Supplementary Material

Contents

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Search syntax.

1) Pubmed

((((stimulation) OR neurostimulation) OR DBS)) AND ((DYT1) OR TOR1A)

2) Embase

(stimulation OR neurostimulation OR dbs) AND (dyt1 OR tor1a) AND [humans]/lim AND [english]/lim

3) Cochrane Library

#1 Stimulation

#2 Neurostimulation

#3 DBS

#4 TOR1A

#5 DYT1

#6 #1 OR #2 OR #3

#7 #4 OR #5

#8 #6 AND #7

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| Supplementary Table 1. Clinical characteristics of the patients excluded. |
| Patients† | Sex | Family history | Age at onset (years) | Region of onset | Body distribution before DBS | Disease duration before dystonia generalization (years) | Disease duration before DBS (years) | Age at DBS (years) | Follow-up after DBS (years) |  | BFMDRS or UDRS | New regions after DBS | Additional DBS surgeries |
| UF | LF | Lx | N | UE | LE | T | 　 | Baseline | Best | Last FU |
| Pt 12 | F | - | 8 | UE | - | - | - | + | + | + | + | 10 years | 24 | 32 | 18 |  | 45 | 7 | 7 |  | Repositioning of bil GPi |
| Pt 13 | M | + | 6 | UE | - | - | - | - | + | + | + | 6 months | 3 | 9 | 1 |  | 43 | 4.5 | 21.5 | N | Bil STN |
| Pt 14 | M | - | 6 | UE | - | - | + | + | + | + | + | 2 months | 2 | 8 | 12 |  | 71 | 7 | 36 | UF, LF | Bil STN |
| Pt 15 | F | + | 6 | LE | - | + | - | + | + | + | + | 4 years | 20 | 26 | 20 |  | 97.5 | 15 | 27 | UF | None |
| Pt 16 | M | + | 7 | UE | - | + | - | + | + | + | + | 9 months | 1.5 | 8.5 | 20 |  | 76 | 0 | 38 | UF | Bil GPi |
| Pt 17 | F | - | 6 | UE | - | - | - | - | + | + | + | 1.5 years | 2 | 8 | 5 |  | 12 | 2 | 8 | UE, LE | None |
| Pt 18 | M | - | 8 | LE | - | - | - | - | + | + | - | Segmental | 49 | 57 | 1 |  | 11 | - | 5 |  | None |
| Regions: UF = Upper face, LF = Lower face, Lx = Larynx, N = Neck, UE = Upper extremity, LE = Lower extremity, T = Trunck; Additional surgery: Bil = Bilateral, GPi = Globus pallidus interna, L = Left, R = Right, STN = Subthalamic nucleus; BFMDRS-M = Burke-Fahn-Marsden Dystonia Rating Scale motor score; DBS = deep brain stimulation; UDRS = Unified Dystonia Rating Scale. |

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| Supplementary Table 2. Changes of BFMDRS/UDRS total scores and subscores |
|  | Baseline |  | Best scores after DBS |  | Long-term follow-ups |
| 　 | Total scores | Face and larynx | Neck | Limbs and trunk | 　 | Total scores | Face and larynx | Neck | Limbs and trunk | 　 | Total scores | Face and larynx | Neck | Limbs and trunk |
| Pt 1 | 55 | 5 | 7 | 43 |  | 32 | 6 | 4 | 22 |  | 57 | 20 | 7 | 30 |
| Pt 2 | 33 | 3 | 0 | 30 |  | 9 | 1 | 0 | 8 |  | 55 | 19 | 0 | 36 |
| Pt 3 | 101.5 | 13.5 | 8 | 80 |  | 35 | 0 | 0 | 35 |  | 71.5 | 11.5 | 6 | 54 |
| Pt 4 | 35.5 | 3.5 | 8 | 24 |  | 23 | 6 | 4.5 | 12.5 |  | 44.5 | 14.9 | 4.5 | 25 |
| Pt 5 | 112 | 24 | 8 | 80 |  | 46.5 | 2 | 2 | 42.5 |  | 93 | 15 | 6 | 72 |
| Pt 6 | 46 | 2 | 3 | 41 |  | 26 | 1 | 0 | 25 |  | 50.5 | 3.5 | 2 | 45 |
| Pt 7 | 64 | 5 | 8 | 41 |  | 44 | 2.5 | 3 | 38.5 |  | 58.5 | 6 | 6 | 46.5 |
| Pt 10 | 32 | 0 | 0 | 32 |  | 2 | 0 | 0 | 2 |  | 24 | 4.5 | 1.5 | 18 |
| Pt 11 | 74 | 6 | 12 | 68 |  | 11 | 4 | 0 | 7 |  | 59 | 15 | 12 | 36 |
| The total scores with the subscores (face and larynx, neck, and limb and trunk) are shown. The data were not available for patient 8 and 9.BFMDRS-M = Burke-Fahn-Marsden Dystonia Rating Scale motor score; DBS = deep brain stimulation; UDRS = Unified Dystonia Rating Scale. |

