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EDITED BY

John Robert Younce,
University of North Carolina at Chapel Hill,
United States

*CORRESPONDENCE

Aparna Wagle Shukla,
✉ aparna.shukla@neurology.ufl.edu

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Beyond the neck: long-term motor and non-motor outcomes of pallidal DBS in craniocervical dystonia

Matthew Remz¹, Pamela Zeilman¹, Samyukta Senthil¹,
Rohini Kumar¹, Isabelle Gerzenshtein¹, Hikaru Kamo^{1,2} and
Aparna Wagle Shukla^{1*}

¹Department of Neurology, Fixel Institute for Neurological Diseases, University of Florida, Gainesville, FL, United States, ²Department of Neurology, Juntendo University School of Medicine, Tokyo, Japan

Pallidal deep brain stimulation (DBS) is effective for treating cranial or craniocervical dystonia, a disorder characterized by abnormal facial expressions, impaired eyelid opening, speech and chewing difficulties, and abnormal head posture, leading to physical disability and functional impairment. However, long-term data on motor, non-motor, and quality-of-life outcomes remain limited with less robust available data than cervical dystonia. In 19 patients with craniocervical dystonia, the Unified Dystonia Rating Scale (UDRS) scores improved by 34.5% at 3–6 months, 51.0% at 1–2 years, 33.3% at 3–5 years, and 43.4% at the most recent follow-up (median of individual improvements). Body region analysis showed that the proportion of patients classified as DBS responders, defined by a ≥ 25 –30% improvement in UDRS scores, ranged from 31.2% to 43.8% at 3–6 months, increasing to a range of 50%–83.3% by the 6–9-year follow-up. Alongside motor improvements, trends in depression and social quality-of-life domain improvements were noted with more robust changes likely limited by sample size and the lack of disease specific metrics. Longer therapy duration appeared to enhance the magnitude of gains. Overall, pallidal DBS improves motor function and daily activities in craniocervical dystonia, with durable and potentially progressive long-term benefits.

KEYWORDS

blepharospasm, craniocervical dystonia, Meige, oromandibular dystonia, DBS, pallidal, globus pallidus internus, long-term

Introduction

Craniocervical dystonia is a form of segmental dystonia affecting the eyes, lower face, jaw, tongue, and neck. Clinical manifestations commonly include impaired eyelid opening (blepharospasm), abnormal facial expressions, speech and mastication difficulties (oromandibular dystonia), and abnormal head posture (cervical dystonia) [1]. Blepharospasm is a common adult-onset focal dystonia characterized by involuntary orbicularis oculi muscle spasms that are usually bilateral, synchronous and symmetrical and excessive blinking (>16 blinks/min) or forceful eye closure [2]. The clinical presentations of oromandibular dystonia include varying combinations of abnormal jaw closing or opening, tongue, or lower face movements [3]. Meige syndrome, named after a French Neurologist Henry Meige, represents a more complex dystonia phenotype in which

blepharospasm is accompanied by oromandibular and occasionally cervical dystonia. These conditions are more prevalent in women than men, with a typical age of onset in the fifth decade of life [1]. Craniocervical dystonia can cause substantial functional impairment, affecting vision, speech, swallowing, and social interaction, and is frequently associated with non-motor symptoms such as anxiety, depression [4], and reduced health related quality-of-life (HRQoL) [5]. While botulinum toxin (BoNT) injections remain first-line therapy [6, 7], pharmacological options may provide suboptimal relief in many patients. Deep brain stimulation (DBS) of the bilateral globus pallidus internus (GPi) is an established and effective treatment for medication-refractory dystonia with many studies reporting good outcomes for focal cervical dystonia [8]. However, data on long-term outcomes encompassing motor and non-motor symptoms, as well as quality-of-life, in craniocervical dystonia remain limited and there is less uniform agreement in the field about its efficacy. We therefore aimed to evaluate long-term outcomes of GPi DBS in craniocervical dystonia and further characterize the durability of motor, non-motor, and HRQoL improvements in patients receiving care at our movement disorders center.

Materials and methods

Study design and data source

This study was a retrospective analysis of a series of craniocervical dystonia patients who underwent DBS at the Fixel Institute for Neurological Diseases, University of Florida. Diagnosis of craniocervical dystonia (blepharospasm, oromandibular dystonia and cervical dystonia) in these patients was established by movement disorders trained neurologists. Patients had been selected for DBS following comprehensive interdisciplinary evaluation and careful risk-benefit assessment [9]. DBS electrodes and the latest pulse generators available at the time of scheduling of DBS were used. Detailed information about surgical procedures and electrode measurements has been described previously [10]. The lateral portion of the sensorimotor posteroventral GPi was targeted to balance the ability to deliver higher stimulation amplitude with capsular side effects with input insights gained from awake surgery with microelectrode recording.

Preoperative clinical data for these patients were available in our database up to 12 months prior to DBS surgery. Postoperative follow-up at our center is scheduled monthly for the first six to 12 months, followed by visits at 12 months and annually thereafter. All patients included in this study had data entered in our database from one or more postoperative follow-up visits.

Data collection and outcome assessment

Data was retrospectively retrieved from the INFORM database, and electronic medical records at the University of Florida. Information was collected using a standardized proforma, with independent verification by two investigators to ensure reliability. INFORM is a clinical-research database that provides information on demographics, clinical, and functional characteristics of patients with movement disorders at the University of Florida. Clinical

information extracted included demographic data, disease characteristics and longitudinal outcome data extracted at prespecified intervals: prior to surgery (baseline or T0) and postoperatively at 3–6 months (T1), 1–2 years (T2), 3–5 years (T3), and the most recent follow-up (T4). Baseline clinical data were derived from records within the 12 months preceding DBS implantation, with dystonia severity assessed under standard treatment conditions during routine clinical evaluations. To accommodate variability in clinical follow-up inherent to routine care, time windows were defined in advance and data were permitted from within 3 months of baseline and T1 visit and within 6 months of the desired timepoint for the T2, T3, T4 visits. For each interval, the clinical encounter closest to the target time point was selected for analysis. If no encounter occurred within the specified window, the corresponding data point was considered missing.

