SSW
SSW
REPORTS

AIDS, BRAINSTEM CVA AND
THE SSW EN ESPANOL

Vol. 13 No. 2 May 1991

BRAINSTEM CVA: A CASE STUDY

John Risey & Jack Katz

A 39 year old woman was seen for a hearing evaluation at the Tulane Medical Center. She complained of hearing loss, but especially of difficulty understanding speech in the presence of background noise. She felt that her hearing had gotten worse since she was seen by the Neurology Department in 1984.

History

The patient had a history of two CVAs on the left side due to a congenital arteriovenous (AV) malformation. Because of its location in the "belly of the pons and upper medulla" it was considered inoperable. Her first CVA was in 1980 when she was 28 years old. It resulted in left cranial nerve involvement as well as right hemiparesis.

She had a second CVA in 1984. It resulted in a right hemiplegia as well as left hemisensory deficit.

Findings

Figure 1 shows the patient's audiogram and speech audiometric results. Reliability was rated as good. Audiometric results show a bilateral sensory-neural hearing loss, poorer in the left ear. Similarly, discrimination for recorded NU6 words (at 70dBHL) was depressed in the left ear (44%).

The SSW test was administered in the ususal manner, except for the reduced presentation level in the left ear (30dBSL) because of patient preference (see Figure 2). The raw SSW (R-SSW) score shows significant impairment in the left ear which is

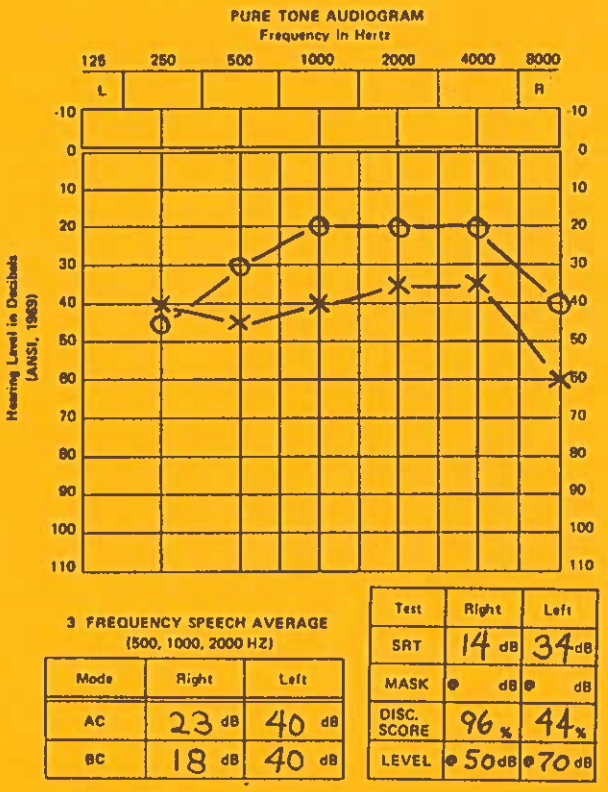

Figure 1. Audiometric results for a patient with an intra-axial lesion in the left side of the lower and mid auditory brainstem. The abnormalities were ipsialteral to the lesion.

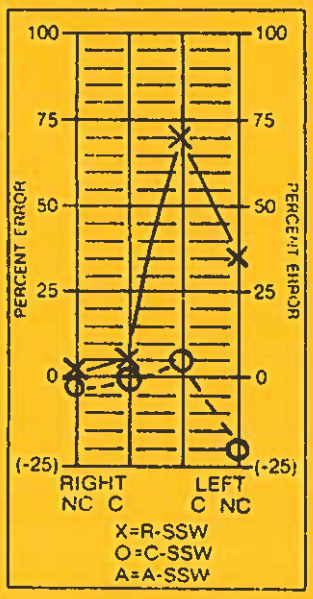

Figure 2. R-SSW and C-SSW results for the patient. The results support involvement of the low-mid brainstem on the left side.

pretty well offset by the SSW correction (C-SSW). The TEC analysis, that is called for in site-of-lesion cases, shows an overcorrected result. There were no response biases.

Interpretation and Comments

We have seen shadow-like audiograms in many brainstem cases as we have found here (Katz, 1970). In addition, a low frequency loss or flat loss is the most typical (but by no means exclusive) pattern that has been observed.

In lesions of the low and middle auditory brainstem, we generally see poor discrimination relative to the puretone findings in the ear ipsilateral to the lesion (as we found in this case). Katz (1976) indicated that the mean discrimination score for Hirsh recorded W-22 words was 14% correct in the ear ipsilateral to low brainstem lesions. For high brainstem cases, the mean score was 64%. The case reported here had a discrimination score of 44%, somewhere in between these two values.

