharp & Hillenbrand, 2008).
- Language includes the following linguistic components:
  - *Phonology*: the ability to analyze words into separate word sounds (*phonemes*) and to assemble phonemes to form words.
  - *Semantics*: the knowledge of word meaning.
  - *Morphology*: the capacity to change words by means of inflections, conjugations, and derivations.
  - *Syntax*: rules that govern word order in a sentence.
  - *Pragmatics*: the use of language as a social tool, and the capacity to deal with language in its situational context, including the interactions between the speaker's and the interlocutor's knowledge and beliefs (e.g., their inferred intents).

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Speech *vs*. Language

Speech

- The motor execution of oral language communication (the act of speaking).
- Speech is produced through complex and coordinated respiratory, laryngeal, velopharyngeal, and articulatory movements (Sharp & Hillenbrand, 2008):
  - Respiration provides the air pressure to initiate sound production through vocal fold vibration at the larynx;
  - Airflow and laryngeal sounds are directed nasally or orally by the velopharynx;
  - Articulators (e.g., tongue, lips, teeth, jaw) shape laryngeal sounds and airflow to create vowels and consonants (*phonemes*).
- Speech production encompasses the areas of voice, fluency, and intelligibility (articulation + resonance).

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Speech disorder *vs*. Language disorder

- An impairment of one or several of the aforementioned linguistic components (phonology, semantics, morphology, syntax, pragmatics) is called a **language** disorder.
- An impairment of one or several of the basic speech production mechanisms is called a **speech** disorder.

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Factors influencing the acquisition of speech and language

- Speech and language acquisition: a natural and self-evident process?

  Yes, on the condition that (a) the child possesses the required physical and intellectual abilities, and (b) his/her environment is sufficiently stimulating.

  Learning by imitating is crucially involved in this process (role of *mirror neurons*?).

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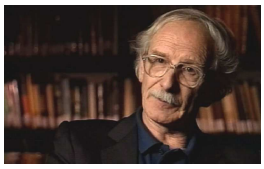
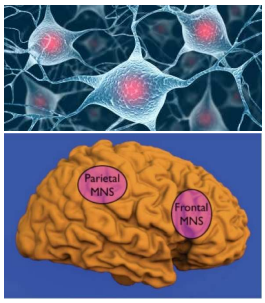
Factors influencing the acquisition of speech and language

- Mirror neuron: a specific type of visuo-motor neuron that discharges both when a monkey executes a motor act and when it observes a similar motor act performed by another individual.
- In a sense, a mirror neuron reflects another primate's behavior, and is equally active as if executing itself the same motor act.
- In humans, the mirror neuron system has been identified in premotor and parietal cortices.
- In 2010, mirror neurons were also reported in the human hippocampus.



Giacomo Rizzolatti



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Factors influencing the acquisition of speech and language

- Mirror neurons play a key role in perceiving, understanding, and interpreting others' actions, and in learning new skills by imitation.

- Mirror neurons are thought to play a role in the anticipation of action outcomes in others and in the apprehension of others' intentions (*theory of mind*), in the ability to enter into others' emotional feelings (*empathy*), and in the acquisition of language –more specifically, the emergence and further development of language production (*speech*).

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Factors influencing the acquisition of speech and language

Language acquisition process = process of interaction between the child and his/her social environment, determined by :

- The child's health and temperament;
- Material circumstances (e.g., housing conditions, family composition);
- The general social and emotional family conditions.

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Factors influencing the acquisition of speech and language

- Medical factors
- Cognitive factors
- Psychosocial factors

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Factors influencing the acquisition of speech and language

Medical factors:

- Hearing
- Morphology of the speech apparatus
- Motility of the speech apparatus
- Neurological and/or psychiatric state

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Factors influencing the acquisition of speech and language

Cognitive factors:

- Global learning ability
- Language sensitivity (“talent for languages”)

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Factors influencing the acquisition of speech and language

Psychosocial factors:

- Stimulating family and school environment (parent-child interaction; home language and school language)
- Housing conditions, family composition and conditions
- Emotional and behavioral development

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Classification of pediatric speech and language disorders

Childhood speech and language disorders can be classified into:

- ✓ ***Developmental*** speech and language disorders
  - *specific* (or primary): no evident, detectable cause.
  - *non-specific* (or secondary): resulting from a demonstrable, unfavorable factor.
- ✓ ***Acquired*** (neurological) speech and language disorders

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Classification of pediatric speech and language disorders

Developmental speech and language disorders

A disorder of speech production and/or linguistic functions resulting from a demonstrated or assumed pathological condition already present before the emergence of language.

Acquired (neurological) speech and language disorders

A speech and/or language disorder caused by a lesion that is sustained after onset of language acquisition, and that disrupts already developed speech/language skills. The pathological process which compromises the functioning of speech and/or language is sustained after a period of language development.

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Children with speech and language impairments

- ***Delayed language development***:
  The child's language system does not match his/her chronological age, but is consistent with that of a younger child.
- ***Deviant language development***:
  The child's language system shows characteristics that do not match any phase of normal language acquisition.

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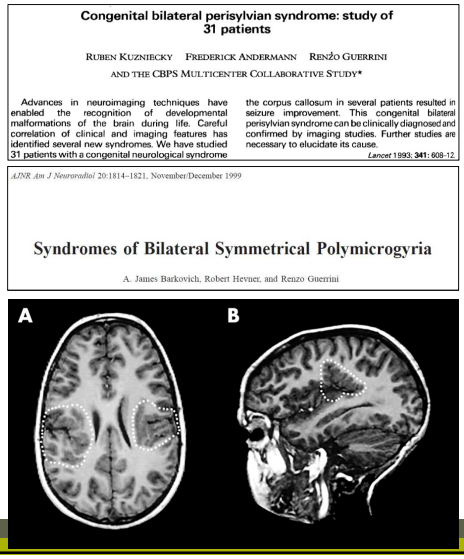
Classification of pediatric developmental speech and language disorders

Examples of secondary S/L disorders

(resulting from a demonstrable, unfavorable factor):

Pre-, peri- or early postnatal brain damage (cf. *congenital dysarthria*).



Congenital bilateral perisylvian syndrome: study of 31 patients

RUBEN KUZNIECKY FREDERICK ANDERMANN RENZO GUERRINI
AND THE CBPS MULTICENTER COLLABORATIVE STUDY*

Advances in neuroimaging techniques have enabled the recognition of developmental malformations of the brain during life. Careful correlation of clinical and imaging features has identified several new syndromes. We have studied 31 patients with a congenital neurological syndrome the corpus callosum in several patients resulted in seizure improvement. This congenital bilateral perisylvian syndrome can be clinically diagnosed and confirmed by imaging studies. Further studies are necessary to elucidate its cause.

Lancet 1993; **341**: 608-12.

AJNR Am J Neuroradiol 20:1814–1821, November/December 1999

Syndromes of Bilateral Symmetrical Polymicrogyria

A. James Barkovich, Robert Hevner, and Renzo Guerrini

A B

Jansen A, Andermann E. J Med Genet 2005: 42: 369-378.

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Classification of pediatric developmental speech and language disorders

Examples of secondary S/L disorders

(resulting from a demonstrable, unfavorable factor):

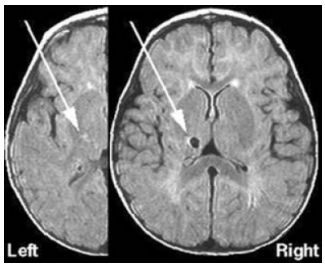
Pre-, peri- or early postnatal brain damage (cf. *congenital aphasia*).

Neurocase, 10(4): 308–315, 2004

1355-4795/04/1004–308$16.00

Language Disorder in a Child with Early Left Thalamic Lesion

Maria Rosa Pizzamiglio[1], Laura Piccardi[1], Marianna Nasti[2], Francesco Tomaiuolo[1] and Umberto Sabatini[1]
[1]IRCCS Fondazione Santa Lucia, Rome, Italy, and [2]Department of Psychology, University of Rome "La Sapienza," Italy



Pizzamiglio MR et al.
Neurocase 2004: 10: 308-315.

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Classification of pediatric developmental speech and language disorders

Examples of secondary S/L disorders

(resulting from a demonstrable, unfavorable factor):

Chromosomal anomalies associated with neurological deficits.