Outcome measures: The Unified Dystonia Rating Scale (UDRS) was administered by fellowship-trained movement disorders specialists at baseline and follow-up intervals, with scores documented in our database. The UDRS evaluates dystonia in 14 body regions such as eyes/upper face, lower face, jaw/tongue, larynx, neck, trunk, and limbs (proximal and distal) by rating both severity (amplitude/intensity) and duration (proportion of time present) of dystonic movements or postures. Severity and duration scores from individual regions are summed to generate a total score (maximal total score of UDRS is 112), with higher scores indicating more severe and widespread dystonia [11]. In our study, in addition to total UDRS scores, we obtained body region-specific UDRS scores categorized into five subdomains: upper face (eyes), lower face (including jaw, tongue, larynx), neck, upper limb (arm and/or hand), and trunk/lower limb (leg and/or foot). For each body region, dystonia duration and motor severity scores were summed to generate regional scores, and the total UDRS score was calculated as the sum across all regions. For the purposes of our study, the primary body regions of interest were the upper face (max score 16), lower face (max score 48), and neck (max score 16) which were analyzed separately in addition to the total UDRS scores.

HRQoL outcomes were assessed at baseline and follow-up time points using the patient-reported Short Form Health Survey-36 (SF-36). The eight SF-36 domains were evaluated and summarized into the total score, Physical Component Summary (PCS), and Mental Component Summary (MCS) [12]. The SF-36 total score was calculated using previously established methods. In addition, non-motor symptoms were assessed using the Beck Depression Inventory (BDI, maximal total score of 63 with higher scores indicating worse depression), Beck Anxiety Inventory (BAI, maximal total score of 63 with higher scores indicating worse anxiety), and the Apathy Scale (AS, maximal score of 42 with higher scores indicating more apathy) described in previous publication [13].

Statistical analysis

Descriptive data for demographics and baseline characteristics were summarized as median values with ranges. Median values and interquartile range (IQR) were calculated at each time point for total and individual body region UDRS scores. Paired comparisons between baseline and each postoperative time point were conducted using the Wilcoxon signed-rank test among patients

with available paired measurements. The % change in UDRS total score (UDRS% change) after surgery was calculated with the following formula: (preoperative UDRS – postoperative UDRS)/preoperative UDRS $\times$ 100. While no specific threshold has been universally established for a clinically meaningful change in UDRS scores, improvements of approximately 25%–30% are generally considered indicative of a DBS responder [14]. While total UDRS could be interpretable as a quasi-continuous measure only when multiple ordinal items are summed across body regions, reporting percentage changes at the level of an individual body region score (ordinal rather than continuous) could be potentially misleading. For example, a score of 4 on the Eye item does not represent “twice as severe” as a score of 2 and calculating percentage changes for a single item is therefore not meaningful. Thus, for individual body regions, we considered meaningful improvement (minimally clinically important difference or MCID) as follows: eyes and upper face (≥ 2 -point drop, representing $\geq 25\%$ improvement of max 8 points for this body region), neck (≥ 2 -point drop, representing $\geq 25\%$ improvement of 8 points), and lower face (lower face, jaw, tongue, and larynx; ≥ 6 -point drop representing $\geq 25\%$ improvement of 24 points for these body regions). We examined the number of individuals achieving these thresholds of improvement in each body region at each of the follow-up time points. Percentage change in SF-36 PCS and MCS was calculated as the difference between the postoperative and preoperative scores, divided by the preoperative (baseline) score, and multiplied by 100 (PCSA and MCSA). Change in individual domains of SF-36, BDI, BAI and AS was noted.

We further examined the longitudinal DBS related improvement in dystonia using a linear mixed-effects model with UDRS score treated as a continuous outcome and a subject-specific random intercept. A similar approach was used for determining longitudinal change in HRQoL and change in each domain was estimated by independently modeling domain score as a continuous outcome with a subject-specific random intercept. We included baseline factors such as age at onset for dystonia, duration of dystonia before DBS, number of body areas affected (eyes, upper face, jaw, tongue, lower face, neck) and sex as covariates in each model. Sensitivity analyses were performed using random patient specific slopes, quadratic fit, and exclusion of variables including disease duration and number of body regions affected to ensure model viability. Visual inspection of residuals and fitted value plots were performed and no flagrant violations of normality of homoscedasticity were appreciated. Confidence intervals were obtained using parametric bootstrapping. To account for multiple comparisons, *p*-values were adjusted using Benjamini–Hochberg false discovery rate (FDR) technique. Statistical significance was assessed at a two-sided FDR-adjusted α level of 0.05.

Statistical analysis was performed with SPSS version 26 and RStudio: Integrated Development Environment for R (Posit Software, PBC, Boston, MA.). Coding for some aspects of statistical modeling was developed with assistance from OpenAI sources.

Ethics, data availability, and reporting guidelines

The study was approved by the Institutional Review Board of the University of Florida (IRB No. 201702279), with a waiver of

informed consent due to the use of de-identified data. The UF INFORM database was independently approved by the Institutional Review Board of the University of Florida (IRB No. 201501166) and all subjects previously provided informed consent in accordance with the Declaration of Helsinki. The datasets analyzed during the current study are not publicly available because of institutional regulations and patient privacy considerations but are available from the corresponding author on request and with appropriate approvals.