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| Supplementary Table 3. Lead positions for the multi-county cohort. |
|  | Original electrodes (x, y, z) | Additional electrodes (x, y, z) |
| Pt 1a | Left GPi: -20.9, 2.4, -5.3Right GPi: 20.5, 0.3, -4.5 | Left STN: -10.9, -2.3, -5.9 |
| Pt 2a | Left GPi: -18.9, 2.0, -2.3Right GPi: 21.8, 4.0, -0.6 | Right GPi: 20.7, 6.9, -6.5 Right STN: 10.3, -0.3, -6.0 |
| Pt 4b | Left GPi: 114.3, 109.9, 99.8Right GPi: 81.3, 111.2, 99.7 | Left GPi: not availableRight GPi: not available |
| Pt 5b | Left GPi: 117.4, 108.8, 100.0Right GPi: 76.7, 108.2, 100.1 | Left GPi: not availableRight GPi: not available |
| Pt 6b | Left GPi: 122.7, 89.6, 80.0Right GPi: 82.0, 90.0, 81.5 | Left GPi: not availableRight GPi: not available |
| Pt 7b | Left GPi: 116.4, 113.3, 93.3Right GPi: 83,4, 112.1, 90.0 | Left GPi: not availableRight GPi: not available |
| Pt 8a | Left GPi: no availableRight GPi: 15.5, 2.2, -5.0 | Left GPi: -19.3, 1.8, -5.6Right GPi: 19.0, 2.8, -5.3 |
| Pt 9a | Left GPi: -18.8, 2.8, -4.0Right GPi: -18.6, 3.3, -4.3 | Left STN: not availableRight STN: not available |
| Pt 10a | Left GPi: -20.9, 2.3, -8Right GPi: 19.3, 1.8, -8.6 | Left STN: -11.5, 2.5, 4.0 |
| Pt 11a | Left GPi: -21.0, 1.0, -4.4Right GPi: 18.3, 2.9, -2.9 | Left STN: 10.5, -3.0, -4.4Right STN: 12.0, -3.0, -8.0 |
| a The anatomical coordinates for the electrode tips relative to the midcommissural point are shown; x (mm), lateral to midline; y (mm), anterior to midcommissural point; z (mm), dorsal to intercommissural plane. b The anatomical coordinates for the upper limit of the second deepest contact (electrode model 3389, Medtronic), which was confirmed to be aligned to the anatomical target. A direct targeting based on MRI with immediate post-operative control were used.Electrode positions for patients 3 were not available. Abbreviations: GPi = globus pallidus internus, STN = subthalamic nucleus |