JSHR, Volume 41, 861–873, August 1998

Language Skills of Children and Adolescents With Down Syndrome: II. Production Deficits

Robin S. Chapman
Hye-Kyeung Seung
University of Wisconsin–Madison
Scott E. Schwartz
Boulder, Colorado, School District
Elizabeth Kay-Raining Bird
Dalhousie University
Halifax, Nova Scotia, Canada

Hypotheses that children and adolescents with Down syndrome show (a) a specific expressive language impairment, (b) a "critical period" for language acquisition, (c) a "simple sentence syntactic ceiling" in production, and (d) deficit in grammatical morphology were investigated cross-sectionally. Conversational and narrative language samples from 47 children and adolescents with Down syndrome (Trisomy 21), aged 5 to 20 years, were compared to those from 47 control children aged 2 to 6 years matched statistically for nonverbal mental age. Children with Down syndrome appear to have a specific language impairment, compared to control children, in number of different words and total words (in the

Down Syndrome

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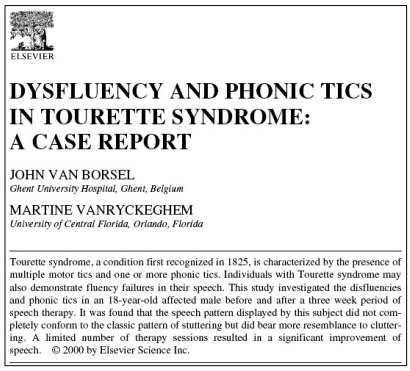
Classification of pediatric developmental speech and language disorders

Examples of secondary S/L disorders

(resulting from a demonstrable, unfavorable factor):

Neurodevelopmental disorders.



ELSEVIER

DYSFLUENCY AND PHONIC TICS IN TOURETTE SYNDROME: A CASE REPORT

JOHN VAN BORSEL
Ghent University Hospital, Ghent, Belgium

MARTINE VANRYCKEGHEM
University of Central Florida, Orlando, Florida

Tourette syndrome, a condition first recognized in 1825, is characterized by the presence of multiple motor tics and one or more phonic tics. Individuals with Tourette syndrome may also demonstrate fluency failures in their speech. This study investigated the disfluencies and phonic tics in an 18-year-old affected male before and after a three week period of speech therapy. It was found that the speech pattern displayed by this subject did not completely conform to the classic pattern of stuttering but did bear more resemblance to cluttering. A limited number of therapy sessions resulted in a significant improvement of speech. © 2000 by Elsevier Science Inc.

J Commun Disord 2000: 33: 227-240.

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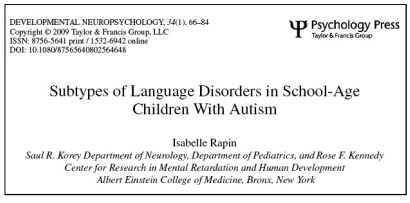
Classification of pediatric developmental speech and language disorders

Examples of secondary S/L disorders

(resulting from a demonstrable, unfavorable factor):

Neurodevelopmental disorders, such as autism and ADHD.

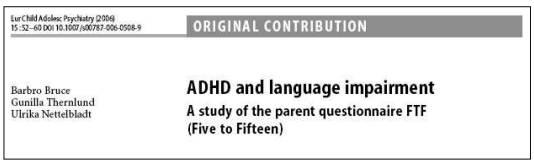


DEVELOPMENTAL NEUROPSYCHOLOGY, 34(1), 66–84
Copyright © 2009 Taylor & Francis Group, LLC
ISSN: 8756-5641 print / 1532-6942 online
DOI: 10.1080/87565640802564648

Psychology Press
Taylor & Francis Group

Subtypes of Language Disorders in School-Age Children With Autism

Isabelle Rapin
Saul R. Korey Department of Neurology, Department of Pediatrics, and Rose F. Kennedy Center for Research in Mental Retardation and Human Development Albert Einstein College of Medicine, Bronx, New York



Eur Child Adolesc Psychiatry (2006)
15 :52–60 DOI 10.1007/s00787-006-0508-9

ORIGINAL CONTRIBUTION

Barbro Bruce
Gunilla Thernlund
Ulrika Nettelbladt

ADHD and language impairment
A study of the parent questionnaire FTF (Five to Fifteen)

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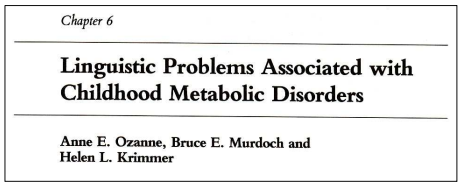
Classification of pediatric developmental speech and language disorders

Examples of secondary S/L disorders

(resulting from a demonstrable, unfavorable factor):

Extremely ill physical condition (anemia, metabolic disorders).

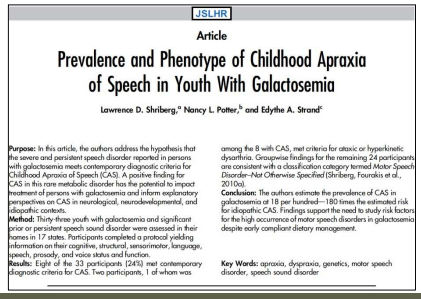


Chapter 6

Linguistic Problems Associated with Childhood Metabolic Disorders

Anne E. Ozanne, Bruce E. Murdoch and Helen L. Krimmer

In: Murdoch BE (ed.), Acquired neurological speech/language disorders in childhood. London, Taylor & Francis, 1990; 199-215.



JSLHR

Article

Prevalence and Phenotype of Childhood Apraxia of Speech in Youth With Galactosemia

Lawrence D. Shriberg,[a] Nancy L. Potter,[b] and Edythe A. Strand[c]

Purpose: In this article, the authors address the hypothesis that the severe and persistent speech disorder reported in persons with galactosemia meets contemporary diagnostic criteria for Childhood Apraxia of Speech (CAS). A positive finding for CAS in this rare metabolic disorder has the potential to impact treatment of persons with galactosemia and inform exploratory perspectives on CAS in neurological, neurodevelopmental, and idiopathic contexts.

Method: Thirty-three youth with galactosemia and significant prior or persistent speech sound disorder were assessed in their homes in 17 states. Participants completed a protocol yielding information on their cognitive, structural, sensorimotor, language, speech, prosody, and voice status and function.

Results: Eight of the 33 participants (24%) met contemporary diagnostic criteria for CAS. Two participants, 1 of whom was among the 8 with CAS, met criteria for ataxic or hyperkinetic dysarthria. Groupwise findings for the remaining 24 participants are consistent with a classification category termed *Motor Speech Disorder–Not Otherwise Specified* (Shriberg, Fourakis et al., 2010a).

Conclusion: The authors estimate the prevalence of CAS in galactosemia at 18 per hundred—180 times the estimated risk for idiopathic CAS. Findings support the need to study risk factors for the high occurrence of motor speech disorders in galactosemia despite early compliant dietary management.

Key Words: apraxia, dyspraxia, genetics, motor speech disorder, speech sound disorder

J Speech Lang Hear Res 2011: 54: 487-519.

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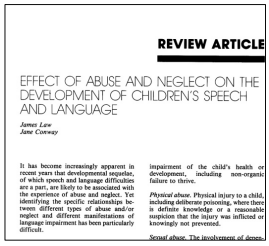
Classification of pediatric developmental speech and language disorders

Examples of secondary S/L disorders

(resulting from a demonstrable, unfavorable factor):

Detrimental environmental factors (deprivation, abuse and neglect, adverse family or social background).



REVIEW ARTICLE

EFFECT OF ABUSE AND NEGLECT ON THE DEVELOPMENT OF CHILDREN'S SPEECH AND LANGUAGE

James Law
Jane Conway

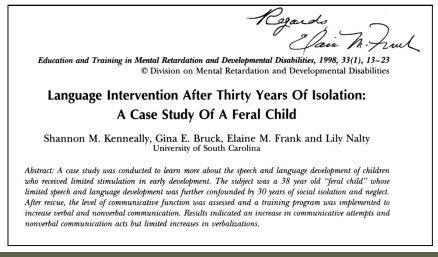
It has become increasingly apparent in recent years that developmental sequelae, of which speech and language difficulties are a part, are likely to be associated with the experience of abuse and neglect. Yet identifying the specific relationships between different types of abuse and/or neglect and different manifestations of language impairment has been particularly difficult.

impairment of the child's health or development, including non-organic failure to thrive.

Physical abuse. Physical injury to a child, including deliberate poisoning, where there is definite knowledge or a reasonable suspicion that the injury was inflicted or knowingly not prevented.

Sexual abuse. The involvement of depen-

Dev Med Child Neurol 1992: 34: 943-948.



Education and Training in Mental Retardation and Developmental Disabilities, 1998, 33(1), 13–23
© Division on Mental Retardation and Developmental Disabilities

Language Intervention After Thirty Years Of Isolation: A Case Study Of A Feral Child

Shannon M. Kenneally, Gina E. Bruck, Elaine M. Frank and Lily Nalty
University of South Carolina

Abstract: A case study was conducted to learn more about the speech and language development of children who received limited stimulation in early development. The subject was a 38 year old "feral child" whose limited speech and language development was further confounded by 30 years of social isolation and neglect. After rescue, the level of communicative function was assessed and a training program was implemented to increase verbal and nonverbal communication. Results indicated an increase in communicative attempts and nonverbal communication acts but limited increases in verbalizations.