Results

Study sample

We identified 19 patients (13 females). The median (range) age of this cohort at first surgery was 66 years (48–83 years) and mean (SD) was 64.8 (8.9) years. The median (range) age at dystonia onset was 59 years (40–72 years) and mean (SD) was 57 (9.3) years. The median (range) duration of follow-up after DBS was 6 years (1–24 years) and mean (SD) was 7.8 (6.1) years. Eight patients remained actively followed at our center. Six patients relocated to another state, two patients were deceased, two discontinued follow-ups for unknown reasons, and one patient had documented travel difficulties that prevented continued follow-up (Table 1). Two of these individuals had exposure to neuroleptic medications prior to onset of symptoms. None of these individuals developed dystonia secondary to structural brain lesions or neurodegenerative diseases. There was no history of prior ablative surgery (e.g., pallidotomy or thalamotomy) for dystonia before DBS implantation or a revision surgery or hardware explantation due to infection or device failure within a year of DBS. Three patients (15.8%) had undergone blepharoplasty, 2 (10.5%) prior eyelid myectomy, and 1 (5.3%) a sling procedure. There was no reported history of tremors in addition to dystonia.

15 patients (78.9%) were using oral medications at baseline, including 11 (57.9%) on benzodiazepines, 2 (10.5%) on baclofen, and 4 (21.1%) on trihexyphenidyl. At the most recent follow-up, 9 patients (47.4%) continued to use oral agents for dystonia. Four patients (21%) were not taking oral medications prior to surgery, whereas eight patients (42%) were not requiring oral medications at the most recent follow-up after DBS. 14 patients (73.6%) received BoNT approximately every 12 weeks prior to surgery, and 11 (57.9%) continued BoNT therapy at the last follow-up (Table 1). Additionally, DBS programming data are summarized with the most recent settings displayed in Table 2.

Motor outcomes measured with UDRS

Among 19 subjects, the median baseline UDRS score was 19 (IQR: 9–20.5). Relative to baseline, median UDRS scores improved at T1 (median 9.5, IQR: 6–19.8, $n = 16$; $p = 0.04$), at T2 (median 8.5, IQR: 4–13, $n = 16$; $p = 0.02$), at T3 (median 5.5, IQR: 2–12.5, $n = 12$; $p = 0.02$), and at T4 (median 6.5, IQR: 4.3–11, $n = 6$; $p = 0.04$). The percentage improvements in UDRS scores (median of individual % improvements) was 34.5% at T1, 51.0% at T2, 33.3% at T3, and 43.4% at T4. UDRS total scores demonstrated significant improvement across all follow-up time points (all *p*-values FDR-

TABLE 1 Demographic and clinical characteristics of individual participants.

S	Sex	Body regions affected	Age at symptom onset in yrs	Disease duration in yrs	Age at DBS in yrs	DBS duration follow-up in yrs	Reason for lost to follow-up	UDRS score before DBS	Medications before DBS	Medications at most recent follow-up	Use of BoNT before DBS		Use of BoNT at most recent follow-up	
											Y/ N	BoNT type and dose in units	Y/ N	BoNT type and dose in units
1	M	Eyes, upper face, lower face, cervical	59	8	67	10	Deceased	19	c	None	N	NA	N	NA
2	F	Eyes, lower face	40	25	65	14	Relocation	19	c	None	Y	OnaBoNT; 400	Y	OnaBoNT; 300
3	M	Eyes, lower face	64	19	83	4	Deceased	19	c	c	N	NA	N	NA
4	F	Eyes	50	2	53	2	Active follow-up +	6	b	b	Y	OnaBoNT; 170	Y	OnaBoNT; 100
5	M	Eyes, upper face, lower face, jaw, tongue, cervical	63	5	68	2.5	Relocation	29	A, c	A, c	N	NA	N	NA
6	M	Eyes, upper face, Lower face, jaw, tongue, cervical	62	2	64	2.5	Relocation	40	None	None	Y	OnaBoNT; 200	Y	OnaBoNT; 200
7	F	Eyes, lower face, cervical	46	5	51	11	Reason not known	8	None	None	Y	OnaBoNT; 200	N	NA
8	F	Eyes, lower face, cervical	66	3	69	3	Reason not known	19	A, c	d	Y	OnaBoNT; 200	Y	OnaBoNT; 200
9	F	Eyes, jaw, lower face, cervical	61	4	65	8	Active follow-up +	9	e	e	Y	OnaBoNT; 200	N	NA
10	F	Eyes, cervical	72	6	78	8	Travel difficulties	19	e	c	Y	OnaBoNT; 200	N	NA
11	F	Eyes, upper face, Lower face, jaw, tongue, cervical	62	5	67	7	Active follow-up +	7	None	None	Y	OnaBoNT; 200	Y	OnaBoNT; 300

(Continued)

TABLE 1 Continued

S	Sex	Body regions affected	Age at symptom onset in yrs	Disease duration in yrs	Age at DBS in yrs	DBS duration follow-up in yrs	Reason for lost to follow-up	UDRS score before DBS	Medications before DBS	Medications at most recent follow-up	Use of BoNT before DBS		Use of BoNT at most recent follow-up	
											Y/ N	BoNT type and dose in units	Y/ N	BoNT type and dose in units
12	F	Eyes, upper face, Lower face, jaw, tongue, cervical	58	8	66	7	Active follow-up +	18	None	None	Y	OnaBoNT; 300	Y	OnaBoNT; 340
13	F	Eyes, jaws, cervical	51	7	58	1.5	Active follow-up +	21	c	c	Y	OnaBoNT; 300	N	NA
14	F	Eyes, cervical	53	16	69	0.5	Relocation	11	c	c	Y	RimaBoNT; 8,500	N	NA
15	M	Eyes, jaws, cervical	62	4	66	5	Relocation	3	a	None	Y	OnaBoNT; 300	N	NA
16	F	Eyes, upper face, Lower face, jaw, tongue, cervical	70	3	73	6	Active follow-up +	9	d	d	Y	OnaBoNT; 100	Y	OnaBoNT; 100
17	F	Eyes, upper face, Lower face, jaw, tongue, cervical	43	12	55	5	Active follow-up +	25	c	c	Y	OnaBoNT; 300	Y	OnaBoNT; 240
18	F	Eyes, upper face, Lower face, jaw, tongue, cervical	59	10	69	4	Relocation	20	c	c	Y	OnaBoNT; 200	Y	OnaBoNT; 100
19	M	Eyes, upper face, Lower face, jaw, tongue, cervical	43	5	48	24	Active follow-up +	24	b	None	Y	OnaBoNT; 200	N	NA

S: subject M: male, F: female, Y: yes, N: n, a: trihexyphenidyl, b: baclofen, c: clonazepam, d: gabapentin, e: lorazepam, BoNT: botulinum toxin, yrs: years, DBS: deep brain stimulation.