With retrocochlear signs an overcorrected score is generally associated with VIII N or low brainstem level involvement. The mean C-SSW score in the affected (ipsilateral) ear was -13% for those with low brainstem lesions. The mean score for VIII N cases was -27 (Katz, 1976). Thus, the score of -8 for this patient was more suggestive of a low brainstem problem. High brainstem cases had a mean C-SSW score of +53 in the affected ear. Thus, this case was somewhere between the low brainstem value and the high brainstem value for both the C-SSW score and for the WDS.

The puretone, WDS and C-SSW scores are consistent with the expected results for low (or more likely lower to middle) auditory brainstem lesion on the left side. This reasonably well describes the location of the AV malformation that was found on angiography and other medical tests. This supports our long standing contention that typically brainstem lesions express themselves primarily in the ipsilateral ear. We have found this to be true with both intra-axial (as in this case) and extra-axial cases. [Perhaps it should be stated that some people claim that low brainstem lesions should be considered "extra-axial", however our findings over the years have shown the deficit to be more ipsilateral whether the lesion was in the low, middle or high brainstem.]

It is interesting to note that the patient was retested on the SSW at 30dBSL in each ear to determine the influence of presentation level on SSW performance. There was no influence. Recall that SSW-Max is about 25 to 50 dBSL, so no major change would be expected. However, 50dBSL is the standard level.

For many years we have noted an interesting phenomenon in brainstem (and perhaps VIII N) cases. The WDS in the poorer ear (ipsilateral to the

lesion) shows poorer than expected results when considering the puretone performance. However, the better ear (contralateral to the lesion) shows better results than expected for the puretone findings, but especially in light of the similarity in the configuration of the patient's two ears.

Summary

A neurology patient who complained of an onset of a hearing problems in noise was found to have a mild-moderate, flat, sensory-neural hearing loss in the left ear. The puretone configuration, the WDS and the C-SSW results were all affected in the left ear, consistent with low-to-mid brainstem involvement on the left side (ipsilateral to the affected ear). Our interpretation was supported by angiography and other medical tests.

References

Katz, J (1970). Audiologic Differential Diagnosis: Cochlea to Cortex. Menorah Medical Journal, 1, 25-38.

Katz, J (1976). Localizing Auditory Disorders of the Brain and Brain Stem. Short Course ASHA Convention, Houston, Tx.

* * * * * * * * *

SSW EN ESPAÑOL

It is here at last, a Spanish Language Version (SLV) of the SSW test. Although the test has been adapted for use in other languages and dialects (e.g., French, Danish, Portuguese), in fact some that are rather exotic (e.g., Japanese, Hebrew, Turkish, Tamil and Cockney English), thus far, a Spanish version has never been developed (so far as I know).

N. Hernan Soto-Ramos and Robert Windham (from Galluadet University) have collaborated to produce a SLV-SSW test. Their procedure follows quite closely the model of the American EC test (although, they chose not to repeat any words on the test). Obviously, the structure of the languages differ in important ways. For example, there are no spondees, per se, in Spanish, so two familiar two-syllable words were were combined to replace the English spondee in the SLV. For example, their "upstairs downtown" item is:

	1	2	3
RE	perro	gordo	
LE		libro	viejo

(or, fat dog, old book)

One question that they had was wether Mr. Soto-Ramos' Chilean Spanish would create a problem for the non-Chilean speakers-listeners of Spanish. Another goal of this research was to establish some tentative norms for adults. They also sought to get some evidence that the test results conformed generally to those that would be expected using the EC list, with normal and non-normal subjects.

A group of 20 native Spanish speaking control cases were tested using a Spanish (bisyllabic) discrimination test (Auditec) and the SLV-SSW that they had developed.

The mean scores for the 20 subjects are comparable to the scores obtained on the EC list when normal General American speaking adults were tested (C-NS-85). The SLV means were within 1-2 percent of the EC norms for each of the four Conditions. Thus, the difficulty level of the two tests appears comparable for normal adult listeners.

Ten of the Spanish speaking subjects were from Chile and 10 were from other Spanish speaking countries. When the results of the two groups were compared, identical means were found. This suggests that the test can be used (at least with normal adult listeners) regardless of the specific Spanish dialect. [In this sense the SLV, because of its use of disyllabic words, is probably more tolerant of various dialects than a monosyllabic test, as we have in English.]