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| Supplementary Table 4. The list of studies which reported individual demographic data and motor outcomes for DYT1 patients. |
| Studies | Total Number of patients reported | Number of DYT1 patients who fulfilled the inclusion criteria |
| Alcindor et al. (2010)e1 | 1 | 1 |
| Anheim et al. (2008) e2 | 2 | 2 |
| Ben-Haim et al. (2016) e3 | 5 | 4 |
| Borggraefe et al. (2008) e4 | 1 | 1 |
| Borggraefe et al. (2010) e5 | 6 | 3 |
| Cersosimo et al. (2008) e6 | 10 | 3 |
| Cheung et al. (2013) e7 | 2 | 1 |
| Cif et al. (2010) e8 | 26 | 26 |
| Dupre et al. (2018) e9 | 1 | 1 |
| Eltahawy et al. (2004) e10 | 15 | 0 |
| Goto et al. (2006) e11 | 3 | 3 |
| Jin et al. (2012) e12 | 1 | 1 |
| Krause et al. (2004) e13 | 10 | 4 |
| Krause et al. (2016) e14 | 8 | 6 |
| Lumsden et al. (2013) e15 | 63 | 4 |
| Markun et al. (2012) e16 | 14 | 13 |
| Mehrkens et al. (2009) e17 | 18 | 3 |
| Mehrkens et al. (2010) e18 | 5 | 3 |
| Miri et al. (2014) e19 | 9 | 3 |
| Miyagi et al. (2013) e20 | 2 | 2 |
| Park et al. (2016) e21 | 36 | 4 |
| Parr et al. (2007) e22 | 4 | 1 |
| Pauls et al. (2017) e23 | 22 | 3 |
| Petrossian et al. (2013) e24 | 14 | 3 |
| Schrader et al. (2011) e25 | 6 | 1 |
| Sensi et al. (2009) e26 | 11 | 1 |
| Sobstyl et al. (2008) e28 | 1 | 1 |
| Sobstyl et al. (2011) e27 | 1 | 1 |
| Starr et al. (2006) e30 | 23 | 5 |
| Starr et al. (2014) e29 | 6 | 5 |
| Tisch et al. (2007) e31 | 15 | 0 |
| Tronnier et al. (2000) e32 | 3 | 1 |
| Tsuboi et al. (2019) e33 | 16 | 4 |
| Tustin et al. (2019) e34 | 60 | 5 |
| Vasques et al. (2009) e35 | 40 | 15 |
| Vidailhet et al. (2007) e36 | 22 | 7 |
| Zorzi et al. (2005) e37 | 12 | 1 |

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| Supplementary Table 5. Suboptimal responders in the historical cohort. |
| References | Sex | Age at onset (years) | Disease duration before DBS (years) | Age at DBS (years) | Follow-up after DBS (years) | Cranial involvement before DBS | Cranial involvement before or after DBS | BFMDRS or UDRS |
| Baseline | Best | Last FU | Percentage improvement at last FU from baseline |
| e33 a | M | 5 | 6 | 11 | 108 | 1 | 1 | 55 | 32 | 57 | -4% |
| e33 a | M | 6 | 2 | 8 | 144 | 1 | 1 | 33 | 9 | 60 | -82% |
| e20 a | M | 7 | 2 | 9 | 12 | 0 | 0 | 78 | 15 | 88 | -13% |
| e20 a | F | 9 | 2 | 11 | 180 | 0 | 1 | 42 | 14 | 60 | -43% |
| e34 a | NA | 10 | 5.75 | 15.75 | 24 | NA | NA | 53 | 16 | 42 | 21% |
| e8 a | M | 8 | 9 | 17 | 96 | 1 | 1 | 35.5 | 23 | 29.5 | 17% |
| e8 a | F | 7 | 8 | 15 | 216 | 1 | 1 | 46 | 25.5 | 35 | 24% |
| e8 a | F | 6.5 | 2.75 | 9.25 | 144 | 1 | 1 | 101.5 | 35 | 95 | 6% |
| e14 a | F | 5 | 2 | 7 | 42 | 1 | 1 | 52 | 16 | 41 | 21% |
| e16 b | M | 5 | 22 | 27 | 28 | 1 | 1 | 74 | 60 | 60 | 19% |
| e8 b | M | 8 | 27.25 | 35.25 | 48 | 1 | 1 | 51 | 23 | 41 | 20% |
| e8 b | M | 6 | 17.5 | 23.5 | 204 | 1 | 1 | 112 | 46.5 | 86 | 23% |
| e36 b | F | 8 | 18 | 26 | 36 | NA | NA | 42 | 29 | 30 | 29% |
| e25 b | M | 12 | 36 | 48 | 26 | 0 | 0 | 33 | 29 | 29 | 12% |
| e23 b | M | 9 | 33 | 42 | 39 | NA | NA | 62 | 62 | 62 | 0% |
| e10 b | M | 6 | 26.7 | 32.7 | 48 | NA | NA | 75 | 67 | 67 | 11% |
| BFMDRS-M = Burke-Fahn-Marsden Dystonia Rating Scale motor score; DBS = deep brain stimulation; NA = not assessed; UDRS = Unified Dystonia Rating Scale.a Patients with short disease duration before DBS in Table 4.b Patients with long disease duration before DBS in Table 4. |

