Brain and Language **80,** 592–602 (2002)
doi:10.1006/brln.2001.2620, available online at http://www.idealibrary.com on IDEAL®

Neuromotor Speech Deficits in Children and Adults with Spina Bifida and Hydrocephalus

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Published online February 20, 2002

Acquired cerebellar lesions are associated with motor speech deficits. Spina bifida with hydrocephalus (SBH) is a neurodevelopmental disorder that involves significant dysmorphology of the cerebellum. Videotaped narratives produced by 40 children and adults with SBH and their 40 age-matched controls were coded for three motor speech deficits: *dysfluency*, *ataxic dysarthria* (articulatory inaccuracy, prosodic excess, and phonatory–prosodic insufficiency) (Brown, Darley, & Aronson, 1970; Darley, Aronson, & Brown, 1969a), and *speech rate.* Individuals with SBH had more motor speech deficits than controls. Dysfluency was related to an interaction between chronological age and SBH. Speech rate was related independently to chronological age and SBH. Ataxic dysarthria was related to the biology of SBH, and was associated with both physical phenotype (level of spinal cord lesion) and medical history (number of shunt revisions). The data show that developmental as well as acquired lesions of the cerebellum disrupt motor speech, and add to the developmental role of the cerebellum in the automatization of motor skills, including speech. © 2002 Elsevier Science (USA)

Key Words: neuromotor disorders; motor speech; cerebellum; spina bifida; hydrocephalus; ataxic dysarthria; dysfluency.

INTRODUCTION

Speech production, a paradigmatic motor act, involves movement sequences that must be finely coordinated and temporally regulated. As part of a neural system that controls motor function, the cerebellum regulates the quality of movement by making it coordinated, automatic, and temporally organized (Hallett & Grafman, 1997; Keele & Ivry, 1991). It is not surprising, therefore, that the cerebellum should have an important role in motor speech production.

This research was supported by National Institute of Child Health and Human Development Grant P01 HD 35946, ''Spina Bifida: Cognitive and Neurobiological Variability''; by project grants to the second author from the Ontario Mental Health Foundation; and by a National Cancer Institute of Canada grant to the fourth author. This research was also funded by Doctoral Research Training Grant No. 9908 from KidsAction Research Society and by a Doctoral Research Training Grant from the Easter Seal Society, Ontario, to the first author. Portions of this research were presented at the Twenty Ninth Annual International Neuropsychological Society Conference, February 14–17, 2001, Chicago, Illinois (*Journal of the International Neuropsychological Society,* vol. 7, no. 2, 2001).

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0093-934X/02 $35.00
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Acquired lesions of the cerebellum in adult life have long been known to affect the regulation of movement, including speech (Holmes, 1956). The motor speech disorders associated with adult cerebellar lesions include ataxic dysarthria, dysfluency, and problems in speech rate.

Cerebellar lesions disrupt the coordination of prosody, articulation, phonation, temporal regulation, and fluency of speech production, particularly of syllables within phrases but also of phonemes within syllables (Kent, Netsell, & Abbs, 1979). The importance of the cerebellum for speech fluency is supported by functional brain imaging studies (Fox et al., 2000).

The motor speech deficits associated with cerebellar lesions, collectively termed *ataxic dysarthria,* are characterized by three clusters of features (Darley, Aronson, & Brown, 1969a; Brown, Darley, & Aronson, 1970). *Articulatory inaccuracy* includes imprecise consonants, irregular articulatory breakdowns, and distorted vowels; it has been associated with inaccuracy of repetitive movements (Brown et al., 1970; Darley et al., 1969a). *Prosodic excess* includes excess and equal stress, prolonged phonemes, slow speech rate, and prolonged intervals between words and syllables; it has been related to motor slowing (Brown et al., 1970; Darley et al., 1969a). *Phonatory–prosodic insufficiency* includes harshness, monopitch, and monoloudness; it may be associated with hypotonia of speech musculature (Brown et al., 1970; Darley et al., 1969a).

Most studies of motor speech deficits have involved adults with acquired cerebellar lesions. Few studies have been conducted with children, despite evidence that developmental and acquired cerebellar compromise is common in childhood, and that severe motor speech deficits caused by acquired cerebellar tumors may be more prevalent in children than in adults (Turgut, 1998).

