Surgical Outcome of Generalized Dystonia: A Single Center Study

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Date of Submission:2nd December 2025 Date of Acceptance: 3rd December 2025 Date of Publication: 15th December 2025

Abstract

Introduction: Dystonias are relatively rare disorders characterized by sustained or intermittent muscle contractions which causes abnormal repetitive movement or postures. Generalized dystonia involve the trunk and two other body regions. It represents not a single disorder but a heterogeneous group of conditions that share similar clinical features yet arise from diverse underlying causes. The etiology of dystonia can be primary, either genetic or idiopathic, or secondary to a wide variety of neurological and systemic disorders.

Materials and Methods: All the patients of generalized dystonia who underwent surgical interventions in Annapurna Neurological Institute and Allied Sciences since January 2016 till December 2023 were included in our study. We performed bilateral pallidotomies or unilateral pallidotomy and contralateral pallidothalamic tractotomy for generalized dystonia. We used ZD Fishers frame and the thermal lesioning machine of Cosman Radiofrequency (RF) generator with the lesioning electrode of 0.75 mm internal diameter and 2 mm exposed tip was used.

Results: There were altogether 17 cases of generalized dystonia. Thirteen cases had bilateral pallidotomy and 4 cases had unilateral pallidotomy with contralateral PTT lesioning. The mean age was 45 ±8 years in Bilateral pallidotomy group and 35±10 years in unilateral Pallidotomy and contralateral PTT lesioning in the same setting. The male: female ratio was 3:1. The mean percentage change in BFMDRS was 60 percent (p value<0.05) in postoperative period in bilateral pallidotomy patients and 70%(p value<0.05) in unilateral Pallidotomy and contralateral PTT lesioning in the same setting. Three cases had relapse . One case of tardive dystonia with repeated bilateral pallidotomy had status dystonicus and finally she got better with intrathecal baclofen pump. Three cases had dysarthria and dysphasia. Two cases had secondary parkinsonism and one case had hypophonia. There was no mortality.

Conclusion: Lesioning surgery is rewarding for generalized dystonia. Unilateral pallidotomy with contralateral PTT lesioning is also satisfactory and may have lesser complications. However, recurrent cases may need DBS or further potential targets.

Keywords: Generalised Dystonia, Lesioning, Movement disorder

Introduction

Dystonia is a hyperkinetic movement disorder in which involuntary contractions of muscles occur.[1] It is characterised by sustained muscle contractions and abnormal trunk, neck, face, arms, and legs postures. It is a painful and disfiguring disease that results in significant physical and social disability, and most forms do not respond to currently available medical therapies.

Generalized dystonia involve the trunk and two other body regions. Generalized dystonia represents not a single disorder but a heterogeneous group of conditions that share similar clinical features yet arise from diverse underlying causes. The etiology of dystonia can be primary, either genetic or idiopathic, or secondary to a wide variety of neurological and systemic disorders.[2] Most cases arise without any identifiable preceding cause and are therefore classified as primary dystonia. In a subset of these primary cases, an underlying genetic mutation can be identified, most commonly involving the DYT1 gene. Although more than 20 DYT genes have now been recognized[3], the majority of primary generalized dystonia cases are not associated with any known mutation and remain idiopathic.

When conservative therapies—such as medications and botulinum toxin injections—fail to provide adequate relief, surgical interventions may be considered for refractory cases. Available surgical options for generalized dystonia include selective peripheral denervation, deep brain stimulation (DBS),

Access this article online

Website: https://www.nepjol.info/index.php/NJN

DOI: https://doi.org/10.3126/njn.v22i24.87067

HOW TO CITE
Gurung, P., Shrestha, R., Dahal, R., Dhungel, R., Dhamala, J., & Pant, B. Surgical Outcome of Generalized Dystonia: A single center study. NJNS. 2025;22(4)33-37

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ISSN: 1813-1948 (Print), 1813-1956 (Online)

and radiofrequency (RF) thermocoagulation.[4–7] Several potential stereotactic targets have been suggested for the treatment of dystonia such as GPi and PTT.[8,9]

The use of globus pallidus pars interna (GPi) DBS for medically refractory dystonia has gained widespread acceptance over the past two decades. This approach provides significant improvement in motor function with a relatively low risk of adverse effects. Pivotal studies by Vidailhet et al.[10] and Kupsch et al.[11] have established strong evidence supporting its effectiveness in primary generalized and segmental dystonia. However, lesioning procedure in the treatment of generalized dystonia is limited. We present here the outcomes of a consecutive series of 17 patients of generalized dystonia patients treated with lesioning.

