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Are we on the right track in DBS surgery for dystonic head tremor? Polymyography is a promising answer

ABSTRACT

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EMG

The clinical benefit of Deep Brain Stimulation (DBS) is associated with electrode positioning accuracy. Intraoperative assessment of clinical effect is therefore key. Evaluating this clinical effect in patients with dystonic head tremor, as opposed to limb tremor, is challenging because the head is fixed in a stereotactic frame. To clinically assess head tremor during surgery, surface electromyography (EMG) electrodes were bilaterally applied to the sternocleidomastoid and cervical paraspinal muscles. This case shows that intraoperative polymyography is an easy and useful tool to assess the clinical effect of DBS electrode positioning.

Deep Brain Stimulation (DBS) is a well-recognized and effective neurosurgical treatment for various movement disorders, such as Parkinson's disease, essential tremor and dystonia. It has been assumed that the clinical benefit of DBS depends on the accurate placement of the electrode tip in the intended target [1,2]. Intraoperative assessment of the clinical DBS effect, which requires the patient to be conscious, can be helpful to achieve this goal [3,4]. It provides the operating team with direct feedback, at a moment when it is still possible to reposition the DBS electrode in case the DBS effect is inadequate. This intraoperative assessment, usually performed by a neurologist specialized in movement disorders, comprises tests focused on change in disease-specific symptoms, such as finger tapping to assess bradykinesia in Parkinson's disease and the finger-to-nose maneuver to assess kinetic tremor of the hands. For patients with dystonic head tremor, the assessment of the clinical effect is challenging, because DBS surgery requires the head of the patient to be completely immobilized in a stereotactic frame [5]. For this patient group, the use of an objective neurophysiology measure, such as electromyography (EMG), might be a solution to overcome this challenge.

Here, we describe a case of a patient affected by idiopathic segmental dystonia, who was severely invalidated by a dystonic head tremor, and who underwent bilateral ventral intermediate nucleus of the thalamus (Vim)-electrode implantation. During the DBS electrode implantation procedure, we applied surface EMG electrodes to the sternocleidomastoid and cervical paraspinal muscles bilaterally. Using surface EMG, we were able to observe a decrease in muscle activity directly after DBS electrode implantation, and a further decrease after test stimulation, providing the surgical team with an effective tool to assess the effect of DBS electrode implantation.

Case description

The patient was a 83-year-old woman affected by idiopathic segmental dystonia for 11 years. Her daily functioning was severely impaired by a dystonic head tremor, a dystonic tremor of the left arm and spasmodic dysphonia. She was treated by intramuscular botulinum toxin injections of the cervical muscles for two years, but without benefit. Her tremor severity had increased markedly during the last year, causing her to be unable to walk without support and limiting her ability to ride a bicycle or drive a car. Given the severity of her dystonic head tremor and its impact on her daily life, she was offered bilateral DBS of the Vim. In this case, Vim was the preferred target over globus pallidus internus (GPi) due to its favorable effects shown in patients with prominent dystonic tremor [6] and spasmodic dysphonia and voice tremor [7].

For placement of the stereotactic frame, the patient was lightly sedated with dexmedetomidine. Analgesia was provided by a targetcontrolled intravenous infusion of remifentanil and local anesthesia. For the latter, a mixture of bupivacaine 0.25% and epinephrine 1:100,000 was used to block the supra-orbital, supra-trochlear and greater auricular nerves, and was also applied to the frame pin sites. The remifentanil infusion was stopped after frame placement. The dexmedetomidine infusion was continued at a rate of 0.3–0.5 mcg/kg/hr until the scalp incisions were made.