Many childhood disorders are associated with cerebellar compromise, including genetic, congenital, behavioral, and acquired conditions. Cerebellar agenesis and/or hypoplasia is associated with Marinesco–Sjorgren Syndrome (Williams, Buchhalter, & Sussman, 1996); ataxia–telangioctasia (Mostofsky, Kunze, Cutting, Lederman, & Denckla, 2000); Fragile X Syndrome (Mostofsky et al., 1998); Dandy–Walker Syndrome (Pellock & Johnson, 1993); Down Syndrome (Jernigan & Bellugi, 1990); and prenatal alcohol exposure (Sowell et al., 1996). Reduced cerebellar size has also been found in male children with Attention Deficit Hyperactivity Disorder (Mostofsky, Reiss, Lockhart, & Denckla, 1998; Berquin et al., 1998).

The posterior fossa is a common site for brain tumors in childhood (Heideman, Packer, Albright, Freeman, & Rorke, 1993), and childhood cerebellar tumors, such as malignant medulloblastomas, are associated with disturbances of neuropsychological function (Dennis, Spiegler, Hetherington, & Greenberg, 1996; Levisohn, Cronin-Golumb, & Schmahmann, 2000; Riva & Giorgi, 2000). More specifically, cerebellar tumors originating in childhood are associated with deficits in both motor speech (Huber-Okrainec, Dennis, Bradley, & Spiegler, 2001a) and perceptual timing (Hetherington, Dennis, & Spiegler, 2000).

Transient cerebellar mutism has been reported following the surgical removal of posterior fossa tumors in some children (e.g., Dennis, 1996; Ferrante, Mastronareli, Acqui, & Fortuna, 1990; Rekate, Grubb, Aram, Hahn, & Ratcheson, 1985; Riva & Giorgi, 2000; van Dongen, Catsman-Berrevoets, & van Mourik, 1994). While mutism occasionally does occur after adult cerebellar lesions (e.g., Çakir, Karakişi, & Koçanogŭllari, 1994; Dunwoody, Alsagoff, & Yuan, 1997; Salvati, Missori, Lunardi, & Orlando, 1991), it is more prevalent in children than in adults (Turgut, 1998). Following posterior fossa surgery in childhood, speech may be fluent, but then a variable period of mutism may emerge that eventually resolves into a dysarthria (Catsman-Berrevoets, van Dongen, & Zwetsloot, 1992; Rekate et al., 1985). In the longer term,

the dysarthria may improve (van Dongen et al., 1994; Catsman-Berrevoets et al., 1992) although it does not resolve completely (Huber-Okrainec, Dennis, Bradley, & Spiegler, 2001b). Most children with transient cerebellar mutism have comorbid hydrocephalus, which, it has been suggested, may intensify the mutism (Turgut, 1998).

Young adult survivors of childhood posterior fossa tumors have implicit timing deficits which are specific to perception of short duration time intervals and do not involve perception of long durations or frequency estimations (Hetherington et al., 2000). Children with ataxia–telangiectasia also show deficits in judging the duration of explicit time intervals (Mostofsky et al., 2000).

Spina bifida with hydrocephalus (SBH) is a common neurodevelopmental disorder. It results from incomplete closure of the neural tube early in gestation, and is associated with developmental anomalies of both the spine and the brain, including cerebellar dysmorphology (Barkovich, 1994). Nearly all children born with SBH exhibit the Arnold–Chiari II malformation, which involves a small posterior fossa, cerebellar dysmorphology, and displacement of the cerebellum into the spinal canal. In the Arnold–Chiari II malformation, the inferior part of the cerebellar vermis herniates downward, while the superior portion is shifted upward. The cerebellar hemispheres are compressed and displaced around the brainstem, and sometimes hypoplasia and agenesis of the lateral cerebellar hemispheres are observed (Barkovich, 1994).