Materials and Methods

Clinical evaluation

This study was approved by the medical ethics committee of Annapurna Neurological Institute and Allied Sciences. The medical ethics committee approved a waiver of consent for the collection of data as part of the routine clinical care and quality control. A retrospective study was conducted which encompasses all patients who underwent surgical intervention for cervical and generalized dystonia at Annapurna Neurological Institute and Allied Sciences between January 2016 and November 2025. A comprehensive review of medical and imaging records was conducted for all participants prior to and following surgical intervention.

Patient data:

Inclusion criteria: All the patients of generalized dystonia who underwent surgical interventions in Annapurna Neurological Institute and Allied Sciences since 2015 were included in our study. Exclusion criteria: All the surgical cases of task specific focal dystonia and cervical dystonia have been excluded. We did bilateral pallidotomy in most of the cases and unilateral pallidotomy with contralateral pallidothalamic tract lesioning for generalized dystonia.

Evaluation:

A systematic analysis of patient demographics, disease duration, and preoperative clinical assessment were executed.

Objective clinical assessment was performed using the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) preoperatively and postoperatively.

Preoperatively, patients underwent MRI to exclude a structural lesion and for surgical planning. All the cases had undergone MRI in sedation due to abnormal posturing.

Surgical procedures

As a part of the protocol, brain MRI (3 T, Siemens) with no spacing and 3-Dimensional Volume reconstruction image was performed and it was obtained in digital imaging communication in medicine (DICOM) standard. The image was loaded in Inomed Planning Software (IPS), where the targeting and trajectory for stereotactic GPi pallidotomy was done (Fig. 1). The anterior commissure (AC) and posterior commissure (PC) were visualized .The primary target was set as 18 to 19.5 mm lateral, 3 mm below (caudal) and 2 mm anterior to the mid-commissural point. Similarly, secondary target was set as 20-21 mm lateral, 1 mm posterior and 3 mm below the mid-commissural line. The target was reconfirmed through the inbuilt Schaltenbrand atlas. We calculated PTT lesioning after visualizing the mammilothalamic tract and from the midpoint of AC-PC line.

Under local anesthesia, stereotactic frame (MRI compatible Zamorano-Dujovny Fisher) frame was applied and computed tomography (CT) scan of head was done (64 slice Siemens with 2 mm slice thickness) with no tilt. These images were retrieved in a DICOM format. Then the MRI and CT images were fused in the workstation and the working coordinates for the surgical procedure was extracted. The patient was then taken to the operating room and the frame was fixed in the Mayfield. The coordinates were set on phantom to reconfirm the accuracy of target. Once confirmed, a burr hole just anterior to coronal suture was made.

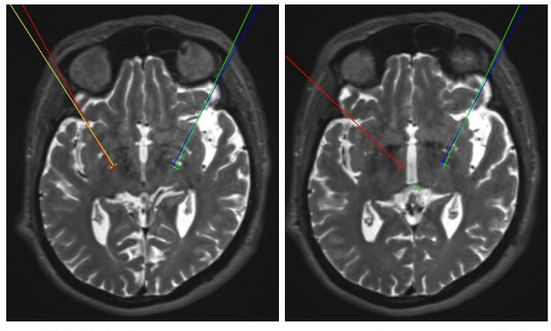

Figure 1a: T2W MRI showing targeting of Bilateral GPi. .1b: T2 W MRI showing targeting of right sided PTT and left sided Gpi.