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Classification of pediatric developmental speech and language disorders

Examples of secondary S/L disorders

(resulting from a demonstrable, unfavorable factor):

Morphology and motility anomalies of the speech apparatus.

Palatoschisis

Folia Phoniatrica et Logopaedica

Original Paper

Folia Phoniatr Logop 2008;60:223–232
DOI: 10.1159/000151242

Published online: August 12, 2008

Speech Outcome regarding Overall Intelligibility, Articulation, Resonance and Voice in Flemish Children a Year after Pharyngeal Flap Surgery

A Pilot Study

Kristiane M. Van Lierde Katrien Bonte Nele Baudonck P. Van Cauwenberge
Els M.R. De Leenheer

Department of Otorhinolaryngology, Head and Neck Surgery and Speech and Language Pathology, Craniofacial Team, University Hospital Gent, Gent, Belgium

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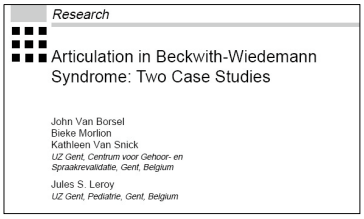
Classification of pediatric developmental speech and language disorders

Examples of secondary S/L disorders

(resulting from a demonstrable, unfavorable factor):

Morphology and motility anomalies of the speech apparatus.



Research

Articulation in Beckwith-Wiedemann Syndrome: Two Case Studies

John Van Borsel
Bieke Morlion
Kathleen Van Snick
UZ Gent, Centrum voor Gehoor- en Spraakrevalidatie, Gent, Belgium

Jules S. Leroy
UZ Gent, Pediatrie, Gent, Belgium

Am J Speech Lang Pathol 2000: 9: 202-213.

Beckwith-Wiedemann Syndrome

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Classification of pediatric developmental speech and language disorders

Examples of secondary S/L disorders

(resulting from a demonstrable, unfavorable factor):

Hearing loss.

Auditory processing disorder.

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Classification of pediatric developmental speech and language disorders

http://www.asha.org/public/hearing/Effects-of-Hearing-Loss-on-Development/

Effects of hearing loss on speech/language development:

- It causes delay in the development of receptive and expressive communication skills (speech and language).
- The language deficit causes learning problems that result in reduced academic achievement.
- Communication difficulties often lead to social isolation and poor self-concept.
- It may have an impact on vocational choices.

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Classification of pediatric developmental speech and language disorders

http://www.asha.org/public/hearing/Effects-of-Hearing-Loss-on-Development/

Specific effects of hearing loss on *vocabulary*

- Vocabulary develops more slowly in children who have hearing loss.
- Children with hearing loss learn concrete words (e.g., *cat*, *table)* more easily than abstract words (e.g., *anger*, *doubt*). They also have difficulty with function words like *the*, *an*, *are*, and *a*.
- The gap between the vocabulary of children with normal hearing and those with hearing loss widens with age. Children with hearing loss do not catch up without intervention.
- Children with hearing loss have difficulty understanding words with multiple meanings. For example, the word *bank* can mean the edge of a stream or a place where we put money.

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Classification of pediatric developmental speech and language disorders

http://www.asha.org/public/hearing/Effects-of-Hearing-Loss-on-Development/

Specific effects of hearing loss on *sentence structure*

- Children with hearing loss comprehend and produce shorter and simpler sentences than children with normal hearing.
- Children with hearing loss often have difficulty understanding and writing complex sentences, such as those with relative clauses ("Do you know the boy whose mother is a nurse?") or passive voice ("The ball was thrown by Mary")
- Children with hearing loss often cannot hear word endings such as /*s*/ or /*ed*/. This leads to misunderstandings and misuse of verb tense, pluralization, non-agreement of subject and verb, and possessives.

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Classification of pediatric developmental speech and language disorders

http://www.asha.org/public/hearing/Effects-of-Hearing-Loss-on-Development/

Specific effects of hearing loss on *speaking*

- Children with hearing loss often cannot hear quiet speech sounds such as “s”, “sh”, “f”, “t”, and “k” and therefore do not include them in their speech. Thus, speech may be difficult to understand.
- Children with hearing loss may not hear their own voices when they speak. They may speak too loudly or not loud enough. They may have a speaking pitch that is too high. They may sound like they are mumbling because of poor stress, poor inflection, or poor rate of speaking.

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Classification of pediatric developmental speech and language disorders

http://www.asha.org/public/hearing/Effects-of-Hearing-Loss-on-Development/

Specific effects of hearing loss on *academic achievement*

- Children with hearing loss have difficulty with all areas of academic achievement, especially reading and mathematical concepts.
- Children with mild to moderate hearing losses, on average, achieve one to four grade levels lower than their peers with normal hearing, unless appropriate management occurs.
- Children with severe to profound hearing loss usually achieve skills no higher than the third- or fourth-grade level, unless appropriate educational intervention occurs early.
- The gap in academic achievement between children with normal hearing and those with hearing loss usually widens as they progress through school.
- The level of achievement is related to parental involvement and the quantity, quality, and timing of the support services children receive.

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Classification of pediatric developmental speech and language disorders

http://www.asha.org/public/hearing/Effects-of-Hearing-Loss-on-Development/

Specific effects of hearing loss on *social functioning*

- Children with severe to profound hearing losses often report feeling isolated, without friends, and unhappy in school, particularly when their socialization with other children with hearing loss is limited.
- These social problems appear to be more frequent in children with a mild or moderate hearing losses than in those with a severe to profound loss.

➢ Reduced psycho-social Quality of Life

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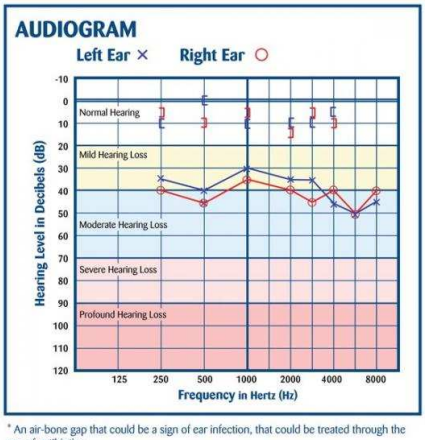
Classification of pediatric developmental speech and language disorders

A fluctuating hearing loss due to repeated ear infections might mean the child doesn't hear consistently and may be missing out on critical speech information.

http://www.chsc.org/Main/Speech-Language-Development-with-Hearing-Loss.aspx



http://www.terracehearing.com/wp-content/uploads/2014/08/audiogram_-_bone_conduction.jpg

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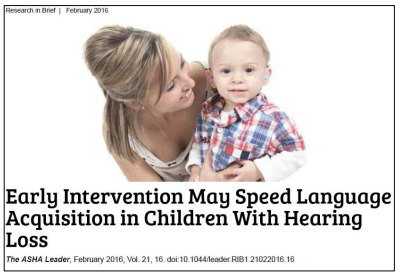
Classification of pediatric developmental speech and language disorders



Research in Brief | February 2016

Early Intervention May Speed Language Acquisition in Children With Hearing Loss

The ASHA Leader, February 2016, Vol. 21, 16. doi:10.1044/leader.RIB1.21022016.16

Children with mild-to-severe hearing loss may develop language skills at a faster rate when they receive high-quality early intervention, according to a new large-scale, longitudinal study.

The Outcomes of Children With Hearing Loss study—by researchers at the University of Iowa, Boys Town National Research Hospital and the University of North Carolina at Chapel Hill, and published in the journal Ear and Hearing—found that children with hearing loss generally have poorer language development than their hearing peers (http://journals.lww.com/ear-hearing/toc/2015/11001), though well-fitting hearing aids increase the likelihood of children closing the gap or significantly improving their language skills.

"The cautionary note from our research is that any degree of hearing loss, even mild, can place children at risk" in terms of learning, speech, language and socialization, says Bruce Tomblin, professor emeritus in the

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Classification of pediatric developmental speech and language disorders

Primary (specific) S/L disorders

Anomalies in speech and/or language acquisition and development, in the absence of an evident, detectable cause (i.e., unfavorable factors cannot be demonstrated).

A genetic predisposition to poor language performance might be involved.

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Developmental language disorders: Dysphasia

Dysphasia (synonyms: *developmental aphasia*; *Specific Language Impairment* [SLI]) refers to abnormal language performance that does not match any expected age norms, and has never shown an arrest at nor a decline from an earlier level of language functioning.