Individuals with SBH show a signature cognitive profile that includes problems in motor function (Hetherington & Dennis, 1999), visual perception (Dennis, Fletcher, Rogers, Hetherington, & Francis, 2002), procedural knowledge (Barnes et al., 2002), and inferencing (Barnes & Dennis, 1998; Dennis, Barnes, & Hetherington, 1999). Individual variability around the cognitive profile (Fletcher, Dennis, & Northrup, 2000) appears (in young adulthood) to be related to factors such as the level of the spinal cord lesion (Dennis et al., in press) and the lifetime number of shunt revisions (Dennis & Barnes, in press).

Motor function is compromised in children and adults with SBH. Children with SBH have a variety of upper limb deficits (Hetherington & Dennis, 1999) that persist into adulthood (Dennis et al., in press) just as deficits in cognitive domains such as mathematics persist into adulthood (Dennis & Barnes, in press). Many of the motor tasks on which SBH individuals are impaired are classical cerebellar tasks, such as the rapid recruitment of temporally organized movements, suggesting that congenital cerebellar dysmorphology in SBH may produce poor automatization of motor skills (Dennis, Hetherington, Spiegler, & Barnes, 1999).

Studies of expressive language in individuals with SBH have focused more on language style than on motor speech characteristics. SBH has been associated with an expressive language style, termed the ‘‘Cocktail Party syndrome,’’ involving superficially fluent but content-poor or empty speech filled with stereotypic phrases (e.g., Taylor, 1961; Dennis, Hendrick, Hoffman, & Humphreys, 1987).

The evidence does suggest that motor speech is compromised in SBH. Some children with SBH show dysarthric characteristics, such as problems in intelligibility, fluency, prosody, vocal intensity, and quality of speech (Murdoch, Ozanne, & Smyth, 1990). Also, children and adolescents with SBH are less fluent and slower than age peers in rapid naming tasks, with the problem being not so much one of naming as of pacing speech production in a smooth, regulated manner (Dennis et al., 1987).

We studied motor speech in children and young adults with SBH. We asked:

- Do individuals with SBH exhibit motor speech deficits in spontaneous narrative speech? Because bilateral cerebellar dysmorphology is an important feature of the Arnold–Chiari II malformation of SBH, we hypothesized that individuals

with SBH would show impairments of motor speech, specifically, dysfluency, ataxic dysarthria, and slowed speech rate.

- Are there age-related motor speech deficits in individuals with SBH? Three alternative hypotheses were entertained: (a) because cerebellar mutism is more common and more pervasive in children than in adults, motor speech deficits might be more prominent in children than in adults with SBH; (b) some cognitive deficits such as those in mathematics occur in both children and adults (e.g., Barnes et al., 2002; Dennis & Barnes, in press), so children and adults with SBH might show similar motor speech deficits; or (c) the longer an individual lives with SBH, the more opportunity for repetitive increases in intracranial pressure and episodes of acute hydrocephalus, so older individuals with SBH might show more motor speech deficits than younger individuals.
- Within SBH groups, is variability in motor speech related to some of the same factors that account for variability in other cognitive functions? We hypothesized that motor speech deficits in both children and adults with SBH would be associated with more shunt revisions and higher spinal lesion levels.

METHODS

Participants

Eighty first-language speakers of English were assigned to four groups on the basis of medical condition and age: children with SBH ($N = 20$); child controls ($N = 20$); adults with SBH ($N = 20$); and adult controls ($N = 20$) (Table 1). SBH participants had been treated for hydrocephalus with a diversionary shunt shortly after birth. Control participants were siblings, relatives, and friends of survivors of posterior fossa tumors. There were no significant age differences between children with SBH and child controls, although adults with SBH were younger than adult controls [$F(1, 38) = 2.1$, $p = .0438$]. Inclusion in the study required each participant to have obtained a Verbal IQ or Performance IQ score of 70 or above on a standard, age-appropriate intelligence test: for SBH children, the Stanford–Binet Intelligence Scale–Fourth Edition (the Vocabulary subtest of Verbal Reasoning, and the Pattern Analysis subtest of Abstract Visual Reason were used) (Thorndike, Hagen, & Sattler, 1986); for child controls, the Wechsler Intelligence Scale for Children—Third Edition (Wechsler, 1991); and for adults with SBH and adult controls, the Wechsler Adult Intelligence Scale—Revised (Wechsler, 1981).

