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Evaluation of outcome of different neurosurgical modalities in management of cervical dystonia

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Abstract

Background: Cervical dystonia is the most common form of focal dystonia and is managed by multiple modalities including repeated botulinum toxin injections, in addition to medical treatment with anticholinergics, muscle relaxants, and physiotherapy. However, surgical interventions could be beneficial in otherwise refractory patients. This study aims to report our experience in the neurosurgical management of cervical dystonia and evaluate patient outcomes using reliable outcome scores for the assessment of patients with cervical dystonia and possible complications. This case series study was conducted on 19 patients with cervical dystonia of different etiologies who underwent surgical management [ten patients underwent selective peripheral denervation, five patients underwent pallidotomy, and four patients underwent bilateral globus pallidus internus (GPi) deep brain stimulation (DBS)] in the period between July 2018 and June 2021 at Ain Shams University Hospitals, Cairo, Egypt. With the assessment of surgical outcomes using the Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) and the Tsui scale 6 months postoperatively.

Results: Surgical management of patients with cervical dystonia of either primary or secondary etiology was associated with significant improvement in head and neck postures after 6 months without major complications associated with the different surgical procedures. The mean improvement in total TWSTRS and Tsui scores were 51.2% and 64.8%, respectively, compared with preoperative scores, while the mean improvement in the TWSTRS subscales (severity, disability, and pain) were 40.2%, 66.9%, and 58.3%, respectively.

Conclusion: Cervical dystonia patients in whom non-surgical options have failed to alleviate their symptoms can be managed surgically leading to significant improvements with minimal adverse effects. However, surgical treatment should be tailored according to several factors including but not limited to the etiology, pattern of dystonic activity, and comorbidities. Therefore, management should be tailored to achieve long-term improvement with minimal risk of complications.

Keywords: Cervical dystonia (CD), Spasmodic torticollis, Selective peripheral denervation, Pallidotomy, Globus pallidus internus (GPi) deep brain stimulation (DBS)

Background

Cervical dystonia (CD) is the most common adult-onset focal dystonia [1–3], characterized by involuntary, intermittent, or sustained muscle contractions causing abnormal neck postures [4, 5]. Epidemiological studies have shown the incidence of CD is from

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0.8 to 1.2 per 100,000 person-years and was 3.5 times greater for women than for men [6–8], and the peak age at onset in the fourth to fifth decade with a mean age of 42 years [7, 9]. Dystonic patterns may be 'simple', with movements limited to one plane, or 'complex', involving more than one plane [10]. Depending on the muscles involved, patients may exhibit torticollis (rotation), laterocollis (tilting), anterocollis (flexion), retrocollis (extension), or a combination of any [11, 12]. Adding to this clinical complexity, CD patients may also present with additional signs and symptoms, such as shoulder elevation, jerk, neck/shoulder pain, and tremors [12–14]. Sensory tricks ('geste antagoniste') often temporarily ameliorate dystonic movements and postures [15]. Commonly used sensory tricks by patients with CD include touching the chin, forehead, or occiput [16].

The definitive mechanism underlying the pathogenesis of CD is unclear and a unifying theory is lacking; however, several factors including genetic and environmental factors such as trauma have been implicated [17–20].

CD is part of either generalized or focal dystonic syndromes which may have a genetic basis, with an identifiable genetic association. Secondary CD may result from central or peripheral trauma, neurodegenerative disease, exposure to dopamine receptor antagonists (tardive dystonia), and any other conditions associated with abnormal functioning of the basal ganglia [21–23]. In the majority of patients, the etiology is not identifiable, and the disorder is often classified as primary cervical dystonia [24].

Therapy for CD is mainly symptomatic [25]. It includes supportive therapy and counseling, physical therapy, pharmacotherapy, chemo-denervation (botulinum toxin, phenol, alcohol), and central and peripheral surgical therapy. The most widely used and accepted therapy for CD is local intramuscular injections with Botulinum Toxin type A and physical therapy can be used as an adjuvant therapy [3]. Pharmacotherapy, including anticholinergics, dopaminergic drugs, and muscle relaxants can be used alone or in combination with other therapeutic interventions [26]. Surgery is usually indicated for patients with CD in whom there is a failure of maximal medical management either due to poor response or occurrence of intolerable side effects, a significant disability that significantly impacts the quality of life, patient should have no cognitive or psychiatric impairment and should be able to fully cooperate with the procedure and long-term follow-up and has no medical contraindications to surgery [5, 8, 24, 27].

Methods

This case series study has been reported in line with the Preferred Reporting Of Case Series in Surgery (PROCESS) 2020 Guidelines [28]. This study has been conducted at the Neurosurgery Department, Ain Shams University Hospitals from July 2018 to June 2021 in Cairo, Egypt.

All patients with cervical dystonia enrolled in this study were regularly seen in our movement disorders clinic before surgery. Our study protocol was reviewed and approved by the ethical board of the neurosurgery department. Informed consent was taken from all the involved patients enrolled in our study. Nineteen patients who had cervical dystonia of different etiologies (primary or secondary), and different patterns (focal or as a part of generalized dystonia) were enrolled in the study as they did not experience adequate relief of their symptoms with non-surgical lines of management.

Patients included in our study were those with primary focal cervical dystonia, or with primary generalized dystonia with predominant cervical dystonia, or secondary dystonia with predominant cervical dystonia who showed poor response/intolerable side effects or complications from oral medical treatment and repeated botulinum toxin injection. While those who were medically unfit for surgery, with severe cognitive or psychiatric impairment, or with severe spinal deformities were excluded.

Bilateral GPi DBS was the first option for all patients, however, this was only afforded by 4 patients in our study population. The main limitation for offering DBS for the remaining population was the lack of financial support for the implant and the procedure, and/or replacement of implantable pulse generators (IPG).