Results

Out of 89 cases of dystonia(17 generalized, 30 Task specific and 42 cervical dystonia), we included 17 cases of generalized dystonia. Out of 17 cases of generalized dystonia, 13 cases had bilateral pallidotomy and 4 cases had unilateral pallidotomy with contralateral PTT lesioning as shown in fig 3a&b. PTT lesioning was started since September 2023.

Patient demographics and outcome are detailed in Table 1. The mean age was 45 ±8 years in Bilateral pallidotomy group and 35±10 years in unilateral Pallidotomy and contralateral PTT lesioning in the same setting. The male: female ratio was 3:1. The mean percentage change in BFMDRS was 65 percent (p value<0.05) in postoperative period in bilateral pallidotomy patients and 70%(p value<0.05) in unilateral Pallidotomy and contralateral PTT lesioning in the same setting.

The mean follow up of bilateral pallidotomy cases were 10 ±2 years. There was relapse in 3 cases after one year follow up. There was permanent dysarthria and dysphagia in 3 cases which were irreversible. One case of recurrent cases eventually required intrathecal baclofen pump.There was also secondary parkinsonism like features after 8 years in two cases. However there was no mortality.

The mean follow up of unilateral Pallidotomy and contralateral PTT lesioning 2 ±1 years. There was hypophonia in only one case. One case had to be operated in General anesthesia as he could not tolerate in local anesthesia. Postoperatively his symptoms got improved. There was no mortality.

Table:1 showing demographics and surgical outcome of generalized Dystonia:

Target	Bilateral Pallidotomy	Unilateral Pallidotomy with contralateral Pallidotomy
Number	13	4
Male: female	3:01	3: 1
Mean age	45±8 years	35±10 years
Mean Change in BFMDRS	65%(p value<0.05)	70%(p value < 0.05)
Follow up	10±2 years	2±1 year
Relapse	3 cases	None
Complications	Dysarthria: 3	Hypophonia:1
	Dysphasia: 3	
	Secondary parkinsonism: 2	

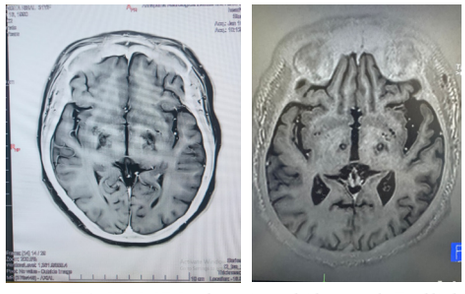

Figure 2 a: T1 W MRI showing bilateral Pallidotomy 2b. T1 W MRI showing right sided PTT lesioning and left sided pallidotomy.

Figure 3: Preoperative and postoperative image of Generalized Dystonia

Figure 4: Preoperative and postoperative image of generalized dystonia

Discussion

Dystonia is a disorder of neural networks, with symptoms emerging from dysfunction within the cortico–basal ganglia circuit. Abnormal signaling from this pathway can lead to changes in cortical excitability and atypical synaptic plasticity.[12] Alongside advances in clinical characterization, progress in genetic testing—particularly next-generation sequencing—has greatly enhanced the diagnosis of primary dystonia. Numerous isolated dystonias and some combined forms are now known to stem from genetic abnormalities, and several causative genes have been identified worldwide, including TOR1A (DYT1), THAP1 (DYT6), and KMT2B (DYT28).[13,14] These diagnostic improvements have facilitated more precise assessment of dystonia phenotypes and have supported more targeted therapeutic decisions, such as the use of DBS and lesioning.

DBS is also a good option, but its high initial cost, hardware-related complications and regular outpatient visits must be considered when implementing it. Stereotactic ablative surgery can be as effective as DBS at less cost.

In most developed countries where DBS is widely accessible, GPi ablation (pallidotomy), an alternative surgical treatment for dystonia, is rarely used. According to limited cohort studies, pallidotomy and GPi-DBS are equally effective in treating dystonia.[15,16] When treating dystonia in those who are against device implantation or who cannot access DBS due to financial or geographic constraints, pallidotomy is particularly crucial. However, single case reports and short cohort studies are the only sources of information now available on the effectiveness of pallidotomy for dystonia. Pallidotomy's effectiveness and safety in treating dystonia are still unclear.