Procedure

A narrative discourse speech sample was elicited using a picture-prompted narrative speech task. Participants were videotaped individually while they told a story from a children's picture storybook (*Frog where are you?*, Mayer, 1969). Each videotaped narrative speech sample was subsequently transcribed.

The number of words in each narrative was measured using a hand counter. All participants told multiword stories (number of words in narrative: SBH children mean 444.5, $SD = 143.4$, range 230–749; control children mean 423.2, $SD = 173.3$, range 149–874; SBH adults mean 585.1, $SD = 151.9$,

TABLE 1
Sample Characteristics

	SBH child ($n = 20$)	Control child ($n = 20$)	SBH adult ($n = 20$)	Control adult ($n = 20$)
Age at test (years)	12.51 (2.20)	12.24 (3.79)	25.38 (4.98)	28.44 (6.09)
Gender (F/M)	(8/12)	(12/8)	(7/13)	(13/7)
Verbal IQ	93 (7.66)	101 (13.68)	97 (8.61)	101 (9.84)
Performance IQ	92 (17.85)	105 (14.79)	85 (10.02)	103 (12.61)

Note. Values are means (*SD*).

range 320–785; adult controls mean 567.6, *SD* = 172.5, range 209–785 words). There were no significant differences in number of words in narratives between SBH and control groups at each age level.

Two speech–language pathologists perceptually judged each videotaped narrative speech sample and coded the number and type of motor speech deficits. The speech–language pathologists rated motor speech independently for every participant, and inconsistencies were later resolved by consensus until full agreement was reached on each narrative speech sample.

Measures

Dysfluency. Dysfluency was coded using a modification of Yaruss' (1998) fluency count coding system. Dysfluency measures were defined and coded as follows:

Blocks (a marked break at the beginning of or within a word);
Prolongations (the prolongation of a sound at the beginning of or within a word);
Part-Word Repetitions (including phoneme and syllable repetitions, e.g., ''The *b*- bees chased the dog'' and ''That looks like a *moo*-moose'');
Word Repetitions (e.g., ''*It it* it is a moose'');
Phrase Repetitions (e.g., ''*The frog was*- the frog was happy'');
Interjections (such as ''uh'' or ''um'' within a phrase);
Word Changes or Unfinished Words (switching from one word to another within a phrase, e.g., ''The boy picked up the *dog* frog'' when the intent was to say ''The boy picked up the frog''; or ''The boy picked up the *do*- frog'' when the intent was to say ''The boy picked up the frog''); and
Phrase Revisions (e.g., ''*The next day when*–the next morning when the boy woke-up he saw that the frog was missing'').

The percentage dysfluency score was the number of dysfluencies divided by the total number of words in the narrative.

Ataxic dysarthria. No pediatric dysarthria scale currently exists, although there is need for such an assessment tool (van Mourik, Catsman-Berrevoets, Yousef-Bak, Paquier, & van Dongen, 1998). The Dysarthria Rating Scale is an acknowledged measure for the differential diagnosis of adult dysarthria (Duffy, 1995, based on Darley et al., 1969a, 1969b). The presence, not the severity, of ataxic dysarthric speech characteristics was judged in each narrative because the presence rather than the severity of ataxic dysarthric speech features is more meaningful for diagnosis (Duffy, 1995).

The ataxic dysarthria speech characteristics (defined by Darley et al., 1969a, 1969b, and Brown et al., 1970) judged were:

Articulatory Inaccuracy (imprecise consonants, irregular articulatory breakdowns, distorted vowels, and repeated phonemes);
Prosodic Excess (excess and equal stress, prolonged phonemes, slow rate, and prolonged intervals). ''Short phrasing'' was also incorporated into the Prosodic Excess category;
Phonatory–Prosodic Insufficiency (harshness, monopitch, and monoloudness).

Also included for analysis in this category was the feature ''strain-strangled speech.''

Speech rate. Speech rate over time was measured (by stopwatch) as total speaking time per minute (excluding intervals between phrases and pausing for page turning from the timing measure, and excluding dysfluencies such as interjections, repeated words and phrases, and phrase revisions in the total word count). Speech rate was the total number of words divided by the total speaking time. Each sample was timed twice by one judge, and the average of the two times was used.