TABLE 2 DBS settings for craniocervical dystonia subjects at most recent follow-up.

Subjects	DBS system	Left lead					Right lead				
		Cathode contact	Anode contact	Amplitude	Pulse width (μs)	Frequency (Hz)	Cathode contact	Anode contact	Amplitude (V/mA)	Pulse width (μs)	Frequency (Hz)
Subject 1	Medtronic 3387 lead; Activa RC IPG	2-	C+	2.7 V	270	185	2-	C+	2.7 V	270	185
Subject 2	Medtronic 3387 lead; Activa PC IPG	1-	C+	3.5 V	150	120	9-	11+	2.5 V	150	120
Subject 3	Medtronic 3387 lead; Activa PC IPG	2-	C+	2.1 V	120	135	9-	C+	2.5 V	120	135
Subject 4	Medtronic 3387 lead; Percept PC IPG	2-	C+	2.2 mA	90	135	10-	C+	2.1 mA	90	135
Subject 5	Medtronic 3387 lead; Activa PC IPG	1-	C+	3.5 V	110	165	9-	C+	3.0 V	110	165
Subject 6	Medtronic 3387 lead; Activa RC IPG	1-, 2-	C+	2.0 V	90	135	2-, 3-	C+	2.0 V	90	135
Subject 7	Medtronic 3387 lead; Activa RC IPG	1-	2+	3.4 V	150	130	10-	C+	2.8 V	150	130
Subject 8	Medtronic 3387 lead; Percept PC IPG	2-	3+	2.4 V	120	125	3-	1+	1.3 V	120	125
Subject 9	Medtronic 3387 lead; Activa RC IPG	0-	1+	1.5 mA	240	120	9-	10+	1.9 mA	240	120
		1-	2+	1.3 mA	240	120	10-	11+	2.0 mA	240	120
Subject 10	Medtronic 3387 lead; Percept PC IPG	2-	C+	4.1 mA	180	70	9-	10+	1.9 mA	180	70
Subject 11	Medtronic 3387 lead; Percept RC IPG	1-, 2-	C+	1.5 mA	120	220	9-, 10-	C+	1.5 mA	180	220
Subject 12	Medtronic 3387 lead; Percept RC IPG	1-	2+	2.1 mA	120	150	9-	10+	2.8 mA	120	150
Subject 13	Medtronic 3387 lead; Percept RC IPG	1-	C+	4.8 mA	110	165	9-	C+	3.2 mA	110	165
Subject 14	Medtronic 3387 lead; Activa PC IPG	1-, 2-	0+	3.0 V	150	130	10-	C+	3.0 V	150	130

(Continued)

TABLE 2 Continued

Subjects	DBS system	Left lead					Right lead				
		Cathode contact	Anode contact	Amplitude	Pulse width (μs)	Frequency (Hz)	Cathode contact	Anode contact	Amplitude (V/mA)	Pulse width (μs)	Frequency (Hz)
Subject 15	Medtronic 3387 lead; Activa PC IPG	2-	C+	2.6 V	90	135	10-	C+	2.6 V	90	135
Subject 16	Medtronic 3387 lead; Activa RC IPG	0-, 1-	C+	2.4 V	110	130	8-, 9-	C+	2.7 V	110	130
Subject 17	Medtronic 3387 lead; Percept RC IPG	0-	C+	1.9 mA	350	125	11-, 10-	C+	1.8 mA	350	125
Subject 18	Medtronic 3387 lead; Percept RC IPG	1-	C+	1.5 mA	150	130	9-	C+	1.3 mA	40	130
Subject 19	Medtronic 3387 lead; Activa RC IPG	1-	C+	1.8 V	130	185	9-	C+	1.8 V	180	185

Subject 9 had interleaving settings. IPG, implantable pulse generator.

TABLE 3 Motor, Quality-of-life and Psychiatric outcomes over time.

