Slow Progression of a Rasmussen’s Case: Neuropsychological, Social and Medical Documentation over Seventeen Years

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The patient is a 25 year old right-handed woman who developed right hemisphere complex partial seizures at age 8. Following EEG telemetry, neuropsychological evaluation imaging and a Wada test revealing left hemisphere speech, she underwent a right frontal lobectomy at age 9, sparing the temporal lobe as well as motor cortex. Left upper extremity motor skills and pathology was consistent with Rasmussen’s encephalitis. Since then, her seizures have been completely controlled despite multiple medications and participation in drug trials. Further neurosurgery (functional hemispherectomy) has been offered on several occasions, but the patient and family have not to date elected this intervention. Serial MRI scans show diffuse volume loss throughout the remaining right hemisphere, including the hippocampal formation.

Follow-up neuropsychological evaluations were conducted when the patient was 10, 13, 18 and 25. Although the patient has grown cognitively since age 8 and acquired some new skills, her intellectual functioning, sequencing, and executive functioning. Selected measures are presented here. Psychosocial inventories completed in the first four evaluations yielded normal profiles. Her last testing was discontinued due to a seizure, and therefore did not include inventories.

Prior to surgery the patient demonstrated better verbal than visual-spatial skills on her intellectual examination, a pattern that has continued over time. Nonverbal IQ scores decreased more quickly than verbal scores. Post-operative memory scores also declined over time, with nonverbal scores decreasing sooner.

Commensurate declines in academic achievement scores were also documented. Arithmetical skills peaked at the 7th grade level at age 13 and decreased to 5th grade level at 18. Single-word reading and spelling skills were maintained at 5th grade level. The patient attended a private Christian school without a special education program, but graduated with accommodations and modifications to the standard curriculum. With her heavy medication load, she slept every afternoon after school and worked for hours on homework in the evenings, thus precluding her participation in after-school social activities.

Over time, the patient’s left hand motor skills declined, both relative to increasing right-hand measures and to previous left-hand skills. Due to a right-hand injury suffered during a seizure, she declined to participate in most motor tasks in the recent testing. Her surgery at age 9 was limited to spare her normal left hand functions, but her physicians now predict that functional hemispherectomy would result in little further loss of motor functions. She now experiences frequent twitches of the left upper extremity. The beginnings of a left visual field deficit were documented in the recent testing.

After her surgery the patient lost her musical talents. In her teenage years she often spoke inappropriately loudly, and social judgment issues emerged. Her peers began to shun her, and she found companionship with younger children and adults.

If you were in the position of having to make these very difficult choices for yourself or for your child, what would you decide to do?