A Syndrome of Neuromotor Speech Deficit and Dysgraphia?

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In the last two years a few patients, including the five to be reported on in this study, have been referred shortly after onset with what is to us an intriguing syndrome of neuromotor speech disorder and dysgraphia. This report summarizes the speech and language findings, neurological examinations, and results of special testing for those patients.

METHOD

Diagnostic Battery. All subjects received a speech and language evaluation, a neurological examination, and either a cat (four subjects) or a brain scan. One subject also came to autopsy. The speech-language evaluation consisted of either the Mayo adaptation of the Minnesota Test for Differential Diagnosis of Aphasia (Schuell, 1965) or the Porch Index of Communicative Ability (PICA) (Porch, 1967); a form of the Token Test (DeRenzi and Vignolo, 1962); the Word Fluency Measure (Borkowski, Benton, and Spreen, 1967); and a standard speech examination, which samples reading, imitation of syllables, words, and sentences, and spontaneous speech (Wertz and Rosenbek, 1971). Three of the five subjects also received LaPointe and Horner's Reading Comprehension Battery for Aphasia (RCBA, 1979). Selected descriptive data appear in Table 1.

Table 1. Selected descriptive data for the 5 subjects.

PATIENT DESCRIPTIVE DATA						
PATIENT	AGE	MPO	PICA OA	TOKEN TEST	RCPM	WFM
A.A.	56	0.7	12.60	49/61	26	24
D.A.	50	0.4	10.53	15/61	11	7
R.E.	63	0.9	10.47	36/61	23	7
F.P.	74	0.3	---	34/61	28	29
S.S.	68	0.2	---	26/61	25	13

Diagnostic Procedures. One of the authors (JCR) screened each of the five subjects for probable neuromotor speech dysfunction and inordinately poor writing. Three other authors (MM, KO, and MC) then listened to speech samples from the five subjects and to speech samples from four other subjects with somewhat similar symptoms. The judges agreed that the five subjects were similar to each other and different from the foils.

One of the authors (KO) then evaluated the speech samples using as a guide a slightly modified version of the perceptual symptoms checklist from the Darley, Aronson, and Brown (1969 a,b) studies of the dysarthrias. The modifications were limited to redefinition of certain types of articulatory errors. This judge's reliability had previously been guaranteed. We also asked Dr. Ray Kent and Dr. Ron Netsell to listen to three of the taped samples, to describe what they heard, and to label it if possible. The writing was evaluated by one author (MM). He first made a list of all the error types in common use for writing analysis. He then tabulated the number of each error type for the five subjects. The samples were then compared to each other and to published samples to aid our hypothesizing about underlying mechanisms. One of the authors (MT) reviewed each neurological examination and special test.

RESULTS

Speech. The speech patterns of these five are not identical; they are similar. In Figures 1 and 2 we have displayed the error types for the five. The solid bar extending all the way across indicates that a symptom was judged to be present for all five patients. Inconsistent sound substitutions, eclipsing, (omission of medial sounds and syllables), and cluster reduction were present in all five. Stuttering, in the form of sound and syllable repetition, was prominent in the speech of four, as was abnormally fast rate. With the exception of harsh voice, also characteristic of four of the five, other symptoms are scattered. Along with Kent and Netsell, we hypothesized that the bulk of errors were neuromotor rather than linguistic and resulted from timing and coordination abnormalities.

Writing. Writing was more impaired than the other language modalities for all five. However, the pattern of writing deficits differed slightly from subject to subject. Two groups seem to have emerged. Three of the subjects (Group I) exhibited the following symptoms. 1) severe tremor or other incoordination in their script, 2) macrographia (immediately following onset which recovered completely by time of discharge), 3) a high percentage of omitted functor words, 4) letter reversals, and 5) letter transpositions.

The other two subjects (Group II) demonstrated 1) perseverations of content words and 2) strings of intelligible letters which were without meaning (jargon words).

Subjects from Groups I and II shared a variety of other writing deficiencies. These errors consisted of 1) reduplication of M, N, W, and U (that is, additions of humps or loops); 2) unlinked or unclosed letters; 3) unintelligible letters, 4) misused content words, including verbal paragraphia; 5) letter omissions; 6) letter substitutions; 7) letter additions, and 8) word and phrase paragraphia. An example of one patient's writing appears in Figure 3.

