EFFECTIVENESS OF BOTULINUM TOXIN TYPE A (BONT-A) IN LONG-TERM TREATMENT OF HEREDITARY SPASTIC PARAPLEGIA IN CHILDREN: REVIEW OF A SERIES OF 63 CASES

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Introduction and Objectives:

BoNT-A has proved to have an important role in the treatment of spasticity produced by nonprogressive disorders, such as cerebral palsy (CP). Hereditary spastic paraplegia (HSP) is a slowly progressive disorder of heterogeneous genetic etiology that is often also accompanied by polyneuropathy. The polyneuropathy is a relative contraindication for BoNT-A treatment.
There are few reports about the use of BoNT-A in HSP, and these are rarely about children.
The objective was to investigate the results of treatment with BoNT-A in a large series of children with HSP.

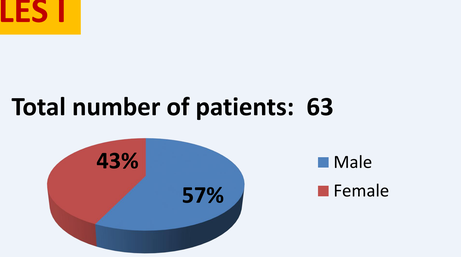
Methods:

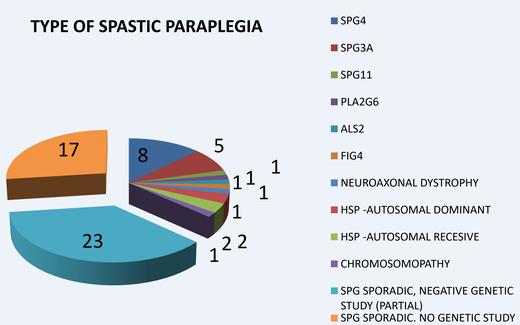
The children were diagnosed using neuroimaging (magnetic resonance imaging), neurophysiology (motor nerve conduction studies, sensory nerve conduction studies, somatosensory evoked potentials, visual evoked potentials, and transcranial magnetic stimulation) and genetic studies, and other etiologies were excluded.
BoNT-A was injected using electromyography, electrostimulation, or palpation and anatomic localization, always with conscious analgesia using nitrous oxide 50%, ethyl chloride spray, or EMLA cream.
All the patients were assessed at each injection and 2-3 months later, using scales of function and spasticity and a questionnaire to collect details of adverse events (AE).
The Physician Rating Scale (PRS) was used to assess the type of foot support, hip adduction, and popliteal angle during gait, and the active foot dorsiflexion.
The spasticity scales used were the Modified Ashworth Scale (MAS) and the Tardieu Scale.

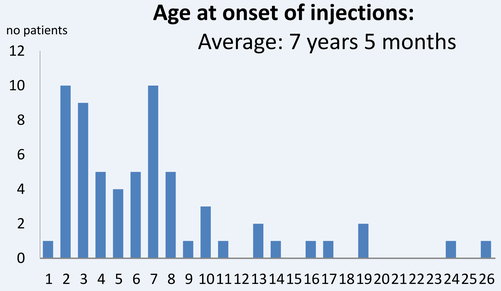
Results:

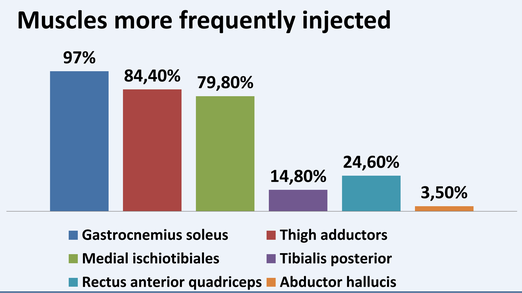
63 cases of HSP were treated. See **group of tables I**: distribution by etiology, age of onset, muscles injected.
The average age at onset was 7y5m, with reinfiltrations every 4,8 m in average.
Clinical features are shown in **group of tables II**. 63,5% of the patients were ambulant (GMFCS I or II).
The total doses/kg bw of BoNT-A injected by session were in the range of recommended dose in childhood spasticity but slightly lower than those used by our team in spasticity due to CP.
Efficacy (table III): All patients improved spasticity in most of the muscles. Concerning to functionality, comparing the first and last examinations 42,2% of patients improved and 4,3% worsened the foot support during gait; 35,5% improved and 5.9% worsened the adduction of hips; 32.2% improved and 8,5% worsened the popliteal angle; and 40,5% improved and 5.1% worsened their active foot dorsiflexion.
The proportion of infiltrations that produced **adverse events (AE)** were 10,8%. **(Group of tables IV).** Out of these 10,8% of AE, they consisted in minimal or slight tiredness or weakness that did not prevented any of the daily activities (80%), moderate weakness in 13,8% and important weakness in 2%. AE lasted less than 5 days in 44% of cases and more than a week in 56%. These data indicate a good profile of safety but less safe than the figures that we have in Cerebral Palsy (3% of AE).

