Chronic cerebellar stimulation and developmental reflexes

RICHARD D. PENN, M.D., AND MARY LIZ ETZEL, M.S., R.P.T.
Departments of Neurosurgery and Physical Therapy, Rush-Presbyterian—St. Luke’s Medical Center, Chicago, Illinois

The changes in motor function in cerebral palsy patients produced by chronic anterior lobe cerebellar stimulation were documented with the Milani-Comparetti developmental scale, which allows comparison between functional gains and reflex patterns. Two patients with marked motor retardation and primitive reflexes were followed serially over several months of stimulation. Using these measures, the efficiency of chronic stimulation can be assessed, although many years will be needed to evaluate its role in treating motor disabilities.

KEY WORDS • electrical stimulation • cerebellum • cerebral palsy • Milani-Comparetti scale • developmental reflexes

CHRONIC electrical stimulation of the anterior cerebellum has been employed recently in an effort to improve motor function in patients with movement disorders and spasticity. The concept stems in part from the finding that decerebrate rigidity in animals decreases when the paleocerebellum is subjected to repeated surface electrical stimuli. The development of chronically implantable receivers and electrode arrays made it possible to stimulate the cerebellum in humans. Cooper and others have reported considerable success in reducing rigidity in cerebral palsy patients.

As with any new surgical procedure, many years will be required to evaluate the worth of the technique. Evaluating cerebellar stimulation is complicated by the difficulty in categorizing cerebral palsy patients and following changes in their motor function. The etiology, location of lesions, and, consequently, manifestations of cerebral palsy are varied, and the terms used to describe the clinical states are inadequate. One approach to the problem of evaluation and classification has come from physical therapists and developmental neurologists. Bobath, a physical therapist, emphasizes that the motor disability can be viewed as being due to the persistence of primitive and widespread reflex patterns of posture and movement that have not been properly inhibited by higher centers of the central nervous system. Seen in this way, motor function is disrupted by obligatory reflex movements and treatment must be directed at overcoming these primitive patterns.

To emphasize the relationship between motor function and reflex testing, Milani-Comparetti devised a developmental testing program for evaluating children with a relatively simple charting system. It has the advantage of quantifying motor function and reflexes in an age-related manner. Descriptive terms such as rigidity, spasticity, dystonia, and paresis are not employed. We have found
Cerebellar stimulation and primitive reflexes

this scale extremely useful in following the changes with chronic cerebellar stimulation and would like to encourage others to employ it so that results can be compared more easily. A further advantage of the scale is that the changes recorded in reflex patterns may help to explain functional improvements and give a clue to their underlying neurophysiological basis.

Clinical Material and Method

Patient Selection

The initial implant group consisted of five patients with cerebral palsy and one adolescent with damage due to trauma (to be reported separately). They were chosen from a large group of cerebral palsy patients on the basis of their severe motor involvement, persistent primitive reflexes, relatively normal intellectual function, and the lack of improvement with standard treatments. All the patients had had considerable physical therapy in the past, some had also had “patterning” exercises. Before surgery, all patients in the chronic cerebellar stimulation study group were given the Milani-Comparetti development test. They also received separate neurological, orthopedic, and speech evaluations, and quantitative reflex tests. If on this initial evaluation it was felt that further orthopedic procedures or physical therapy would be of value, an implant operation was not performed. Once selected, the patients and parents were informed of the risks of surgery including the possibility of damage to the cerebellum with chronic stimulation and the unpredictability of the results.

Implantation of Stimulating System

The chronic stimulating system was implanted as described by Davis. Two sets of platinum electrodes on Silastic sheets were placed close to the midline anteriorly over the cerebellum through small suboccipital craniotomies. The electrode wires were tunneled underneath the skin to one or two subcutaneous receivers. Stimulation parameters were controlled externally using an adjustable transmitter* with one or two antennae that lay on the skin directly over the receivers. Stimulation was carried out only during the waking hours since spasticity and abnormal movements markedly decrease in sleeping cerebral palsy patients. Control periods without stimulation were occasionally tried. Controls could not be run as single- or double-blind experiments since most of the patients could tell by some sensation whether or not the stimulation was on.

