

REFERENCES

1. Tarsy D, Simon DK. Dystonia. *N Engl J Med* 2006;355:818-829.
2. Geyer HL, Bressman SB. The diagnosis of dystonia. *Lancet Neurol* 2006;5:780-790.
3. Jankovic J. Treatment of dystonia. *Lancet Neurol* 2006;5:864-872.
4. Hamani C, Moro E. Surgery for other movement disorders-dystonia, tics. *Current Opinion Neurology* 2007;20:470-476.
5. Benabid AL, Deuschl G, Lang AE, Lyons KE, Rezai AR. Deep Brain Stimulation for Parkinson's Disease. *Mov Disord* 2006;Suppl 14:S168-170.
6. Borggraefe I, Mehrkens JH, Telegravciska M, Berweck S, Bötzel K, Heinen F. Bilateral pallidal stimulation in children and adolescents with primary generalized dystonia-Report of six patients and literature based analysis of predictive outcomes variables. *Brain Dev* 2010;32:223-228.
7. Vasques X. Factors predicting improvement in primary generalized dystonia treated by pallidal deep brain stimulation. *Mov Disord* 2009;24:846-853.
8. Alterman RL, Tagliati M. Deep brain stimulation for torsion dystonia in children. *Childs Nerv Syst* 2009;23:1033-1040.
9. Isaias IU, Alterman RL, Tagliati M. Outcome predictors of pallidal stimulation in patients with primary dystonia: the role of disease duration. *Brain* 2008;131:1895-1902.
10. Coubes P, Roubertie A, Vayssiere N, Hemm S, Echenne B. Treatment of DYT1-generalised dystonia by stimulation of the internal globus pallidus. *Lancet* 2000;355:2220-2221.
11. Coubes P, Cif L, El Fertit H, et al. Electrical stimulation of the globus pallidus internus in patients with primary generalized dystonia: long-term results. *J Neurosurg* 2004;101:189-194.
12. Halbig TD, Gruber D, Kopp UA, et al. Pallidal stimulation in dystonia: effects on cognition, mood, and quality of life. *J Neurol Neurosurg Psychiatry* 2005;76:1713-1716.
13. Cersosimo MG, Raina GB, Piedimonte F, et al. Pallidal surgery for the treatment of primary generalized dystonia: Long-term follow-up. *Clin Neurol Neurosurg* 2008;110:145-150.
14. Eltahawy HA, Saint-Cyr J, Giladi N, Lang AE, Lozano AM. Primary dystonia is more responsive than secondary dystonia to pallidal interventions: outcome after pallidotomy or pallidal deep brain stimulation. *Neurosurgery* 2004;54:613-619;discussion 619-21.
15. Valdeoriola F, Regidor I, Mínguez-Castellanos A, et al. Efficacy and safety of pallidal stimulation in primary dystonia: results of the Spanish multicentric study. *J Neurol Neurosurg Psychiatry* 2010;81:65-69.
16. Holloway KL, Baron MS, Brown R, et al. Deep Brain Stimulation for Dystonia: A Meta-Analysis. *Neuromodulation* 2006;9:253-261.
17. Kiss ZHT, Doig-Beyaert K, Eliasziw M, et al. The Canadian multicenter study of deep brain stimulation for cervical dystonia. *Brain* 2007;130:2879-2886.
18. Hung SW, Hamani C, Lozano AM, et al. Long-term outcome of bilateral pallidal deep brain stimulation for primary cervical dystonia. *Neurology* 2007;68(6):457-459.
19. Yianni J, Bain P, Giladi N, et al. Globus pallidus internus deep brain stimulation for dystonic conditions: a prospective audit. *Mov Disord* 2003;18:436-442.
20. Bereznai B, Steude U, Seelos K, Bötzel K. Chronic high-frequency globus pallidus internus stimulation in different types of dystonia: a clinical, video, and MRI report of

- six patients presenting with segmental, cervical, and generalized dystonia. *Mov Disord* 2002;17:138-144.
21. Tonomura Y, Kataoka H, Sugie K, et al. Atlantoaxial rotatory subluxation associated with cervical dystonia. *Spine* 2007;32:E561-564.
 22. Muta D, Goto S, Nishikawa S, et al. Bilateral pallidal stimulation for idiopathic segmental axial dystonia advanced from Meige syndrome refractory to bilateral thalamotomy. *Mov Disord* 2001;16:774-777.
 23. Opherk C, Gruber C, Steude U, Dichgans M, Bötzel K. Successful bilateral pallidal stimulation for Meige syndrome and spasmodic torticollis. *Neurology* 2006;66:E14.
 24. Capelle HH, Weigel R, Krauss JK. Bilateral pallidal stimulation for blepharospasm-omromandibular dystonia (Meige syndrome). *Neurology* 2003;60:2017-2018.
 25. Houser M, Waltz T. Meige syndrome and pallidal deep brain stimulation. *Mov Disord* 2005;20:203-205.
 26. Foote KD, Sanchez JC, Okun MS. Staged deep brain stimulation for refractory craniofacial dystonia with blepharospasm: case report and physiology. *Neurosurgery* 2005;56:E415;discussion E415
 27. Castelnau P, Cif L, Valente EM, et al. Pallidal stimulation improves pantothenate kinase-associated neurodegeneration. *Ann Neurol* 2005;57:738-741.
 28. Albanese A., Barnes MP, Bathia KP, et al. A systematic review on the diagnosis and treatment of primary (idiopathic) dystonia and dystonia plus syndromes: report of an EFNS/MDS-ES Task Force. *Eur J Neurol* 2006;13:433-444.
 29. Draganski B, Thun-Hohenstein C, Bogdahn U, Winkler J, May A. "Motor circuit" gray matter changes in idiopathic cervical dystonia. *Neurology* 2003;61:1228-1231.
 30. Delmaire C, Vidailhet M., Elbaz A, et al. Structural abnormalities in the cerebellum and sensorimotor circuit in writer's cramp. *Neurology* 2007;69:376-380.
 31. Burkhard PR, Vingerhoets FJ, Berney A, Bogousslavsky J, Villemure JG, Ghika J. Suicide after successful deep brain stimulation for movement disorders. *Neurology* 2006;63:2170-2172.
 32. Foncke EM., Schuurman PR, Speelman JD. Suicide after deep brain stimulation of the internal globus pallidus for dystonia. *Neurology* 2006;66:142-143.
 33. Damier P, Thobois S, Witjas T, et al. Bilateral deep brain stimulation of the globus pallidus to treat tardive dyskinesia. *Arch Gen Psychiatry* 2007;64:170-176.
 34. Gruber D, Trottenberg T, Kivi A, et al. Long-term effects of pallidal deep brain stimulation in tardive dystonia. *Neurology* 2009;73:53-58.
 35. Kosel M, Sturm V, Frick C, et al. Mood improvement after deep brain stimulation of the internal globus pallidus for tardive dyskinesia in a patient suffering from major depression. *J Psychiatr Res* 2007;41:801-803.
 36. Jahanshahi M, Czernecki V, Zurowski M. Neuropsychological and neuropsychiatric issues in DBS for dystonia. **In this *Mov Disord* Suppl.**
 37. Kupsch A, Benecke R, Müller J, et al. Pallidal deep-brain stimulation in primary generalized or segmental dystonia. *N Engl J Med* 2006;355:1978-1990.
 38. Halbig TD, Gruber D, Kopp UA, Schneider GH, Trottenberg T, Kupsch A. Pallidal stimulation in dystonia: effects on cognition, mood, and quality of life. *J Neurol Neurosurg Psychiatry* 2005;76:1713-1716.
 39. Pillon B, Ardouin C, Dujardin K, et al. Preservation of cognitive function in dystonia treated by pallidal stimulation. *Neurology* 2006;66:1556-1558.
 40. Kleiner-Fisman G, Liang GS, Moberg PJ, et al. Subthalamic nucleus deep brain stimulation for severe idiopathic dystonia: impact on severity, neuropsychological status, and quality of life. *J Neurosurg* 2007;107:29-36.