Fifteen patients underwent destructive procedures; unilateral GPi pallidotomy was performed for those patients who suffered from cervical dystonia associated with asymmetrical truncal or hemidystonia, while those who were suffering from only cervical dystonia, were operated for selective peripheral denervation of dystonic neck muscles.

A paired sample two-sided *t*-test with alpha level at 0.05 was used to conduct the statistical analysis of this study along with descriptive statistics using Microsoft Excel. The paired *t*-test was used since there is only one group and we are evaluating and comparing the difference between the means of the scores preoperatively versus 6 months postoperatively. During our analysis, we were calculating the *p*-value, the mean difference between preoperative and postoperative treatment, and the 95% confidence interval. The null hypothesis states that there is no significant difference between the scores preoperatively and 6 months postoperatively.

Interventions used in our study population: *(A) Selective peripheral denervation:* the principle of selective peripheral denervation is to denervate dystonic muscles and preserve other muscles that do not participate in dystonia. In cases where selective denervation of posterior neck muscles was planned, the procedure was done as first described by Claude M. Bertrand [29]. In cases where selective denervation of the sternocleidomastoid muscle (SCM) was planned, the procedure was done with the aid of intraoperative electrical stimulation to identify the spinal accessory nerve branch supplying the SCM or the trapezius to be sectioned according to the preoperative plan for each patient. *(B) GPi DBS:* Patients are operated upon under general anesthesia without a muscle relaxant. Leksell stereotactic frame G (Elekta Instruments AB, Stockholm, Sweden) was used, and planning software (Framelink; Medtronic Inc., Minneapolis, MN, USA) for target and trajectory planning of the posteroventral part of the GPi. Microelectrode recording (MER) was performed in all cases using 2 microelectrodes per side and a multidrive system (LeadPoint; Medtronic, Inc., Minneapolis, MN, USA), followed by the insertion of a permanent quadripolar electrode (model 3389; Medtronic Inc., Minneapolis, MN, USA) under intraoperative fluoroscopy. Electrodes were implanted bilaterally in all patients and then connected to IPG (Activa PC; Medtronic Inc., Minneapolis, MN, USA) implanted in the left sub-clavicular region. Postoperative CT scans were performed routinely in all patients to ensure the adequate site of lead position and to detect any intracranial complications which may occur intraoperatively with the most important being intracerebral hematoma. Programming was performed 2–4 weeks postoperatively. *(C) Pallidotomy:* Stereotactic frame placement, CT scan image acquisition, preoperative targeting, and trajectory are done in the same steps as DBS. A monopolar radiofrequency probe and lesion generator (Neuro N 50 lesion generator, Stockert GmbH, Freiburg, Germany) were used for stimulation and lesioning. After confirming that macrostimulation did not affect the internal capsule a test lesion is done at 45 °C for 10 s followed by a permanent lesion done at 60 to 70 °C for 60 s after patient re-evaluation for the absence of new deficit. Another 2 lesions are performed using similar parameters after the electrode was withdrawn for 2 mm and 4 mm above the target, respectively, resulting in 3 contiguous lesions.

The effect of the surgery was evaluated by the reduction in the TWSTRS score [30] and Tsui score [31] 6 months postoperatively relative to preoperative scores. Criteria for evaluating the clinical results according to TWSTRS score and Tsui score are illustrated in Table 1 [32] and Table 2 [33], respectively.

Table 1 Criteria for evaluating the clinical results according to TWSTRS score changes [32]

Outcome rating	% reduction in the TWSTRS score
Excellent	75–100%
Moderate	25–75%
No relief	< 25%

Table 2 Criteria for evaluating the clinical results according to Tsui score changes [33]

Outcome rating	Score
Excellent	Decrease of ≥ 10 points, or final score of 0
Good	Decrease by 5–9 points
Fair	Decrease by 3–4 points
No change	Change of ± 2 points
Worse	Increase by ≥ 3

Results

Nineteen patients (10 males and 9 females) were included in this study. There characteristics are shown in Table 3. The mean (SD) age at onset, age at surgery, and duration of symptoms being 14.1 (14.2), 21.2 (11.6), and 6.6 (5.6) years, respectively. Only four patients were diagnosed with primary cervical dystonia, while the remaining 15 patients were of secondary etiology, either due to perinatal insult, trauma, or antidepressant drug intake. Three patients were found to have associated swallowing difficulty, while seven patients have a variable degree of head tremors, and abnormal neck movements were found to be partially or completely relieved by sensory tricks in about half of our study population. Eight patients have received prior botulinum toxin therapy and all the patients had a history of physical therapy and a trial of medical treatment, but with a limited or no benefit and their symptoms recurred soon after. The incidence of different patterns of cervical dystonia observed among the study population is illustrated in Table 4 and Fig. 1. The type of surgical intervention performed in our study population is illustrated in Table 5.

The primary outcome measure includes TWSTRS and Tsui scores improved from a mean (SD) of 49.1 (11.4) and 11.7 (3.8) before surgery to 24.4 (13.8) and 4.2 (3.5), respectively, at 6 months postoperatively ($p < 0.0001$). The TWSTRS severity, disability, and pain scores improved from 22.3 (3.0), 15.6 (7.2), and 11.0 (3.5) before surgery, to 13.6 (4.3), 6.3 (7.5), and 4.6 (3.3), respectively, at 6 months postoperatively (Fig. 2).