In addition to the control subjects, two atypical cases were reported. One of them was a 16-year old female with a history of learning disabilities. Her 8CNs were as follows: 0 0 1 2 0 5 2 1. As you can see she demonstrated a Type A-LC pattern if we use the EC criteria for response bias. While this is not a proper approach, we do not have any other criteria to use. It is interesting therefore that we should find a reasonable interpretation. The SSW-gram is shown below:

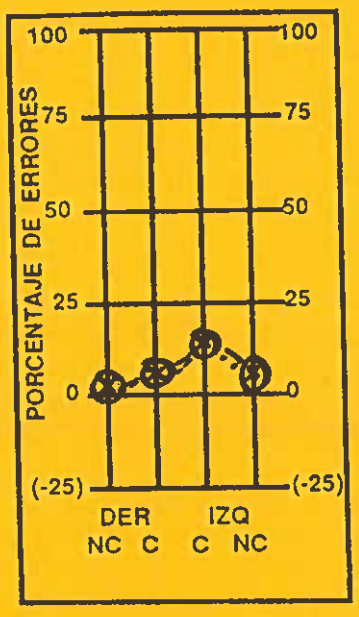

Figure 1. SSW-gram for a 16-year old learning disabled girl on the SLV-SSW test.

The Spanish Language Version of the SSW test by Soto-Ramos and Windham (1991) is available from Auditec of St. Louis (330 Selma Ave., St. Louis, MO). Needless to say, in order to use the SLV-SSW it is necessary that the evaluator be fluent in Spanish.

* * * * * * * * * *

DEAR ACKIE

Dear Ackie:

Most of my central cases involve auditory processing problems and not site-of-lesion considerations. Thus, 3 or 4 times a year when I get a referral for a CANS evaluation I get up tight when it comes to inter-preting the results. Can you help me?

I.M. Shaking (not my real name)

Dear I.M.S.:

Next time you have a site-of-lesion case, pull out this issue of SSW REPORTS and ask yourself these 10 questions. For each one circle whether the answer is yes (Y), no (N) or I don't know (?). This should help reduce the options and strengthen your feelings about certain sites.

IS THERE EVIDENCE OF:

1.	CNS dysfunction	Y	N	?
2.	AR dysfunction	Y	N	?
3.	Hi brainstem dysfunction	Y	N	?
4.	Corpus callosum/ant. com.	Y	N	?
5.	Anterior cerebral dysfun.	Y	N	?
6.	Posterior temporal dysf.	Y	N	?
7.	Lo brainstem dysfunction	Y	N	?
8.	Right side CNS disorder	Y	N	?
9.	Left side CNS disorder	Y	N	?
10.	Other (e.g., cerebellum, thalamus etc.)	Y	N	?

No problem. Try it out on a few cases in the Workshop Manual (p. M-1)

Ackie

SUDDEN ONSET OF SPEECH AND HEARING PROBLEMS: A PATIENT WITH AIDS

David Johnson and Ann Biller
Hennepin Co. Medical Ctr, Minneapolis

Literature Review. HIV positive patients with/without AIDS have been reported to have sudden peripheral hearing loss (Hart et al., 1989; Real et al., 1987; Timon and Walsh, 1989), sudden balance problems (Hart et al., 1989), and abrupt facial nerve weakness (Brown et al., 1988). Unlike previous studies, the case reported here demonstrates a central basis for his complaints.

Presenting Complaint. A 30 year old homosexual male presented with sudden onset right ear pain and inability to hear, as well as decreased speech fluency. History revealed an +HIV homosexual male with oral thrush and disease of the lymph nodes. His medical history shows rheumatic fever at 2 to 3 years of age, meningitis at 4 to 5 years of age, and years of substance abuse. It cannot be determined whether any or all of these symptoms could have influenced our results.

Evaluations and Findings. Motor speech assessment by the speech-language pathologist revealed mild to moderate flaccid dysarthria secondary to right VIIth and XIIth cranial nerve involvement. This was characterized by reduced range and strength of lingual movements, overall decreased speaking rate which was utilized spontaneously for preserving intelligibility, and decreased pitch with hoarse vocal quality. Results of the Boston Diagnostic Aphasia Examination were normal.

Audiometric results show normal hearing bilaterally, however, the right ear was decreased by 5 to 20 dB, on a sensorineural basis, relative to the left ear (see Figure1). Discrimination scores were good bilaterally. The tympanogram on the right had

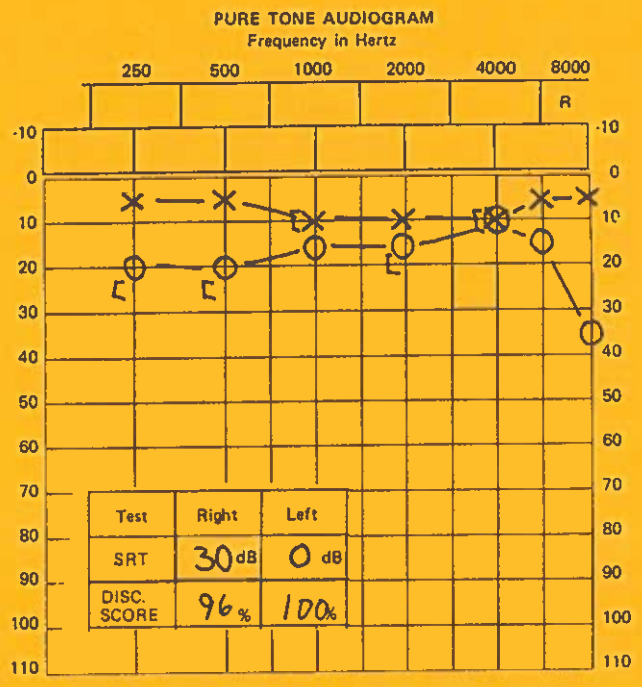

Figure 1. Audiometric results for a 30-year old person with AIDS. The patient complained of hearing loss and pain in the right ear.