41. Dauer WT, Burke RE, Greene P, Fahn S. Current concepts on the clinical features, aetiology and management of idiopathic cervical dystonia. *Brain* 1998;121:547-560.
42. Pettigrew L, Jankovic J. Hemidystonia: a report of 22 patients and a review of the literature. *J Neurol Neurosurg Psychiatry* 1985;48:650-657.
43. Chuang C, Fahn S, Frucht SJ. The natural history and treatment of acquired hemidystonia: report of 33 cases and review of the literature. *J Neurol Neurosurg Psychiatry* 2002;72:59-67.
44. Vidailhet M, Vercueil L, Houeto JL, et al. Bilateral deep-brain stimulation of the globus pallidus in primary generalized dystonia. *N Engl J Med* 2005;352:459-567.
45. Mueller J, Skogseid IM, Benecke R, et al. Pallidal deep brain stimulation improves quality of life in segmental and generalized dystonia: Results from a prospective, randomized sham-controlled trial. *Mov Disord* 2008;23:131-134.
46. Zorzi G, Marras C, Nardocci N, et al. Stimulation of the globus pallidus internus for childhood-onset dystonia. *Mov Disord* 2005;20:1194-1200.
47. Manji H, Howard RS, Miller DH, et al. Status dystonicus: the syndrome and its management. *Brain* 1998;121:243-252.
48. Pretto TE, Dalvi A, Kang UJ, et al. A prospective blinded evaluation of deep brain stimulation for the treatment of secondary dystonia and primary torticollis syndromes. *J Neurosurg* 2008;109:405-409.
49. Vidailhet M, Vercueil L, Houeto JL, et al. Bilateral, pallidal, deep-brain stimulation in primary generalized dystonia: a prospective 3 year follow-up study. *Lancet Neurol* 2007;6:223-229.
50. Ostrem JL, Marks WJ Jr., Volz MM, Heath SL, Starr PA. Pallidal deep brain stimulation in patients with cranial-cervical dystonia (Meige syndrome). *Mov Disord* 2007;22:1885-1891.
51. Krause M, Fogel W, Tronnier V, et al. Long-term benefit to pallidal deep brain stimulation in a case of dystonia secondary to pantothenate kinase-associated neurodegeneration. *Mov Disord* 2006;21:2255-2257.
52. Umemura A, Jaggi JL, Dolinskas CA, Stern MB, Baltuch GH. Pallidal deep brain stimulation for longstanding severe generalized dystonia in Hallervorden-Spatz syndrome. Case report. *J Neurosurg* 2004;100(4):706-709.
53. Shields DC, Sharma N, Gale JT, Eskandar EN. Pallidal stimulation for dystonia in pantothenate kinase-associated neurodegeneration. *Pediatr Neurol* 2007;37:442-445.
54. Sako W, Goto S, Shimazu H, et al. Bilateral deep brain stimulation of the globus pallidus internus in tardive dystonia. *Mov Disord* 2008;23:1929-1931.
55. Eltahawy HA, Feinstein A, Khan F, Saint-Cyr J, Lang AE, Lozano AM. Bilateral globus pallidus internus deep brain stimulation in tardive dyskinesia: a case report. *Mov Disord* 2004;19:969-972.
56. Trottenberg T, Paul G, Meissner W, Maier-Hauff K, Taschner C, Kupsch A. Pallidal and thalamic neurostimulation in severe tardive dystonia. *J Neurol Neurosurg Psychiatry* 2001;70:557-559.
57. Trottenberg T, Volkmann J, Deuschl G, et al. Treatment of severe tardive dystonia with pallidal deep brain stimulation. *Neurology* 2005;64:344-346.
58. Zhang JG, Zhang K, Wang ZC, Ge M, Ma Y. Deep brain stimulation in the treatment of tardive dystonia. *Chin Med J (Engl)* 2006;119:789-792.
59. Evidente VG, Lyons MK, Wheeler M, et al. First case of X-linked dystonia-parkinsonism (Lubag) to demonstrate a response to bilateral pallidal stimulation. *Mov Disord* 2007;22:1790-1793.

60. Martinez-Torres I, Limousin P, Tisch S, et al. Early and marked benefit with GPi DBS for Lubag syndrome presenting with rapidly progressive life-threatening dystonia. *Mov Disord* 2009;24:1710-1712.
61. Cif L, Valente EM, Hemm S, et al. Deep brain stimulation in myoclonus-dystonia syndrome. *Mov Disord* 2004;19:724-727.
62. Trottenberg T, Meissner W, Kabus C, et al. Neurostimulation of the ventral intermediate thalamic nucleus in inherited myoclonus-dystonia syndrome. *Mov Disord* 2001;6:769-771.
63. Magarinos-Ascone CM., Regidor I, Martinez-Castrillo JC, Gomez-Galan M, Figueiras-Mendez R. Pallidal stimulation relieves myoclonus-dystonia syndrome. *J Neurol Neurosurg Psychiatry* 2005;76:989-991.
64. Vercueil L, Pollak P, Fraix V, et al. Deep brain stimulation in the treatment of severe dystonia. *J Neurol* 2001;248:695-700.
65. Krauss JK, Loher TJ, Weigel R, Capelle HH, Weber S, Burgunder JM. Chronic stimulation of the globus pallidus internus for treatment of non-DYT1 generalized dystonia and choreoathetosis: 2-year follow up. *J Neurosurg* 2003;98:785-792.
66. Vidailhet M, Yelnik J, Lagrange C, et al. Bilateral pallidal deep brain stimulation for the treatment of patients with dystonia-choreoathetosis cerebral palsy: a prospective pilot study. *Lancet Neurol* 2009 Aug;8(8):709-717.
67. Katsakiori PF. Deep brain stimulation for secondary dystonia: results in 8 patients. *Acta Neurochir (Wien)* 2009;151:473-478;discussion 478.
68. Bronte-Stewart H. Surgical therapy for dystonia. *Curr Neurol Neurosci Rep* 2003;3:296-305.
69. Diamond A, Shahed J, Azher S, Dat-Vuong K, Jankovic J. Globus pallidus deep brain stimulation in dystonia. *Mov Disord* 2006;21:692-695.
70. Tagliati M., Shils J, Sun C, Alterman R. Deep brain stimulation for dystonia. *Expert Rev Med Devices* 2004;1:33-41.
71. Wang S, Liu X, Yianni J, et al. Use of surface electromyography to assess and select patients with idiopathic dystonia for bilateral pallidal stimulation. *J Neurosurg* 2006;105:21-25.
72. Yianni J, Wang SY, Liu X, et al. A dominant bursting electromyograph pattern in dystonic conditions predicts an early response to pallidal stimulation. *J Clin Neurosci* 2006;13:738-746.
73. Krauss JK, Yianni J, Loher TJ, Aziz TZ. Deep brain stimulation for dystonia. *J Clin Neurophysiol* 2004;21:18-30.
74. Das K, Benzil DL, Rovit RL, Murali R, Couldwell WT. Irving S. Cooper (1922-1985): a pioneer in functional neurosurgery. *J Neurosurg* 1998;89:865-873.
75. Benabid AL, Koussie A, Benazzouz A, et al. Deep brain stimulation of the corpus luyssi (subthalamic nucleus) and other targets in Parkinson's disease. Extensions to new indications such as dystonia and epilepsy. *J Neurol* 2001;248(Suppl)III37-47.
76. Chou KL, Hurtig HI, Jaggi JL, Baltuch GH. Bilateral subthalamic nucleus deep brain stimulation in a patient with cervical dystonia and essential tremor. *Mov Disord* 2005;20:377-80.
77. Fukaya C, Katayama Y, Kano T, et al. Thalamic deep brain stimulation for writer's cramp. *J Neurosurg* 2007;107:977-982.
78. Katayama Y, Fukaya C, Kobayashi K, Oshima H, Yamamoto T. Chronic stimulation of the globus pallidus internus for control of primary generalized dystonia. *Acta Neurochir Suppl* 2003;87:125-128.

