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High Frequency Bilateral Globus Pallidus Interna Deep Brain Stimulation Can Improve Both Chorea and Dysarthria in Chorea-acanthocytosis

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Chorea-acanthocytosis (ChAc) is the most common of the neuroacanthocytosis syndromes, which refer to a group of rare heterogeneous neurodegenerative disorders associated with red cell acanthocytes in peripheral blood smears [1]. To date, treatment options for ChAc have been largely symptomatic and include oral medications as well as botulinum-toxin injections, with an unsatisfactory symptom amelioration [2]. In the past decade, globus-pallidus-interna deep-brain-stimulation (GPi-DBS) has been increasingly adopted as a potential treatment for ChAc [3] (detailed in Table s3). Although the outcomes described have been variable, GPi-DBS significantly controlled chorea in most cases [2]. In contrast, dysarthria has largely been refractory to GPi-DBS and has not been reported with high frequency stimulation. We report a ChAc patient who got significant improvement in chorea and dysarthria with high frequency GPi-DBS.

A 43-year-old female was referred to our center with a five-year-history of progressive dysphagia, dysarthria, abnormal hyperkinetic movements, involuntary tongue protrusion, lip biting, and teeth grinding. As her symptoms progressed, she developed increasing difficulty in eating and communicating verbally. Three years after symptom onset, choreic-dyskinesias had spread to the trunk and limbs. Subsequently she was unable to independently perform any activities of daily living with frequent falls. Despite modest benefit from tiapride (100 mg, two times per day), her symptoms continued to gradually progress over the next two years. Upon examination, she was noted to have severe impairments in speech and swallowing due to orolingual hyperkinesia, and actually, her verbal output was unintelligible at that point. Unfortunately, tiapride did not provide any additional benefit. In order to confirm the diagnosis, we performed diagnostic workup including peripheral blood smear, which showed more than 30% acanthocytes, and scanning electron microscopy (Fig. 1A). In addition, brain MR imaging revealed bilateral atrophic change in the head of caudate nuclei on the T2-weighted image (online-suppl., Fig. s1).

After interdisciplinary discussion with the patient and her family members in details, we decided that he was eligible for bilateral GPi-DBS for symptomatic treatment of choreic-dyskinesia in an attempt to improve her functional capacity and quality of life. Informed consent was obtained and surgical planning was utilized to develop the following electrode implantation coordinates (Left/Right) relative to the anterior commissure: 8.2/7.7-mm vertical, 19.1/17.4-mm lateral, and 12.5/13.5-mm axial. Once the ideal locations had been confirmed, quadripolar electrodes (L302, PINS Medical, China) were implanted in the bilateral GPi. We performed an initial three-days' period of neurological testing on externalized leads to confirm the effects of DBS with an external pulse generator, and then a permanent implanted pulse generator (IPG) was connected to the leads and implanted on the surface of the pectoralis under general anesthesia. Four weeks after the second operation, DBS settings were optimized and chronic stimulation was initiated. The final optimized programmed parameters were as follows: Left Lead-Amplitude, 2.2-V; Pulse-Width, 60-μs; Frequency, 130-Hz, Case (+) and Contact 1 (−); Right Lead-Amplitude, 2.2-V; Pulse-Width, 60-μs; Frequency, 130-Hz, Case (+) and Contact 5 (−). Postoperative CT of 0.625-mm-slice-thickness was obtained and fused with pre-operative MRI to confirm the positioning of the leads (Fig. 1B) with active contacts (the coordinates were in Table s4).

After 12 months of continuous GPi-DBS stimulation, total Unified Huntington's Disease Rating Scale score was reduced from 61 to 31. In addition, we noticed a Chorea sub-score reduction from 24 to 11, the dysarthria sub-score from 3 to 1 (Fig. 1C, detailed in suppl, Table. s1, s2). Also, Burke-Fahn-Marsden Dystonia Rating Scale score was improved from 44 to 20, with reductions in the movement and disability sub-scores of 10 and 14, respectively. We observed a marked improvement of anxiety and depression (suppl., Fig. s2). Most importantly, her dysarthria has been significant improved clinically. She was able to walk without any assistance, communicate with families, as well as manage almost activities of daily living independently (suppl-video). Her symptoms remained stable at the time of last follow-up examination.

