REFRACTORY LANCE-ADAMS SYNDROME: PHARMACOTHERAPY MANAGEMENT AND IATROGENIC COMPLICATIONS

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BACKGROUND
Lance–Adams syndrome (LAS) is a chronic posthypoxic myoclonus that may appear after a period of cerebral hypoxia. Many different antiepileptic drugs (AED) have been used for the symptomatic control of LAS. In the absence of response to classic AED, it is necessary to consider new off-label therapeutic options which may cause unpredictable adverse events.

PURPOSE
To present the pharmacotherapy approach to a case of refractory LAS, describing treatment related adverse events and its management.

MATERIAL AND METHODS
Clinical information was collected from the electronic medical records looking for evidence for the use of perampanel, 5-hydroxytryptophan (5-HT) and sodium oxybate in LAS.

RESULTS

AT ADMISSION
- Treated with levetiracetam, sodium valproate.
- Sedated with propofol and sodium thiopental.
- Myoclonus was not controlled: piracetam, zonisamide (to reduce the use of sedative drugs) and clonidine were added to the treatment, without improvement.

DURING HOSPITALIZATION
- Sodium oxybate was added, but it was discontinued because of the risk of respiratory arrest.
- 5-HT also was added with no significant outcome and severe diarrhea as an adverse event.
- Perampanel (24 mg/day - maximum daily dose doubled) was added to the treatment achieving myoclonus improvement.
- Simultaneously, the patient had behavioral disorders that were linked with perampanel treatment needing addition of risperidone.

AT DISCHARGE
LAS control was achieved and the patient was discharged with levetiracetam, gabapentin, perampanel and risperidone treatment.

CONCLUSION
- The refractory nature of LAS forced the medical team to use off-label drugs and supratherapeutic doses, with increased frequency of adverse events.
- The drug related events were identified and properly managed, allowing treatment continuation and ensuring patient improvement.