79. Yianni J, Bain PG, Gregory RP, et al. Post-operative progress of dystonia patients following globus pallidus internus deep brain stimulation. *Eur J Neurol* 2003;10:239-247.
80. Moro E, Lang AE, Strafella AP, et al. Bilateral globus pallidus stimulation for Huntington's disease. *Ann Neurol* 2004;56:290-294.
81. Hebb MO, Garcia R, Gaudet P, Mendez IM. Bilateral stimulation of the globus pallidus internus to treat choreathetosis in Huntington's disease: technical case report. *Neurosurgery* 2006;58:E383.
82. Fasano A, Mazzone P, Piano C, et al. GPi-DBS in Huntington's disease: results on motor function and cognition in a 72-year-old case. *Mov. Disord* 2008;23:1289-1292.
83. Guehl D, Cuny E, Tison F, et al. Deep brain pallidal stimulation for movement disorders in neuroacanthocytosis. *Neurology* 2007;68:160-161.
84. Taira T, Kobayashi T, Hori T. Disappearance of self-mutilating behavior in a patient with Lesch-Nyhan syndrome after bilateral chronic stimulation of the globus pallidus internus. Case report. *J Neurosurg* 2003;98:414-416.
85. Roze E, Paschke E, Lopez N, et al. Dystonia and parkinsonism in GM1 type 3 gangliosidosis. *Mov Disord* 2005;20:1366-1369.

Pre-operative Evaluations

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ABSTRACT

The preoperative evaluation in dystonia aims at characterizing the severity and topography of motor symptoms in patients, who have previously been selected for deep brain stimulation (DBS). Commonly used scales for clinical assessment are the Burke Fahn Marsden (BFM) scale for generalized dystonia and the Toronto Western Spasmodic Torticollis Scale (TWSTRS) for cervical dystonia. Motor assessment is completed by quality of life and functional scales, such as the SF-36 or PDQ-39. Validated rating scales for cranial or upper limb dystonia are lacking. In common clinical practice, these outcome measures can be administered in an open-label fashion since double blind assessment is only required for ascertaining new treatment indications or research purposes. The same measures are to be used postoperatively to reevaluate outcome after DBS. Brain MRI is required to confirm diagnosis and assess structural abnormalities. Other imaging techniques, particularly functional imaging are used for research purposes.

INTRODUCTION

The preoperative evaluation is a crucial step in the management of patients with dystonia who are candidate for deep brain stimulation (DBS). Issues related to the inclusion/exclusion criteria for DBS surgery have been detailed in a previous chapter¹ and will not be discussed again. Before entering preoperative workup, each patient should be classified along with the three axes of aetiology, age of onset and spread of dystonia;² this will allow identifying the most appropriate tools for assessment. The preoperative evaluation aims at characterizing the severity and topography of motor symptoms and their impact on activities of daily living (ADLs) and social activities, and it provides a baseline to serve as a reference for mid- and long-term postoperative evaluations. The quality and accuracy of the preoperative assessment and the choice of assessment tools is crucial as will affect all subsequent postoperative comparisons. The preoperative phase also includes a number of steps related to the assessment of the surgical risk and the determination of the surgical trajectory. This paper will review the evidence for the application and evaluation of the clinical scales to be used for preoperative and postoperative evaluations of dystonia patients undergoing DBS.

METHODS

Search Strategy

The literature search was performed using PubMed, CINAHL and the Cochrane Collaborative databases initially from 1980 to January 2008 using the terms: dystonia AND deep brain stimulation; pallidal stimulation AND dystonia; subthalamic stimulation AND dystonia; thalamic stimulation AND dystonia; secondary dystonia AND DBS; neurodegenerative diseases AND DBS. The search was combined with the one used for neuropsychology, neuropsychiatry, microelectrode recording, neuroimaging, electrophysiology, surgical techniques, complications and targeting. Only English-language publications involving human subjects' were considered. A total of 235 papers were retrieved. To facilitate the committees' work, the articles were divided in 3 groups, which often overlapped: pre-operative, intra-operative and post-operative. A PDF file was created for each paper obtained from the search and put in a CD that was mailed to the members. During the writing phase additional 71 articles were added to update the search, covering the period from January 2008 to September 2009.

Process of Generating Clinical Recommendations

The Consensus Committee members of the Task Force included neurologists, neurosurgeons, neurophysiologists, psychiatrists, neuropsychologists, nurses and mid-level practitioners with expertise and experience in DBS. The experts were also chosen from different countries in Asia, Europe, North and South America, to provide a more comprehensive contribution to the Task Force. The authors of each chapter were selected taking into account their specific expertise in the field. The steering committee prepared a list of questions related to pre-operative, intra-operative and post-operative issues and established two chairs responsible for each of these 3 areas (subcommittees). These chairs then assigned a few questions to be addressed by each member of the subcommittees. The answers to the questions had to be formulated after reviewing the available literature (provided on CD) and combining their expertise. Since the level of evidence for most of the DBS studies was low, the responses were organized following the template previously used for the Special Supplement on DBS for Parkinson's disease (PD): 1) available data, 2) conclusions, 3) pragmatic recommendations, and 4) points to be addressed.³ A first document was prepared from this

initial work and was reviewed and discussed by the entire Task Force group during a one-day meeting. During this meeting the Task Force members provided further feedback and agreed on additional refinements of the whole document adding the comments and remarks collected during the meeting. Special attention was paid to formulate pragmatic recommendations in absence of available studies. A second version of the project was sent to the entire working committee for final approval. The Executive Committee then met again to refine the Special Issue document before submission.