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| Supplementary Table 6. Good responders in the historical cohort. |
| References | Sex | Age at onset (years) | Disease duration before DBS (years) | Age at DBS (years) | Follow-up after DBS (years) | Cranial involvement before DBS | Cranial involvement before or after DBS | BFMDRS or UDRS |
| Baseline | Best | Last FU | Percentage improvement at last FU from baseline |
| e9 | M | 15 | 3 | 18 | 15 | 0 | 0 | 46 | 1 | 1 | 98 |
| e3 | F | 6 | 7 | 13 | 102 | NA | NA | 81 | 0 | 0 | 100 |
| e3 | M | 8 | 3 | 11 | 24 | 0 | 0 | 17 | 2 | 2 | 88 |
| e3 | M | 8 | 3 | 11 | 12 | 1 | 1 | 107 | 48 | 48 | 55 |
| e12 | F | 5 | 2 | 7 | 30 | 0 | 0 | 34 | 3 | 3 | 91 |
| e36 | F | 9 | 9 | 18 | 36 | NA | NA | 25 | 0 | 1 | 96 |
| e36 | M | 7 | 29 | 36 | 36 | NA | NA | 59.5 | 27.5 | 27.5 | 54 |
| e36 | M | 8 | 32 | 40 | 36 | NA | NA | 61 | 17 | 17 | 72 |
| e36 | M | 5 | 17 | 22 | 36 | NA | NA | 95.5 | 41 | 43 | 55 |
| e36 | F | 7 | 23 | 30 | 36 | NA | NA | 58.5 | 4.5 | 4.5 | 92 |
| e36 | M | 9 | 21 | 30 | 36 | NA | NA | 44.5 | 28 | 28 | 37 |
| e13 | M | 10 | 17.7 | 27.7 | 24 | NA | NA | 70 | 9.5 | 10 | 86 |
| e13 | M | 9 | 12.5 | 21.5 | 47 | NA | NA | 66.5 | 23.5 | 24.5 | 63 |
| e13 | M | 10 | 8.8 | 18.8 | 52 | NA | NA | 77 | 25 | 35 | 55 |
| e2 | NA | 10 | 38 | 48 | 12 | 0 | 0 | 82.5 | 37 | 37 | 55 |
| e2 | NA | 8 | 7 | 15 | 12 | 0 | 0 | 74.5 | 10 | 10 | 87 |
| e19 | F | 7 | 4 | 11 | 12 | NA | NA | 41 | 3 | 3 | 93 |
| e19 | F | 7 | 7 | 14 | 11 | NA | NA | 44 | 14 | 14 | 68 |
| e19 | F | 8 | 11 | 19 | 11 | NA | NA | 20 | 4 | 4 | 80 |
| e19 | M | 9 | 12 | 21 | 11 | NA | NA | 46 | 16 | 16 | 65 |
| e7 | M | 11 | 4 | 15 | 72 | 0 | 0 | 64 | 0 | 4 | 94 |
| e18 | M | 8 | 4 | 12 | 101 | 0 | 0 | 55 | 7 | 7 | 87 |
| e18 | F | 12 | 2 | 14 | 100 | 1 | 1 | 65 | 6 | 6 | 91 |
| e18 | F | 7 | 1 | 8 | 50 | 0 | 0 | 60 | 4 | 5 | 92 |
| e18 | F | 7 | 13 | 20 | 90 | NA | NA | 32 | 6 | 7 | 78 |
| e4 | F | 8 | 1 | 9 | 14 | 0 | 0 | 58 | 1 | 1 | 98 |
| e24 | M | 11 | 1.8 | 12 | 80 | 0 | 0 | 37 | 4 | 4 | 89 |
| e24 | M | 14 | 3 | 17 | 37 | 0 | 0 | 30 | 0 | 0 | 100 |