Medical variables for SBH participants. The number of shunt revisions was established from medical records and/or direct parent enquiry. This provides a measure of the stability of the hydrocephalus condition over time.

During gestational development, the neural tube closes in stages and at multiple sites (Van Allen et al., 1993). On the basis of medical records, spinal lesion level was coded as *Upper* (closure failure at or above L1, within Van Allen's Closure site 1), or *Lower* (closure failure at or below L2, within Van Allen's Closure site 5).

RESULTS

Dysfluency

The Dysfluency results are shown in Table 2. An ANOVA was conducted on the dysfluency results with condition and age group as between-group factors. Individuals with SBH showed more overall dysfluency than controls [$F(1, 76) = 21.8$,

TABLE 2
Dysfluency

	SBH child (n = 20)	Control child (n = 20)	SBH adult (n = 20)	Control adult (n = 20)
Overall dysfluency (%)	4.7 (2.6)	3.1 (1.2)	5.5 (4.7)	1.3 (0.9)
Blocks	0.1 (0.4)	0.0 (0.0)	0.0 (0.0)	0.0 (0.0)
Prolongations	0.5 (0.9)	0.0 (0.0)	0.5 (0.8)	0.1 (0.3)
Part-word repetitions	1.6 (2.0)	1.4 (1.8)	1.9 (1.4)	0.1 (0.5)
Word repetitions	2.5 (3.1)	1.2 (1.6)	3.8 (3.9)	1.1 (1.6)
Phrase repetitions	2.5 (3.0)	1.8 (2.2)	3.5 (3.4)	0.2 (0.7)
Interjections	3.9 (4.4)	2.9 (2.8)	12.9 (23.1)	2.5 (3.7)
Word changes	3.9 (3.3)	2.4 (2.2)	3.0 (3.5)	1.2 (1.0)
Phrase revisions	6.4 (5.1)	3.5 (2.6)	7.2 (7.9)	1.9 (2.9)

Note. Values are means (*SD*).

$p = < .0001$]; specifically, they showed more prolongations [$F(1, 76) = 10.3$, $p = .0019$], part-word (phoneme and syllable) repetitions [$F(1, 76) = 8.1$, $p = .0057$], word repetitions [$F(1, 76) = 9.5$, $p = .0028$], phrase repetitions [$F(1, 76) = 12.4$, $p = .0007$], interjections [$F(1, 76) = 4.5$, $p = .0364$], word changes or unfinished words [$F(1, 76) = 7.6$, $p = .0072$], and phrase revisions [$F(1, 76) = 12.7$, $p = .0006$]. These results were qualified by a significant interaction between condition and age group such that, compared to adult controls, young adults with SBH were overall more dysfluent [$F(1, 76) = 4.3$, $p = .0408$], and showed more part-word repetitions [$F(1, 76) = 4.6$, $p = .0360$], phrase repetitions [$F(1, 76) = 4.9$, $p = .0305$], and interjections [$F(1, 76) = 3.1$, $p = .0830$].

Ataxic Dysarthria

The Ataxic Dysarthria results are shown in Table 3. An ANOVA was conducted with condition and age group as between-group factors. Results indicated that individuals with SBH exhibited significantly more ataxic dysarthric characteristics than controls [$F(1, 76) = 40.7$, $p < .0001$]: they had more Articulatory Inaccuracies [$F(1, 76) = 21.6$, $p < .0001$]; more Phonatory–Prosodic Insufficiency [$F(1, 76) = 15.4$, $p = .0002$]; and more Prosodic Excess [$F(1, 76) = 42.1$, $p < .0001$]. These results were qualified by an interaction between condition and age group for the Prosodic Excess variable [$F(1, 76) = 6.0$, $p = .0165$], such that young adults with spina bifida had significantly more prosodic excess than children with spina bifida.