Over the past decade, deep brain stimulation (DBS) targeting the internal globus pallidus (GPi) has become the leading treatment option for patients with severe primary dystonia that responds poorly to medication.[10,11] Numerous retrospective case series and prospective clinical trials have demonstrated the short-term effectiveness of pallidal DBS in primary dystonia; however, only a limited number of studies have evaluated its long-term outcomes in larger patient cohorts.[17,18]

Isaias et al evaluated that pallidal DBS is a safe and effective treatment for medically refractory primary generalized dystonia PGD, with improvement sustained for up to 8 years in 1 patient without significant changes in stimulation variables.[6] Martinez et al demonstrated that 80 consecutive patients with dystonia who underwent neurosurgical management by means of intrathecal pump implantation, pallidotomy or deep brain stimulation (GPi or VIM) had mean improvement in BFM-DRS score among patients with primary and secondary dystonia,87.54% and 42.21%, respectively.[19]

Vidailhet et al reported that bilateral pallidal stimulation provides sustained motor benefit after 3 years.[20] Mild long-term improvements in quality of life and attention were also observed. Volkmann et al reported that 3 years and 5 years after surgery, pallidal neurostimulation continues to be an effective and relatively safe treatment option for patients with severe idiopathic dystonia.[21] Similarly, Park et al reported GPi DBS is a safe and efficient therapeutic method for treatment of dystonia patients to improve both movement and disability.[22]

Horisawa et al. (2021) reported an open-label pilot study demonstrating that Vo MRgFUS significantly improved

focal hand dystonia in 10 patients, with mild dysarthria observed in only one patient as the sole adverse event over a 12-month follow-up period.[23] Similarly, another pilot study by Horisawa et al in 2025 suggests that FUS pallidothalamic tractotomy may be an effective treatment option for patients with cervical dystonia.[24] The TWSTRS at 6 months (29.9 ± 16.0, range: 3-55) was significantly improved by 43.4% ($P < 0.001$) from baseline. The BFMDRS-Neck scales at 6 months (4.2 ± 2.8) were significantly improved by 38.2% ($P < 0.001$) from baseline. Simiarly, Galimova et al reported that MRgFUS is efficient and sufficiently safe for symptomatic treatment in pharmacoresistant cervical dystonia patients.[25] They reported that MRgFUS achieved a 70.6% improvement of the TWSTRS score at the last observation. However we could not find the result of MRgFUS on generalized Dystonia.

Previous studies has shown that bilateral pallidotomy carries significantly greater risks compared to unilateral procedures. When we did bilateral pallidotomy we made one lesion bigger than the other. Similarly while performing the second pallidotomy, we ask the patient to swallow water from the straw in order to stop the procedure if there are any complications. There was a case of early internal capsule infarction following GPi lesioning leading to hemiparesis[26], transient dysarthria in three cases, dysphagia in two cases and hypophonia in two cases. To minimize complications linked to bilateral pallidal surgeries, the pallidothalamic tract as a potential target for dystonia treatment has been proposed.[27] A combined approach—unilateral pallidotomy with contralateral pallidothalamic tractotomy—resulted in a 74.3% improvement in BFMDRS scores among 11 dystonia patients, without inducing parkinsonism.[27] As we started to do PTT lesioning since 2023 there are only four cases of Unilateral Pallidotomy and contralateral PTT lesioning. The result is slightly better than bilateral pallidotomies with lesser complications and the follow up is also lesser. We suggest that it is better to do right sided PTT lesioning and left sided pallidotomy. Further research is warranted to validate the safety and effectiveness of this combined ablative strategy.

Conclusion

Lesioning surgery is rewarding for generalized dystonia. Recurrent cases may need DBS or further potential targets. In bilateral pallidotomies, smaller lesion in the opposite side may decrease the risk of complications. Unilateral pallidotomy and contralateral PTT lesioning in generalised dystonia is also rewarding and it should be also promoted than bilateral pallidotmies.

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