| e24 | M | 9 | 3 | 12 | 39 | 0 | 0 | 40 | 13 | 13 | 68 |
| e22 | M | 8 | 7 | 15 |  | 1 | 1 | 66 | 8 | 8 | 88 |
| e26 | F | 10 | 13 | 23 | 83 | 0 | 2 | 33 | 7 | 7 | 79 |
| e21 | M | 8 | 4 | 12 | 60 | NA | NA | 30 | 9.5 | 9.5 | 68 |
| e21 | M | 9 | 8 | 17 | 36 | NA | NA | 40.5 | 6 | 6 | 85 |
| e21 | M | 7 | 3 | 10 | 12 | NA | NA | 32.5 | 6.5 | 6.5 | 80 |
| e21 | M | 7 | 25 | 32 | 12 | NA | NA | 50 | 24 | 24 | 52 |
| e11 | M | 7 | 14 | 21 | 12 | 0 | 0 | 68.5 | 0 | 0 | 100 |
| e11 | M | 12 | 15 | 27 | 12 | 0 | 0 | 3 | 0.5 | 0.5 | 83 |
| e11 | M | 9 | 6 | 15 | 12 | 0 | 0 | 61 | 3 | 3 | 95 |
| e16 | M | 11 | 1 | 12 | 45 | 0 | 2 | 34 | 1 | 1 | 97 |
| e16 | F | 8 | 9 | 17 | 46 | 1 | 1 | 51.5 | 0 | 0 | 100 |
| e16 | M | 7 | 10 | 17 | 26 | 1 | 1 | 90 | 44 | 44 | 51 |
| e16 | M | 9 | 6 | 15 | 11 | 0 | 2 | 58 | 14 | 14 | 76 |
| e16 | M | 12 | 5 | 17 | 28 | 1 | 1 | 49.5 | 22 | 22.5 | 55 |
| e16 | F | 8 | 3 | 11 | 77 | 0 | 2 | 46 | 2 | 2 | 96 |
| e16 | M | 8 | 9 | 17 | 24 | 1 | 1 | 48.5 | 10 | 10 | 79 |
| e16 | F | 7 | 3 | 10 | 36 | 0 | 2 | 78 | 0 | 11.5 | 85 |
| e16 | F | 7 | 4 | 11 | 32 | 0 | 2 | 32 | 2 | 2 | 94 |
| e16 | F | 4 | 19 | 23 | 48 | 1 | 1 | 22 | 12 | 12 | 45 |
| e16 | M | 7 | 15 | 22 | 28 | 1 | 1 | 69.5 | 42 | 42 | 40 |
| e16 | F | 9 | 2 | 11 | 15 | 0 | 2 | 22 | 3 | 3 | 86 |
| e29 | M | 7 | 2 | 9 | 12 | 1 | 1 | 44 | 4 | 4 | 91 |
| e29 | M | 8 | 3 | 11 | 12 | 0 | 2 | 68 | 0 | 0 | 100 |
| e29 | M | 13 | 2 | 15 | 12 | 1 | 1 | 22.5 | 1 | 1 | 96 |
| e29 | F | 7 | 4 | 11 | 12 | 0 | 2 | 36.5 | 0 | 0 | 100 |
| e29 | M | 6 | 1 | 7 | 12 | 0 | 2 | 20 | 9 | 9 | 55 |
| e29 | NA | 8 | 9 | 17 | 15 | 0 | 2 | 52 | 0 | 0 | 100 |
| e29 | NA | 7 | 10 | 17 | 17 | NA | NA | 90 | 44 | 44 | 51 |
| e29 | NA | 9 | 6 | 15 | 14 | NA | NA | 58 | 14 | 14 | 76 |
| e29 | NA | 12 | 5 | 17 | 11 | 0 | 2 | 49.5 | 22 | 22 | 56 |
| e14 | M | 6 | 10 | 16 | 119 | NA | NA | 74.5 | 34 | 35.5 | 52 |
| e14 | F | 7 | 5 | 12 | 156 | NA | NA | 41 | 9 | 24 | 41 |
| e14 | M | 7 | 1 | 8 | 20 | 1 | 1 | 50 | 1.5 | 1.5 | 97 |
| e14 | F | 10 | 7 | 17 | 20 | NA | NA | 36 | 17 | 20 | 44 |
| e6 | F | 6 | 4 | 10 | 93 | NA | NA | 67 | 34 | 45 | 33 |