TABLES I

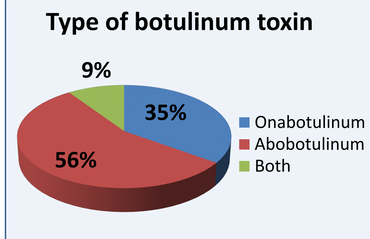


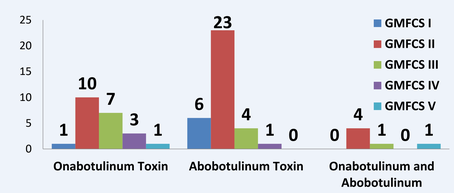


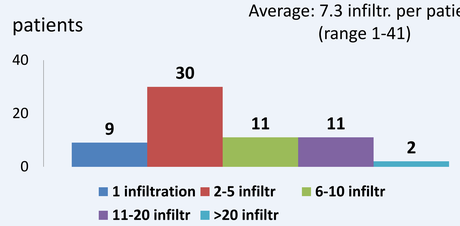


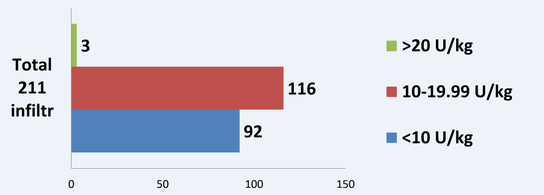


TABLES II

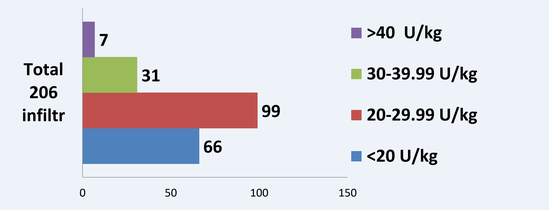






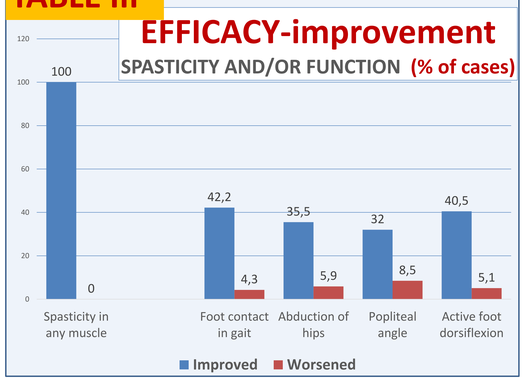


Average 263 U by session (10.63 U/kg)

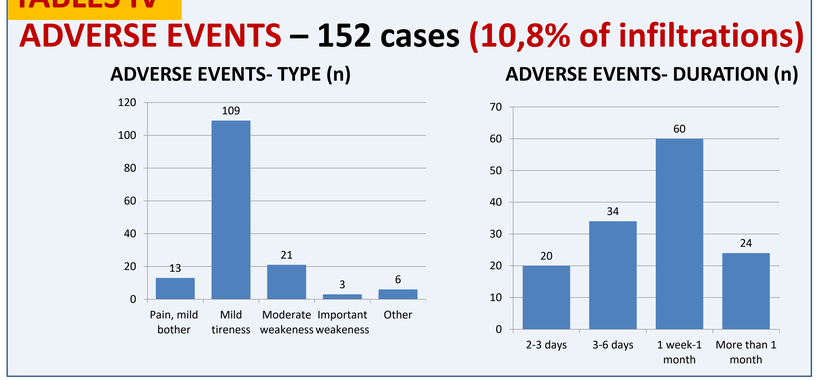


Average 742 U by session (23.57 U/kg)

TABLE III



TABLES IV



CONCLUSIONS:

- BoNT-A is a useful and safe treatment of spasticity in HSP, both in the short and long term.
- It improved spasticity in all cases, and some degree of functionality in more than a third of cases.
- The doses injected must be carefully calculated and are lower than those used for non-progressive disorders.
- AEs are infrequent and mild, but must be expected in a proportion of cases.