Two cases were chosen for illustration because of their marked primitive reflex patterns and the clear changes found upon serial examination.

Illustrative Case Reports

Case 1

This 7-year-old boy with cerebral palsy secondary to prematurity (1 lb 9 oz) was evaluated because of his severe motor disabilities. He was mildly retarded, markedly dysarthric, and unable to walk or crawl on all fours. His primary means of locomotion was rolling over and pulling with his right arm. He could feed, but not dress himself. At 3 years of age, he had had several major motor seizures, including one episode of status epilepticus, and since then had been well controlled on Dilantin and phenobarbital.

Examination. He was a cooperative boy who could follow instructions but was unable to perform many simple motor commands. He was quadriparetic with minimal involvement of his right arm and hand, but had marked spasticity and rigidity in the lower extremities and flexion posturing of the left arm with cortical thumbing. Deep tendon reflexes were difficult to elicit because of the increased tone, but were hyperactive; Babinski signs were present bilaterally. Developmental testing (Fig. 1) showed retention of obligatory primitive reflexes such as asymmetrical tonic neck and foot grasping reactions, as well as absent protective and equilibratory responses. He was chosen for surgery because of his lack of improvement in spite of considerable physical therapy over 2 years, his marked spasticity, and his parents’ understanding of the risks of surgery and ability to maintain the electrical equipment.

Stimulator Implantation. Cerebellar stimulating electrode arrays were inserted over the anterior lobes bilaterally. Stimulation was begun the sixth postoperative day at

*Transmitter manufactured by Avery Laboratories, Inc., 145 Rome Street, Farmingdale, New York 11735.
R. D. Penn and M. L. Etzel

FIG. 1. Case 1. Pre- and postoperative results of Milani-Comparetti developmental tests.

20 Hz and 8 V, with 7 minutes on/off cycles for approximately 10 hours each day. The first major change in motor function was an immediate decrease in the tone of the left arm with lessening of the abnormal posturing. Over the next week, tone began to decrease in the lower extremities (Fig. 1).

The pictures in Fig. 2, taken after 30 weeks of stimulation, show inhibition of the asymmetrical tonic neck reflex, the ability to sit with equilibrium reactions, and standing posture in parallel bars using the previously nonfunctional left arm for support. By 30 weeks, he was able to crawl on all fours, walk...
Cerebellar stimulation and primitive reflexes

in parallel bars, and use his left arm and hand for support and crude grasping functions. He could not perform fine movements on the left nor did he have the balance to walk with a walker or crutches. His hyperactive deep tension reflexes were easier to demonstrate, and clonus and Babinski responses were present. As the number of days of stimulation accumulated, he could be left off the stimulation for hours and finally for 24 to 48 hours until his rigidity and abnormal posturing began to reappear.

Case 2

This 23-year-old man suffered anoxic damage at birth due to cord strangulation. He had markedly delayed developmental milestones but with great assistance from his parents, he learned to read and communicate at a high school level. Most of his day was spent in his wheel chair; he was unable to crawl, walk, feed, or dress himself. Gross motor movements were markedly interfered with because of persistence of the tonic reflexes and mild athetosis. Tone was abnormal, fluctuating in the arms but generally increased in the legs. Deep tendon reflexes were difficult to demonstrate and Babinski responses were not present. Speech was slow and dysarthric. Cranial nerve and sensory function was normal. He had received several years of extensive physical therapy as a teenager but because of lack of improvement, it was finally stopped. The severe motor involvement coupled with normal intelligence and a great desire to improve made him a suitable candidate for stimulation.

Stimulator Implantation. After hospital testing, a dual set of electrodes was implanted over the anterior lobes of the cerebellum to allow separate stimulation of the left or right sides. The external transmitter was set to produce alternate activation from side to side in a 7-minute cycle; voltage was adjusted to a level just below that which produced a headache, 6 V, rate 20 Hz, and pulse width 500 μsec. Initially with stimulation little change was seen but the patient stated he was more “peaceful” and “less tense.” After several weeks, tone decreased in the legs and his parents reported he could now put on his socks, turn the pages of the newspaper, but could not feed himself. By 18 weeks, he was able to remove his jacket, advance along parallel bars, sit erect, maintain a position on all fours and begin to help with transfers. His speech was less dysarthric and serial recordings showed greater intelligibility. The developmental reflexes (Fig. 3) paralleled his improvement in function.

