

**Conclusions:** is evident the high degree of sequelae in our population, mostly in the motor área. The mortality was lower than international reports. Is necessary to find new methods to prevent or ameliorate the impairment of children affected with the disease.

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#### Late Breaking Posters 1

##### Acute allergic reaction following injection of generic, government-provided botulinum toxin a (GGP-BTA) for hemifacial spasm

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**Background:** Allergic reactions after administration of Botulinum Toxin A are rare, unpredictable complications and may vary from rash to anaphylaxis. These have been reported with the use of commercially available formulations, but not with generic formulations. These are probably due to allergic reactions to albumin, lactose or gelatin use in the formulation of BTA.

**Objective:** To report the case of a patient who developed facial angioedema one hour after receiving intramuscular Generic, Government-provide Botulinum Toxin A (GGP-BTA).

**Patient and method:** Case report.

**Result:** A patient with left hemifacial spasm, who had already been treated with commercially available BTA (Botox®) with no significant side-effects, was treated with intramuscular injections of GGP-BTA on the affected muscles of the left side and additional injections in the periocular region of the right side for aesthetic symmetry. About one hour following the procedure he developed a purplish-red oedema of the eyelids, as well as the left nasolabial fold (Figure). He was treated at first with IV Methylprednisolone, followed by oral 60 mg qd of Prednisone. As there was minor improvement with corticosteroids, following evaluation by the Dermatology team, he received intramuscular Adrenaline and was started on oral Diphenhydramine with resolution of the angioedema.

**Conclusion:** Although rare, allergic reactions to commercially available BTA may vary from rash to life-threatening anaphylaxis. We reported a case of facial angioedema following injection of GGP-BTA, with resolution after adequate systemic treatment with anti-allergic medications.

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#### Late Breaking Posters 1

##### Phenotypic description of Rett syndrome patients with MECP2 gene mutation

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**Introduction:** there are several clinical phenotypes related to mutations in MECP2 gene, being Rett syndrome in females the most frequent in its classical and variant forms, and syndromes in males going from encephalopathy to mental disability.

**Objective:** to establish the clinical phenotype and evolution of patients with mutations in MECP2 gene and to describe its relation to the phenotype.

**Materials and methods:** descriptive and retrospective study of patients with mutations in MECP2 gene encountered in our Service between 2005-2009. Cases with positive mutations are studied from the protocol applied to girls with Rett syndrome phenotype.

**Results and discussion:** 11 girls with Rett syndrome phenotype, 9 classical forms and 2 variants. The 9 girls with the classical form had normal initial development, regression between 6 to 18 months, stereotypies emergence, 7 lost hand propositivity, 6 had epilepsy, 7 acquired microcephaly, 4 achieved gait and none had language. 6 patients had frequent mutations R255X, R168X y R306C with similar clinical severity. The 2 patients with variant forms had normal development until 3 years old, slow progression of language, normocephaly, epilepsy, late stereotypies, preserved gait, with R294X and TRUNC293 mutations.

**Conclusion:** phenotypes of MECP2 gene mutations manifest as usually is described in females with Rett syndrome. Most are classical forms, with the exceptional gait achievement as highlight. The most frequent mutation is R255X and the severity of the phenotype is not related with a mutation in particular.

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#### Late Breaking Posters 1

##### Allocentric neglect strongly associated with egocentric neglect during intraoperative eletrophysiological stimulation in low grade glioma

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#### Abstract

The authors show a case of 34 year old female patient with a tumor - Astro grade II in supramarginal gyrus and and angular gyrus on the right side, who presented as initial clinical picture a partial motor and complex seizure, of difficult control. During the craniotomy the angular area was stimulated with 3 volts, with a monopolar tip, as well as the supramarginal gyrus, during intraoperative neuropsychological testing. The response was curious, being allocentric neglet strongly associated with egocentric neglect. There is an established tradition for considering these two phenomena as both behaviorally and anatomically dissociable. However, several studies and some theoretical work have been suggested that these rather reflect two aspects of a unitary underlying cause. An consistent and important work of Yue et al. (2012), suggests that allocentric behavioral deficits might appear only in conjunction with egocentric deficits as well as there would an large corresponding overlap for the anatomical regions associated with egocentric and with allocentric neglect. The authors discuss how different anatomical and behavioral findings can be explained in a unified physiologically plausible framework, whereby allocentric and egocentric effects interact.