Generalized dystonia

Generalized dystonia refers to dystonia that affects multiple muscle groups throughout the body.

Generalized dystonia typically affects muscles in the torso and limbs, and sometimes the neck and face.

Generalized dystonia typically begins during childhood or adolescence, often without additional neurological symptoms.

However, dystonia in children can be associated with more complex neurological or metabolic diagnoses.

Dystonia may result from changes in certain genes (including DYT1/TOR1 and DYT6/THAP1), birth injury, exposure to certain drugs, head injury, infection, and other secondary causes.

Treatment often requires a combination of therapies including oral medications, botulinum neurotoxin injections, surgical interventions, and/or complementary therapies.

The aim of a study was to evaluate the use of a novel technique for baclofen delivery using an intrathecal catheter inserted through a lumbar laminotomy with the tip placed at the cisterna magna (supraspinal baclofen (SSB)) for the treatment of severe generalized secondary dystonia.

A cohort study of six individuals (4M/2F, mean±SD = 15±4.86 years) with generalized dystonia unresponsive to oral medications were treated with SSB and followed clinically for 8 years. Intrathecal catheter tips were positioned under fluoroscopic guidance just above the level of the foramen magnum, at the cisterna magna.

Five of the 6 patients experienced sustained benefit with SSB; the group mean modified Fahn-Marsden scale scores decreased from 95 to 55 ($t = 3.29$, $p = 0.02$). One patient suffered a complex pump pocket infection and therefore underwent pump explantation.

Supraspinal baclofen may be an effective method for infusing baclofen into the ventricular system of the brain for the treatment of secondary dystonia which is unresponsive to oral therapy [1)].

Deep brain stimulation (DBS) is an established treatment for generalized dystonia. However, the DBS device is sometimes removed owing to hardware complications. Here, we present four cases of generalized dystonia treated with radiofrequency lesioning through the DBS electrodes.

The patients were three men and one woman (34 to 44 years old). They had undergone DBS for generalized dystonia. However, they subsequently developed complications, such as infection, and removal of the devices became necessary. As stopping the stimulation caused recurrence of uncontrollable symptoms, radiofrequency lesioning was performed through the DBS electrodes under local anesthesia, and the DBS systems were removed under local or generalized anesthesia thereafter. The procedures performed were as follows: two patients had bilateral pallidotomy, one patient had unilateral pallidotomy, and one patient had pallidotomy and ipsilateral thalamotomy. As a result, in four patients, the dystonic symptoms did not worsen even after removal of the DBS systems

during a follow-up period of 1 to 12 years. However, one patient had a small hemorrhage and two patients showed recurrence of dystonia.

Radiofrequency lesioning with DBS electrodes is feasible in cases of generalized dystonia when the DBS leads have to be removed [2].

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2)
Takeda N, Hoeisawa S, Taira T, Kawamata T. Radiofrequency Lesioning through Deep Brain Stimulation Electrodes in Patients with Generalized Dystonia. World Neurosurg. 2018 Apr 18. pii: S1878-8750(18)30771-X. doi: 10.1016/j.wneu.2018.04.055. [Epub ahead of print] PubMed PMID: 29679783.