increased compliance. Acoustic reflexes were normal bilaterally and reflex decay was absent.

The SSW was presented approximately 50dBSL. Because of limited time only 20 items were administered. A large number of errors was noted in the RNC and RC conditions, with normal performance on the left side. The severe combined TEC (i.e., T=33/S, E=61/S, C=76/S) was suggestive of a left auditory reception (AR) involvement. However, it is interesting to note that for a L-AR case, WDS was quite good and performance in the left ear also surprisingly was good. [Generally L-AR cases have about 10% poorer WDS than this in the RE and the LE on the SSW is depressed to a greater degree (Katz & Pack, 1975)].

The use of a half test precludes the evaluation of Ear and Order Effects, however, there was a suggestion of an anterior Ear Effect (12/16). There was also one reversal (NS).

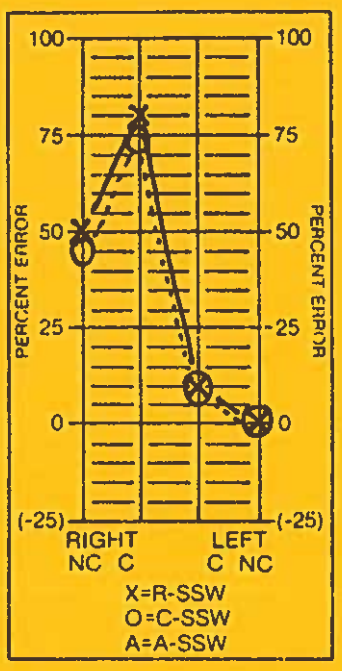

Figure 2. SSW-gram showing a severe combined TEC score.

Electroencephalography showed "intermittent polymorphic slowing, delta and theta, with left mid to anterior temporal involvement with background slowing, consistent with a mild to moderately abnormal EEG showing diffuse cerebral dysfunction with superimposed left temporal dysfunction".

Radiologic evaluation showed the CT scan to be within normal limits, but the MRI showed a lesion in the left insula extending medially into the extreme capsule, claustrum, and external capsule. There was involvement of the posterior aspect of the anterior limb of the internal capsule, genu, retrolenticular limb, as well as the sublenticular limb of the internal capsule. MRI data were thus compatible with left insular and basal ganglia lesion with involovement of adjacent medial, temporal white matter. There was thickening of the left parietal cortex suspicious for early signs of a coronal lesion (often associated with AIDS).

Interpretation and Conclusion. The patient was thought to be demonstrating a number of early AIDS central nervous system characteristics (e.g., VIIth nerve --facial involvement; cochlear versus VIIIth nerve--decreased hearing; XIIthe nerve--tongue control; as well as base of brain--central auditory processing problem). We will continue to follow the patient for AIDS-related emerging health care problems.

[Some of the peculiarities noted in this patient, compared to other AR cases might suggest that the ascending pathways in the thalamus and basal ganglia may interrupt the information reaching the L-AR region but not actually damaging Heschl's gyrus and the important decoding centers around it. One support for this notion is the lack of Wernicke's aphasia, or aphasia of any sort. Of course, the MRI provides anatomical evidence that these regions are spared as well.]

Update on Health Status after Four Months. The patient was subsequently hospitalized several times and last discharged for Home Health Care services in addition to regular care by a relative. In addition to AIDS, the diagnosis of progressive multifocal leukoencephalopathy was established.

References

Brown MM, Thompson A, Goh BT, Forster GE, Swash M: Bell's palsy and HIV infection. J Neurol Neurosurg Psychiatry 1988, 51:425-6.

Hart CW, Colely CG, Schupbach J, Dal Canto MC, Coppleson LW: Neurotologic findings of a patient with acquired immune deficiency syndrome. Ear Hear 1989, 10:68-76.

Real R, Thomas M, Gerwin JM: Sudden hearing loss and acquired immunodeficiency syndrome. Otolaryngol Head Neck Surg 1987, 97:409-12.

Timon CI, Walsh MA: Sudden sensorineural hearing loss as a presentation of HIV infection. J Laryngol Otol 1989, 103:107-2.