SECTION 1

Methods of Assessments

a. Descriptions and interest of the different scales for dystonia

Available Data

Motor scales

Generalized/segmental dystonia

The Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS)⁴ was introduced to assess generalized dystonia patients. It is composed of a motor part assessing the dystonic movements and a part assessing the consequent disability. The motor subscale evaluates two clinical features of dystonia (severity and provoking factors) in eight body regions (eyes, mouth, neck and the four limbs) and one functional area (speech and swallowing). Severity ranges from 0 (no dystonia) to 4 (severe dystonia). The provoking factors assess the situation under which dystonia occurs and range from 0 (no dystonia) to 4 (dystonia at rest). These two features, severity and provoking factors, are multiplied and then scores are summed, except for the eyes mouth and neck which are halved prior to summing as they are considered regions of “lower weight”. The resulting maximum total score on the BFM severity is 120.⁴ The BFMDRS was clearly designed to assess patients with severe generalized dystonia, and has limitations when applied to milder or non-generalized cases. These limitations include the fact that arms and legs are given one rating each, without distinguishing proximal and distal components, the combination of functional features (such as speech and swallowing) with the inspection of dystonia in other body regions, and the arbitrary reduction of the weight in the cranial/cervical region.

The BMFDRS clinimetric properties were assessed in a study of 10 patients with dystonia rated by 4 different examiners: the overall reliability, inter-rater agreement and concurrent validity were demonstrated for the BMFDRS total score but not analyzed for each different body regions and area of function.⁴ After the first encouraging effort, the BFMDRS was not further systematically developed and tested as a multicenter instrument.

The BFMDRS section on disability assesses the consequences induced by the dystonia in ADL (speech, handwriting, feeding, eating/swallowing, hygiene, dressing and walking), and the total maximum score is 30.

The Unified Dystonia Rating Scale (UDRS) was designed to overcome limitations of the BFMDRS. It includes a more detailed assessment of separate body areas with specific ratings for proximal and distal limbs, and does not mix bodily inspection with functional variables, such as speech and swallowing.⁵ In addition, the UDRS rates duration similarly to the duration factor previously validated for the Toronto Western Spasmodic Torticollis Scale

(TWSTRS).⁵ Furthermore, the UDRS weights the different body regions equally. Fourteen body areas are evaluated: eyes and upper face, lower face, jaw and tongue, larynx, neck, trunk, shoulder/proximal arm (right and left), distal arm/hand (right and left), proximal leg (right and left), distal leg/foot (right and left). For each of these, the UDRS requires rating the severity and duration. Severity rating is specific for each body region and varies from 0 (no dystonia) to 4 (extreme dystonia); duration also ranges from 0 to 4 and assesses whether dystonia occurs at rest or with action, and whether it is predominantly of maximal or sub maximal intensity. The total UDRS score is the sum of the severity and duration factors, with a maximum total of 112. The severity score is expressed as a percentage of the maximum amplitude of the physiological movement, which indicates that this scale is more appropriate to rate “mobile” versus “fixed” dystonia.

The Global Dystonia Rating Scale (GDS) evaluates the severity of dystonia in the same 14 body areas as the UDRS.⁴ The GDS is a Likert-type scale with ratings of 0-10 (from 0, no dystonia, to 10, severe dystonia). There are no modifying factors in the GDS, and the total score is the sum of all the body area scores with a maximum of 140. The GDS is a very simple scale that allows a quick rating of dystonia but does not give precise indications about its clinical aspects (mobile vs. fixed ; disability...). On the other hand each body part has a similar weight, which has the advantage not to minimize any features of dystonia. The other advantage of this scale is its ease of use.

A comparison of the internal consistency and reliability of the BFMDRS, UDRS and GDS was performed by 25 dystonia experts using a standardized videotape protocol.⁵ All 3 scales showed excellent internal consistency and good correlation among raters. The inter-rater agreement was excellent being lowest for eyes, jaw, face, and larynx. There was higher inter-rater consistency for motor severity than for the ratings of modifying factors (duration in the UDRS and provoking factors in the BFM). Seventy-four percent of the raters found the GDS the easiest to apply against 38% for the BFM and only 5 % for the UDRS.⁵

The Global Outcome Scale (GOS) scores the global improvement of the dystonia after a therapeutic intervention. The improvement is rated from 4 (marked) to 0 (no effect).⁶ The GOS is a very simple but imprecise scale that does not differentiate the improvement of each body part. Because of these limitations the scale is rarely employed.⁶

For tardive dyskinesia, which encompasses dystonia and other movement disorders (particularly chorea, myoclonus and tremor), composite scales appear more appropriate, such as the Abnormal Involuntary Movement Scale (AIMS) or the Extrapyramidal Symptoms Rating Scale (ESRS).⁷⁻⁸ The ESRS is divided into four subscales and four clinical global impression severity subscales. These consist in a questionnaire of drug-induced extrapyramidal symptoms, an examination of parkinsonism and akathisia, an examination of dystonia, an examination of dyskinesia and a clinical global impression severity scales for tardive dyskinesia, parkinsonism, dystonia and akathisia.⁸ The AIMS contains 7 items assessing the severity of abnormal movements in different body locations. This scale also includes a global judgment of the severity, consequences and patient’s awareness of abnormal movements. It has been observed that the ESRS and the AIMS have a high degree of concordance.⁹

Cervical dystonia

The Tsui Torticollis Rating Scale was the first rating scale specifically designed for cervical dystonia (CD).¹⁰ It contains six items and is designed for video assessment. This scale evaluates the amplitude and duration of neck involuntary movements in the neck, elevation of shoulder and head tremor.

The TWSTRS⁵ was developed to provide clinical investigators with a better instrument to assess the severity and disability of CD, which is the most common form of focal or segmental dystonia. The TWSTRS was developed in 1990 and consists of 22 items. The total TWSTRS is comprised of 3 separate subscales: motor severity, disability and pain due to CD. The motor severity scale consists of 10 items assessing the severity of head posture in several axes of movement (turning, tilting, anterocollis, retrocollis, shoulder elevation), the effect of sensory tricks, range of motion, and duration of dystonia. The score for motor severity subscale ranges from 0 (no symptoms) to 35 (severe CD). The TWSTRS subsection for motor severity has been validated for inter-rater reliability and validity and a teaching tape has been developed to ensure consistency across raters for multi-center trials.¹¹⁻¹² The disability subscale consists of 7 items assessing the effect of CD on work performance, activities of daily living, driving, reading, watching television, conducting activities outside home, and social embarrassment. The maximal score for the disability subscale is 32. The pain subscale consists of 5 items to assess CD related pain at its maximal, minimal and usual level, and to indicate the duration of pain during a day, and disability due to pain. The maximum score for the pain subscale is 20. The total TWSTRS is the sum of the three subscale scores, with a maximum value of 87. The total TWSTRS has been used extensively as an outcome variable in clinical trials of pharmacological and surgical interventions.¹³⁻²⁰

It has been shown that there is a good correlation between the scores obtained with the TWSTRS and the Tsui scale.²¹ The metric properties of the total TWSTRS and of severity subscales were investigated. Factor analysis showed that 18 of the 22 items of the total TWSTRS fall into three clinically distinct and relevant factors, (1) motor severity, (2) disability and (3) pain.¹¹ These domains correspond to the three subscales of the total TWSTRS, and each measures a separate aspect of CD. The item for social embarrassment did not load on any factor.¹² Three additional items (sensory trick, lateral shift and sagittal shift) did not load on one factor. There are two possible explanations for this inconsistency. First, the range of scores available for these items is limited to absence/presence (lateral and sagittal shift) or to 0-2 (sensory tricks). Second, it has been observed that the observation of sensory tricks is a clinical feature relevant to the diagnosis rather than to clinical signs. Furthermore, the TWSTRS does not clearly assess dystonic tremor, as well as complex combination of phasic and tonic dystonic features.