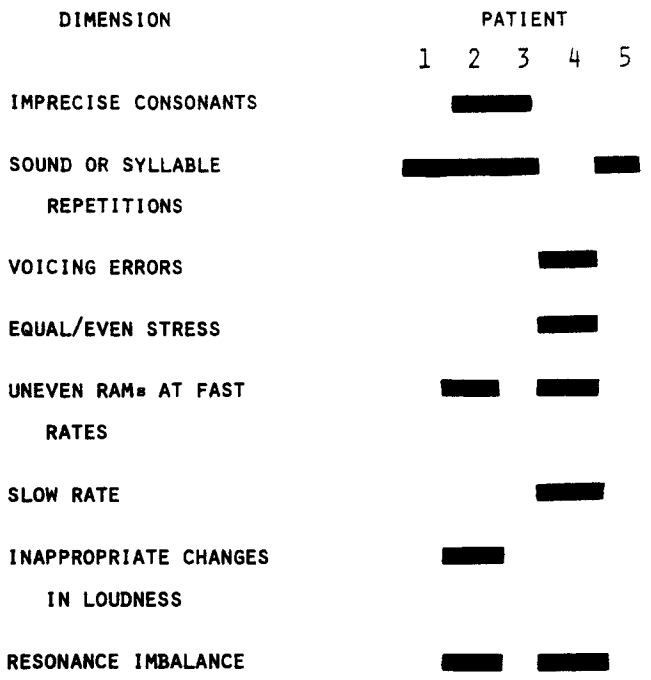

Figure 1. Error types for each of the 5 subjects.

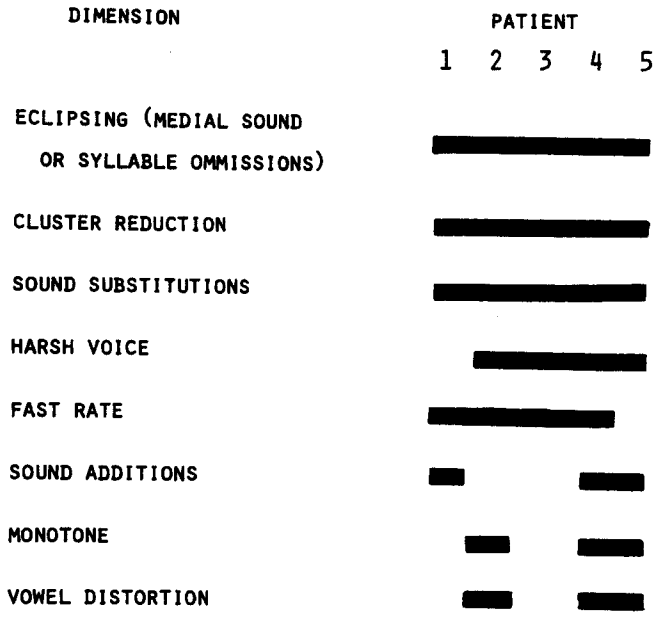

Figure 2. Error types for each of the 5 subjects.

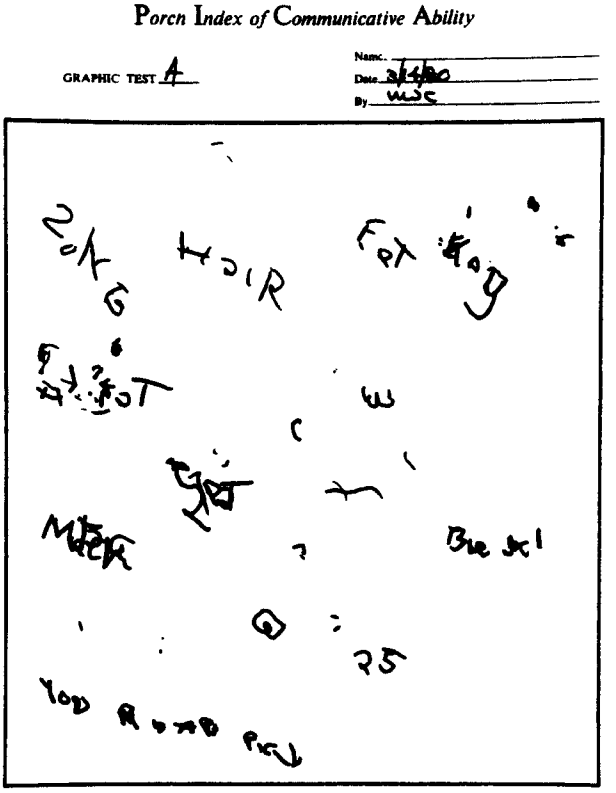

Figure 3. Representative writing sample obtained from Subject 3.

Other Language Performance. These subjects had aphasic deficits in auditory and reading comprehension and in word finding. For example, scores on the Token Test ranged from 15/61 to 49/61. The 15 is above the 30th percentile, the 49 is above the 80th for left-hemisphere damaged patients (Wertz and Lemme, 1974). More extensive speech and language data are available upon request. To be emphasized here, however, is that each subject's writing was more severely involved than his comprehension and oral language.

Neurological. All subjects had sudden onset of neurological deficit, but all were alert and responsive when first evaluated. All showed motor deficits involving the right lower face, the right arm, and the right leg, with the arm being the most severely involved. These findings were combined with right-sided hyper-reflexia and right Babinski signs. Two of the five subjects had normal sensory examinations. One had mild dimunition of primary sensation on the right side; the other two subjects had deficits in right-sided cortical sensation (graphesthesia and stereognosis). The motor findings suggested a lesion most likely involving the pre-central gyrus of the frontal lobe or fibers departing from that area. The sensory findings suggested that three of the subjects also had involvement of the parietal

lobe or of the underlying white matter. Only one was suspected of having a visual field deficit and this was not confirmed on a subsequent examination. All five improved and three of them did so within a few days. Cat and brain scans failed to demonstrate a single area of involvement. One cat scan was normal. One showed a deep frontal-parietal lesions, one a deep frontal-temporal lesion, and the brain scan revealed a deep parietal lesion. The subject who came to autopsy was found to have had a hypertensive hemmorrhage involving the left caudate nucleus, the internal capsule and the lenticular nuclei. The locus of his lesion appears in Figure 4.