Outcome measures	Baseline or T0 median (IQR)	3–6 months or T1 median (IQR)	1–2 years or T2 median (IQR)	3–5 years or T3 Median (IQR)	Most recent follow-up or T4 median (IQR)	3–6 months vs. baseline	1–2 years vs. baseline	3–5 years vs. baseline	Most recent follow-up yrs vs. baseline
UDRS									
Total	19 (9–20.5)	9.5 (6–19.8)	8.5 (4–13)	5.5 (2–12.5)	6.5 (4.3–11)	0.03*	0.01*	0.01*	0.04*
Eyes & upper face item	5 (4–6.5)	4.5 (3–5.3)	2.5 (2–4)	2 (0–3)	2.5 (0.5–3.8)				
Lower face item	6 (2.5–11.5)	4 (0–10)	2 (0–6.3)	1 (0–3.3)	1.5 (0–3)				
Neck item	4 (1–5.5)	4 (2–4)	1 (0–2.3)	0 (0–2)	1 (0–2)				
SF–36 HRQoL									
Physical function	55 (45–75)	70 (50–82.5)	65 (55–80)	70 (60–77.5)	75 (62.5–80)	0.65	0.68	0.65	0.83
Role physical	25 (0–62.5)	0 (0–62.5)	25 (0–75)	25 (6.3–50)	50 (0–100)	0.83	0.77	0.77	0.77
Bodily pain	51 (31–92)	52 (41–73)	52 (41–74)	51 (41–68.5)	51.5 (43.5–68.5)	0.70	0.84	0.84	0.70
General health	57 (42.5–72)	57 (46–77)	57 (45–72)	52 (40–67)	52 (46.8–70.8)	0.90	0.90	0.90	0.90
Physical component summary	52.5 (36.9–62.3)	52.3 (47.6–60.8)	53.3 (34.8–69.3)	51.3 (44.8–66.3)	63.5 (44.3–68.1)	0.72	0.39	0.41	1
Vitality	50 (22.5–55)	45 (32.5–50)	45 (35–55)	42.5 (31.3–55)	57.5 (41.25–70)	0.81	0.81	0.81	0.81
Social function	50 (38–75)	63 (50–75)	63 (38–88)	69 (41–88)	69 (53.3–75)	0.53	0.5382	0.5382	0.5775
Role emotional	67 (0–100)	67 (0–100)	67 (0–100)	100 (39.75–100)	83.5 (67–100)	1	1	1	1
Mental health	68 (56–80)	64 (56–72)	64 (56–84)	58 (52–80)	58 (53–81)	1	1	1	1
Mental component summary	55.8 (33.6–74.3)	60.5 (38.9–72)	50.5 (37.5–73.3)	61 (48.6–73.6)	66.6 (60.8–80.3)	0.39	0.58	0.4973	1
Psychological scales									
BDI	8.5 (4–11.3)	9 (5.8–11.5)	8.5 (4–11.5)	10.5 (6–14.3)	7 (5–7.5)	0.6098	0.1976	1	1
BAI	6 (1–8.5)	6 (4–10)	7 (2–12)	8 (2.5–12)	2 (1–3)	1	0.9902	1	1
AS	9 (8–10.8)	13 (10–18)	12 (7.5–16)	11 (6.25–16.25)	13 (8–14)	0.2722	0.2807	0.2722	N/A

*Denotes statistically significant FDR, corrected p-values with Wilcoxon test <0.05. This test was not run for eyes and upper face, lower face and neck UDRS, items. See manuscript for data reflecting the number of individuals (%) at each time point for Eyes and upper face (≥2-point improvement) Lower Face that includes jaw and tongue (≥6-point improvement) Neck (≥2-point improvement). IQR: interquartile range, yrs: years, mos: months, BDI: beck depression inventory, BAI: beck anxiety inventory, AS: apathy scale.

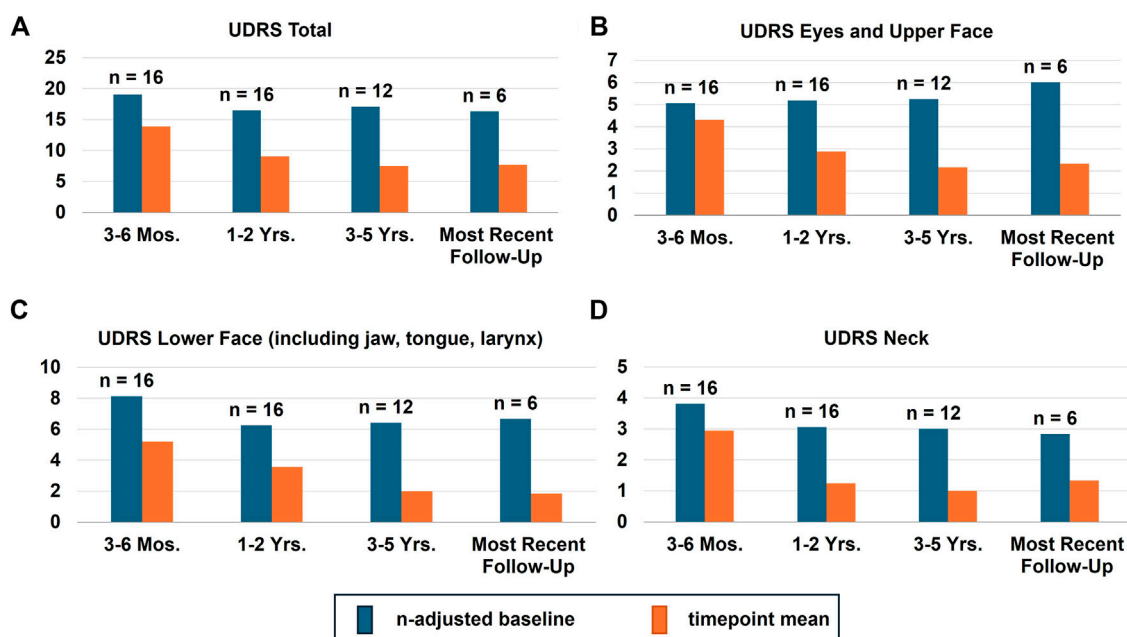


FIGURE 1

Bars represent mean values for longitudinal UDRS scores measured at 3–6 months, 1–2 years, 3–5 years, and the most recent follow-up. (A) UDRS total score; (B) UDRS eyes and upper face body region score; (C) UDRS lower face body region score (lower face, jaw, tongue, larynx); and (D) UDRS cervical body region score. Blue bars represent values before DBS and orange bars represent values after DBS. Bars represent mean data from individuals with available baseline and corresponding follow-up time points. The number of individuals is displayed above each bar.

adjusted). Full data, including median and IQRs, are available across all time points in Table 3. Figure 1 shows UDRS data (mean) from individuals with available baseline and corresponding follow-up time points.