Focal dystonias

The clinical evaluation of focal dystonias is often difficult.

A scale of 0 (normal) to 4 (worst) has been proposed to rate the severity of blepharospasm and oromandibular dystonia, but the inter-rater reproducibility was poor.²²⁻²³ In a recent study the metric properties of the Jankovic Rating Scale (JRS) and a self-rating patient response outcome scale (the Blepharospasm Disability Index, BSDI) have been compared in blepharospasm patients.²⁴ The internal consistency, and retest reliability of the BSDI were good and the scores obtained using both scales were well correlated. Therefore, these authors suggest that JRS and BSDI can be both used to reliably assess blepharospasm in treatment trials.

For task-specific dystonias, the Writer's Cramp Rating Scale (WCRS) was developed for patients with writer's cramp.²⁵ The WCRS is divided into 3 subscales, respectively studying the dystonic posture, the latency for dystonia to occur and the presence of writing tremor.²⁵ Although this scale is easy to use and has sufficient inter-rater reliability it remains largely unused.

Quality of life scales

The assessment of quality of life is crucial to determine the impact of the surgery on ADL. Most of the studies assessing this outcome measure have used the Short-Form Health Survey (SF-36) or the Parkinson's Disease Questionnaire 39 (PDQ-39).²⁶⁻³³ The SF-36 scale assesses the general and mental health, the physical and social functioning, the physical and emotional roles, the pain and vitality.³⁴ The scores on each subscale are comprised between 0 (worst) to 100 (best). The PDQ-39 scale was originally designed for Parkinson's disease³⁵ but has also been employed for dystonia. It is divided into 7 sections: mobility, activities of daily living, emotional wellbeing, stigma, cognition, communication and bodily discomfort.

The Cervical Dystonia Impact Profile (CDIP-58) has been developed for CD. It measures the health impact of the disease from patient's perceptions.³⁶ This scale is divided into 8 sections (head and neck symptoms, pain and discomfort, upper limb activities, walking, sleep, annoyance, mood and psychosocial functioning). This composite scale appears more sensitive than the SF-36 or TWSTRS to measure the functional outcome of a treatment such as botulinum toxin.³⁷ However, its use remains rare compared to the SF 36 for example.

Conclusions

For generalized and cervical dystonia, the two most accepted and used rating scales are the BFMDRS and TWSTRS, respectively. For other focal dystonias, there are no generally agreed upon scales. The currently available rating scales have several limitations. The BFMDRS scale uses weighting factors that can minimize the real impact of eyes, mouth and neck dystonia. In addition, other associated movement disorders, such as tremor or myoclonus, are not considered in most of the available dystonia scales. Moreover, the available current scales do not sufficiently discriminate more mobile (phasic) dystonic movements from more fixed (tonic) dystonic postures.

Pragmatic Recommendations

The features of dystonia should be monitored before DBS using the most appropriate among the available dystonia scales. The choice of which scale to use should depend upon the dystonia type, according to the topography rather than the etiology of dystonia. For generalized dystonia, the total BFMDRS is recommended. For focal dystonias, the BFMDRS may not always be appropriate. As an alternative, the GDS provides a rapid assessment; although this scale has been less used than the BFMDRS, it can be easily applied in the clinical setting. The UDRS may also be used although its implementation is more difficult. For cervical dystonia, the TWSTRS, including subscales for severity, disability and pain, is recommended. The available scales have been designed to assess patients with primary dystonia and do not always capture complex dystonia phenotypes, such as those observed in dystonia-plus or in secondary dystonias.

Given these limitations, it is recommended that a limited number of expert evaluators is charged to rate patients with dystonia and that standardized videos are performed during each assessment.⁴

The impact of surgery on quality of life is a crucial outcome issue that may provide results divergent from the motor assessment.

Points to be addressed

New more comprehensive scales should be developed: they should also accurately measure tonic postures and phasic movements. Finally, there is a need for uniform training for the BFMDRS and UDRS. Uniform training is available for the TWSTRS, although it has not been shown whether such training improves inter-rater reliability. For other focal dystonias,

although several scales exist, their internal consistency and reliability have been poorly studied and their use remains incidental. Thus there is a clear need for specific scales that objectively quantify the effect of DBS in focal dystonias.

b. Clinical use of the scales for dystonia

1. Should standardized evaluation be performed pre-operatively and post-operatively? How? When?

Available data

Motor assessment

Post-operative objective and subjective assessments have been compared to the preoperative condition in a number of publications, encompassing clinical series, case control studies, cohort studies and single case reports.^{20,26,28-33,38-75} There are only 6 controlled trials that evaluate the effects of GPi DBS in a blinded fashion.^{28-29,31-32,38,77} One of these studies³¹ reach a Class I level of evidence while the 5 others reach a Class II/III level of evidence in the classification proposed by the American Academy of Neurology (Table 1).⁷⁶ These trials provide a clear demonstration of the benefit of DBS for the primary generalized and tardive dystonias and also for CD.^{28-29,31-32,38} Favorable outcome has also been reported for PKAN.³⁹ In these studies a videotaped assessments scored by independent blinded raters allowed controlled evaluations of the effects of the surgery.^{28-29,31-32,38-39} It is notable that data on the benefit of DBS in dystonia reported by open studies are in keeping with the findings reported by controlled studies.

A number of practical issues have been addressed by the available studies. Preoperatively the assessment is most often performed between within the last month and the last week preceding the surgery.^{20,26,28-33,38-75} The time interval between surgery and the first post-operative evaluation is usually comprised between 3 and 12 months.^{20,26,28-33,38-75}

Management of the patients does not require more frequent controls and the first preoperative evaluation is aimed at assessing any acute effects of stimulation on dystonia and threshold for stimulation-induced side effects. Most of the studies have clearly shown that the improvement starts within the first hours or days after beginning the stimulation, and then progresses. Most of the benefit is usually obtained after 3 to 6 months.^{20,26,28-33,38-75} The improvement first affects the phasic signs and later the tonic ones.²⁸ Some additional improvement can occur later but, usually, to a less extent and slower. Some studies however have shown an additional 10-30 % improvement of the dystonia between 1 year and 1.5 year.^{41,73-74} The post-operative outcomes will be discussed in detail in another paper on this same issue.⁷⁸

Quality of life assessment

The quality of life (QoL) assessment is usually performed when the patients have the pre-operative motor assessment, i.e. from 1 month to one week before surgery.²⁶⁻³³ The interval between surgery and the post-operative evaluation of QoL is generally between 3 and 18 months.²⁶⁻³³ QoL usually improves significantly after GPi DBS in generalized and segmental dystonia, and CD.²⁶⁻³³

Conclusions

Validated motor and disability scales are widely used to assess patients before surgery in all the published studies. Most of the time evaluations have been done in open label fashion.