The disorder that hinders the normal development of language functions is not adequately accounted for by:

- Intellectual disability
- Motor or sensory defects (e.g. deafness)
- Frank neurological problems
- Severe emotional and/or behavioral disturbances
- Major environmental and/or language deprivation

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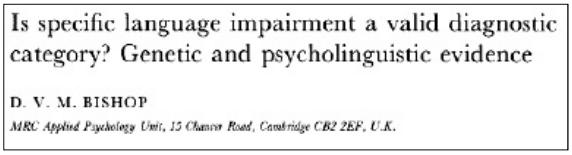
Developmental language disorders: Dysphasia = SLI

"The terminology used to refer to such children has varied over the past few decades: 'developmental aphasia' and 'developmental dysphasia' (…) More neutral diagnostic terms with fewer medical overtones, such as 'developmental language disorder' or '**specific language impairment**' (SLI) are now usually preferred." (Bishop, 1994).



Is specific language impairment a valid diagnostic category? Genetic and psycholinguistic evidence

D. V. M. BISHOP

MRC Applied Psychology Unit, 15 Chaucer Road, Cambridge CB2 2EF, U.K.

Phil Trans R Soc Lond B 1994: 346: 105-111.

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Developmental language disorders: Dysphasia prevalence

Prevalence

Morley (1972)	3-yr-old 7-yr-old	± 10-20 % ± 1.5-3 %	Silva (1980)	3-yr-old	± 8%
			Beitchman et al. (1986)	< 6-yr-old	± 3-10%

- Dysphasia is not a delayed but a *deviant* development of language.
- Is difficult to differentiate from a delayed language development at a younger age.
- Late talking toddlers –who erroneously received a diagnosis of dysphasia at a younger age– naturally catch up with their peers at a later age (*delayed* language development).

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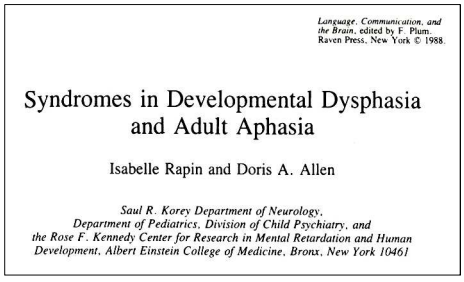
Developmental language disorders: Dysphasia syndromes



Language, Communication, and the Brain, edited by F. Plum. Raven Press, New York © 1988.

Syndromes in Developmental Dysphasia and Adult Aphasia

Isabelle Rapin and Doris A. Allen

Saul R. Korey Department of Neurology, Department of Pediatrics, Division of Child Psychiatry, and the Rose F. Kennedy Center for Research in Mental Retardation and Human Development, Albert Einstein College of Medicine, Bronx, New York 10461

In: F. Plum (Ed.), Language, communication, and the brain. New York: Raven Press, 1988; pp. 57-75.

Updates:

Rapin I. J Child Psychol Psychiat 1996: 37: 643-655.

Rapin I. & Dunn M. Brain Dev 2003: 25: 166-172.

Rapin I. et al. Dev Neuropsychol 2009: 34: 66-84.

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Developmental language disorders: Dysphasia syndromes (Rapin & Dunn, 2003)

Table 2
Allen and Rapin clinically defined language disorder subtypes in preschool children [23,24]

A. Mixed receptive/expressive disorders	Impaired phonologic decoding which affects all subsequent processing of language. Expression sparse and dysfluent. Language acquisition through the visual channel often unaffected
Verbal auditory agnosia	Phonologic decoding so profoundly impaired that the children understand no language and therefore are non-verbal or virtually so
Phonologic–syntactic subtype	Comprehension impaired but equal to or superior to language production. Expressive language sparse, in rudimentary, poorly articulated sentences, vocabulary impoverished
B. Higher order processing disorders	Comprehension and formulating of discourse impaired, phonology and syntax may be delayed but are not a primary deficit
Lexical syntactic subtype	Severe word finding deficit resulting in dysfluent language, syntax often immature. Expression may start as fluent jargon
Semantic pragmatic subtype	Expressive language fluent, echolalic, often verbose and scripted, with verbal perseveration, unusual word choices, and impaired conversational use of language. Comprehension more impaired than production
C. Expressive disorders	Comprehension normal or near normal
Verbal dyspraxia	Extremely dysfluent expression in the face of normal or near normal comprehension. Although verbal dyspraxia may be associated with oromotor deficits and overall clumsiness, these motor deficits are not severe enough to account for the profoundly impaired expressive deficit which is postulated to be at the level of retrieval of the commands for verbal expression
Phonologic programming subtype	The children are fluent and unintelligible, or they have small distorted expressive vocabularies and simplified syntax.

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Developmental language disorders: Semantic-pragmatic language disorder

- E.g., autism spectrum disorder (ASD), Asperger syndrome, Williams syndrome, hydrocephalus +/- spina bifida (*cocktail party syndrome*), non-verbal learning disorder (+ *hyperlexia*), …
- Fluent, verbose and scripted, grammatically complex language, but with poor sensitivity to the communicative situation.
- Difficulty understanding irony and empathy.
- Difficulty comprehending inferential meaning.
- Pragmatic deficits mild in SLI, universal and often blatant in ASD.

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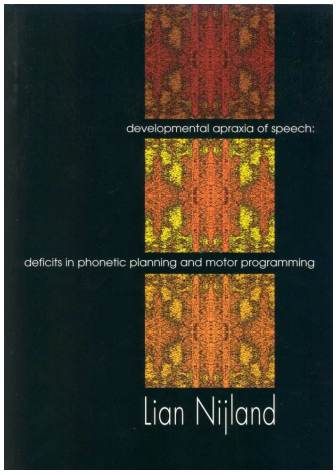
Developmental speech-sound disorder: Verbal dyspraxia (apraxia of speech)



PhD dissertation, KU Nijmegen, 2003.

- Poorly understood, severe childhood speech-sound disorder.
- Characterized by deficits in timing and sequencing the movements required for speech production.
- Limited repertoire of phonemes, and reduced articulatory precision with increasing word length and speaking rate.

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Developmental speech-sound disorder: Verbal dyspraxia (apraxia of speech)

Developmental apraxia of speech (DAS) is a neurologically based speech disorder that can be understood as a deficit in planning and regulating motor actions of voluntary and complex sequential speech movements, in the absence of dysarthria, mental retardation, hearing loss, receptive language impairments, and orofacial malformation. Its defining characteristics are: unintelligible speech due to a large number of consonant errors (especially substitutions and omissions), inconsistency of speech errors, articulatory abnormalities like groping behavior, and abnormal prosody.

Nijland L. Developmental apraxia of speech: deficits in phonetic planning and motor programming. Catholic University Nijmegen, PhD dissertation, 2003.

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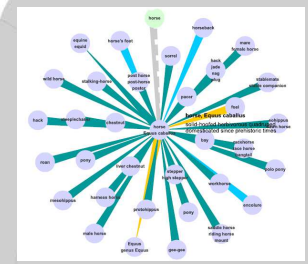
Schema…ion of the cognitive p…g



- Early v… of the…
- Acce… (or me…
- Access… auditory…
- Programming the… movements (lips, tongue, jaw, palate, etc) and their sequence;
- Articulating the word by implementing the planned movements.

Hillis AE, Course N° 2DS.008, American Academy of Neurology, 53rd Annual Meeting, 2001.

↓

…ral Representation

↓

Semantics

↓

Auditory Word Form

↓

Motor Planning

↓

Articulation

↓

« horse »

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Schematic representation of the cognitive processes underlying naming

- Early visual processing ⇒ recognition of the picture;
- Accessing the semantic representation (or meaning);
- Accessing the phonological (or auditory) form of the word;
- Programming the articulatory movements (lips, tongue, jaw, palate, etc) and their sequence;
- Articulating the word by implementing the planned movements.

Hillis AE, Course N° 2DS.008, American Academy of Neurology, 53rd Annual Meeting, 2001.

↓

Structural Representation

↓

Semantics

↓

Auditory Word Form

↓

Motor Planning

↓

Articulation

↓

« horse »

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Schematic representation of the cognitive processes underlying naming

- Early visual processing ⇒ recognition of the picture;
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- Accessing the phonological (or auditory) form of the word;
- Programming the articulatory movements (lips, tongue, jaw, palate, etc) and their sequence;
- Articulating the word by implementing the planned movements.

Hillis AE, Course N° 2DS.008, American Academy of Neurology, 53rd Annual Meeting, 2001.

↓

Structural Representation

↓

Semantics

↓

Auditory Word Form

↓

Motor Planning ⇒ Apraxia of Speech

↓

Articulation

↓

« horse »

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Developmental language disorders: Dysphasia = SLI

"*Specific language impairment* (SLI) is diagnosed when **for no obvious reason** a child fails to develop normal language but has adequate nonverbal intelligence, physical ability, and hearing." (Bishop & Adams, 1992).

