Emergency Department Management of Pediatric Patients Receiving Intrathecal Baclofen

Algorithm 1. Patient presents to ED with questionable intrathecal baclofen WITHDRAWAL

**Inclusion Criteria**
- Patients presenting to the ED with questionable intrathecal baclofen (ITB) withdrawal

**Exclusion Criteria**
- Patients with indwelling pumps delivering any medication other than baclofen
- Patients receiving only oral baclofen
- Patients receiving intrathecal baclofen who present to a department other than the ED

**Symptoms of baclofen withdrawal:**
- Increased tone with or without:
  - Uncontrolled tone despite rescue baclofen doses
  - Fever
  - Pruritus
  - Agitation
  - Increased seizures
  - Rhabdomyolysis

**Criteria for admission:**
- Cardiorespiratory instability
- Uncontrolled tone despite rescue baclofen doses
- Concern for serious infection
- Clear indication of withdrawal after diagnostics and discussion with Rehab On Call provider

**Consider ICU for:**
- Need for IV benzodiazepine
- Increased seizures

**Quick Links**
*Enteral Baclofen Dosing*
*Differential Diagnoses of ITB Withdrawal*
*Sepsis Clinical Pathway*
Algorithm 2. Patient presents to ED with questionable intrathecal baclofen OVERDOSE

Symptoms of baclofen overdose:
- Decreased tone or flaccid paralysis
- Decreased level of consciousness
- Nausea/vomiting
- Hypotension
- Bradycardia, tachycardia, or other cardiac abnormalities
- Respiratory depression
- Seizures

Inclusion Criteria
- Patients presenting to the ED with questionable intrathecal baclofen (ITB) overdose

Exclusion Criteria
- Patients with indwelling pumps delivering any medication other than baclofen
- Patients receiving only oral baclofen
- Patients receiving intrathecal baclofen who present to a department other than the ED

Quick Links
- Differential Diagnoses of ITB Overdose
- Sepsis Clinical Pathway
TARGET POPULATION

Inclusion Criteria

- Patients receiving intrathecal baclofen who present to the Emergency Department

Exclusion Criteria

- Patients with indwelling pumps delivering any medication other than baclofen
- Patients receiving only oral baclofen
- Patients receiving intrathecal baclofen who present to a department other than the Emergency Department

DEFINITIONS

- ITB: intrathecal baclofen

INITIAL EVALUATION

- What other medications or substances are being taken by the patient, how much and when? Are any of these possibly sedating?
- When was the patient’s last ITB pump refill?
- When is the patient’s Low Reservoir Alarm Date or next scheduled refill?
- Are there any audible alarms emanating from the pump?
- If there was initially concern for withdrawal, has the patient taken their rescue oral baclofen dose? Were there any effects from that dose?
- Has the patient been ill or experienced other significant symptoms?
CLINICAL MANAGEMENT OF POTENTIAL INTRATHECAL BACLOFEN WITHDRAWAL (ALGORITHM 1)

Noxious stimuli or infection (e.g. pneumonia, otitis media, constipation, hangnail, pressure injury, etc.) can cause an increase in tone and should be considered prior to diagnosing the patient’s presentation as ITB withdrawal.

Signs | Symptoms of Potential ITB Withdrawal:
- Increased spasticity, dystonia, clonus or spasms
- Fever
- Tachycardia
- Pruritus
- Paresthesia
- Increased seizures
- Agitation, anxiety, irritability
- Rhabdomyolysis

Note: If the patient does not have any of the signs/symptoms listed above, or is found with any potentially noxious stimuli or infection, their condition is most likely not related to ITB withdrawal. The underlying problem needs to be addressed.