TABLE 3
Ataxic Dysarthria

	SBH child (n = 20)	Control child (n = 20)	SBH adult (n = 20)	Control adult (n = 20)
Total number of ataxic dysarthric features	2.5 (2.5)	0.5 (0.6)	3.6 (2.5)	0.5 (0.8)
Articulatory inaccuracy[a]	0.7 (1.0)	0.2 (0.4)	1.1 (1.1)	0.0 (0.0)
Prosodic excess[b]	0.8 (1.1)	0.1 (0.2)	1.6 (1.1)	0.0 (0.0)
Phonatory–prosodic insufficiency[c]	1.1 (1.0)	0.2 (0.4)	1.0 (1.0)	0.5 (0.8)

Note. Values are means (*SD*).
[a] Total possible number of features: 4.
[b] Total possible number of features: 5.
[c] Total possible number of features: 3.

TABLE 4
Speech Rate

	SBH child (n = 20)	Control child (n = 20)	SBH adult (n = 20)	Control adult (n = 20)
Speech rate (words/min)	154.0 (26.8)	171.8 (27.3)	167.2 (38.6)	215.3 (22.7)

Note. Values are means (*SD*).

Speech Rate

The Speech Rate results are shown in Table 4. An ANOVA was conducted on speech rate with condition and age group as between-group factors. Individuals with SBH had significantly slower speech rates than controls [$F(1, 76) = 25.1$, $p < .0001$]. Age also affected speech rate [$F(1, 76) = 18.5$, $p < .0001$], and control adults had faster speech rates than control children. There was an interaction between clinical group and age such that adults with SBH had significantly slower speech rates than control adults [$F(1, 76) = 5.3$, $p = .0240$]. In fact, the speech rates (words per minute) of adults with SBH were like those of the control children (SBH adult mean 167.2; child control mean 171.8; SBH child mean 154.0; and adult control mean 215.3).

Medical Variables

An ANOVA on motor speech scores for the SBH individuals using number of shunt revisions and age group as between-group factors showed that number of shunt revisions was not related to the percentage dysfluency or speech rate scores, but that a greater number of shunt revisions was associated with more dysarthric features [$F(1, 35) = 9.7$, $p = .0037$]. There were no significant interactions between number of shunt revisions and age group.

An ANOVA on motor speech scores using spinal lesion level (upper, lower, and controls) and age group as between-group factors showed that spinal lesion level was unrelated to dysfluency or speech rate. For the ataxic dysarthric features measure, there was a significant interaction between lesion level and age group [$F(1, 36) = 4.6$, $p = .0392$]. Children with SBH who had upper lesions had more dysarthric features than either children with lower lesions or controls; adults with SBH had a higher number of dysarthric features than controls regardless of lesion level (Fig. 1). A chi-square analysis showed that there was no significant association between shunt group and spinal lesion level for children or adults with SBH.

DISCUSSION

Motor speech deficits are evident in individuals with SBH, even those individuals who narrated full, superficially fluent stories at least 100 words in length. In comparison to age-matched controls, individuals with SBH were more dysfluent, had more ataxic dysarthric features, and had a slower speech rate.

Speech involves both motor and timing functions, both of which appear to be impaired in individuals with SBH. In addition, the different motor speech impairments may have an additive effect on function. Speech dysfluencies contribute to the inefficiency of speech rate by slowing the number of words produced, which attenuates the information in the narrative per unit of time. For example, in the dysfluent sentence ‘‘*The boy put on*—the boy put on his shirt and went outside’’ (where the dysfluent phrase ‘‘*the boy put on*’’ is a phrase repetition and therefore is not counted),