Table 3 Demographic and clinical characteristics of cervical dystonia patients

***n* of patients**	**19**
Gender	
Male	10 (19)
Female	9 (19)
Follow-up period, months	6
Mean age at onset ± SD, years	14.1 ± 14.2
Mean disease duration ± SD, years	6.6 ± 5.6
Mean age at surgery ± SD, years	21.2 ± 11.6
Diagnosis	
1ry dystonia	4 (19)
2ry dystonia	15 (19)
Etiology of 2ry dystonia	
Perinatal insult	12 (19)
Trauma	2 (19)
Antidepressant drug intake	1 (19)
Concomitant symptoms	
Geste antagoniste	9 (19)
Tremors	7 (19)
Swallowing difficulty	3 (19)
Trial of medical treatment	
Yes	19
No	0
Trial of botulinum neurotoxin injection	
Yes	8
No	11

SD standard deviation

Table 4 Incidence of different patterns of head movement in cervical dystonia observed among the study population

Pattern of cervical dystonia	***n* (%)**
Torticollis	12 (63)
Laterocollis	14 (74)
Anterocollis	4 (21)
Retrocollis	2 (10.5)
Sagittal shift	1 (5)
Lateral shift	2 (10.5)

Table 6 shows the mean (SD) percentage reduction in the postoperative score relative to the preoperative score in TWSTRS severity, TWSTRS disability, TWSTRS pain, TWSTRS total score, and Tsui score among our all-study population which was 40.2% (16.4), 66.9% (31.5), 58.3% (22.7), 51.2% (19.8), and 64.8% (22.3), respectively. The highest percentage of reduction was noted in the disability subscale, followed by the pain subscale, then the severity subscale. The percentage of reduction was greater in the Tsui score relative to the TWSTRS score. The P-value across all scores preoperatively and postoperatively was < 0.0001, indicating that the difference in scores due to the surgery was statistically significant (Table 7).

Comparing the percentage of reduction in the postoperative TWSTRS score relative to the preoperative score, only one patient was rated as an excellent outcome with more than 75% reduction in the TWSTRS score and this patient has undergone bilateral GPi DBS, 16 patients were rated as a moderate outcome with 25–75% reduction in the TWSTRS score, and two patients were rated as no relief with less than 25% reduction in the TWSTRS score were both patients had secondary cervical dystonia and one was operated for DBS while the other operated for pallidotomy.

Clinical results evaluated according to the postoperative decrease in the Tsui score were rated as an excellent outcome in six patients with a decrease of ≥ 10 points, good outcome in nine patients with a decrease by 5–9 points, fair outcome in three patients with a decrease by 3–4 points, no change in only one patient with a change of ± 2 points, and no one was rated as a worse outcome in our population.

Comparing the outcome of different surgical interventions used in our study, the mean (SD) percentage of postoperative reduction in the TWSTRS severity, TWSTRS disability, and TWSTRS pain score was highest in those who underwent selective peripheral denervation at 44.1% (9.2), 76.8% (24.1), and 62.6% (12.8), respectively. Among the different surgical interventions, the mean (SD) percentage reduction in the postoperative total TWSTRS and Tsui score relative to the preoperative score was highest among those who underwent selective peripheral denervation with a mean value of 55.4% (6.6) and 71.5% (5.7), respectively (Fig. 3).

In our study population, no mortality was reported during the first 6 months. Complications occurred in about 30% of our study population, either intraoperatively or postoperatively. This occurred in three patients who underwent selective peripheral denervation, one patient underwent pallidotomy, and two patients underwent GPi DBS. In patients who underwent selective peripheral denervation, one patient developed periauricular neuralgia and weakness of the neck muscles, the second patient developed wound collection/seroma, and the third patient developed hypoesthesia over C2 on the right side, all of which were treated conservatively and improved gradually without any residual effect. In a patient operated for unilateral pallidotomy, the patient postoperatively developed speech and swallowing abnormalities, which improved over time but with a residual deficit. While in patients operated for GPi DBS, the first

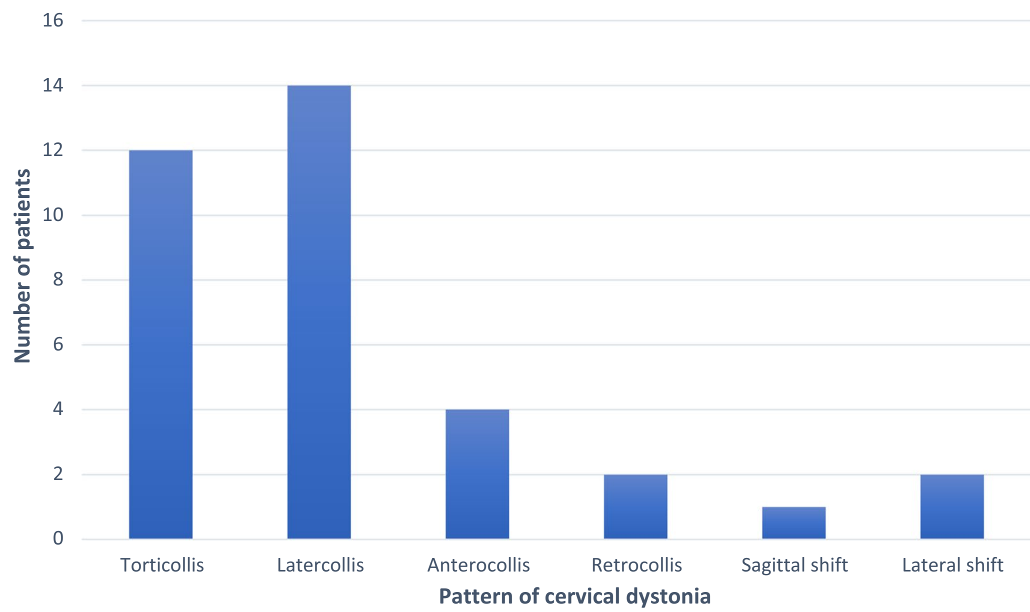

Fig. 1 Incidence of different patterns of head movement in cervical dystonia observed among the study population