Discussion

Four of the five cerebral palsy patients undergoing chronic anterior lobe cerebellar stimulation have shown progressive improvement of developmental reflexes. The most striking changes were seen in Cases 1 and 2. The usefulness of serial testing of these reflexes should be clear. At the initial evaluation, the abnormally powerful tonic labyrinthine and tonic neck reflexes accounted for much of the poor motor function. Categorizing the patients only according to spasticity, rigidity, paralysis, or athetosis would have missed much of the reason for their motor disability. Coincident with the decreasing influence of the tonic reflexes and the appearance of equilibrium responses with stimulation, both

FIG. 2. Photographs taken after 30 weeks of cerebellar stimulation show inhibited asymmetrical tonic neck reflex (upper), sitting balance with equilibrium reactions (lower left), and standing posture in parallel bars using both hands for support (lower right).
patients showed useful functional gains such as crawling or improved gross motor control of the upper extremities. Developmental testing allows correlation of the improved motor function with the changed reflex patterns.

A change in developmental reflexes does not necessarily mean that significant functional gains will be forthcoming. For example, one patient who relied on extensor thrust for support had considerably more difficulty maintaining a standing position when stimulation had decreased the reflex. The patient relied on the abnormal reflex for useful function. For inhibition of tonic reflexes to be of value, the capacity to develop higher level...
Cerebellar stimulation and primitive reflexes

motor control must be present. This capacity for improved function cannot be known until the reflexes are decreased, and this poses a major problem in patient selection for the surgeon.

Physical therapy is a necessary adjunct to electrical stimulation in the postoperative period. Abnormal reflexes are inhibited to initiate more normal movement and promote postural control. When possible, righting and equilibrium responses are used to promote new patterns of movement. It should be emphasized that such treatment programs had been tried in all patients before electrical stimulation and there had been no significant progress. Furthermore, if stimulation was interrupted, rigidity would recur and once again interfere with motor function. Postoperative physical therapy does not by itself appear to account for the functional improvements seen.

Can the developmental progress in these two patients be accounted for simply by a decrease in tone in the extremities? For the emergence of equilibrium and righting responses, one might argue in the affirmative. The rigid hypertonic limbs are not able to move, so the response, although possibly present, is not seen. On the other hand, for such reflexes as the symmetrical or asymmetrical tonic neck to be inhibited, some mechanism other than a decreased tone must be involved. Precisely what neurophysiological mechanism would account for such suppression of primitive reflexes is unknown.

Considerable experimental information suggests that the anterior lobe of the cerebellum exerts an inhibitory influence on postural tone and modifies postural reflexes. In the simplest scheme, the Purkinje cells of the medial anterior lobe inhibit Deiters nucleus, which in turn has monosynaptic excitatory effects on spinal alpha and gamma motor neurons. Therefore, firing of Purkinje cells induced by surface stimulation would be expected to reduce motor neuron excitation. Such a simplified view, while apparently useful, in fact is probably misleading. Animal experiments stimulating the anterior lobe have demonstrated many different excitatory as well as inhibitory effects on the neurons of the brain-stem nuclei and the reticular formation. In man, we do not even have proof that surface stimulation fires Purkinje cells. Maybe just the opposite is true and stimulation disrupts cerebellar function. An added problem is that in patients with cerebral palsy, the location of pathological damage to the nervous system is usually unknown. In short, until more basic studies are performed in man and animals, theories proposing to explain on a neurophysiological basis the postural and tone changes seen in our patients are certainly premature. The important thing for the moment is to be able to document accurately the changes with chronic cerebellar stimulation, and see if in time useful functional gains are made and maintained.

References


Address reprint requests to: Richard D. Penn, M.D., Department of Neurosurgery, Rush-Presbyterian — St. Luke's Medical Center, 1753 West Congress Parkway, Chicago, Illinois 60612.