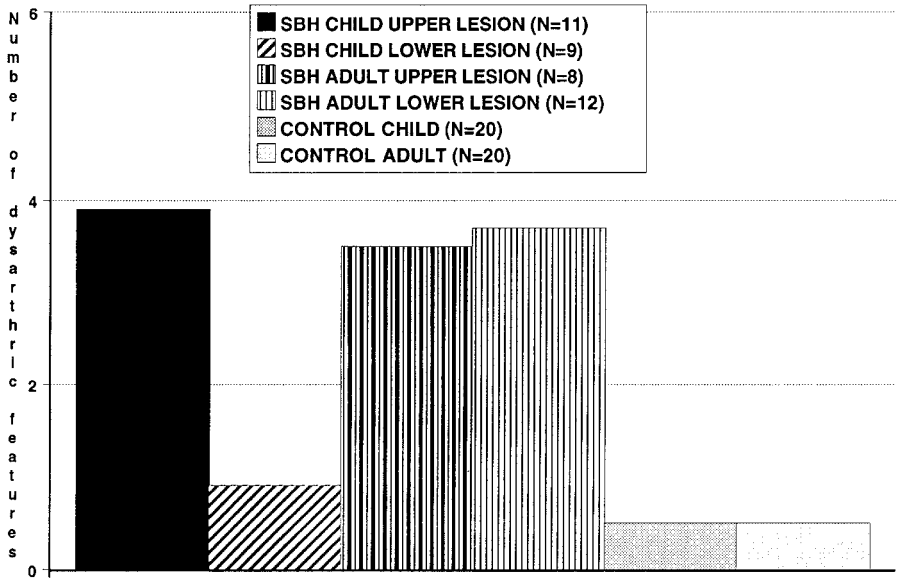

FIG. 1. Interaction effects for number of dysarthric features between spinal lesion level and age group.

the word count would be nine words: ''*The boy put on* (not counted) (1) The (2) boy (3) put (4) on (5) his (6) shirt (7) and (8) went (9) outside'' rather than 13 words.

The effects of SBH on motor speech were not stable through the lifespan. Instead, they were associated with variability in physical phenotype, chronological age, and aspects of medical history. Each measure, however, showed a different pattern. Dysfluency was related to an interaction between chronological age and SBH. Speech rate was related independently to chronological age and SBH. Ataxic dysarthria was related to SBH in a complex manner: it showed main effects of SBH, rather than interactions, and it was associated with both physical phenotype and medical history.

Spinal lesion level is a biological constant throughout life, established in the embryo under genetic control (genetic heterogeneity in the form of mutation in the gene regulating folate and homocysteine metabolism occurs in the mothers of children with upper but not lower spinal lesions, Volcik et al., 2000). The effects of the physical SBH phenotype, level of spinal lesion, were observed only in children; SBH children with upper spinal lesions showed more dysarthric features than either child controls or SBH children with lower spinal lesions. It is not clear whether the effect of spinal lesion level on motor speech is a direct effect on the mechanics of speech production, or a more indirect effect mediated through other influences on breathing, motor control, or the brain systems that control the timing and production of speech.

Increasing chronological age produced a motor speech disadvantage. As a group, adults with SBH showed more motor speech deficits than both their age controls and SBH children. For some motor speech functions, the long-term effect of SBH appears to be a developmental arrest; for example, the developmental improvements in speech rate and fluency seen in controls do not seem to occur in individuals with SBH, so that the speech rate of 24-year-old individuals with SBH is like that of younger, typically developing children.

The greater frequency and type of motor speech deficits in SBH adults may represent the cumulative effect of longstanding clinical or subclinical hydrocephalus on motor and timing functions. In support of this, a greater number of shunt revisions was associated with more ataxic dysarthric features, and adults with SBH showed

many features of ataxic dysarthria regardless of spinal lesion level. In adulthood, progressive damage to motor control of speech might occur as a function of age and medical complications involved in episodes of increased intracranial pressure.

The fact that adults with SBH have more motor speech difficulties than children, combined with the fact that spinal lesion level is more important for motor speech in childhood than in adulthood, suggests that factors associated with living with SBH for many years, with attendant clinical and subclinical increases in intracranial pressure, may override spinal lesion effects that can be seen in childhood. These data are consistent with the proposal that brains with congenital dysmorphologies have limited ability to cope with the challenges of aging and minor repetitive brain insults such as increases in intracranial pressure (Dennis, Spiegler, & Hetherington, 2000).

Acquired lesions of the cerebellum, particularly bilateral lesions, are known to affect motor speech in both children and adults. In this article, we show that congenital malformations of the cerebellum associated with SBH are also associated with motor speech deficits in both childhood and young adulthood. The data are consistent with the proposed role of the cerebellum in the automatization of a variety of learned motor behaviors, including speech production. One question yet to be examined is the relationship between perceptual timing deficits in SBH and motor speech, which would provide more information about how congenital cerebellar dysmorphologies in SBH affect the neural computations involved in timing. While bilateral cerebellar dysmorphology is characteristic of SBH, the type and degree of dysmorphology vary among individuals. Another question to be examined is individual variability in motor speech as it relates to cerebellar dysmorphology.

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