Differential Diagnoses of ITB Withdrawal:
- Noxious pain (e.g. fracture, constipation, wound, infection)
- Autonomic dysreflexia (bradycardia with hypertension, lack of increased spasticity)
- Malignant hyperthermia (after anesthesia, familial disorder)
- Serotonergic syndrome (selective serotonin reuptake inhibitor [SSRI] overdose, myoclonus, elevated liver function tests [LTFs])
- Neuroleptic malignant syndrome (use of dopamine blocking neuroleptic drugs or abrupt withdrawal of dopamine agonist)
- Sepsis
- Meningitis

Monitoring
- Place on cardiorespiratory monitors
- Vital signs every 2 hours
- Pain assessment/re-assessment
- Bladder scan every 4 hours and straight catheterization for bladder volume calculated to be greater than or equal to [(patient’s age + 2) x 30 mL] or greater than or equal to 400 mL. An indwelling foley is not recommended in the initial management in order to allow for further monitoring of signs of urinary retention
- If rhabdomyolysis, start IV fluids and monitor renal function and CK

Consults
Contact the Rehabilitation Medicine provider on call for any questions or concerns of diagnostics and management of intrathecal baclofen withdrawal.
CLINICAL MANAGEMENT OF POTENTIAL INTRATHECAL BACLOFEN OVERDOSE (ALGORITHM 2)

The signs/symptoms of ITB overdose are dose dependent on the amount of baclofen being delivered. There is no specific antidote for ITB overdose. If suspicion exists for overdose, contact the Rehabilitation Medicine provider.

**Signs | Symptoms of Potential ITB Overdose:**
If patient does not have any of the signs/symptoms listed below, their condition is most likely not related to ITB.

- Decreased tone or flaccid paralysis
- Decreased level of consciousness
- Nausea/vomiting
- Hypotension
- Bradycardia, tachycardia, or other cardiac abnormalities
- Respiratory depression
- Seizures

**Differential Diagnoses of ITB Overdose:**
- Sepsis
- Increased intracranial pressure (i.e. intracranial bleed, VP shunt malfunction)
- Hypoglycemia
- Electrolyte imbalance
- Overdose of oral baclofen or a sedating medication or substance other than baclofen

**Monitoring**
- Place on cardiorespiratory monitors
- Vital signs every 2 hours
- Pain assessment/re-assessment

**Consults**
If suspicion exists for overdose, contact the Rehabilitation Medicine provider.
THERAPEUTICS

Withdrawal

- Enteral baclofen may be administered as “rescue doses”; monitor for effect. Dosing can be variable from 10-20 mg per dose every 6 hours as needed based on patient response. The following includes dosing stratified by age:
  - 2-7 years old: starting dose 10 mg per dose, then titrate to effect
  - 8 years old or greater: starting dose 20 mg per dose, then titrate to effect
- Diazepam via PO (0.2 to 0.3 mg/kg/dose q6h) or IV (0.04 to 0.3 mg/kg/dose every 2 to 4 hours, Max 0.6 mg/kg, Max 10 mg in a single dose within an 8-hour period) could also be administered for tone reduction.
- Cyproheptadine 4 to 8 mg orally or via G-tube every 6 to 8 hours is effective in symptomatically treating pruritus associated with intrathecal baclofen withdrawal.
- Consider aggressive bowel clean out with administration of softeners and laxatives orally and per rectum if there is suspicion for constipation.

LABORATORY STUDIES | IMAGING

- First-line laboratory studies that are recommended include CK, Comprehensive Metabolic Panel, CBC. High CK levels would not necessarily be diagnostic.
- Second-line laboratory studies that could be considered include CRP, ESR, CSF Cytospin [Cell Count & Differential], and CSF culture, particularly if there is concern for an underlying infection contributing to the presentation. If CSF studies are indicated, a specimen may be obtained via a side-port access performed by a Rehabilitation Medicine provider.
- See the sepsis clinical pathway if concerned for sepsis.

Radiology

- Consider brain imaging to rule out ventriculoperitoneal (VP) shunt malfunction.
- Anteroposterior (AP) and lateral views of the spine allow for visualization of the intrathecal catheter, and determination of whether there has been migration of the catheter tip. Current catheter is not radiopaque at the connector site. Visualization of the bowel with the spine x-ray can also help determine stool burden.
- If a spine x-ray is not obtained, a one-view abdominal x-ray is recommended for determining stool burden and need for an aggressive bowel clean-out since constipation is a common trigger for increased spasticity.
REFERENCES


CLINICAL Pathway

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REVIEW I REVISION SCHEDULE
Scheduled for full review on October 22, 2023

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