

# Intention Myoclonus in Paediatric Patients Following Severe Cardiopulmonary Failure: A Report of Three Cases

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## ABSTRACT

We describe the development of intention myoclonus following severe cardiopulmonary failure in three paediatric patients. Symptoms occurred during the withdrawal of midazolam and fentanyl, which were used for prolonged sedation, and resolved spontaneously. Because the three patients had concomitant brain injury secondary to cardiopulmonary failure-related hypoxia, we propose that the combination of hypoxic brain injury and sedative withdrawal may predispose to intention myoclonus. (**Critical Care and Resuscitation 2002; 4: 104-106**)

**Key words:** Intention myoclonus, withdrawal syndrome, cerebral hypoxia, child

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Neurological complications are an infrequent but significant problem in the paediatric intensive care unit. We encountered three patients that demonstrated intention myoclonus upon discontinuation of prolonged, continuous intravenous sedation with midazolam and fentanyl. All patients had concomitant hypoxic brain injury secondary to severe cardiopulmonary failure, suggesting a role of hypoxic injury in the pathogenesis of this neurological disorder.

## CASE REPORTS

### *Patient 1*

A 7-day-old female was admitted to the paediatric intensive care unit (PICU) with right ventricular failure due to congenital pulmonary stenosis. The patient underwent mechanical ventilation with midazolam and fentanyl used for continuous sedation. During management, the patient suffered severe transient circulatory failure due to a pericardial tamponade with the mean

arterial blood pressure (MAP) decreasing to < 30mmHg. Following surgical correction of the tamponade and stabilisation of the cardiovascular status, the patient remained mechanically ventilated. Sedation with midazolam and fentanyl was employed for a total of 23 days. Seven days after the discontinuation of sedation, the patient developed limb tremors with touching stimulus. The tremors subsequently extended to the entire body and persisted for several minutes. These involuntary movements persisted for 13 days before resolving spontaneously.

### *Patient 2*

A 10-month-old male was administered to the PICU following bi-directional cavopulmonary shunt surgery. During weaning from cardiopulmonary bypass the patient experienced severe hypoxia (SpO<sub>2</sub> < 50 %) and systemic hypotension (MAP < 30mmHg). Postoperatively, the patient underwent mechanical ventilation using fentanyl, midazolam and vecuronium for continuous

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sedation for a total of 13 days. One day following withdrawal of these agents, the patient demonstrated limb and eye tremors triggered by changes in the patient's position. Computed tomography imaging revealed diffuse brain atrophy, and single photon emission computed tomography (SPECT) imaging demonstrated hypoperfusion of the right frontal lobe. These involuntary movements spontaneously remitted after 2 months.

#### Patient 3

A 2-year-old female was admitted to the PICU following cardiopulmonary resuscitation for apnea, severe cyanosis and bradycardia due to acute respiratory failure from bacterial pneumonia. The patient underwent mechanical ventilation using fentanyl, midazolam and vecuronium for a total of 9 days. One day after the withdrawal of sedatives and muscle relaxant, the patient developed tremors, limb rigidity and ocular convergence triggered by crying. These involuntary movements recurred with a frequency of once every twenty minutes. Computed tomography imaging demonstrated progressive brain atrophy, and SPECT revealed decreases in blood flow to the frontal lobes, right parietal lobe and grey matter. The involuntary movements spontaneously remitted after 9 days, but mild general muscular rigidity persisted.

#### DISCUSSION

In the three cases described, the prolonged and continuous administration of intravenous midazolam and fentanyl may have been responsible for the development of a "withdrawal syndrome".<sup>1,2</sup> The total amounts of midazolam and fentanyl in these patients were particularly high, ranging from 37 - 39 mg/kg and 99 - 700 µg/kg, respectively (table 1) and were abruptly discontinued.

**Table 1. The duration and dose of the sedative agents**

Patient	1	2	3
Age	7 d	0.83 yr	2 yr
Weight (kg)	3.2	5.4	6.7
Duration of sedation (days)	23	13	9
Total sedative dose:			
midazolam (mg/kg)	37	39	37
fentanyl (µg/kg)	99	771	139

However, the neurological findings in these patients were limited to intention myoclonus (i.e. an intermittent syndrome of unidirectional contractions and relaxations of a group of muscles triggered by stimulation such as

light, sound and touch). As other withdrawal symptoms were not present, we believe that these symptoms were not typical of a "withdrawal syndrome".<sup>1,2</sup> As all three patients had a history of significant cardiopulmonary failure we believe that hypoxic brain injury probably played a significant role in the pathogenesis of these symptoms. Two of the three patients demonstrated diffuse atrophic/hypoperfusion brain changes in brain image studies performed after the occurrence of the involuntary movement, supporting this hypothesis.

Intention myoclonus after hypoxic brain damage is also known as Lance-Adams syndrome.<sup>3,4</sup> Although there are no previous reports of this phenomenon occurring in children, their symptoms satisfy the clinical criteria described for this syndrome. However, as myoclonic symptoms may be suppressed with sedatives, distinguishing between a withdrawal syndrome and Lance-Adams syndrome in these cases may be difficult. Nevertheless, discriminating between the two entities is useful, as medical therapy differs. For example, withdrawal syndrome is often treated with the administration of benzodiazepines or barbiturates, both of which usually fail to relieve the symptoms associated with intention myoclonus.<sup>3,4</sup> The treatment for Lance-Adams syndrome may include clonazepam, which can increase the level of 5-hydroxyindoleacetic acid (5-HIAA in the cerebrospinal fluid (low cerebrospinal fluid levels of 5-HIAA is often found in patients with intention myoclonus).<sup>5</sup> However, we did not use clonazepam in our patients because it can increase airway secretion retention and hence worsen respiratory failure in children with severe cardiopulmonary disease. Although spontaneous remission from both withdrawal syndrome<sup>1</sup> and Lance-Adams syndrome can be expected,<sup>3,4</sup> an early diagnosis of the specific disorder will allow treatment to be individualised, thereby decreasing the severity and duration of the disorder.

In summary, we describe three paediatric patients who developed transient involuntary movements following long-term sedation in the setting of severe cardiopulmonary insufficiency with brain damage. Intensivists should place particular attention to the appearance of neurological complications in patients with hypoxic brain injury following the discontinuation of long-term sedation.

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