Pragmatic recommendations

Validated scales (see previous section) should be used to assess patients with dystonia within few weeks before surgery. The benefit should be evaluated at 3-6 month after surgery and further evaluations should be scheduled at yearly intervals. Videotaped assessments are recommended.

Points to be addressed

The ideal time-frame to assess the efficacy of DBS in different forms of dystonia needs to be better defined. It remains also to be specified if this should differ for primary generalized or focal forms or for secondary dystonias.

2. Should evaluation in the OFF stimulation condition be performed in routine or research protocol? How long and when?

Available data

Evaluations are rarely performed in OFF stimulation condition.^{28-29,38,49,51,59,64,75} However, assessments without stimulation may provide important information on the immediate effect of stimulation, the delay of reoccurrence of the clinical signs and possibly further worsening of pre-operative motor conditions. OFF stimulation studies thus allow better comparison with the preoperative motor condition and may show evidence of underlying disease progression.

The duration of the stimulation wash-out period preceding assessment may be variable. This has been specifically studied by Grips et al.,⁵¹ who showed that most of the phasic motor symptoms in patients with segmental dystonia reoccurred within 4 hours after switching off bilateral GPi DBS, while the tonic signs may take much longer to worsen. In the Vidailhet's et al.²⁸ study of generalized dystonia patients the maximum tolerated duration of the OFF stimulation period was 7 hours. In a single case study in Lesch-Nyhan dystonia the stimulator could be switched off for 1 month.⁶⁴ By contrast, tardive dystonia and cervical dystonia may worsen very quickly after the stimulator is switched off.^{38,75} This indicates that the effects observed after switching off stimulation may depend on the etiology of dystonia.

Furthermore, it has to be taken into account that severe worsening of dystonia may be life threatening in severe generalized cases; this can be prevented by careful observation of patients during this period.

Conclusions

Evaluations in the OFF stimulation condition have been performed in few studies, which provide interesting data concerning the post-effect duration of DBS in dystonia.

Pragmatic recommendations

A reasonable duration of the OFF period may be of around 3 to 4 hours although this does not lead to the worst off condition. In routine clinical setting, OFF stimulation evaluation is not acceptable because of the risk of reoccurrence of severe dystonia manifestations.

Points to be addressed

It is unclear whether the time course of motor signs reoccurrence after DBS switch-off depends on the etiology of dystonia. This needs to be addressed by specific studies.

SECTION 2

Role of Imaging

Is there any role for pre-operative imaging (brain MRI, PET)?

a. Morphological imaging

Available Data

Conventional MR Imaging

Brain imaging is mandatory in order to determine the aetiology of dystonia and should be performed before considering any patient for surgical treatment.¹ In primary dystonia there are no major structural abnormalities as seen with brain CT or MRI. However, some detailed MRI studies indicate changes of gray mater density in the motor circuit or changes of basal ganglia volume.^{2,79-81} One study with conventional MRI showed T2 bilateral abnormalities in the lentiform nucleus in primary cervical dystonia.⁸² However, the abnormalities were only detected on calculated T2 values; no obvious signal changes could be recognized on visual inspection of T2-weighted images.⁸² Recently, structural abnormalities were shown in the cerebellum and sensorimotor circuit in writer's cramp.⁸³ Using voxel-based morphometry, gray matter decrease was found in the hand area of the left primary sensorimotor cortex, bilateral thalamus and cerebellum. However, such changes were not visualized on conventional images. The main aim of conventional structural MR images of the brain in surgical candidates is to determine the feasibility of surgical implantation and the technical approach independently of the search for the cause of the dystonia. Surgeons will use this brain MRI to rule out major surgical contra-indications such as brain tumors, severe vascular changes or malformations and to visualize the target structures. Some secondary dystonias such as PKAN, post-stroke dystonia, neuroacanthocytosis or inborn errors of metabolism are associated with severe basal ganglia damage that can have an impact on the choice of the target of implantation and on the expected results.⁸⁴⁻⁸⁷ In most of the published series the brain MRI sequences are not described.

Non conventional MR imaging

Brain MR spectroscopy revealed no abnormal N-acetylaspartate/creatine (NAA/Cr) and lactate/creatine ratios in patients with focal hand dystonia, while it has been shown that NAA/Cho and NAA/Cr were significantly lower in patients with spasmodic torticollis.⁸⁸⁻⁸⁹

There are some reports on diffusion tensor images (DTI) indicating abnormal fractional anisotropy and mean diffusivity in cervical dystonia and idiopathic dystonia.⁹⁰⁻⁹¹

Conclusions

Brain MRI is required for the aetiological diagnosis of dystonia. At the preoperative evaluation stage brain MRI is used to ensure that no focal lesions may interfere with the implantation. Other imaging modalities such as fMRI, MR spectroscopy and DTI are used only for research purpose, and thus not useful for routine preoperative evaluation.

Pragmatic Recommendations

Brain MRI should be performed in every patient considered for DBS in order to ascertain if there are structural lesions that may be causative of dystonia or interfere with the surgical procedure. Functional MRI, MR spectroscopy and DTI are not necessary in general clinical

practice of DBS and do not influence surgical procedure or outcome. Therefore they should be done in specialized centers for research on movement disorders.

Points To Be Addressed

Morphological brain MRI is required before DBS in dystonia for every patient. However, the sequences to be used may differ from a center to another. It would be useful to define a common protocol that could be applied in every center aiming at implanting patients with dystonia. The contribution of new MRI sequences also needs to be clarified.

b. Functional imaging

Available Data

The pathophysiology of dystonia is complex and not fully understood. Electrophysiological and functional imaging studies have shown an excess of brain activation, a loss of cortico-cortical inhibition and a lack of the selectivity of brain activation.⁹² More precisely, functional imaging studies have shown overactivity of the dorsolateral prefrontal cortex, premotor and anterior cingulate cortex, cerebellum and putamen in patients with primary and secondary dystonia.⁹²⁻⁹⁵ In primary dystonia (generalized or focal) a decrease of rCBF is usually seen in the primary motor cortex.⁹³⁻⁹⁷ On the other hand, in secondary dystonia rCBF is often increased in the primary motor cortex.⁹⁸ fMRI studies performed in writer's cramp and Meige's syndrome have demonstrated an altered somatotopic representation, which contributes to the loss of functional selectivity of muscle activity.⁹⁹ In tardive dystonia an increase in regional cerebral blood flow has been found in the prefrontal cortex (areas 8 and 11), the anterior cingulate and the lateral premotor cortex.¹⁰⁰ Other PET or SPECT studies in tardive dystonia patients have looked at the modifications of the post-synaptic dopaminergic system. In patients studied after long-term neuroleptic treatment withdrawal, an upregulation of dopaminergic D2 receptors has been observed using PET and [11C]-Raclopride, a D2 receptor ligand.¹⁰¹ Notably, these studies concerned patients with severe tardive dystonia, and they are in agreement with the suspected role of dopamine receptor trafficking in the occurrence of this pathology.¹⁰¹ In contrast, other studies showed normal dopamine D2 receptor density and / or affinity in TD.¹⁰²

Conclusions

PET functional imaging has clearly demonstrated that the abnormal movements and postures in dystonia are related to a widespread excess of brain activation, whatever is the cause of the dystonia.