Journal of Speech and Hearing Research, Volume 35, 119–129, February 1992

Comprehension Problems in Children With Specific Language Impairment: Literal and Inferential Meaning

D. V. M. Bishop
C. Adams*
Department of Psychology
University of Manchester, England

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Developmental language disorders: Dysphasia (SLI)

Folia Phoniatrica et Logopaedica

Report

Folia Phoniatr Logop 2013;65:68–77
DOI: 10.1159/000353896

Published online: August 12, 2013

Developmental Language Disorders: Challenges and Implications of Cross-Group Comparisons

Susan Ellis Weismer

Department of Communication Sciences and Disorders/Waisman Center, University of Wisconsin-Madison, Madison, Wisc., USA

SLI is typically defined as a developmental language disorder that is associated with **no known** sensory, **neurological**, intellectual, or emotional deficits.

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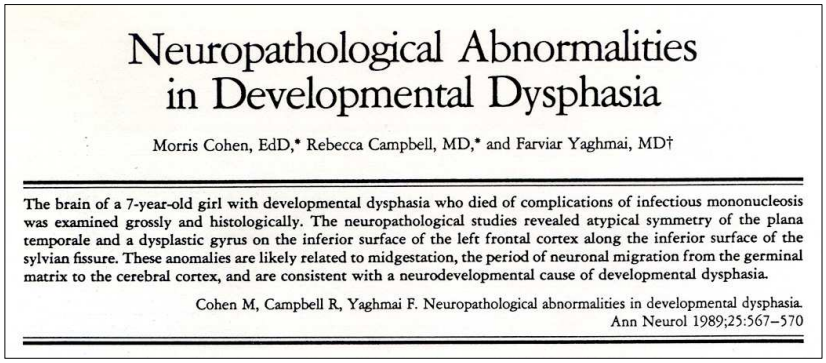
Dysphasia (SLI): No (known) neurological disorder?



Neuropathological Abnormalities in Developmental Dysphasia

Morris Cohen, EdD,* Rebecca Campbell, MD,* and Farviar Yaghmai, MD†

The brain of a 7-year-old girl with developmental dysphasia who died of complications of infectious mononucleosis was examined grossly and histologically. The neuropathological studies revealed atypical symmetry of the plana temporale and a dysplastic gyrus on the inferior surface of the left frontal cortex along the inferior surface of the sylvian fissure. These anomalies are likely related to midgestation, the period of neuronal migration from the germinal matrix to the cerebral cortex, and are consistent with a neurodevelopmental cause of developmental dysphasia.

Cohen M, Campbell R, Yaghmai F. Neuropathological abnormalities in developmental dysphasia. Ann Neurol 1989;25:567–570

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Dysphasia (SLI): No (known) neurological disorder?

Eur J Pediatr (1998) 157: 849–852

NEUROPEDIATRICS

S. Preis · V. Engelbrecht · Y. Huang · H. Steinmetz

Focal grey matter heterotopias in monozygotic twins with developmental language disorder

Received: 17 September 1997 / Accepted: 2 March 1998

Abstract We describe 9-year-old monozygotic male twins with a developmental language disorder of the phonologic-syntactic type and learning difficulties. High-resolution MRI revealed bilateral parieto-temporal grey matter heterotopias in both twins, on the left more than on the right, and more pronounced in the more affected twin. This suggests a causal relationship between the heterotopias and the neuropsychological findings in this twin pair.

Conclusion Neuronal migration defects and ensuing focal heterotopias may be causally related to developmental language disorders.

Key words Focal heterotopia · Neuronal migration · Developmental language disorder · Monozygotic twins

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Dysphasia (SLI): No (known) neurological disorder?

Am. J. Hum. Genet. 64:157–164, 1999

Quantitative-Trait Locus for Specific Language and Reading Deficits on Chromosome 6p

Javier Gayán,[1] Shelley D. Smith,[2] Stacey S. Cherny,[1,3] Lon R. Cardon,[4] David W. Fulker,[1,3] Amy M. Brower,[2] Richard K. Olson,[1] Bruce F. Pennington,[5] and John C. DeFries[1]

[1]Institute for Behavioral Genetics, University of Colorado, Boulder, Colorado; [2]Center for Hereditary Communication Disorders, Boys Town National Research Hospital, Omaha, Nebraska; [3]Social, Genetic, and Developmental Psychiatry Research Centre, Institute of Psychiatry, London; [4]Wellcome Trust Centre for Human Genetics, University of Oxford, Oxford; and [5]Department of Psychology, University of Denver, Denver

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Dysphasia (SLI): No (known) neurological disorder?

Am. J. Hum. Genet. 65:1215–1221, 1999

HUMAN GENETICS '99
Functional and Structural Brain Abnormalities Associated with a Genetic Disorder of Speech and Language

Kate E. Watkins,[1] David G. Gadian,[2] and Faraneh Vargha-Khadem[3]

[1]Cognitive Neuroscience Unit, Montreal Neurological Institute, McGill University, Montreal, and [2]Radiology and Physics Unit and [3]Cognitive Neuroscience Unit, Institute of Child Health, University College London Medical School, London

"KE family"

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Dysphasia (SLI): No (known) neurological disorder?

- KE family, large three-generation pedigree.
- Half of the members are affected by a severe speech/language disorder.
- Transmitted as an autosomal dominant monogenic trait.
- Responsible gene (FOXP2) on long arm of chromosome 7 (7q31).

Brain (2002), **125**, 465–478

MRI analysis of an inherited speech and language disorder: structural brain abnormalities

K. E. Watkins,[1] F. Vargha-Khadem,[1] J. Ashburner,[3] R. E. Passingham,[4] A. Connelly,[2] K. J. Friston,[3] R. S. J. Frackowiak,[3] M. Mishkin[5] and D. G. Gadian[2]

[1]*Developmental Cognitive Neuroscience Unit and* [2]*Radiology and Physics Unit, Institute of Child Health,* [3]*Wellcome Department of Cognitive Neurology, Institute of Neurology, University College London Medical School, London,* [4]*Department of Experimental Psychology, University of Oxford, Oxford, UK and* [5]*Laboratory of Neuropsychology, National Institutes of Mental Health, Bethesda, Maryland, USA*

Correspondence to: Kate Watkins, Cognitive Neuroscience Unit, Montreal Neurological Institute, 3801 University Street, Montreal, QC, Canada H3A 2B4
E-mail: kwatkins@bic.mni.mcgill.ca

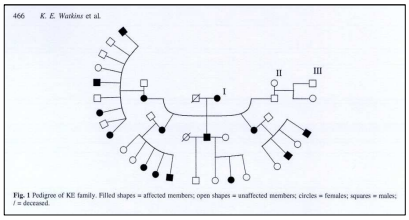

Fig. 1 Pedigree of KE family. Filled shapes = affected members; open shapes = unaffected members; circles = females; squares = males; / = deceased.

Brain 2002: 125: 465-478.

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Dysphasia (SLI): No (known) neurological disorder?

Human Brain Mapping 18:194–200(2003)

Bilateral Brain Abnormalities Associated With Dominantly Inherited Verbal and Orofacial Dyspraxia

Emma Belton,[1]* Claire H. Salmond,[1] Kate E. Watkins,[2] Faraneh Vargha-Khadem,[1] and David G. Gadian[3]

[1]*Developmental Cognitive Neuroscience Unit, Institute of Child Health, University College London, Great Ormond Street Hospital for Children, NHS Trust, London, United Kingdom*
[2]*Cognitive Neuroscience Unit, Montreal Neurological Institute, Montreal, Canada*
[3]*Radiology and Physics Unit, Institute of Child Health, University College London, Great Ormond Street Hospital for Children, NHS Trust, London, United Kingdom*

Abstract: The KE family is a large three-generational pedigree in which half of the members suffer from a verbal and orofacial dyspraxia in association with a point mutation in the FOXP2 gene. This report extends

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Dysphasia (SLI): No (known) neurological disorder?

European Journal of Human Genetics (2015), 1–5

www.nature.com/ejhg

SHORT REPORT

A highly penetrant form of childhood apraxia of speech due to deletion of 16p11.2

Evelina Fedorenko*[,1,9], Angela Morgan[2,3,9], Elizabeth Murray[2], Annie Cardinaux[4], Cristina Mei[2], Helen Tager-Flusberg[5], Simon E Fisher[6,7] and Nancy Kanwisher[4,8]

Childhood apraxia of speech is a rare motor speech disorder that affects the production, sequencing, and timing of sounds, syllables, and words. It is distinct from other speech (e.g., stuttering) and language (e.g., SLI) disorders.

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Dysphasia (SLI): No (known) neurological disorder?

