Posthypoxic action myoclonus (the Lance-Adams syndrome)

Christopher Marcellino [1], Eelco FM Wijdicks [2]

[1]Departments of Neurology and Neurologic Surgery, Mayo Clinic, Rochester, Minnesota, USA
[2]Neurology, Mayo Clinic College of Medicine and Science, Rochester, Minnesota, USA

Correspondence to
Dr Eelco FM Wijdicks; wijde@mayo.edu

Accepted 26 March 2020

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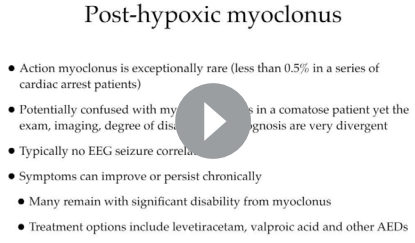
To cite: Marcellino C, Wijdicks EFM. *BMJ Case Rep* 2020;**13**:e234332. doi:10.1136/bcr-2020-234332

DESCRIPTION

A 62-year-old man was resuscitated after an in-hospital pulseless electrical activity (PEA) cardiac arrest. He awakened after 24 hours but after another 24-hour interval, he had worsening action and stimulus-induced myoclonus.[1–3] He was able to relax fully without movement but any volitional movement initiated a burst of appendicular arrhythmic jerks (video 1). The disabling myoclonus was significantly muted with levetiracetam and valproate. No cerebellar lesions were found on CT or MRI.

Action myoclonus is exceptionally rare (less than 0.5% in a series of patients who have a cardiac arrest) and is potentially confused with myoclonus status in a comatose patient, yet the examination, imaging, degree of disability, time of onset and prognosis are very divergent.[4] Typically, there is no electroencephalogram (EEG) seizure correlate.[3] The symptoms can improve or persist chronically, and many remain with significant disability from myoclonus. Medication treatment options include levetiracetam, valproic acid and other anti-epileptic drugs (AEDs).



video 1. Pretreatment and posttreatment demonstration of subacute action myoclonus after cardiac arrest, with narration and reference text. The disabling myoclonus was completely muted with levetiracetam and valproate within an additional 72 hours. No acute findings were found on CT or MRI, and had mild confusion but no other focal deficits. Myoclonus occurring after hypoxic brain injury from cardiac arrest, characterised by abrupt irregular muscle contractions. (1) Acute: starting within 48 hours after the arrest (when isolated, sometimes terms acute Lance-Adams syndrome). (2) Chronic: Lance-Adams syndrome, which may start from days to weeks after arrest and progressively worsen, with or without other neurological symptoms.

Learning points

- Myoclonus occurring after hypoxic brain injury from cardiac arrest, characterised by abrupt irregular muscle contractions. (1) Acute: starting within 48 hours after the arrest (when isolated, sometimes terms acute Lance-Adams syndrome). (2) Chronic: Lance-Adams syndrome, which may start from days to weeks after arrest and progressively worsen, with or without other neurological symptoms.
- Potentially confused with myoclonus status in a comatose patient, yet the examination, imaging, degree of disability and prognosis are very divergent.
- Typically, no EEG seizure correlates.

Twitter Christopher Marcellino @chrismarcellino

Contributors CM: draft of the manuscript, clinical care of the patient, video recording and editing. EFMW: revised the manuscript for intellectual content, clinical care of the patient. EFMW is also a current BMJ fellowship subscriber.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Next of kin consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

ORCID iDs
Christopher Marcellino http://orcid.org/0000-0002-8046-5902
Eelco FM Wijdicks http://orcid.org/0000-0001-9807-9172

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