Pragmatic Recommendations

Despite their important application elucidating the pathophysiology of dystonia, functional imaging studies have no clear role at present in routine clinical practice.

Points to be addressed

None.

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REFERENCES

1. Bronte-Stewart H, Valldeoriola F, Merello M, et al. Inclusion and exclusion criteria for DBS in Dystonia. *In this Mov Disord Suppl 2010.*
2. Albanese A, Barnes MP, Bhatia KP, et al. A systematic review on the diagnosis and treatment of primary (idiopathic) dystonia and dystonia plus syndromes: report of an EFNS/MDS-ES Task Force. *Eur J Neurol* 2006;13:433-444.
3. Benabid AL, Deuschl G, Lang AE, Lyons KE, Rezai AR. Deep Brain Stimulation for Parkinson's Disease. *Mov Disord* 2006;21(Suppl14):S168-170.
4. Burke RE, Fahn S, Marsden CD, Bressman SB, Moskowitz C, Friedman J. Validity and reliability of a rating scale for the primary torsion dystonias. *Neurology* 1985;35:73-77.
5. Comella CL, Leurgans S, Wu J, Stebbins GT, Chmura T. Dystonia Study Group. Rating scales for dystonia: a multicenter assessment. *Mov Disord* 2003;18:303-312.
6. Eltahawy HA, Saint-Cyr J, Poon YY, Moro E, Lang AE, Lozano AM. Pallidal deep brain stimulation in cervical dystonia: clinical outcome in four cases. *Can J Neurol Sci* 2004;31:328-332.
7. Guy E. Abnormal Involuntary Movement Scale. 1976. Rockville, MD, National Institute of Mental Health. ECDEU Assessment of Manual for Psychopharmacology: revised 1976.
8. Chouinard G, Ross-Chouinard A, Annable L, Jones B. The extrapyramidal symptom rating scale. *Can J Neurol Sci* 1980;7:233.
9. Gharabawi GM, Bossie CA, Lasser RA, Turkoz I, Rodriguez S, Chouinard G. Abnormal Involuntary Movement Scale (AIMS) and Extrapyramidal Symptom Rating Scale (ESRS): cross-scale comparison in assessing tardive dyskinesia. *Schizophr Res* 2005;77:119-128.
10. Tsui JK, Eisen A, Stoessl AJ, Calne S, Calne DB. Double-blind study of botulinum toxin in spasmodic torticollis. *Lancet* 1986;2:245-247.
11. Consky E, Lang A. Clinical assessment of patients with cervical dystonia. In: Jankovic J, Hallett M, eds. *Therapy with botulinum toxin*. New York: Marcel Dekker, 1994:211-237.
12. Comella CL, Stebbins GT, Goetz CG, Chmura TA, Bressman SB, Lang AE. Teaching tape for the motor section of the Toronto Western Spasmodic Torticollis Scale. *Mov Disord* 1997;12:570-575.
13. Ford B, Louis ED, Greene P, Fahn S. Outcome of selective ramisectomy for botulinum toxin resistant torticollis. *J Neurol Neurosurg Psychiatry* 1998;65:472-478.
14. Brashear A, Lew MF, Dykstra DD, et al. Safety and efficacy of NeuroBloc (botulinum toxin type B) in type A-responsive cervical dystonia. *Neurology* 1999;53:1439-1446.

15. Brin MF, Lew MF, Adler CH, et al. Safety and efficacy of NeuroBloc (botulinum toxin type B) in type A-resistant cervical dystonia. *Neurology* 1999;53:1431-1438.
16. Kiss ZH, Doig K, Eliasziw M, Ranawaya R, Suchowersky O. The Canadian multicenter trial of pallidal deep brain stimulation for cervical dystonia: preliminary results in three patients. *Neurosurg Focus* 2004;17:E5.
17. Comella CL, Jankovic J, Shannon KM, et al. Dystonia Study Group. Comparison of botulinum toxin serotypes A and B for the treatment of cervical dystonia. *Neurology* 2005;65:1423-1429.
18. Truong D, Duane DD, Jankovic J, et al. Efficacy and safety of botulinum type A toxin (Dysport) in cervical dystonia: results of the first US randomized, double-blind, placebo-controlled study. *Mov Disord* 2005;20:783-791.
19. Meyer C. Outcome of selective peripheral denervation for cervical dystonia. *Stereotact Funct Neurosurg* 2001;77:44-47.
20. Bittar RG, Yianni J, Wang S, et al. Deep brain stimulation for generalised dystonia and spasmodic torticollis. *J Clin Neurosci* 2005;12:12-16.
21. Tarsy D. Comparison of clinical rating scales in treatment of cervical dystonia with botulinum toxin. *Mov Disord* 1997;12:100-2.
22. Jankovic J, Schwartz K, Donovan DT. Botulinum toxin treatment of cranial-cervical dystonia, spasmodic dysphonia, other focal dystonias and hemifacial spasm. *J Neurol Neurosurg Psychiatry* 1990;53:633-639.
23. Defazio G, Lepore V, Abbruzzese G, et al. Reliability among neurologists in the severity assessment of blepharospasm and oromandibular dystonia: a multicenter study. *Mov Disord* 1994;9:616-621.
24. Jankovic J, Kenney C, Grafe S, Goertelmeyer R, Comes G. Relationship between various clinical outcome assessments in patients with blepharospasm. *Mov Disord* 2009;24:407-413.
25. Wissel J, Kabus C, Wenzel R, et al. Botulinum toxin in writer's cramp: objective response evaluation in 31 patients. *J Neurol Neurosurg Psychiatry* 1999;61:172-175.
26. Bereznai B, Steude U, Seelos K, Bötzel K. Chronic high-frequency globus pallidus internus stimulation in different types of dystonia: a clinical, video, and MRI report of six patients presenting with segmental, cervical, and generalized dystonia. *Mov Disord* 2002;17:138-144.
27. Diamond A, Jankovic J. The effect of deep brain stimulation on quality of life in movement disorders. *J Neurol Neurosurg Psychiatry* 2005;76:1188-1193.
28. Vidailhet M, Vercueil L, Houeto JL, et al. Bilateral deep-brain stimulation of the globus pallidus in primary generalized dystonia. *N Engl J Med* 2005;352:459-467.
29. Vidailhet M, Vercueil L, Houeto JL, et al. Bilateral, pallidal, deep-brain stimulation in primary generalised dystonia: a prospective 3 year follow-up study. *Lancet Neurol* 2007;6:223-229.
30. Hälbig TD, Gruber D, Kopp UA, Schneider GH, Trottenberg T, Kupsch A. Pallidal stimulation in dystonia: effects on cognition, mood, and quality of life. *J Neurol Neurosurg Psychiatry* 2005;76:1713-1716.