Am. J. Hum. Genet. 70:384–398, 2002

A Genomewide Scan Identifies Two Novel Loci Involved in Specific Language Impairment*

The SLI Consortium*

Approximately 4% of English-speaking children are affected by specific language impairment (SLI), a disorder in the development of language skills despite adequate opportunity and normal intelligence. Several studies have indicated the importance of genetic factors in SLI: a positive family history confers an increased risk of development, and concordance in monozygotic twins consistently exceeds that in dizygotic twins. However, like many behavioral traits, SLI is assumed to be genetically complex, with several loci contributing to the overall risk. We have compiled 98 families drawn from epidemiological and clinical populations, all with probands whose standard language scores fall ⩾1.5 SD below the mean for their age. Systematic genomewide quantitative-trait–locus analysis of three language-related measures (i.e., the Clinical Evaluation of Language Fundamentals–Revised [CELF-R] receptive and expressive scales and the nonword repetition [NWR] test) yielded two regions, one on chromosome 16 and one on 19, that both had maximum LOD scores of 3.55. Simulations suggest that, of these two multipoint results, the NWR linkage to chromosome 16q is the most significant, with empirical P values reaching 10^{-3}, under both Haseman-Elston (HE) analysis (LOD score 3.55; $P = .00003$) and variance-components (VC) analysis (LOD score 2.57; $P = .00008$). Single-point analyses provided further support for involvement of this locus, with three markers, under the peak of linkage, yielding LOD scores >1.9. The 19q locus was linked to the CELF-R expressive-language score and exceeds the threshold for suggestive linkage under all types of analysis performed—multipoint HE analysis (LOD score 3.55; empirical $P = .00004$) and VC (LOD score 2.84; empirical $P = .00027$) and single-point HE analysis (LOD score 2.49) and VC (LOD score 2.22). Furthermore, both the clinical and epidemiological samples showed independent evidence of linkage on both chromosome 16q and chromosome 19q, indicating that these may represent universally important loci in SLI and, thus, general risk factors for language impairment.

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Dysphasia (SLI): No (known) neurological disorder?

NEWS & VIEWS

Late talking toddlers may have special genes

A twin study suggests that different genetic and nongenetic factors operate in the development of typical versus atypical vocabulary skills.

JEFFREY W. GILGER

OF THE MANY complex human traits, language is among the most interesting in reference to the nature–nurture debate. As a trait unique to humans, spoken language has all of the characteristics needed to fuel questions concerning the relative importance of nature and nurture. Most agree that language has been an adaptive ability in human evolution, implying that its roots have some sort of species-specific innate basis. So it is not surprising that a genetic component to individual differ-

within the normal range.

The hypothetical normal curve for continuously varying traits such as IQ, height and vocabulary is shown in Fig. 1*a*. Classic quantitative genetics assumes that such distributions primarily reflect the additive effects of multiple factors, namely many different genes and many different nongenetic influences. If the linguistic skills of

averaging of 'positive' and 'negative' factors determines a person's place on this scale, all else being equal. In a large population, with the opportunity for all the possible mixtures of factors, a normal curve results.

If, however, the continuum of vocabulary skills does not represent a simple continuum of the multiple factors, then the abilities of individuals at different levels may actually be a consequence of different etiologies. In this case, the general popu-

Nature Medicine 1998: 4: 892-893.

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Dysphasia (SLI): No (known) neurological disorder?

Reprinted from
Brain & Development 1992;14: 216–25

Epileptic Electroencephalographic Abnormalities and Developmental Dysphasias: A Study of 32 Patients

Bernard Echenne, MD, Renée Cheminal, MD, François Rivier, MD, Christian Negre, MD, Jacques Touchon, MD and Michel Billiard, MD

Electrical Status Epilepticus during slow Sleep (ESES)
Continuous Spikes and Waves during slow Sleep (CSWS)

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Dysphasia (SLI): No (known) neurological disorder?

Sleep EEG and developmental dysphasia

A Picard* MD, Service de Neurologie et de Rééducation Infantile;
F Cheliout Heraut MD, Service d'Explorations Fonctionnelles;
M Bouskraoui MD, Service de Neurologie et de Rééducation Infantile;
M Lemoine MD, Service d'Explorations Fonctionnelles;
P Lacert MD, Service de Neurologie et de Rééducation Infantile;
J Delattre MD, Service d'Explorations Fonctionnelles;
Hôpital Raymond Poincaré, 92380 Garches, France.

**Correspondence to first author.*

Developmental dysphasia is a disorder which severely affects the acquisition of oral language skills. This disorder occurs in children with normal intelligence who have neither auditory nor motor deficiencies, psychological problems nor difficulties with social communication (Rapin and Allen 1983).

While the underlying causes remain unknown, epilepsy has been suggested as a possible factor, as it is in Landau–Kleffner syndrome (Landau and Kleffner 1957, Deonna 1993). However, there are some important differences between this syndrome and developmental dysphasia. Landau–Kleffner syndrome is an acquired oral language disorder that affects both language comprehension and expression; in addition, paroxysmal abnormalities are frequently seen on EEG, especially within the temporal regions. When recorded over sleeping periods these abnormalities tend to increase during slow-wave sleep. Though some authors have reported dysphasic children with paroxysmal irregularities either during standard waking EEG (Sato and Dreifuss 1973, Maccario et al. 1982) or during sleep (Echenne et al. 1992, Duvelleroy-Hommet et al. 1995), these EEG anomalies affect only a certain number of infants with developmental dysphasia. To examine the relation between paroxysmal

Dev Med Child Neurol 1998: 40: 595-599.

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Dysphasia (SLI): No (known) neurological disorder?

Epilepsia, 50(Suppl. 7):21–24, 2009
doi: 10.1111/j.1528-1167.2009.02213.x

FIFTY YEARS OF LANDAU-KLEFFNER SYNDROME

Specific language impairment versus Landau-Kleffner syndrome

*Catherine Billard, †Joel Fluss, and *Florence Pinton

*Centre de Référence pour les Troubles des Apprentissage, Hôpital Bicêtre, France; and †Unité de Neuropédiatrie, Hôpital des Enfants, Genève, Switzerland

SUMMARY

The occurrence of sleep electroencephalography (EEG) abnormalities in some children with specific language impairment (SLI), the various forms of language dysfunction patterns seen in children with benign childhood epilepsy with centrotemporal spikes (BECTS), and finally the acquired aphasia in Landau-Kleffner syndrome (LKS) indicate a large spectrum of interactions between language and epilepsy. As such, the question is whether SLI and LKS should rather be considered along a continuum or as two entirely distinct entities. In addition, the rationale for using antiepileptic medications in rare forms of SLI is discussed.

KEY WORDS: Epilepsy, Landau-Kleffner syndrome, Centrotemporal spikes, Language development disorder.

Epilepsia 2009: 50 (Suppl. 7): 21-24.

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Dysphasia (SLI): No (known) neurological disorder?

Review

Nocturnal epileptiform EEG discharges, nocturnal epileptic seizures, and language impairments in children: Review of the literature

G.M. Overvliet [a,b,c,*], R.M.H. Besseling [e], J.S.H. Vles [c,d], P.A.M. Hofman [e], W.H. Backes [e], M.H.J.A. van Hall [c], S. Klinkenberg [d], J. Hendriksen [c], A.P. Aldenkamp [a,b,c]

[a] Department of Neurology, Maastricht University Medical Center, Maastricht, The Netherlands
[b] Research School of Mental Health and Neuroscience, Maastricht University Medical Center, Maastricht, The Netherlands
[c] Epilepsy Center Kempenhaeghe, Heeze, The Netherlands
[d] Department of Child Neurology, Maastricht University Medical Center, Maastricht, The Netherlands
[e] Department of Radiology, Maastricht University Medical Center, Maastricht, The Netherlands

Epilepsy & Behavior 2010: 19: 550-558.

"We suggest a spectrum or continuum of nocturnal epileptiform activity and language impairment that ranges from SLI at one end to LKS and ESES at the most affected end".

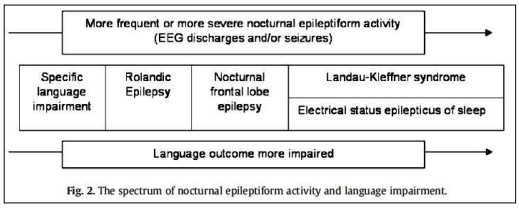

Fig. 2. The spectrum of nocturnal epileptiform activity and language impairment.

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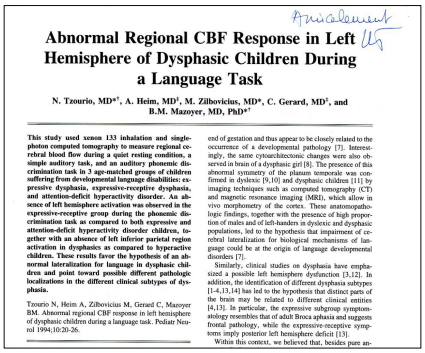
Dysphasia (SLI): No (known) neurological disorder?



