# A Case of Lance-Adams Syndrome: The Clinical Process of the Evidence-Based Clinician

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**Abstract**

Evidence-based practice (EBP) has become a hallmark of modern medicine. Diving into the literature is an essential part of clinical decision-making. The management of rare disorders and disease almost always prompts a search, but the answers to important clinical questions can be challenging and rarely straightforward. The majority of us have endured the "EBP Class" of our academic careers, drilling the hierarchy of research evidence and whether strict inclusion/exclusion criteria were considered, etc. We frequently see our medical colleagues turn to high powered trials and meta-analyses, but such works are few and far between. Single-subject design, case studies, case series, and reviews of such cannot be discredited, especially within a profession that is relatively new in the broad scheme of medicine. The following case study details a clinician's process through the identification, appraisal, and application of relevant literature in the management of a rare disorder: Lance-Adams syndrome. Proposed assessment considerations are intended to help the reader's approach to diagnosis and intervention.

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***Learning Objectives***

1) Define Lance-Adams Syndrome and identify its associated communication and swallowing impairments.
2) State the roles of the interdisciplinary team appropriate for individuals with Lance-Adams Syndrome.
3) Summarize appropriate diagnostic considerations for swallowing and communication difficulties in individuals with Lance-Adams Syndrome.

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**A** 33-year-old male with newly diagnosed Lance-Adams syndrome (LAS) was referred to the speech-language pathology service to address communication and swallowing. The patient was admitted to a teaching hospital after being stabilized at an outside facility following heroin overdose. During the overdose, the patient went into cardiac arrest, and his spouse started cardio-pulmonary resuscitation. The patient was transported by ambulance to his community hospital where he remained unconscious and intubated with mechanical ventilation for four days. Continuous electroencephalography suggested status epilepticus. The patient was transferred to the teaching hospital two weeks thereafter. Following the transfer, he was awake and alert and resumed a regular-textured oral diet, ordered by the medical team.

Records to this point were scattered. It was not until the patient was admitted to the teaching facility that clinical features consistent with LAS were observed. A chest radiograph revealed atelectasis, or reduced lung inflation, of the right lower lobe and developing pneumonia. Repeated radiograph one month later was normal. Repeated head imaging was also normal. The patient told his medical team that he was having trouble feeding himself and "getting [his] words out", prompting our referral.

The patient had a history of hepatitis C virus, substance abuse disorder (including alcohol), post-traumatic stress disorder, anxiety, depression, and tinnitus. The patient served four years in the United States Air Force after graduating from high school and was currently unemployed. He had four children and was living with his spouse prior to admission. The patient described his relationship as "rocky".

Inpatient medications included clonazepam, a benzodiazepine, and valproic acid, an antiepileptic drug. Both were prescribed to treat the myoclonic dyskinesia described later. Mirtazapine, an atypical antidepressant, and sertraline, a selective serotonin reuptake inhibitor, were both used to treat depression and generalized

anxiety disorder (off-label). Clonidine was prescribed (off-label) to address opioid withdrawal. Heparin was prescribed as an anticoagulant. Thiamine was used to prophylactically treat thiamine-deficiency in the context of a known history of alcohol abuse.

**Supporting Research Evidence**

A search for best available evidence led to the databases PubMed, Google Scholar, and ASHA Wire. Combinations of relevant search terms were used: *Lance-Adams syndrome, speech, swallowing, cognition, language, post hypoxic myoclonus, hyperkinetic, patient reported outcome.* The search returned a series of case studies and systematic reviews. The following describes those deemed most pertinent to the case under discussion.

LAS was first described in 1963 (Lance & Adams, 1963). Four patients sustained anoxic encephalopathy following either cardiac arrest or a prolonged airway obstruction. Similar disturbances in movement were observed amongst the patients after regaining consciousness. Volitional movement induced an arrhythmic course or fine jerking of a single or group of muscles. Movements were approximately one-fifth of a second in duration and labeled *action* or *intention* myoclonus. Voluntary motion was initiated well, but became quickly interrupted. Movement became increasingly chaotic and fragmented as it continued. Severity of the myoclonus was found to be grossly proportional to the precision of the task. Lance and Adams (1963) described the relevant examples of "conveying food to the mouth" and "drinking from a glass" (p. 117) as particularly difficult activities. Myoclonic movements ceased when patients relaxed. All four patients consistently maintained an ataxic gait.

Lance and Adams (1963) hypothesized that the underlying neuropathophysiology included a repetitive firing of thalamocortical fibers, stemming from the ventrolateral thalamic nucleus. The ventrolateral thalamic nucleus plays a large role in mediating motor function. This area receives input from both the cerebellum and basal ganglia and projects to the primary motor and premotor cortices (Siegel & Sapru, 2015). Abnormal firings through these projections are relayed to the corticobulbar and corticospinal tracts, producing action myoclonus (Duffy, 2013). Autopsies from Lance and Adams's other deceased patients revealed that the globus pallidus, hippocampus, deep folia of the cerebellum, and deep layers of the cerebral cortex were structurally damaged by similar anoxic brain injuries. These areas, as well as the Purkinje cells of the cerebellum, were hypothesized to be damaged in LAS specifically.

Lance and Adams's (1963) paper is certainly dated, but they provide a descriptive review. Their description continues to be referenced as a standard definition of the syndrome in the most recent literature. The four patients that are described closely resemble the current case, making this evidence pertinent to diagnostic considerations.

