Juvenile Parkinsonism Treated with Bilateral Pallidotomies
—Case Report—

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Abstract
A 47-year-old female diagnosed as having juvenile parkinsonism at age 38 years developed progressive motor fluctuation while receiving levodopa medication. She underwent right posteroventral pallidotomy which achieved only a transient effect on the wearing-off. An additional procedure on the left pallidum resulted in long-lasting relief of the parkinsonian symptoms.

Key words: juvenile parkinsonism, pallidotomy

Introduction
Juvenile parkinsonism is a clinical subclassification of Parkinson's disease defined as parkinsonian symptoms manifesting in a patient under 40 years old which respond remarkably to L-dopa.10 There are at least two types of juvenile parkinsonism, but the common characteristics are marked daily fluctuation in motor response, paucity of severe tremor, and drug-induced dyskinesia in the extremities.a,7,10 Posteroventral pallidotomy is an effective treatment for drug-resistant bradykinesia, rigidity, tremor, and drug-induced dyskinesia of Parkinson's disease,5,6,8 but treatment of juvenile parkinsonism with posteroventral pallidotomy has not been described in detail.

We report the successful treatment of a 47-year-old female with juvenile parkinsonism using bilateral posteroventral pallidotomies.

Case Report
This 47-year-old female was admitted to the neurosurgical unit of Tottori University Hospital on July 21, 1993, because she could no longer support herself. She had first noted difficulty in moving her right extremities at the age of 38 years. Juvenile parkinsonism was diagnosed by a neurologist in 1983. She was treated with a small dosage of levodopa, which achieved complete relief of the signs and symptoms. However, 5 years later, serious motor fluctuation or wearing-off associated with changes in the plasma levodopa concentration appeared. In addition, choreoathetotic movements of her bilateral extremities bothered her during the “on” period. At the age of 47 years, the total duration of daily “off” periods was over 7 hours when she was receiving 700 mg/day of carbidopa-levodopa and 7.5 mg/day of bromocriptine. Administration of carbidopa-levodopa at 800 mg/day precipitated severe drug-induced dyskinesia. Other drugs did not influence the motor fluctuation.

On admission she was almost normally active during the “on” period except for the bothersome choreoathetotic movements of her bilateral extremities. In contrast, bradykinesia, rigidity, and frozen gait during the “off” period were so severe that she could not walk at all unless aided.

On July 25, she underwent right pallidotomy
under local anesthesia. The target point was defined by ventriculography with Leksell's stereotactic system. The target coordinates were: 2 mm in front of the midcommissural point, 6 mm below the intercommissural line, and 21 mm lateral to the midline of the third ventricle. The final lesion site was based on impedance recording and electrical stimulation as described by Laitinen et al.\textsuperscript{1,6} The pallidotomy abolished the motor fluctuation and dyskinesia of the right extremities completely without complications, but the effect lasted only for 4 days. The symptoms recurred gradually except for the dyskinesia of the right extremities. The "off" period elongated to that seen preoperatively over the next 6 months.

On March 21, 1994, she underwent left pallidotomy in the expectation of abolishing the motor fluctuation and dyskinesia of the left extremities. The method was the same as that of the right pallidotomy. Postoperatively, she had immediate and complete relief of her symptoms without any complications. There were no symptoms during the 7 days after the operation, but she showed moderate wearing-off for the next 3 weeks. The parkinsonian symptoms during the "off" period were essentially the same as those of the preoperative period. The motor response fluctuation then faded out during the following 7 days (Fig. 1). Magnetic resonance (MR) imaging 6 months after the operation showed lesions of $6 \times 6 \times 3$ mm in the lateral part of the right internal pallidum and $8 \times 8 \times 4$ mm contralaterally (Fig. 2).

The effects of the pallidotomies have continued for more than 11 months after the second operation. The "off" period now totals 5–30 min/day mainly at night. She is normally active with a regimen of 600–700 mg/day of carbidopa-levodopa and 10 mg/day of bromocriptine.

Fig. 1 Graph showing the maximal duration of the daily "off" period before and after the operations. arrows: the first and second pallidotomies.

Fig. 2 MR image (repetition time 3800 msec, echo time 90 msec) 6 months after the second procedure showing the lesions of the bilateral posterovertral pallidotomies limited to the posterovertral region of the bilateral internal pallidums (arrowheads).

### Discussion

Posterovertral pallidotomy was developed by Leksell in the 1950s\textsuperscript{9} and revived by Laitinen in the 1980s.\textsuperscript{5,6} The procedure can abolish all parkinsonian symptoms.\textsuperscript{5,6,8} Present understanding of the physiology of the basal ganglia justifies this procedure as a treatment for Parkinson's disease.\textsuperscript{1,2} However, some patients do not respond well to the intervention. For example, patients with pure akinesia or severe motor fluctuations are difficult to treat with ordinary pallidotomy.

In our patient, the efficacy of the first procedure persisted only for 4 days although the lesion had been made at the appropriate site. In contrast, the effects of the second pallidotomy have lasted for more than 11 months except for a transient exacerbation possibly due to the effect of posterovertral pallidotomy on the external pallidum. The perifocal edema due to the larger lesion in the left pallidum may have affected the indirect striopallidal pathway.\textsuperscript{9} The mechanism by which the bilateral pallidotomies resulted in the stable motor response in our patient is unclear.

Central dopamine synthetic and storage capacity cannot buffer fluctuations in plasma levodopa concentrations in patients who manifest wearing-off.\textsuperscript{3} The suppression of the thalamus may be easily reversed even with degenerated dopaminergic fibers if more GABAergic neurons at the exit of the internal
pallidum are affected by pallidotomy. Moreover, bilateral posteroventral pallidotomies can be more effective than the unilateral procedure against motor fluctuation if wearing-off is a result of degeneration of bilateral dopaminergic fibers. Therefore, if a parkinsonian patient showing serious levodopa-related fluctuation does not respond to unilateral posteroventral pallidotomy, an additional contralateral procedure is recommended.

References

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