Table 5 Types of surgical intervention used in the management of the patients

Surgical intervention	*n* (%)
Selective peripheral denervation	10 (52.6)
Selective denervation of SCM/trapezius/both	7 (36.8)
Selective denervation of SCM/trapezius/both + myotomy	2 (10.5)
Selective peripheral denervation C1–C4	1 (5.3)
Pallidotomy	5 (26.3)
GPi DBS	4 (21.1)

SCM sternocleidomastoid, *GPi* globus pallidus internus

patient intraoperatively developed focal seizures with secondary generalization, which was aborted by midazolam, followed by regular intake of levetiracetam. Postoperative CT revealed only pneumocephalus which was treated conservatively, while the second patient developed low-grade infection over the IPG, which was manifested by redness and tenderness, and was managed with wound debridement and postoperative intravenous antibiotics with complete resolution of infection.

Discussion

Surgical management of patients with cervical dystonia of either primary or secondary etiology was associated with significant improvement in head and neck postures after 6 months.

Our results regarding the mean percentage of improvement in the TWSTRS total, severity, disability, and pain score obtained among those patients operated for DBS were nearly similar to those obtained from other studies [34–36] and this was consistent with a recently published meta-analysis by Tsuboi and colleagues [37]. However, Eltahawy and colleagues [38] reported a higher percentage of improvement in the total TWSTRS score which is attributed to the very high percentage of improvement in the pain scale by 91% which in turn was reflected by functional improvement and greater reduction in disability score. Volkmann, J. and colleagues [39] reported lower results, but this may be attributed to the large study population in the study (57 patients) and the type of study design.

Regarding the percentage of improvement in the total TWSTRS score among those who underwent selective peripheral denervation, there was a wide range of results, ranging from about 30% as reported by Münchau and colleagues [40] to about 73% as reported by Wang and colleagues and Jang and colleagues [41, 42]. To our knowledge, this great variability in results is probably attributed to a wide variety in surgical techniques used even within the same study, as the selective denervation is related to the involved muscle group/groups, which is fashioned independently in each pattern of cervical dystonia. It is also probably related to the incidence of occurrence of incomplete denervation or re-innervation or change in the muscular activation pattern due to progression of dystonic symptoms involving other muscle groups that became evident postoperatively.

Regarding the percentage of improvement in the total TWSTRS score among those who underwent

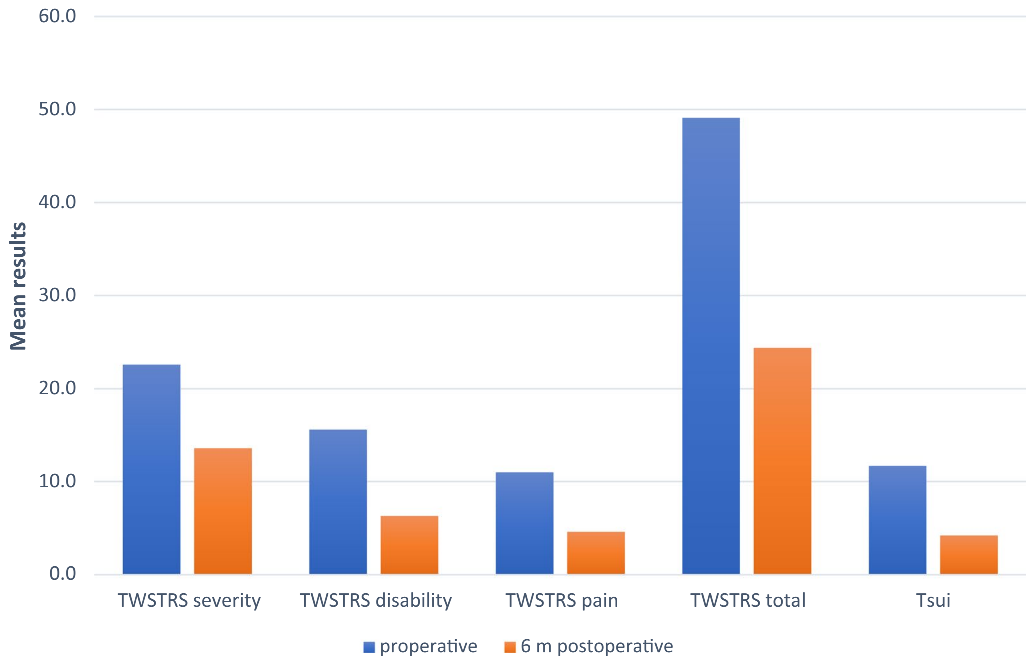

Fig. 2 Comparison between the mean value of the preoperative and 6-month postoperative results according to TWSTRS severity, disability, pain, and total score, and Tsui score

Table 6 Mean ± SD % of improvement in TWSTRS severity, disability, pain, and total score, and Tsui score among our all-study population, patients who underwent selective peripheral denervation, pallidotomy, and DBS

	All patients (mean ± SD)	Selective peripheral denervation (mean ± SD)	Pallidotomy (mean ± SD)	DBS (mean ± SD)
TWSTRS severity	40.2 ± 16.4	44.1 ± 9.2	31.8 ± 17.6	40.7 ± 27.8
TWSTRS disability	66.9 ± 31.5	76.8 ± 24.1	52.7 ± 35.2	60.1 ± 42.6
TWSTRS pain	58.3 ± 22.7	62.6 ± 12.8	55.3 ± 30.6	51.1 ± 34.5
TWSTRS total	51.2 ± 19.8	55.4 ± 6.6	42.9 ± 25.8	51.1 ± 34.1
Tsui	64.8 ± 22.3	71.5 ± 5.7	55.4 ± 25.4	60.0 ± 41.1