Abnormal Regional CBF Response in Left Hemisphere of Dysphasic Children During a Language Task

N. Tzourio, MD*†, A. Heim, MD‡, M. Zilbovicius, MD*, C. Gerard, MD†, and B.M. Mazoyer, MD, PhD*†

This study used xenon 133 inhalation and single-photon computed tomography to measure regional cerebral blood flow during a quiet resting condition, a simple auditory task, and an auditory phonemic discrimination task in 3 age-matched groups of children suffering from developmental language disabilities: expressive dysphasia, expressive-receptive dysphasia, and attention-deficit hyperactivity disorder. An absence of left hemisphere activation was observed in the expressive-receptive group during the phonemic discrimination task as compared to both expressive and attention-deficit hyperactivity disorder children, together with an absence of left inferior parietal region activation in dysphasics as compared to hyperactive children. These results favor the hypothesis of an abnormal lateralization for language in dysphasic children and point toward possible different pathologic localizations in the different clinical subtypes of dysphasia.

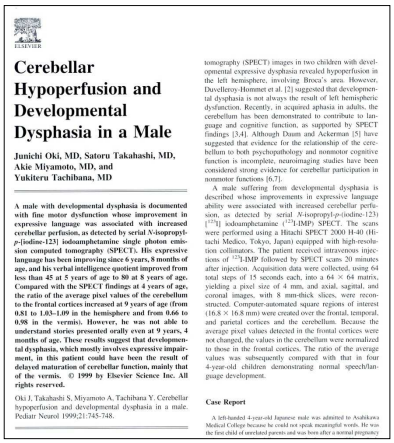
Tzourio N, Heim A, Zilbovicius M, Gerard C, Mazoyer BM. Abnormal regional CBF response in left hemisphere of dysphasic children during a language task. Pediatr Neurol 1994;10:20-26.

end of gestation and thus appear to be closely related to the occurrence of a developmental pathology [7]. Interestingly, the same cytoarchitectonic changes were also observed in brain of a dysphasic girl [8]. The presence of this abnormal symmetry of the planum temporale was confirmed in dyslexic [9,10] and dysphasic children [11] by imaging techniques such as computed tomography (CT) and magnetic resonance imaging (MRI), which allow in vivo morphometry of the cortex. These anatomopathologic findings, together with the presence of high proportion of males and of left-handers in dyslexic and dysphasic populations, led to the hypothesis that impairment of cerebral lateralization for biological mechanisms of language could be at the origin of language developmental disorders [7].

Similarly, clinical studies on dysphasia have emphasized a possible left hemisphere dysfunction [3,12]. In addition, the identification of different dysphasia subtypes [1-4,13,14] has led to the hypothesis that distinct parts of the brain may be related to different clinical entities [4,13]. In particular, the expressive subgroup symptomatology resembles that of adult Broca aphasia and suggests frontal pathology, while the expressive-receptive symptoms imply posterior left hemisphere deficit [13].

Within this context, we believed that, besides pure an-

Pediat Neurol 1994: 10: 20-26.



Cerebellar Hypoperfusion and Developmental Dysphasia in a Male

Junichi Oki, MD, Satoru Takahashi, MD, Akie Miyamoto, MD, and Yukiteru Tachibana, MD

A male with developmental dysphasia is documented with fine motor dysfunction whose improvement in expressive language was associated with increased cerebellar perfusion, as detected by serial *N*-isopropyl-*p*-[iodine-123] iodoamphetamine single photon emission computed tomography (SPECT). His expressive language has been improving since 6 years, 8 months of age, and his verbal intelligence quotient improved from less than 45 at 5 years of age to 80 at 8 years of age. Compared with the SPECT findings at 4 years of age, the ratio of the average pixel values of the cerebellum to the frontal cortices increased at 9 years of age (from 0.81 to 1.03–1.09 in the hemisphere and from 0.66 to 0.98 in the vermis). However, he was not able to understand stories presented orally even at 9 years, 4 months of age. These results suggest that developmental dysphasia, which mostly involves expressive impairment, in this patient could have been the result of delayed maturation of cerebellar function, mainly that of the vermis. © 1999 by Elsevier Science Inc. All rights reserved.

Oki J, Takahashi S, Miyamoto A, Tachibana Y. Cerebellar hypoperfusion and developmental dysphasia in a male. Pediatr Neurol 1999;21:745-748.

tomography (SPECT) images in two children with developmental expressive dysphasia revealed hypoperfusion in the left hemisphere, involving Broca's area. However, Duvelleroy-Hommet et al. [2] suggested that developmental dysphasia is not always the result of left hemispheric dysfunction. Recently, in acquired aphasia in adults, the cerebellum has been demonstrated to contribute to language and cognitive function, as supported by SPECT findings [3,4]. Although Daum and Ackerman [5] have suggested that evidence for the relationship of the cerebellum to both psychopathology and nonmotor cognitive function is incomplete, neuroimaging studies have been considered strong evidence for cerebellar participation in nonmotor functions [6,7].

A male suffering from developmental dysphasia is described whose improvements in expressive language ability were associated with increased cerebellar perfusion, as detected by serial *N*-isopropyl-*p*-(iodine-123) (^{123}I) iodoamphetamine (^{123}I-IMP) SPECT. The scans were performed using a Hitachi SPECT 2000 H-40 (Hitachi Medico, Tokyo, Japan) equipped with high-resolution collimators. The patient received intravenous injections of ^{123}I-IMP followed by SPECT scans 20 minutes after injection. Acquisition data were collected, using 64 total steps of 15 seconds each, into a 64 × 64 matrix, yielding a pixel size of 4 mm, and axial, sagittal, and coronal images, with 8 mm-thick slices, were reconstructed. Computer-automated square regions of interest (16.8 × 16.8 mm) were created over the frontal, temporal, and parietal cortices and the cerebellum. Because the average pixel values detected in the frontal cortices were not changed, the values in the cerebellum were normalized to those in the frontal cortices. The ratio of the average values was subsequently compared with that in four 4-year-old children demonstrating normal speech/language development.

Case Report

A left-handed 4-year-old Japanese male was admitted to Asahikawa Medical College because he could not speak meaningful words. He was the first child of unrelated parents and was born after a normal pregnancy

Pediat Neurol 1999: 21: 745-748.

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Dysphasia (SLI): No (known) neurological disorder?

J Autism Dev Disord (2010) 40:300–316
DOI 10.1007/s10803-009-0872-7

ORIGINAL PAPER

Cerebellum, Language, and Cognition in Autism and Specific Language Impairment

Steven M. Hodge · Nikos Makris · David N. Kennedy · Verne S. Caviness Jr. · James Howard · Lauren McGrath · Shelly Steele · Jean A. Frazier · Helen Tager-Flusberg · Gordon J. Harris

Normal controls: Left-sided IFG > Right-sided IFG and Right-sided cerebellar lobule VIIIA > Left-sided cerebellar lobule VIIIA.

SLI subjects: opposite pattern = larger Right-sided IFG and Left-sided VIIIA → *reversed asymmetry*.

Abstract We performed cerebellum segmentation and parcellation on magnetic resonance images from right-handed boys, aged 6–13 years, including 22 boys with autism [16 with language impairment (ALI)], 9 boys with Specific Language Impairment (SLI), and 11 normal controls. Language-impaired groups had reversed asymmetry relative to unimpaired groups in posterior-lateral cerebellar lobule VIIIA (right side larger in unimpaired groups, left side larger in ALI and SLI), contralateral to previous findings in inferior frontal cortex language areas. Lobule VIIA Crus I was smaller in SLI than in ALI. Vermis volume, particularly anterior I–V, was decreased in language-impaired groups. Language performance test scores correlated with lobule VIIIA asymmetry and with anterior vermis volume. These findings suggest ALI and SLI subjects show abnormalities in neurodevelopment of fronto-corticocerebellar circuits that manage motor control and the processing of language, cognition, working memory, and attention.

Keywords Autism · Specific language impairment · Cerebellum · Broca's area · Asymmetry

Introduction

Autism is a neurodevelopmental disorder displaying deficits in social interaction and communication skills, repetitive behaviors, and stereotyped interests (APA 1994). Language deficits range from absence of functional lan-

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Dysphasia (SLI): No (known) neurological disorder?

Is a definition of dysphasia (SLI) that includes “no known neurological deficits” still tenable ?

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Dysphasia (SLI): No (known) neurological disorder?

If a developmental language disorder is associated with a cytoarchitectonic anomaly present at birth, such as cortical dysplasia resulting from a neuronal migration disorder, is it dysphasia (SLI) or “congenital” aphasia ?

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Developmental language disorders: Congenital aphasia

Neurology, 10, 1960

Congenital aphasia

A clinicopathologic study

William M. Landau, M.D., Robert Goldstein, Ph.D. and Frank R. Kleffner, Ph.D.

CONGENITAL APHASIA has been recognized sporadically in the neurologic literature for well over half a century.[1–5] Several authors have been especially concerned with the labeling of the phenomenon, for the term aphasia is usually applied to a condition in which speech and language are affected after they develop nor-

scoliosis developed during the first year of life. He sat at 14 months and did not walk until 5 years. However, he was toilet trained at 18 months.