Based on clinical observation and palpation, surface electrodes were applied bilaterally over the sternocleidomastoid and cervical paraspinal muscles after positioning the patient on the operating table. Thereafter, we reviewed the report of the preoperative detailed surface EMG assessment and verified that these muscles were indeed responsible for the dystonic movement. At baseline, obvious EMG bursts (~300 ms) were observed, most prominently in the sternocleidomastoid muscle. The right-sided muscles showed more burst-activity than the left-sided muscles. After inserting the left DBS electrode, burst-activity decreased markedly in both the sternocleidomastoid and cervical paraspinal muscles. The decrease in burst-activity was most conspicuous in the contralateral muscles, but a milder decrease was also observable in the ipsilateral muscles. High-frequency test stimulation was carried out in two steps. First 2,0 V and subsequently 4,0 V stimulation was applied over the DBS electrode, using a pulse width of 60 µs and a frequency of 185 Hz. At each step we observed a further burst-activity decrease (Fig. 1). After implanting the right DBS electrode, burst-activity was almost completely suppressed, indicating a satisfactory DBS effect, and



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Fig. 1. Polymyography data obtained at 4 time-points. Surface EMG burst-activity. A: before DBS electrode insertion. B: after left DBS electrode insertion. C: during 2,0 V DBS test stimulation. D: during 4,0 V DBS test stimulation. Upper trace: sternocleidomastoid muscle, lower trace: cervical paraspinal muscles. The surface EMG-activity decreased at each step.

making further assessment of EMG-burst activity irrelevant. If doubts exist on whether the therapeutic effect or a placebo effect is observed during this phase, blinded testing could be implemented.

A CT scan confirmed adequate positions of the DBS electrode tips, after which general anesthesia was induced and maintained with a combination of propofol and remifentanil. The internal pulse generator (IPG) was then implanted and connected to the DBS electrodes.

At the neurosurgical ward a microlesion effect was present immediately postoperatively (see Supplementary Video). We observed a drastic reduction of the head tremor, which was consistent with the polymyography results during surgery, confirming correct positioning of the DBS electrodes. The tremor of the left arm and spasmodic dysphonia also improved. Two weeks after surgery the IPG was first programmed and switched on, with good suppression of all dystonic symptoms, including the head tremor. After several weeks the head tremor gradually returned, while the dystonic arm tremor and spasmodic dysphonia remained adequately suppressed. The DBS parameters are currently being adjusted to obtain optimal clinical effects.

Supplementary video related to this article can be found at https://d oi.org/10.1016/j.parkreldis.2021.11.013.

This case report shows that intraoperative polymyography during DBS implantation surgery for patients with head tremor facilitates optimal DBS electrode placement and is easy to apply. It provided useful information indicating that the electrode had been correctly positioned, which was corroborated by neuroimaging and the presence of a clear microlesion effect. Intraoperative polymyography could aid in deciding whether or not to revise the electrode position or to move to a different target. Of note, in this particular case, repositioning to the GPi would have been an option in case of absent burst-activity reduction during peroperative Vim stimulation [6,7]. Also, intraoperative polymyography can be used in other tremors, such as essential tremor with a neck or head component. Further research is needed to determine whether the use of intraoperative polymyography in DBS-surgery for dystonic head tremor is associated with a better long-term DBS outcome.

Authorship statement

Naomi I. Kremer: Conceptualization, Investigation, Project administration, Visualization, Writing – original draft; D.L. Marinus Oterdoom: Writing - review & editing; Anthony R. Absalom: Writing - review & editing; David W. ten Cate: Investigation, Resources; J. Marc. C. van Dijk: Writing - review & editing; Martje E. van Egmond: Conceptualization, Investigation, Supervision, Writing - review & editing; Gea Drost: Conceptualization, Investigation, Supervision, Writing - review & editing.

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Declaration of competing interest

Anthony R. Absalom: has no conflicts relevant to this work. His research group/department has received payments for sponsor-initiated research or consultancy advice from The Medicines Company (Parsippany, NJ, USA), Janssen (Beerse, Belgium), Becton Dickinson (Eysins, Switzerland), Paion (Aachen, Germany), Terumo (Tokyo, Japan), Ever Pharma (Unterach am Attersee, Austria), Rigel Pharmaceuticals (San Francisco, USA) and Philips BV (Eindhoven, The Netherlands). Martje E. van Egmond: has no conflicts relevant to his work. She participated in a training program sponsored by Medtronic. All other authors have no interests to declare.

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