SD standard deviation

Table 7 Comparison of the preoperative and 6 months postoperative results according to TWSTRS severity, disability, pain, and total score, and Tsui score

	Preoperative score		Postoperative score		*p*-value
	Mean ± SD	95% CI	Mean ± SD	95% CI	
TWSTRS severity score	22.3 ± 3.0	21.2–24.1	13.6 ± 4.3	11.5–15.7	˂ 0.0001
TWSTRS disability score	15.6 ± 7.2	12.1–19.1	6.3 ± 7.5	2.7–9.9	˂ 0.0001
TWSTRS pain score	11.0 ± 3.5	9.3–12.7	4.6 ± 3.3	3.0–6.2	˂ 0.0001
TWSTRS total score	49.1 ± 11.4	43.6–54.6	24.4 ± 13.8	17.8–31.1	˂ 0.0001
Tsui score	11.7 ± 3.8	9.9–13.6	4.2 ± 3.5	2.5–5.9	˂ 0.0001

CI confidence interval, *SD* standard deviation

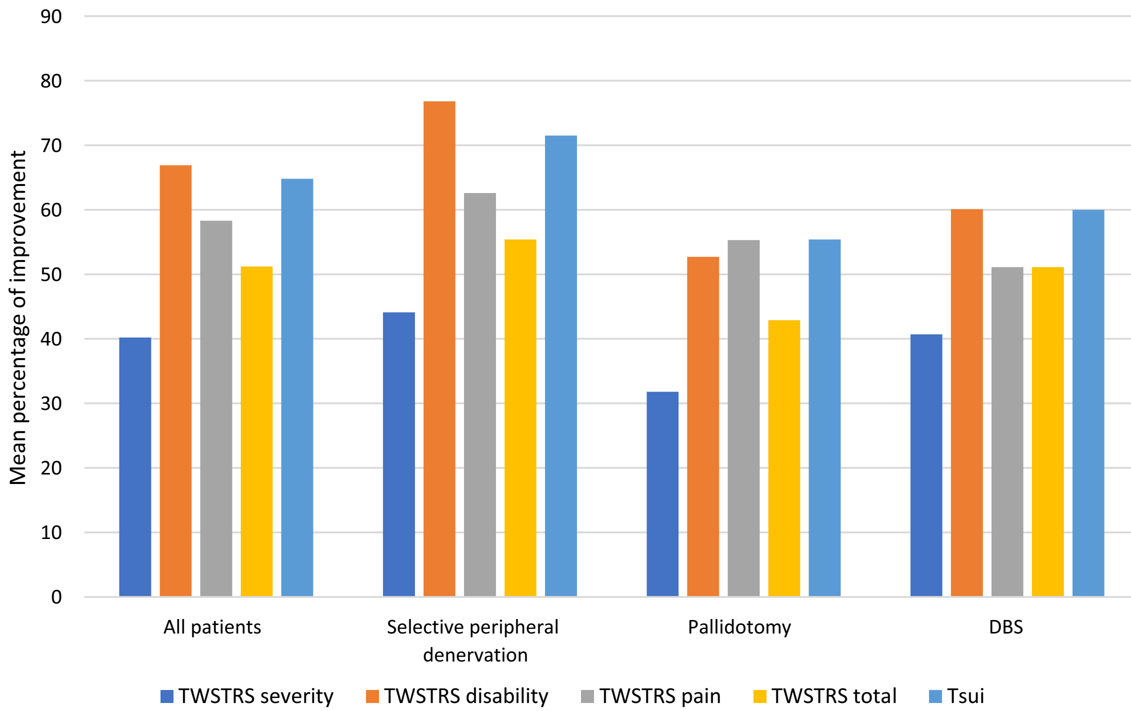

Fig. 3 Mean percentage of improvement in TWSTRS severity, disability, pain, and total score, and Tsui score among our all-study population, patients who underwent selective peripheral denervation, pallidotomy, and DBS

pallidotomy, our results were nearly congruent to those reported by Horisawa and colleagues in a retrospective observational study carried out on 25 patients with asymmetrical botulinum toxin-resistant cervical dystonia [43]. In our series, those operated for pallidotomy were manifesting with asymmetrical dystonic phenotype, either segmental or truncal with predominant unilateral cervical dystonia thus causing symptoms improvement after the unilateral lesioning procedure. Unfortunately, this may not be considered a surgical option in those patients presenting with bilateral symmetrical abnormalities as in anterocollis and retrocollis cases, in which bilateral lesioning is not applicable due to the high incidence of morbidity [27, 43]. Bilateral pallidotomy may result in permanent speech disturbance [44–46], transient lethargy/somnolence/stupor [44, 47–49], and transient hemiparesis [47, 49].

In this study, the only patient who was rated as an excellent outcome with more than 75% reduction in the TWSTRS score has underwent bilateral GPi DBS, and this is consistent with the evidence suggesting that patients with cervical dystonia have bilateral basal ganglia dysfunction regardless of their clinical manifestations, thus supporting the rationale for bilateral surgical intervention [8, 50, 51]. While those two patients were rated as having no relief with less than 25% reduction in the TWSTRS score both patients had secondary cervical dystonia and one was operated for DBS while the other operated for pallidotomy. Eltahawy and colleagues and Capelle and colleagues both reported that the response to pallidal surgery is dependent on the etiology, were improvement among patients with secondary dystonia undergoing functional stereotactic procedures will respond less well compared to the improvement in patients with primary dystonia, and this is attributed to the structural lesions in patients with secondary dystonia [45, 52].

