

Challenges and scope of movement disorder surgery in Nepal



Resha Shrestha¹ , Pritam Gurung² , Basant Pant³ 

^{1,2,3}Department of Neurosurgery, Annapurna Neurological Institute and Allied Sciences, Maitighar, Kathmandu, Nepal

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Movement disorders (MD) are clinical syndromes with either excess or paucity of voluntary or involuntary movements without weakness or spasticity. Hypokinetic MD includes primary and secondary Parkinsons disease and Parkinsons syndrome and hyperkinetic MD includes dystonia, tremors, choreas and dyskinesias. Although medical treatment is the mainstay of treatment modality, in selected cases surgical treatment is a safe option.

Parkinson's disease (PD) is a neurodegenerative disorder characterized primarily by loss of dopamine neurons in the substantia nigra. The clinical features comprises of slowly progressive asymmetric resting tremor, cogwheel rigidity and bradykinesia. Other non motor feayures include anosmia, constipation and depression. Additional non motor features in later stages are autonomic dysfunction, pain and cognitive decline.¹ In estimates based on health care utilization, PD incidence ranges from 5/100,000 to over 35/100,000 new cases yearly. Incidence increases 5 to 10 fold from the sixth to the ninth decades of life.^{1,2} In Nepal, with the total population of 30 million as of 2021, the new cases ranges from 1500 to 10500 per year and with growing ageing population the incidence is going to increase. The age standardized prevalence rate of PD in Nepal is 50 to 60 per 100000 population.³

Medical therapy is the mainstay of treatment for PD with dopaminergics drugs (levodopa), Enzyme inhibitors like(MAO-B inhibitors, COMT inhibitors), Anticholinergics and Amantadines. Surgical treatment of PD has evolved since 1950s as ablative procedures like pallidotomy and thalamotomy. Deep brain stimulation (DBS) surgery which was started in 1997 was pioneered by Benabid. In Nepal, DBS for PD was started in 2014 in Annapurna Neurological Institute and Allied Sciences with the support of Prof Takaomi Taira from Japan.⁴ Till date, 30 DBS surgeries have been performed with the overall result of 65 % improvement in Unified Parkinsons Disease Rating score (UPDRS) in early follow up. The main hindrance for DBS surgery is the cost itself with the tentative cost of 18000\$. The National Health insurance system does not support for DBS Surgery. However Nepal Government is providing 1 lakh Nepali Rs (800\$)n to the patients of PD if treated in government hospital.

The ablative surgeries for PD was also started in 2014 in Annapurna Neurological Institute and Allied Sciences and 45 pallidotomies have been done for PD. There is 70% improvement in UPDRS in early follow up. We use radiofrequency lesioning with Cosman RF generator. This is a cost effective surgical treatment (which costs around 1500 \$) for Lower Middle Income Countries (LMIC) in Nepal and it can be popularized after proper training to the individuals. With the growing trend of DBS for PD in the global scenario, ablative surgeries like pallidotomies has its place and should not be abandoned. With the recent FDA approval of Magnetic Resonance guided Focal Ultrasound therapy (MRFUG), for PD in 2018, many centers are still going back to ablative procedures. The hindernce of MRFUG is the cost in our country and we believe that ultimate result is the same and RF lesioning is more cost effective.

Dystonia comprises movement disorder in which there are sustained or intermittent muscle contractions. This causes abnormal, often repetitive, movements, postures, or both. They are typically patterned, twisting and may be tremulous. The incidence of generalized dystonia is 2 per million person per year and focal dystonia is 24 per million per person. The prevalence is 3.4 per 100,000 and focal dystonia is 30 per 100,000.⁵ In Nepal, around 60 persons can have generalized dystonia per year and about 750 people can have focal dystonia

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Address for correspondence:

Dr Resha Shrestha
Department of Neurosurgery
Annapurna Neurological Institute And Allied Sciences, Maitighar,
Kathmandu, Nepal
E-mail: reshkums@hotmail.com

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per year. The population prevalence is 1020 and 9000 respectively. The genetic classification is based on the loci of genes involved (DYT1 through DYT15) that include autosomal dominant, autosomal recessive and X-linked causes of primary dystonia, dystonia-plus syndromes and paroxysmal dystonias. The genetic testing of dystonia is still in primitive phase in Nepal and hence is difficult to categorize it. The commonest focal dystonia are Writers cramp and Musicians cramp. Medical treatment of dystonia include dopamin, anticholinergics (trihexphenidyl), benzodiazepines (diazepam, clonazepam), dopamine depletors (tetrabenazine), gamma aminobutyric acidergics (baclofen), atypical neuroleptics (clozapine, olanzapine) and even typical neuroleptics (e.g., haloperidol) in very disabled patients. Botox injection is also a feasible treatment for focal dystonia.

Generalized dystonia is very difficult to treat medically. Before 1999 all cases of Dystonias were treated by ablative procedures like pallidotomies, but nowadays, DBS has become the mainstream surgical procedure for virtually all types of dystonia, owing mainly to its safety profile compared with pallidotomy, especially when bilateral. Some studies have stated that due to promotion of DBS by industries, there is lack of training in ablative surgery for emerging neurosurgeons leading to decrease in pallidotomy procedure.^{6,7}

In Nepal, we have done pallidotomies in 25 cases of generalized dystonia with 80 percent change in Burkey Fahr Mardson Dystonia Rating Score (BFMDRS) ($P < 0.05$). There was recurrence in 5 cases and these recurrent cases might benefit from DBS. However none of the recurrent cases could afford DBS surgeries. We have also done 15 cases of VOA Thalamotomy in Focal hand dystonia. The majority of them are writers cramp followed by musicians cramp and typist cramp. Our result is 85% improvement in Writers cramp rating score ($p < 0.05$) with only one recurrence. There is even better response in largest series of 171 cases of focal hand dystonia cases.⁸ We believe that DBS is not necessary for Focal hand dystonia cases.

Essential tremor is the most common movement disorder and is characterized by rhythmic oscillation of agonist and antagonist muscle groups. ET affects people of all ages and has a bimodal distribution of age at onset, respectively peaking at the second and sixth decades.⁹

Although ET is generally regarded as a benign neurological disorder, a wide range of symptoms and comorbidities can lead to impaired quality of life, disability and social handicap. The overall prevalence of ET in the general population was 0.32% and was higher in males.¹⁰ In 2020, the number of people affected by ET globally was 24.91 million. In Nepal the prevalence of ET is about 100000.

The medical treatment of ET is mainly beta blockers like Propranolol and anti seizure medicines like primidone, Gabapentin and Clonazepam. Patients who do not benefit from medical treatment can opt for surgical treatment. Thalamotomy of VIM (Ventral intermediate) region of thalamus can reduce tremor to more than 90%. In Nepal, we have done 20 cases of VIM thalamotomy by radiofrequency electrode. Our result is 75% improvement in Clinical Rating Scale for Tremor CRST Score ($p < 0.05$). This is very few in number compared to the overall prevalence in the country. Proper referral of the cases should be done. DBS of Posterior Subthalamic Area (PSA) and VIM DBS can also reduce the tremor by 65-80%.¹¹ In Nepal, we believe that DBS for essential tremor might not be feasible due to cost effectiveness. Another treatment modalities for ET is focused ultrasound thalamotomy, and radiosurgical thalamotomy. There is no MRFUG center and gamma knife center in Nepal at this moment.

Conclusion

There are many cases of Movement disorders of which surgical treatment options are effective in selective patients. Appropriate referral of such cases will help to address such conditions properly.

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