Regarding regional changes in UDRS scores (Table 3), in the eyes and upper face region, 7/16 patients (43.8%) at T1, 12/16 (75.0%) at T2, 9/12 (75.0%) at T3, and 5/6 (83.3%) at T4 achieved a ≥ 2 -point reduction. In the lower face subdomain, 6/16 patients (37.5%) at T1, 7/16 (43.8%) at T2, 6/12 (50%) at T3, and 3/6 (50%) at T4 achieved a ≥ 6 -point reduction. For the neck region, 5/16 patients (31.3%) at T1, 9/16 (56.3%) at T2, 7/12 (58.3%) at T3, and 3/6 (50%) at T4 achieved a ≥ 2 -point reduction. Given the small sample size, statistical analysis was not performed for the change in individual body regions. Additionally, among the 9 of 19 patients who continued follow-up in our clinic and were clinically interviewed, 6 reported subjective improvements. These included reduced involuntary movements; improved eyelid control and head movements; improved mastication; decreased pain and tightness in the jaw and neck muscles; and enhanced performance of daily activities (e.g., eating, personal care, and driving), all of which were meaningful to them and contributed to improved quality-of-life.

Longer duration of DBS therapy was associated with a significant reduction in UDRS score, indicating improvement over time ($\beta = -0.10$ points per month, $p = 0.00018$ [FDR corrected], Supplementary Figure S1). This association remained robust after adjustment for sex, disease duration, age at onset, and body areas involvement, and in sensitivity analyses excluding disease duration and number of body areas affected. Our model showed that number of body areas affected predicts a higher UDRS score ($\beta =$

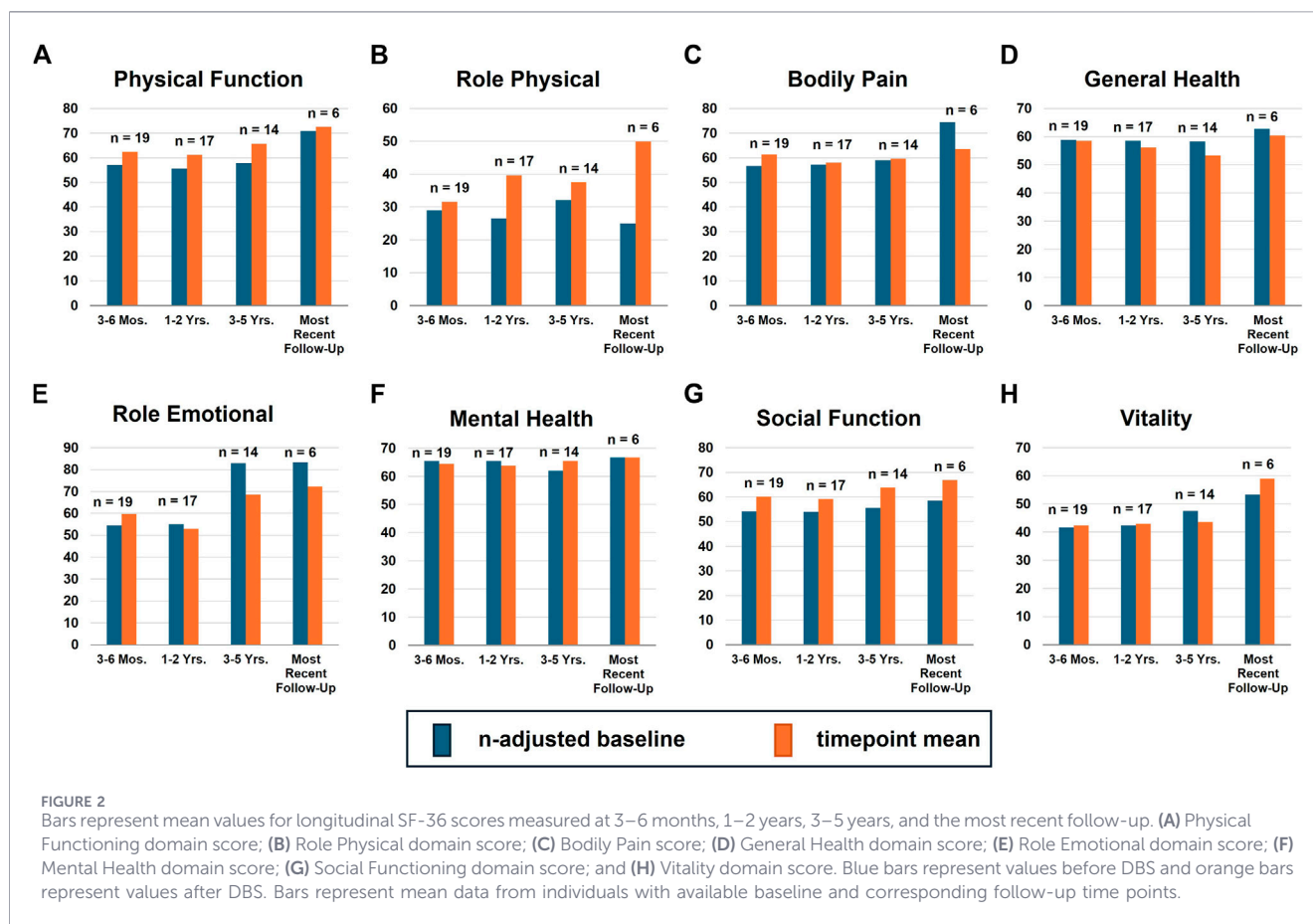
4.86, $p = 0.0043$) as did longer duration of disease prior to DBS ($\beta = 0.43634$, $p = 0.039$), but the effects of other co-variables on modeled UDRS score were not significant. Because some patients may initially have benefits with programming that plateau over time, and we were interested in understanding the longer-term effects of pallidal DBS in this context, we performed quadratic modelling to undervalue this initial programming effect and clarify longer term benefit. While this disclosed a mildly better fit based on likelihood ratio testing with ANOVA ($\chi^2 = 4.78$, $p = 0.028$), the linear term remained significant, and the primary conclusions were unchanged.