It should be noted that the aim of management is not the cure of the disease, but to ameliorate symptoms, social and functional disability, and to decrease the incidence of complications encountered in the cervical spine such as cervical spondylosis, disc herniation, cervical radiculopathy, cervical myelopathy, vertebral subluxation, fractures, muscle contractures, and fixed bony deformity. Due to the elective nature of the procedures, whatever the treatment strategy decided it should be attempted to keep the potential for adverse events to a minimum. This was consistent with our results as adverse effects occurred in six patients in our study group. Three of them occurred in patients who underwent selective peripheral denervation and are commonly reported in the literature [41, 42, 53]. Interestingly, they were only transient with gradual improvement over time. Only one patient developed weakness in the neck muscle associated with neck drop and was managed conservatively with intense physiotherapy for 2 months, which resulted in gradual improvement and good control of neck movements. In the patient operated for unilateral pallidotomy on the right side, the patient was right-handed with truncal dystonia, with predominant cervical dystonia over the left side and was planned for staged bilateral pallidotomy, however, the patient postoperatively developed speech and swallowing abnormalities, although the patient did not improve significantly postoperatively, contralateral pallidotomy was omitted. While in patients operated for GPi DBS, the first patient developed intraoperative seizures. The second patient developed a low-grade infection over the pulse generator and was operated for sub-clavicular wound debridement with copious irrigation with saline and antibiotics without removal of the device, followed by postoperative intravenous antibiotics and repeated wound dressing resulting in complete resolution of infection.

In our study, only one patient was operated for selective peripheral denervation from C1–4 on one side without involving the SCM, while others were operated for selective denervation of the SCM/trapezius/ or both, with or without muscle sectioning, without denervation of the posterior neck muscle. This resulted in 50% of these patients requiring botulinum toxin injection in the first 6 months postoperatively and this may have caused false high results in our study. This suggests that those patients would benefit from staged surgery which will require another operation to be planned for the remaining dystonia muscles which were not denervated in the first operation.

Bilateral GPi DBS can be recommended as the first line of management for patients with cervical dystonia as cervical dystonia patients have bilateral basal ganglia dysfunction, thus justifying their improvement when undergoing bilateral surgical intervention as in GPi DBS, as bilateral pallidotomy is not recommended owing to the associated high morbidity and complications. However, the main limitation for offering bilateral GPi DBS was lacking financial support for the implant and the procedure and/or replacement of IPG.

The aim of this study was not to compare the postoperative outcome between DBS, pallidotomy, and selective peripheral denervation, however, different surgical options included in our study were associated with significant improvement in patients' symptoms with minimal adverse effect. A systematic review and meta-analysis conducted by Ravindran, K. and colleagues comparing the outcome of DBS against selective peripheral denervation in cervical dystonia patients, suggested that both surgical options are effective in treatment for cervical dystonia, with DBS effective at reducing TWSTRS severity and disability score and a minimal effect on pain reduction [54]. Although each intervention has its pros and cons, and patients may respond better to one intervention over the other, identification of those patients who may benefit from each intervention has not been done.

This study has several limitations. One is the study design in which there is no control group to analyze and compare variable outcomes of different surgical approaches. Another limitation is the relatively small sample size ($n = 19$) and short follow-up period (6 months) which did not highlight the long-term outcome postoperatively. The heterogenicity in our study population together with the variable and individualized surgical options did not allow comparative analysis of the long-term efficacy of the different procedures in our study. Rating scales assessments were not performed in a blinded fashion but were performed at routine 6-month follow-up clinic visits. Our study was a non-randomized non-blinded study, which may suffer from selection, classification, and confounding biases.

Conclusions

Cervical dystonia can be effectively treated surgically without major complications. Patients that are refractory to conservative medical treatment may be potential surgical candidates. In contrast with botulinum toxin which provides only short-term relief of symptoms and require repeated injections, surgical interventions can result in a long-term improvement in abnormal dystonic activity. Moreover, patients with muscle contractures may not improve with botulinum toxin injection, but may benefit from surgery when denervation is combined with myotomy. Surgical modalities in terms of bilateral GPi DBS, unilateral GPi pallidotomy, and selective peripheral nerve denervation(s) are applicable and of accepted outcome in selected individuals. The treatment plan and selection of the appropriate surgical option is a major challenge, and usually depends on several factors, including the age of the patient, clinical type of cervical dystonia, etiology of cervical dystonia, previous exposure to medications, previous trials of botulinum toxin injection, medical condition, prior surgery, cost, and patient's preference, therefore therapy must be individualized. However, the prognostic factors determining favorable outcomes for which patient will respond better to a specific type of surgery have not been determined. Therefore, further randomized, blinded, studies with a longer follow-up period are needed to allow the identification of patient characteristics that favors one surgical option over the other, and the long-term effect of each surgical option.

Abbreviations
CD: Cervical dystonia; CSF: Cerebrospinal fluid; CT: Computed tomography; DBS: Deep brain stimulation; GPe: Globus pallidus externus; GPi: Globus pallidus internus; IPG: Implantable pulse generators; MER: Microelectrode recording; MRI: Magnetic resonance imaging; SCM: Sternocleidomastoid muscle; SD: Standard deviation; TWSTRS: Toronto Western Spasmodic Torticollis Rating Scale.

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Author contributions
MA is the primary author who was responsible for data gathering, analysis, and writing of the manuscript. AN, SM, AG, ZY assisted in providing and facilitating access to cases with required consent. WG and AR helped in the overall study design and manuscript review. All authors have read and approved the manuscript.

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Availability of data and materials
All the aforementioned data and results of the statistical analysis are available with the authors and ready to be shared with approved personnel upon request.

Declarations

Ethical approval and consent to participate
All patients were informed about the benefits and the risks of the intended procedure. A written informed consent was taken from all the participants or their legal guardians. The study was conducted with the approval of the Research Ethics Committee, Faculty of Medicine, Ain-Shams University (FMASU REC). Institutional Review Board No. is FWA 000017585.