31. Kupsch A, Benecke R, Müller J, et al. Deep-Brain Stimulation for Dystonia Study Group. Pallidal deep-brain stimulation in primary generalized or segmental dystonia. *N Engl J Med* 2006;355:1978-1990.
32. Kiss ZH, Doig-Beyaert K, Eliasziw M, Tsui J, Haffenden A, Suchowersky O. Functional and Stereotactic Section of the Canadian Neurosurgical Society; Canadian Movement Disorders Group. The Canadian multicentre study of deep brain stimulation for cervical dystonia. *Brain* 2007;130:2879-2886.
33. Blahak C, Wöhrle JC, Capelle HH, et al. Health-related quality of life in segmental dystonia is improved by bilateral pallidal stimulation. *J Neurol* 2008;255:178-182.
34. Ware JE Jr, Sherbourne CD. The MOS 36-item short-form health survey (SF-36). I. Conceptual framework and item selection. *Med Care* 1992;30:473-483.
35. Peto V, Jenkinson C, Fitzpatrick R. PDQ-39: a review of the development, validation and application of a Parkinson's disease quality of life questionnaire and its associated measures. *J Neurol* 1998;245(Suppl 1):S10-14.
36. Cano SJ, Warner TT, Linacre JM, et al. Capturing the true burden of dystonia on patients: the Cervical Dystonia Impact Profile (CDIP-58). *Neurology* 2004;63:1629-1633.
37. Cano SJ, Hobart JC, Edwards M, et al. CDIP-58 can measure the impact of botulinum toxin treatment in cervical dystonia. *Neurology* 2006;67:2230-2232.
38. Damier P, Thobois S, Witjas T, et al. French Stimulation for Tardive Dyskinesia (STARDYS) Study Group. Bilateral deep brain stimulation of the globus pallidus to treat tardive dyskinesia. *Arch Gen Psychiatry* 2007;64:170-176.
39. Castelnau P, Cif L, Valente EM, et al. Pallidal stimulation improves pantothenate kinase-associated neurodegeneration. *Ann Neurol* 2005;57:738-741.
40. Diamond A, Shahed J, Azher S, Dat-Vuong K, Jankovic J. Globus pallidus deep brain stimulation in dystonia. *Mov Disord* 2006;21:692-695.
41. Krauss JK, Pohle T, Weber S, Ozdoba C, Burgunder JM. Bilateral stimulation of globus pallidus internus for treatment of cervical dystonia. *Lancet* 1999;354:837-838.
42. Krauss JK, Loher TJ, Pohle T, et al. Pallidal deep brain stimulation in patients with cervical dystonia and severe cervical dyskinesias with cervical myelopathy. *J Neurol Neurosurg Psychiatry* 2002;72:249-256.
43. Krauss JK, Loher TJ, Weigel R, Capelle HH, Weber S, Burgunder JM. Chronic stimulation of the globus pallidus internus for treatment of non-dYT1 generalized dystonia and choreoathetosis: 2-year follow up. *J Neurosurg* 2003;98:785-792.
44. Tronnier VM, Fogel W. Pallidal stimulation for generalized dystonia. Report of three cases. *J Neurosurg* 2000;92:453-456.
45. Coubes P, Roubertie A, Vayssiere N, Hemm S, Echenne B. Treatment of DYT1-generalised dystonia by stimulation of the internal globus pallidus. *Lancet* 2000;355:2220-2221.
46. Coubes P, Cif L, El Fertit H, et al. Electrical stimulation of the globus pallidus internus in patients with primary generalized dystonia: long-term results. *J Neurosurg* 2004;101:189-194.

47. Alterman RL, Miravite J, Weisz D, Shils JL, Bressman SB, Tagliati M. Sixty hertz pallidal deep brain stimulation for primary torsion dystonia. *Neurology* 2007;69:681-688.
48. Cersosimo MG, Raina GB, Piedimonte F, Antico J, Graff P, Micheli FE. Pallidal surgery for the treatment of primary generalized dystonia: Long-term follow-up. *Clin Neurol Neurosurg* 2008;110:145-150.
49. Goto S, Yamada K. Long term continuous bilateral pallidal stimulation produces stimulation independent relief of cervical dystonia. *J Neurol Neurosurg Psychiatry* 2004;75:1506-1507.
50. Goto S, Yamada K, Shimazu H, et al. Impact of bilateral pallidal stimulation on DYT1-generalized dystonia in Japanese patients. *Mov Disord* 2006;21:1785-1787.
51. Grips E, Blahak C, Capelle HH, et al. Patterns of reoccurrence of segmental dystonia after discontinuation of deep brain stimulation. *J Neurol Neurosurg Psychiatry* 2007;78:318-320.
52. Ostrem JL, Marks WJ Jr, Volz MM, Heath SL, Starr PA. Pallidal deep brain stimulation in patients with cranial-cervical dystonia (Meige syndrome). *Mov Disord* 2007;22:1885-1891.
53. Starr PA, Turner RS, Rau G, et al. Microelectrode-guided implantation of deep brain stimulators into the globus pallidus internus for dystonia: techniques, electrode locations, and outcomes. *J Neurosurg* 2006;104:488-501.
54. Opherk C, Gruber C, Steude U, Dichgans M, Bötzel K. Successful bilateral pallidal stimulation for Meige syndrome and spasmodic torticollis. *Neurology* 2006;66:E14.
55. Tisch S, Zrinzo L, Limousin P, et al. Effect of electrode contact location on clinical efficacy of pallidal deep brain stimulation in primary generalised dystonia. *J Neurol Neurosurg Psychiatry* 2007;78:1314-1319.
56. Vercueil L, Pollak P, Fraix V, et al. Deep brain stimulation in the treatment of severe dystonia. *J Neurol* 2001;248:695-700.
57. Yianni J, Bain PG, Gregory RP, et al. Post-operative progress of dystonia patients following globus pallidus internus deep brain stimulation. *Eur J Neurol* 2003;10:239-247.
58. Zorzi G, Marras C, Nardocci N, et al. Stimulation of the globus pallidus internus for childhood-onset dystonia. *Mov Disord* 2005;20:1194-1200.
59. Muta D, Goto S, Nishikawa S, et al. Bilateral pallidal stimulation for idiopathic segmental axial dystonia advanced from Meige syndrome refractory to bilateral thalamotomy. *Mov Disord* 2001;16:774-777.
60. Magariños-Ascone CM, Regidor I, Martínez-Castrillo JC, Gómez-Galán M, Figueiras-Méndez R. Pallidal stimulation relieves myoclonus-dystonia syndrome. *J Neurol Neurosurg Psychiatry* 2005;76:989-991.
61. Capelle HH, Weigel R, Krauss JK. Bilateral pallidal stimulation for blepharospasm-omandibular dystonia (Meige syndrome). *Neurology* 2003;60:2017-2018.
62. Chuang C, Fahn S, Frucht SJ. The natural history and treatment of acquired hemidystonia: report of 33 cases and review of the literature. *J Neurol Neurosurg Psychiatry* 2002;72:59-67.