HRQoL outcomes measured with SF-36 tool

With regards to the HRQoL, median Physical Component Summary (PCS) scores were 52.5 (IQR: 36.9–62.3) at baseline, 52.3 (IQR: 47.6–60.8) at T1, 53.3 (IQR: 34.8–69.3) at T2, 51.3 (IQR: 44.8–66.3) at T3 and 63.5 (IQR: 44.3–68.1) at T4. Median Mental Component Summary (MCS) scores were 55.8 (IQR: 33.6–74.3) at baseline, 60.5 (IQR: 38.9–72) at T1, 50.5 (IQR: 37.5–73.3) at T2, 61.0 (IQR: 48.6–73.6) at T3 and 66.6 (IQR: 60.8–80.3) at T4. Individual domain scores are shown in Figure 2.

Following FDR-adjusted analysis, the physical and mental component summary scores did not differ significantly across time points. Likewise, statistical comparisons across the eight individual domains of quality-of-life did not yield any statistically significant differences. However, social functioning demonstrated a directional trend toward improvement based on median values.

In independent mixed effect linear regression models with domains of the MOS-36 as the dependent variable, a longer duration of DBS therapy was associated with improvement in



multiple HRQoL domains in unadjusted analyses. The Role Physical domain ($\beta = 2.63$ points per month, $p = 0.011$), Vitality domain ($\beta = 2.021$ points per month, $p = 0.049$), and Social functioning domain ($\beta = 3.01$, $p = 0.0042$) improved over time, while Physical Functioning, Bodily Pain, General Health, Role Emotional and Mental Health domains did not vary significantly with time. Notably, baseline UDRS and number of body regions explained variance in the physical functioning and role emotional domains in unadjusted analyses. After correction for multiple comparisons, social functioning and role physical functioning domain scores ($p = 0.034$ and 0.044 respectively) remained significantly correlated, though other domains did not.

Non-motor outcomes measured with BDI (depression), BAI (anxiety) and AS (apathy)

The median BDI scores were 8.5 (IQR 4–11.3) at baseline, 9 (IQR 5.8–11.5) at T1, 8.5 (IQR 4–11.5) at T2, 10.5 (IQR 6.0–14.3) at T3, and 7.0 (IQR: 5.0–7.5) at T4. Median BAI scores were 6 (IQR 1.0–8.5) at baseline and 6 (IQR 4.0–10.0) at T1, 7.0 (IQR 2.0–12.0) at T2, 8.0 (IQR 2.5–12.0) at T3, and 2 (IQR 1.0–3.0) at T4, respectively. Median (apathy) AS scores were 9.0 (IQR: 8.0–10.8) at baseline and 13.0 (IQR 10.0–18.0), 12 (IQR 7.5–16.0), 11 (IQR 6.3–16.3), and 13.0 (IQR 8.0–14.0) at T1 to T4, respectively.

In Wilcoxon signed-rank paired analyses comparing each follow-up time point to baseline, none of these comparisons reached the threshold for statistical significance after FDR-adjustment.

Discussion

Pallidal DBS was associated with a 50%–70% improvement in craniocervical dystonia, comparable to prior studies in cervical dystonia and generalized dystonia [15]. Regional analysis suggested that the greatest number of patients had a meaningful response in the eyes, followed by the lower face and neck. Longer DBS duration was associated with continued motor improvement, with UDRS scores demonstrating sustained or further gains even 5 years after surgery, indicating a gradual and protracted trajectory toward maximal symptom control. These findings are clinically meaningful given the chronic and often disabling nature of symptoms in blepharospasm and Meige syndrome. Although improvements in HRQoL did not reach statistical significance, social functioning analysis showed a trend toward improvement, and model-based analysis suggested progressive gains over time. Together, these motor and HRQoL findings are important for preoperative counseling, as they help set realistic expectations regarding the potentially delayed time to maximal benefit and the durability of DBS effects. Our modest sample size limited statistical power to detect changes in depression, anxiety, or apathy.

Most studies to date that reported encouraging long term efficacy data with pallidal DBS in craniocervical dystonia are among Asian cohorts [16–21]. Across these cohorts, GPi DBS was noted to typically lead to 50%–70% improvement in dystonia severity, though principally measured using the Burke Fahn Marsden Dystonia Rating Scale (BFMDRS), in contrast to our

use of the UDRS. For example, Horisawa and colleagues reported approximately 60% improvement in BFMDRS motor scores following GPi DBS in patients with Meige syndrome [22]. Similarly, Zheng et al. demonstrated ~65% improvement at long-term follow-up in a cohort treated with pallidal stimulation [23]. Some DBS studies targeted the subthalamic nucleus (STN) for the treatment of craniocervical dystonia [20]. Comparative studies evaluating the efficacy of STN versus GPi DBS generally found no significant differences in clinical outcomes for craniocervical dystonia. This was illustrated in two recent studies, from Wu, G and colleagues retrospectively and Wu, Y and colleagues prospectively with both cohorts located in China [21, 24]. In the randomized, prospective trial involving 62 patients, GPi DBS resulted in 54.9%, 57.3%, and 59.7% improvements in BFMDRS movement scores at 3, 6, and 12 months, respectively, with outcomes comparable to those observed with STN DBS [24]. It is notable that the score for neck involvement in these comparative studies was a median of 0 in the prospective and mean of 0.13 in the retrospective cohort respectively [21, 24], starkly different from our study and highlighting phenotypic variability between geographic regions and centers.

Importantly, emerging data suggests that outcomes are influenced by identifiable prognostic factors. Shorter disease duration has been associated with superior long-term motor improvement, supporting earlier surgical referral [25]. Our regression analysis supports this, with disease duration emerging as an independent factor correlated to higher UDRS scores. In addition, greater engagement of the GPi sensorimotor subregion, reflected by larger volumes of activated tissue within this territory, has been identified as an independent predictor of better motor outcomes [24]. However, a greater impairment in speech and swallowing abilities noted preoperatively is accompanied by poorer DBS outcomes [24].