Consent for publication
Not applicable.

Competing interests
The authors declare that they have no competing interests.

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References
1. Krauss JK. Deep brain stimulation for treatment of cervical dystonia. Acta Neurochir Suppl. 2007;97(Pt 2):201–5.
2. Stacy M. Idiopathic cervical dystonia: an overview. Neurology. 2000;55(12 Suppl 5):S2–8.
3. Albanese A, Abbruzzese G, Dressler D, Duzynski W, Khatkova S, Marti MJ, et al. Practical guidance for CD management involving treatment of botulinum toxin: a consensus statement. J Neurol. 2015;262(10):2201–13.
4. Samotus O, Lee J, Jog M. Personalized botulinum toxin type A therapy for cervical dystonia based on kinematic guidance. J Neurol. 2018;265(6):1269–78.
5. De Pauw J, Mercelis R, Hallemans A, Van Gils G, Truijen S, Cras P, et al. Postural control and the relation with cervical sensorimotor control in patients with idiopathic adult-onset cervical dystonia. Exp Brain Res. 2018;236(3):803–11.
6. LaHue SC, Albers K, Goldman S, Lo RY, Gu Z, Leimpeter A, et al. Cervical dystonia incidence and diagnostic delay in a multiethnic population. Mov Disord. 2020;35(3):450–6.
7. Norris SA, Jinnah HA, Espay AJ, Klein C, Brüggemann N, Barbano RL, et al. Clinical and demographic characteristics related to onset site and spread of cervical dystonia. Mov Disord. 2016;31(12):1874–82.
8. Oropilla JQ, Kiss ZHT. Central procedures for cervical dystonia. In: Lozano AM, Gildenberg PL, Tasker RR, editors. Textbook of stereotactic and functional neurosurgery. Heidelberg: Springer, Berlin Heidelberg; 2009. p. 1871–84.
9. Defazio G, Jankovic J, Giel JL, Papapetropoulos S. Descriptive epidemiology of cervical dystonia. Tremor Other Hyperkinet Mov (N Y). 2013;3:tre-03-193-4374-2.
10. Misra VP, Trosch RM, Maisonobe P, Om S. Spectrum of practice in the routine management of cervical dystonia with abobotulinumtoxinA: findings from three prospective open-label observational studies. J Clin Mov Disord. 2018;5:4.
11. Espay AJ, Trosch R, Suarez G, Johnson J, Marchese D, Comella C. Minimal clinically important change in the Toronto Western Spasmodic Torticollis Rating Scale. Parkinsonism Relat Disord. 2018;52:94–7.
12. Reichel G. Cervical dystonia: a new phenomenological classification for botulinum toxin therapy. Basal Ganglia. 2011;1(1):5–12.
13. Albanese A, Bhatia K, Bressman SB, DeLong MR, Fahn S, Fung VS, et al. Phenomenology and classification of dystonia: a consensus update. Mov Disord. 2013;28(7):863–73.
14. Jankovic J, Tsui J, Bergeron C. Prevalence of cervical dystonia and spasmodic torticollis in the United States general population. Parkinsonism Relat Disord. 2007;13(7):411–6.
15. Patel N, Hanfelt J, Marsh L, Jankovic J. Alleviating manoeuvres (sensory tricks) in cervical dystonia. J Neurol Neurosurg Psychiatry. 2014;85(8):882–4.
16. Sarasso E, Agosta F, Piramide N, Bianchi F, Butera C, Gatti R, et al. Sensory trick phenomenon in cervical dystonia: a functional MRI study. J Neurol. 2020;267(4):1103–15.
17. Skogseid IM. Dystonia–new advances in classification, genetics, pathophysiology and treatment. Acta Neurol Scand Suppl. 2014;198:13–9.
18. Dauer WT, Burke RE, Greene P, Fahn S. Current concepts on the clinical features, aetiology and management of idiopathic cervical dystonia. Brain. 1998;121(Pt 4):547–60.
19. Abbruzzese G, Pelosin E, Avanzino L. Physiology of dystonia. In: Kanovsky P, Bhatia KP, Rosales RL, editors. Dystonia and dystonic syndromes. Vienna: Springer; 2015. p. 13–25.
20. Cassidy A. Pathophysiology of idiopathic focal dystonia. ACNR. 2010;10:14–8.
21. LeDoux MS, Brady KA. Secondary cervical dystonia associated with structural lesions of the central nervous system. Mov Disord. 2003;18(1):60–9.
22. Molho ES, Feustel PJ, Factor SA. Clinical comparison of tardive and idiopathic cervical dystonia. Mov Disord. 1998;13(3):486–9.
23. O'Riordan S, Hutchinson M. Cervical dystonia following peripheral trauma–a case-control study. J Neurol. 2004;251(2):150–5.
24. Velickovic M, Benabou R, Brin MF. Cervical dystonia pathophysiology and treatment options. Drugs. 2001;61(13):1921–43.
25. van den Dool J, Visser B, Koelman JH, Engelbert RH, Tijssen MA. Cervical dystonia: effectiveness of a standardized physical therapy program; study design and protocol of a single blind randomized controlled trial. BMC Neurol. 2013;13:85.
26. Bledsoe IO, Viser AC, San LM. Treatment of dystonia: medications, neurotoxins, neuromodulation, and rehabilitation. Neurotherapeutics. 2020;17(4):1622–44.
27. Moro E, Gross RE, Krauss JK. What's new in surgical treatment for dystonia? Mov Disord. 2013;28(7):1013–20.
28. Agha RA, Sohrabi C, Mathew G, Franchi T, Kerwan A, O'Neill N, et al. The PROCESS 2020 Guideline: updating consensus Preferred Reporting Of CasESeries in Surgery (PROCESS) guidelines. Int J Surg. 2020;84:231–5.
29. Bertrand CM. Selective peripheral denervation for spasmodic torticollis: surgical technique, results, and observations in 260 cases. Surg Neurol. 1993;40(2):96–103.
30. Consky E, Basinski A, Belle L, Ranawaya R, Lang A. The Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS): assessment of validity and inter-rater reliability. Neurology. 1990;40(suppl 1):445.