| e6 | M | 24 | 4 | 28 | 35 | NA | NA | 16.5 | 4 | 5 | 70 |
| e6 | F | 5 | 4 | 9 | 33 | NA | NA | 45 | 21 | 21 | 53 |
| e27 | F | 14 | 3 | 17 | 24 | 0 | 0 | 25 | 5 | 5 | 80 |
| e28 | M | 7 | 17 | 24 | 60 | 0 | 0 | 112 | 11 | 23 | 79 |
| e34 | NA | 6.5 | 0.8 | 7.3 | 24 | NA | NA | 50 | 6 | 22.5 | 55 |
| e34 | NA | 10.25 | 3.65 | 13.9 | 24 | NA | NA | 57 | 9 | 9 | 84 |
| e34 | NA | 9 | 4.75 | 13.75 | 12 | NA | NA | 25 | 10 | 10 | 60 |
| e34 | NA | 8 | 3.75 | 11.75 | 24 | NA | NA | 59.5 | 10 | 12.5 | 79 |
| e8 | F | 7.5 | 3 | 10.5 | 120 | 1 | 1 | 81 | 0 | 0 | 100 |
| e8 | F | 6 | 20 | 26 | 120 | 1 | 1 | 97.5 | 10 | 10 | 90 |
| e8 | F | 7 | 7 | 14 | 108 | 1 | 1 | 63 | 0 | 5 | 92 |
| e8 | M | 7 | 1.5 | 8.5 | 240 | 0 | 1 | 76 | 0 | 38 | 50 |
| e8 | M | 9 | 4 | 13 | 90 | 0 | 0 | 51 | 0 | 15 | 71 |
| e8 | F | 11.5 | 2 | 13.5 | 90 | 0 | 0 | 24 | 0 | 15.5 | 35 |
| e8 | F | 9 | 4.5 | 13.5 | 90 | 1 | 1 | 37.5 | 4 | 21 | 44 |
| e8 | M | 8 | 5 | 13 | 84 | 0 | 0 | 37 | 0 | 4 | 89 |
| e8 | M | 7 | 6 | 13 | 84 | 1 | 1 | 65.5 | 6 | 8.5 | 87 |
| e8 | M | 6.5 | 35 | 41.5 | 84 | 1 | 1 | 84.5 | 28.5 | 29 | 66 |
| e8 | M | 6 | 7 | 13 | 72 | 1 | 1 | 69 | 0 | 2 | 97 |
| e8 | F | 7 | 1.5 | 8.5 | 72 | 0 | 0 | 35.5 | 0 | 0 | 100 |
| e8 | F | 7 | 43 | 50 | 66 | 1 | 1 | 28 | 1 | 4.5 | 84 |
| e8 | F | 20 | 5 | 25 | 66 | 0 | 0 | 38 | 0 | 2 | 95 |
| e8 | F | 8 | 28.5 | 36.5 | 63 | 1 | 1 | 67 | 10.5 | 16 | 76 |
| e8 | F | 6.5 | 12.5 | 19 | 60 | 0 | 0 | 64 | 0 | 0 | 100 |
| e8 | F | 9 | 56.5 | 65.5 | 51 | 0 | 0 | 27.5 | 10.5 | 10.5 | 62 |
| e8 | M | 8 | 40 | 48 | 48 | 1 | 1 | 61.5 | 21 | 25 | 59 |
| e8 | F | 9 | 3 | 12 | 45 | 0 | 0 | 40 | 0 | 5 | 88 |
| e8 | F | 6 | 3 | 9 | 42 | 0 | 0 | 42 | 3 | 3 | 93 |
| e8 | F | 9 | 1.5 | 10.5 | 36 | 1 | 1 | 60 | 0 | 0 | 100 |
| e33 | F | 12 | 12 | 24 | 156 | 1 | 1 | 50 | 6 | 23 | 54 |
| e33 | M | 8 | 24 | 32 | 108 | 1 | 1 | 44 | 0 | 0 | 100 |
| e15 | M | 11.2 | 3 | 14.2 | 12 | NA | NA | 24 | 5 | 5 | 79 |
| BFMDRS-M = Burke-Fahn-Marsden Dystonia Rating Scale motor score; DBS = deep brain stimulation; NA = not assessed; UDRS = Unified Dystonia Rating Scale. |

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