Indeed, GPi DBS with durable benefits as demonstrated in our case series is traditionally the preferred brain target for treating generalized and segmental dystonia. The GPi has a relatively large nuclear volume and allows for broader tolerable stimulation settings. The posterolateral–ventral portion of the GPi is widely considered an effective site for alleviating dystonia symptoms. Consistent with the known somatotopic organization of the GPi, the more ventral, lateral, and posterior regions are associated with orofacial and cervical representations, whereas the more dorsal, medial, and anterior regions correspond to lower limb representations [26]. While the mechanisms by which pallidal DBS treat craniocervical dystonia have not been characterized yet, our findings of continued improvement over a protracted time course may support underlying neuroplastic effects [27, 28]. Because the other principal movement disorder treated with GPi DBS is Parkinson's disease, which is neurodegenerative and progressive, it is difficult to say how this potentially building benefit over time reflects on broader mechanisms of the therapeutic effects of DBS in general.

In a recent larger long-term series [29], Hao et al. reported improvements in motor function accompanied by enhancement in HRQoL measures at 1-year follow-up, without significant worsening of non-motor or psychiatric features. Similarly, Zheng et al. demonstrated significant improvement in SF-36 scores at final follow-up (mean follow-up 62.9 ± 24.8 months), indicating sustained gains in overall HRQoL albeit following STN DBS [23]. In our cohort, overall HRQoL improvements did not reach statistical

significance, likely reflecting the modest sample size and the multidomain structure of the SF-36, which captures broad aspects of HRQoL. Improvements in segmental dystonia may translate into only modest changes in overall SF-36 scores, particularly as patients age and accumulate additional comorbidities. Nevertheless, we observed trends toward improvement over time in several domains, particularly those related to physical health and social functioning. The consistent reduction in motor severity, especially in functionally disabling features such as blepharospasm, jaw dystonia, and cervical involvement, likely translates into meaningful benefits in social interaction, visual function, speech, and daily activities.

Another important observation from our study relates to the use of adjunctive therapies following DBS. While botulinum toxin dosing remained largely unchanged in this cohort, the proportion of patients requiring oral medications decreased by approximately half after DBS alongside improvements in clinical scores. This reduction in pharmacologic burden may have meaningful implications for treatment burden and quality-of-life, particularly given the cumulative effects of long-term polypharmacy in this population. Regarding programming parameters for GPi DBS, the pulse widths used in our study were higher ($156.6 \pm 70.6 \mu\text{s}$) compared with those reported in a recent large study [24] ($84.6 \pm 12.5 \mu\text{s}$), whereas stimulation frequency was similar ($139.2 \pm 33.4 \text{ Hz}$ vs. $139.2 \pm 18.9 \text{ Hz}$).

Our study has several limitations. The retrospective design introduces potential selection and detection bias, and the sample size was modest. Programming strategies and adjunctive therapy adjustments were not fully standardized, reflecting real-world practice but potentially contributing to variability in outcomes. In addition, while follow-up was long term, attrition over time may influence observed trajectories. Importantly, it is possible that patients with longer available follow up durations are also more likely to be those who improve with continued care and thus continue to interact with the clinic for further data collection. The data presented is derived from a single center with significant DBS expertise and individual factors related to lead placement and programming which may affect generalizability of our results. The UDRS is inherently biased toward the assessment of generalized dystonia and may be less sensitive to segmental dystonia. Further, there was not standardization of botulinum toxin therapy timing and collection of objective scores in this retrospective cohort. Patients were optimized on non-surgical therapies prior to proceeding to brain surgery implying a mild effect of this intervention on our analyses, but this does limit interpretation of UDRS scores. It is notable that the scores analyzed here were collected at DBS programming visits which are scheduled for the same day as botulinum toxin injections whenever possible. As a result, scores were most likely to be collected at the nadir of toxin efficacy as a matter of routine delivery of care at our center. HRQoL was assessed using the SF-36, which is not disease-specific for dystonia. As a result, some patients reported subjective improvements impacting their quality-of-life that were not captured in the HRQoL scores. Future large, prospective multicenter studies with standardized outcomes and postoperative care are needed to refine patient selection and optimize stimulation strategies. Quality-of-life metrics specific to cranial, cervical, or craniocervical dystonia were also not employed.

In conclusion, DBS appears to provide sustained long-term benefits for patients with craniocervical dystonia, with durable symptom control, a potential improvement in HRQoL, generally stable effects on psychiatric features of dystonia, and a possible reduction in adjunctive treatment burden. These findings receiving care at a DBS center in US mirror those found in Asian studies, and there is a signal for improvement even on a protracted time scale. Further studies examining optimal stimulation targets using connectomic approaches, as well as investigations of neurophysiologic signals such as local field potentials, may offer additional insights into the mechanisms of DBS and help refine strategies to optimize and sustain therapeutic outcomes in craniocervical dystonia.

Data availability statement

The data analyzed in this study is subject to the following licenses/restrictions: The datasets analyzed during the current study are not publicly available because of institutional regulations and patient privacy considerations but are available from the corresponding author on request and with appropriate approvals. Requests to access these datasets should be directed to AS, aparna.shukla@neurology.ufl.edu.

Ethics statement

The studies involving humans were approved by the University of Florida Institutional Review Board. The studies were conducted in accordance with the local legislation and institutional requirements. Written informed consent for participation was not required from the participants or the participants' legal guardians/next of kin in accordance with the national legislation and institutional requirements.

Author contributions

MR: Conception, Organization, Execution, Writing of the first draft, Review and critique, Writing of the final manuscript. PZ: Organization, Review and critique. SS: Organization, Review and critique. RK: Organization, Review and critique. IG: Organization,

Review and critique. HK Organization, Review and critique, Execution. AS: Conception, Organization, Execution, Writing of the first draft, Review and critique, Writing of the final manuscript. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The author(s) declared that this work was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Generative AI statement

The author(s) declared that generative AI was not used in the creation of this manuscript.

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Supplementary material

The Supplementary Material for this article can be found online at: <https://www.frontierspartnerships.org/articles/10.3389/dyst.2026.16756/full#supplementary-material>

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