31. Tsui JK, Eisen A, Stoessl AJ, Calne S, Calne DB. Double-blind study of botulinum toxin in spasmodic torticollis. Lancet. 1986;2(8501):245–7.
32. Hua C, Pu B, Liu K, Huang Z, Li C, Zhao C, et al. New rhizotomy procedure for primary spasmodic torticollis. J Craniofac Surg. 2018;29(5):1338–40.
33. Taira T, Kobayashi T, Takahashi K, Hori T. A new denervation procedure for idiopathic cervical dystonia. J Neurosurg. 2002;97(2 Suppl):201–6.
34. Kiss ZH, Doig-Beyaert K, Eliasziw M, Tsui J, Haffenden A, Suchowersky O. The Canadian multicentre study of deep brain stimulation for cervical dystonia. Brain. 2007;130(Pt 11):2879–86.
35. Skogseid IM, Ramm-Pettersen J, Volkmann J, Kerty E, Dietrichs E, Røste GK. Good long-term efficacy of pallidal stimulation in cervical dystonia: a prospective, observer-blinded study. Eur J Neurol. 2012;19(4):610–5.
36. Hung S, Hamani C, Lozano A, Poon Y, Piboolnurak P, Miyasaki J, et al. Long-term outcome of bilateral pallidal deep brain stimulation for primary cervical dystonia. Neurology. 2007;68(6):457–9.
37. Tsuboi T, Wong JK, Almeida L, Hess CW, Wagle Shukla A, Foote KD, et al. A pooled meta-analysis of GPi and STN deep brain stimulation outcomes for cervical dystonia. J Neurol. 2020;267(5):1278–90.
38. 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(3):328–32.
39. Volkmann J, Mueller J, Deuschl G, Kühn AA, Krauss JK, Poewe W, et al. Pallidal neurostimulation in patients with medication-refractory cervical dystonia: a randomised, sham-controlled trial. Lancet Neurol. 2014;13(9):875–84.
40. Münchau A, Palmer J, Dressler D, O'Sullivan J, Tsang K, Jahanshahi M, et al. Prospective study of selective peripheral denervation for botulinum-toxin resistant patients with cervical dystonia. Brain. 2001;124(Pt 4):769–83.
41. Jang KS, Park HK, Joo WI, Ji C, Lee KJ, Choi CR. Selective peripheral denervation for the treatment of spasmodic torticollis. J Korean Neurosurg Soc. 2005;37(5):350–3.
42. Wang J, Li J, Han L, Guo S, Wang L, Xiong Z, et al. Selective peripheral denervation for the treatment of spasmodic torticollis: long-term follow-up results from 648 patients. Acta Neurochir (Wien). 2015;157(3):427–33.
43. Horisawa S, Fukui A, Kohara K, Kawamata T, Taira T. Unilateral pallidotomy in the treatment of cervical dystonia: a retrospective observational study. J Neurosurg. 2019;1–7.
44. Yoshor D, Hamilton WJ, Ondo W, Jankovic J, Grossman RG. Comparison of thalamotomy and pallidotomy for the treatment of dystonia. Neurosurgery. 2001;48(4):818–26.
45. 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(3):613–21.
46. Loher TJ, Capelle HH, Kaelin-Lang A, Weber S, Weigel R, Burgunder JM, et al. Deep brain stimulation for dystonia: outcome at long-term follow-up. J Neurol. 2008;255(6):881–4.
47. Lin JJ, Lin SZ, Chang DC. Pallidotomy and generalized dystonia. Mov Disord. 1999;14(6):1057–9.
48. Cubo E, Shannon KM, Penn RD, Kroin JS. Internal globus pallidotomy in dystonia secondary to Huntington's disease. Mov Disord. 2000;15(6):1248–51.
49. Teive HA, Sá DS, Grande CV, Antoniuk A, Werneck LC. Bilateral pallidotomy for generalized dystonia. Arq Neuropsiquiatr. 2001;59(2-B):353–7.
50. Magyar- Lehmann S, Antonini A, Roelcke U, Maguire RP, Missimer J, Meyer M, et al. Cerebral glucose metabolism in patients with spasmodic torticollis. Mov Disord. 1997;12(5):704–8.
51. Naumann M, Pirker W, Reiners K, Lange KW, Becker G, Brücke T. Imaging the pre- and postsynaptic side of striatal dopaminergic synapses in idiopathic cervical dystonia: a SPECT study using [123I] epidepride and [123I] beta-CIT. Mov Disord. 1998;13(2):319–23.
52. Capelle H-H, Krauss JK. Functional stereotactic procedures for treatment of secondary dystonia. In: Lozano AM, Gildenberg PL, Tasker RR, editors. Textbook of stereotactic and functional neurosurgery. Heidelberg: Springer, Berlin Heidelberg; 2009. p. 1835–56.
53. Bergenheim AT, Nordh E, Larsson E, Hariz MI. Selective peripheral denervation for cervical dystonia: long-term follow-up. J Neurol Neurosurg Psychiatry. 2015;86(12):1307–13.
54. Ravindran K, Ganesh Kumar N, Englot DJ, Wilson TJ, Zuckerman SL. Deep brain stimulation versus peripheral denervation for cervical dystonia: a systematic review and meta-analysis. World Neurosurg. 2019;122:e940–6.

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