In 1952, at age 6, the patient was studied at the Johns Hopkins Hospital Cardiac Clinic. He was observed then to be poorly nourished

Neurology 1960: 10: 915-921.

Staudt M et al. Neurology 2001: 57: 122-125.

Dysphasia is not synonymous with *congenital aphasia*.

Congenital aphasia: A language disorder that is a consequence of early and –mostly– extensive lesions in the thalamo-cortical projection system which have occurred before language acquisition.

Due to such demonstrable structural lesions, children with congenital aphasia fail to develop normal language functions.

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Congenital aphasia: a contested concept

Language, Communication, and the Brain, edited by F. Plum. Raven Press, New York © 1988.

Language Sequelae of Unilateral Brain Lesions in Children

Dorothy M. Aram

Department of Pediatrics, Case Western Reserve University and Rainbow Babies and Children's Hospital, Cleveland, Ohio 44106

In: F. Plum (Ed.), Language, communication, and the brain. New York: Raven Press, 1988; pp. 171-197 .

Although significantly poorer performance was identified in early left-lesioned children than in controls across a range of syntactic and lexical language tasks, these deficits were mild, and often subtle, **with few children remaining clinically aphasic** (p. 195).

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Congenital aphasia: a contested concept

PLASTICITY, LOCALIZATION AND LANGUAGE DEVELOPMENT

Elizabeth Bates
University of California, San Diego

In: S.H. Broman & J.M. Fletcher (Eds.), The changing nervous system: Neurobehavioral consequences of early brain disorders. New York: Oxford University Press, 1999; pp. 214-253 .

- In the absence of other complications, infants with congenital, unilateral brain damage usually go on to acquire **language abilities that are well within the normal range** (p. 214).
- Children with a history of early brain injury typically perform below neurologically intact age-matched controls on a host of language tasks, but **they are not aphasic**, despite early damage of a sort that often leads to irreversible aphasia when it occurs in an adult (p. 214).

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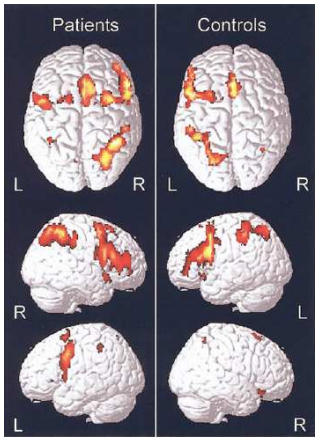
Congenital aphasia: a contested concept

Left-hemispheric (LH) brain lesions acquired early in life can induce language organization in the undamaged right hemisphere (RH). This study addresses the anatomical correlates of language processing in the RH of such individuals. Five hemiparetic patients with left periventricular brain lesions of pre- and perinatal origin were included, in whom fMRI during a word generation task had yielded predominantly RH activation; five age- and sex-matched healthy right-handers served as controls. The patterns of activation in the RH of patients showed a striking similarity with the LH patterns of the normal controls, and voxel-wise comparison failed to detect significant differences. This demonstrates that in patients with early LH damage, RH recruitment for language occurs in brain areas homotopic to the LH regions involved in language processing under normal circumstances.



Staudt M et al. NeuroImage 2002: 16: 954-967.

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Congenital aphasia: a contested concept

Neurocase, 2013
Vol. 19, No. 3, 209–231, http://dx.doi.org/10.1080/13554794.2011.654226
Routledge

Is a lone right hemisphere enough? Neurolinguistic architecture in a case with a very early left hemispherectomy

Laura Danelli[1], Giuseppe Cossu[2], Manuela Berlingeri[1], Gabriella Bottini[3,4], Maurizio Sberna[5], and Eraldo Paulesu[1,6]

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[3]Center of Cognitive Neuropsychology, Niguarda Ca' Granda Hospital, Milan, Italy
[4]Psychology Department, University of Pavia, Pavia, Italy
[5]Neuroradiology Department, Niguarda Ca' Granda Hospital, Milan, Italy
[6]IRCCS Galeazzi, Milan, Italy

"A lone right hemisphere may not be sufficient to guarantee full blown linguistic competences after early left hemispherectomy".

- The patient's fMRI patterns for several, basic linguistic tasks were similar to those observed in the dominant hemisphere of controls, suggesting that his language network confirms to a left-like blueprint of the linguistic network.
- Stronger right prefrontal activations compared to controls may represent the neurofunctional reflection of compensatory mechanisms necessary to achieve adequate language performance.

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Developmental language disorders: Congenital aphasia

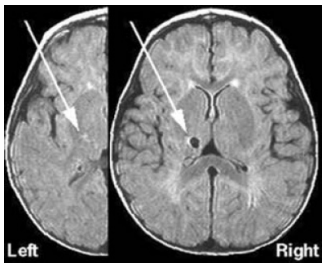
- Girl with early left thalamic vascular damage (before 9 months of age) who subsequently developed a language disorder.
- At 3 years and 8 months, her language was poor and unintelligible and showed phonetic, phonological and morpho-syntactic disorders. She did not exhibit any signs of mental retardation.
- This paper describes the effect of a thalamic injury in the earliest phases of language acquisition in a child who showed consistent phonological disorders.
- This case seems to confirm early hemispheric specialization and the importance of a timely therapy.

Neurocase, 10(4): 308–315, 2004

1355-4795/04/1004–308$16.00
Psychology Press

Language Disorder in a Child with Early Left Thalamic Lesion

Maria Rosa Pizzamiglio[1], Laura Piccardi[1], Marianna Nasti[2], Francesco Tomaiuolo[1] and Umberto Sabatini[1]
[1]IRCCS Fondazione Santa Lucia, Rome, Italy, and [2]Department of Psychology, University of Rome "La Sapienza," Italy



Pizzamiglio MR et al.
Neurocase 2004: 10: 308-315.

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Developmental motor speech disorders: Congenital dysarthria

Dysarthria

- A collective name for a group of related speech disorders that are due to disturbances in muscular control of the speech mechanism resulting from damage of the central or peripheral nervous system, and impairing any of the basic motor processes involved in the execution of speech.
- It can affect respiration, phonation, resonance, articulation, and prosody, either singly or in combination.

Darley FL et al. Motor Speech Disorders. Philadelphia: WB Saunders, 1975.

Duffy JR. Motor Speech Disorders: Substrates, Differential Diagnosis, and Management. St. Louis: Elsevier Mosby, 2005.

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Schematic representation of the cognitive processes underlying naming

- Early visual processing ⇒ recognition of the picture;
- Accessing the semantic representation (or meaning);
- Accessing the phonological (or auditory) form of the word;
- Programming the articulatory movements (lips, tongue, jaw, palate, etc) and their sequence;
- Articulating the word by implementing the planned movements.

Hillis AE, Course N° 2DS.008, American Academy of Neurology, 53rd Annual Meeting, 2001.

↓

Structural Representation

↓

Semantics

↓

Auditory Word Form

↓

Motor Planning

↓

~~**Articulation**~~ ⇒ **Dysarthria**

↓

« horse »

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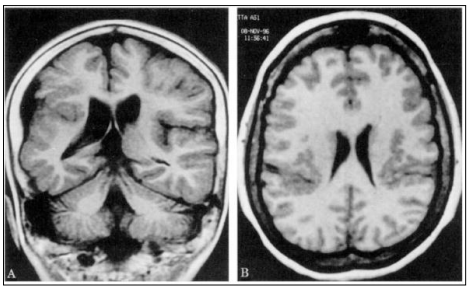
Developmental motor speech disorders: Congenital dysarthria

Malformations of cortical development (e.g., bilateral perisylvian polymicrogyria).

Congenital bilateral perisylvian syndrome: congenital diplegia of the facial, pharyngeal, and masticatory muscles, epilepsy, and mental retardation.



Borgatti R et al. Neurology 1999: 52: 1910-1913.

Clinical speech/language features:

- Moderate to severe **dysarthria** and nasal speech (+ drooling, dysphagia).
- Understanding commensurate with intelligence and always better than speech might suggest.
- Tongue movements consistently restricted, with very limited protrusion and lateral movements.

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Pediatric developmental speech and language disorders

The need for a transdisciplinary diagnostic and therapeutic approach:

- *Medical factors*: hearing; morphology and motility of the speech apparatus; neurological and/or psychiatric state.
- *Cognitive factors*: global learning ability; language sensitivity ("talent for languages").
- *Psychosocial factors*: stimulating family and school environment (parent-child interaction; home language and school language); housing conditions, family composition and conditions; emotional and behavioral development.

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Pediatrische spraak- en taalstoornissen: besluit

Bishop (2000):

"Rather than assuming that brain anomaly leads deterministically to disorder, it seems more appropriate to regard atypical brain structure as a risk factor that makes it more likely that the child will develop a disorder, but the nature, severity, and persistence of disorders are likely to depend on non-biological factors."