Supplementary video related to this article can be found at https://doi.org/10.1016/j.parkreldis.2019.01.008.

Our findings regarding the magnitude of improvement in chorea and dystonia achievable with DBS are in keeping with prior reports [2–5]. However, previously published literature indicated that there was no improvement or even worsening of dysarthria following high frequency DBS in patients with ChAc [4]. In contrast, this case had significant clinical improvement in dysarthria with a reduction in

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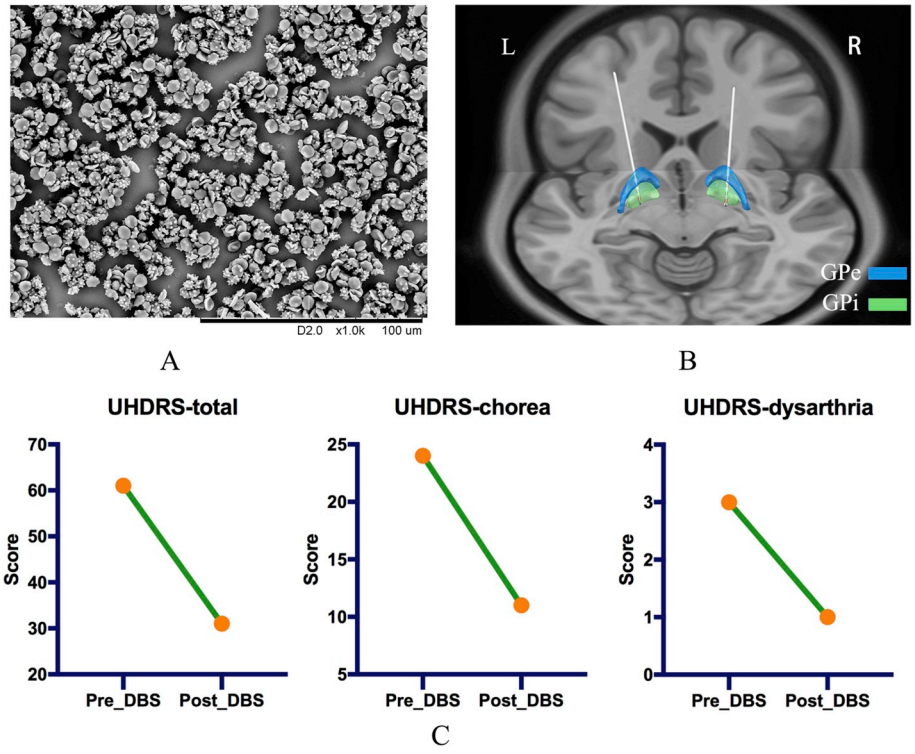

Fig. 1. A. Scanning electron microscopy of peripheral blood clearly showed more than 30% acanthocytes, with abnormal erythrocytes with spiky projections. B. Three dimensional reconstructed positions of leads-contacts in the GPi, in which we adopted a monopole-stimulation model, and the active contacts were highlighted in red colour. Left, Case (+) and Contact-1 (−); Right, Case (+) and Contact-5 (−). C. The changing of UHDRS pre- and post-DBS-operation, and all demonstrated an obvious improvement with the reduced scores. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

UHDRS dysarthria sub-score from 3 to 1. On the other hand, dysarthria improvement was reported in only three case reports previously [3–5]. It should be noted that, in these publications, dysarthria improvement was found solely by low frequency DBS (40 and 60 Hz). Due to rare ChAc cases, there is currently no consensus regarding optimal stimulation parameters for managing these patients. This is to our knowledge the first published description of improved dysarthria with high frequency (130 Hz) bilateral GPi-DBS stimulation.

Conflicts of interest

The authors have no conflicts of interest to disclose.

Author contributions

Kai-Liang Wang, acquisition of data, analysis of imaging data, complete the manuscript.

Dan Xu, acquisition of data, analysis of data.

Jian-Guo Zhang, Fan-Gang Meng, management of the patient, performing the DBS operation and clinical data analysis.

Christopher W. Hess, Wei Hu, data analysis, critical revision of manuscript for intellectual content.

Notes

The work described has not been submitted elsewhere for publication. Informed written consent was obtained from the patient, and all procedures were approved by the ethics committee and the neuromodulation committee at Beijing Tiantan Hospital.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.parkreldis.2019.01.008.

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