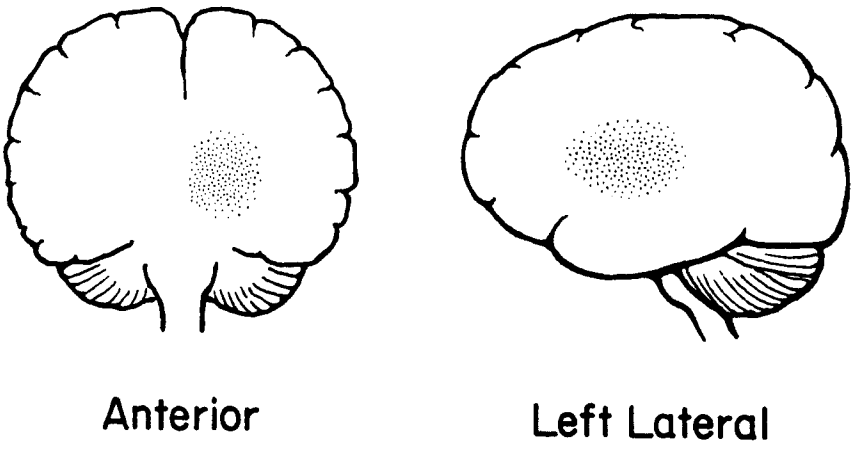

Figure 4. Locus of lesion for Subject 2.

DISCUSSION

These subjects are noteworthy for their combination of neuromotor speech difficulty and dysgraphia. Their speech errors, while varied, give one the overall impression of impaired timing and impaired coordination. Along with Dr. Kent and Dr. Netsell, we were impressed with the similarity of these subjects' symptoms to traditional ataxic dysarthria. They were inconsistent and generally poorly modulated. Because they also made a variety of inconsistent substitutions, we dubbed them, in one of our less lucid moments, ataxo-praxics. The term is foolish; what it suggests may not be. Cortex and cerebellum interact through a series of loops described generally by Brodal (1972) and in greater detail by Allen and Tsukahara (1974). It is probable that this sensorimotor system or set of systems acts in controlling the timing and coordination of speech movements and that lesions to frontal-parietal cortex, to the cerebellar hemispheres, or to the white matter tracts connecting the two may result in some common speech symptoms. As noted previously, we could not establish a single site of lesion for our five. We were not surprised; their symptoms were not identical. We had the strong suspicion, however, that in four of the five, subcortical white matter was involved. Our hypothesis is that such subcortical lesions, depending on their loci, may create a speech and language deficit that is not classical

aphasia, nor apraxia, nor ataxic dysarthria; but which has characteristics of all three, depending on the exact locus and extent of lesion.

Chedru and Geschwind (1972) classified writing disorders into linguistic, motor, and spatial types. Our five subjects made some linguistic errors such as letter and word substitutions. They also made errors such as unintelligible letters and unlinked and unclosed letters more consistent with disturbed motor control mechanisms. The three subjects in Group I evidenced a constellation of linguistic errors such as omitted functor words and errors of motor coordination and sequencing that somewhat resembled the agrammatic speech of the Broca's aphasic speaker. Group II did not have the motor control errors to the degree that Group I did, and they did not omit functor words. Instead, this group substituted content words and used jargon words. Such errors are more frequently associated with posterior lesions and Wernicke's aphasia. Neither group had one of the traditional aphasic syndromes, however. We remain unsure of how the writing errors are to be explained.

An important question is whether we have rediscovered one of Benson's (1979) three subcortical aphasic syndromes - aphasia of Marie's quadrilateral space, thalamic aphasia, or striatal aphasia. Our subjects most closely resemble the description for striatal aphasia, and it will be recalled that the autopsied subject had an appropriately placed lesion. But even he does not fit Benson's description very closely. The other four did not appear to have the catastrophic neurologic symptoms (including mutism) that are reported to result from striatal - usually hemorrhagic - lesions. Nonetheless, we cannot rule out the possibility that our subjects are part of the distribution of striatal aphasic speakers. If they are, this report expands on both the speech and writing characteristics that Benson (1979), Naeser, Alexander, Helm, Levine, Laughlin, and Geschwind (In Press) and others have mentioned. In subsequent studies it will be important to control for time of testing and to make careful, frequent checks on progress, because our impression is that the time one sees such a patient has a significant influence on what one sees.

Why should we worry about identifying such patients? The syndrome may prove to have localizing significance. Patients with the syndrome may have a better prognosis than some other patients. Treating the syndrome may require specific methods.

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