Gupta and Caviness (2016) conducted the most recent systematic review of the LAS literature. The authors concluded that the neuropathophysiology and the neuroanatomy implicated remains poorly understood. Imaging studies collectively identified a wide variety of damaged neuroanatomy, including the basal ganglia, parietal lobes, and hippocampus. Some findings were even transient, resolving after repeating their imaging studies weeks later. Seizures were often observed in the acute comatose stage following a hypoxic event. Onset of LAS symptoms was observed days to weeks thereafter. LAS was found to be associated with dysarthria, dysphagia, seizures, cognitive deficits, and gait disturbances. Unfortunately, these impairments of significant interest to the speech-language pathologist were not well described. The collective literature indicated LAS responded well to benzodiazepines and anticonvulsant drugs. The syndrome typically persisted indefinitely.

Lee and Lee (2011) published a case report describing a patient strikingly similar to the current case. A 32-year-old male went into cardiac arrest after attempting to hang himself. He was successfully resuscitated and transferred to his community hospital. Computed tomography of the head was normal. Electroencephalography revealed status epilepticus. On day-12 of his hospitalization the patient regained consciousness, but he displayed action myoclonus. Performance on the Mini-Mental State Examination (Folstein et al., 1975) improved from a total score of 19 out of 30 (moderate impairment) to 22 out of 30 (mild impairment) by discharge. Coupled with pharmacologic intervention, rehabilitation focused on voluntarily slowing volitional movement. This approach successfully reduced myoclonic jerking. By discharge, the patient was able to walk 20 meters with the assistance of a rolling walker. His dysarthria persisted.

**Possible Impairments**

Understanding the basic underlying nature of LAS and its known consequences can guide the evidence-based

clinician. The case history and review of relevant external evidence suggests diagnostic hypotheses implicating oropharyngeal swallowing, motor speech, and cognitive-communication.

**Swallowing**

The literature identified action myoclonus, a dyskinesia, as the primary feature of LAS. Dyskinesias, like those in LAS, may result in dyscoordination of the oropharyngeal swallow (Stierwalt, 2013). The muscle relaxant and sedative properties of the benzodiazepine (clonazepam) and anticonvulsant (valproic acid) could contribute to both dyscoordination and inattention. Atypical antidepressants (mirtazapine) may also cause sedation. Side effects are often amplified in the setting of liver dysfunction (Carl & Johnson, 2006). Lance and Adams (1963) described relatively fine motor movement resulting in proportionally worse myoclonic jerking. The ability to self-feed would be impacted.

**Speech**

Motor speech impairment is very likely to be observed, specifically hyperkinetic dysarthria. Experts suggest dysarthria associated with action myoclonus may have a greater functional effect on speech production, as compared to the more familiar palatopharyngeal myoclonus (Duffy, 2013). Aronson, O'Neill, and Kelly described their own four cases of LAS (as cited in Duffy, 2013). Their patients demonstrated fluctuations in phonation, and the aberrant phonatory function synced with action myoclonic movement of the lips. Sudden voice arrests, generally slow rate of speech, and articulatory imprecision when rate increased would therefore likely be observed in the current case (Duffy, 2013). Errors would be highly variable and increase in occurrence with longer utterances.

**Cognitive-Communication**

The etiology of LAS suggests cognitive-communication impairment should also be considered. Plans for intervention may be directly impacted by such findings. Studies sporadically implicated structures of the basal ganglia and cerebellum among other areas. Both the basal ganglia and cerebellum have been found to play a role in cognitive functioning (Middleton & Strick, 2000). A review of long-term consequences associated with hypoxic-ischemic encephalopathy indicated a range of severity of cognitive impairments has been observed (Khot & Tirshwell, 2006). A reversible metabolic encephalopathy has been reported after relatively brief periods of circulatory arrest. This transient difficulty, characterized by a short period of confusion, or at least improvement in functioning, as described in the case by Lee and Lee (2011), may be expected in the current case.

**Conclusions: Diagnostic Considerations**

The comprehensive speech-language pathology evaluation will vary amongst clinicians. The following points are intended to assist the evidence-based clinician shape the assessment plan in consideration of the literature.

Clinical examination of the oropharyngeal swallow is an essential start. The gross impact of the action myoclonus and medication effects on feeding and swallowing must be considered prior to a potentially complicated transport to a radiology suite (videofluoroscopic swallow study) or more invasive nasendoscopy (fiberoptic endoscopic evaluation of swallowing). Severe onset of jerking movements while eating and drinking may impede a clinician's ability to capture a meaningful instrumental assessment. The clinical exam can also provide opportunity to trial optimal posturing and plans for feeding during an anticipated instrumental assessment. Medication changes may be made frequently, and clinical re-assessment will be necessary.

Assessment of cognitive-communication skills should consider medication effects, medication changes, and the action myoclonus itself. As mentioned previously, many drugs have sedating effects and may directly contribute to inattention. Selection and administration of standardized measures must consider the difficulties that movement of the upper extremities pose (e.g., trail-making task).

An interdisciplinary approach will be essential to maximizing this patient's outcomes. Referral to and close collaboration with allied rehabilitation providers should be of high priority. The American Speech-Language-Hearing Association supports our role as "Case Manager" in individuals with cognitive-communication impairments (ASHA, 2005). The speech-language pathologist may assist with establishing the team and the effective communication amongst the following essential members. Specific implications for the speech-language pathologist are proposed:

- *Physicians*. Close collaboration will clarify medication effects and changes. This may incorporate *Pharmacists.* Plans for the patient's disposition, ordered by the physician, must consider all team members' observations.
- *Occupational Therapists*. Adaptive feeding and writing devices will directly affect the speech-

language pathologist's assessments and interventions.

- *Physical Therapists*. Optimal positioning recommendations and management of action myoclonus of the upper extremities may improve ability to capture meaningful evaluations (e.g., videofluoroscopic swallow study).
- *Mental Health Team/Social Workers*. Establishing substance abuse treatment is essential to maximize long-term success. Counseling should consider the transition into a sudden, permanent disability and its effect on mental well-being. ♦

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