Emergency Management of Pediatric Patients Receiving Intrathecal Baclofen

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

1. Patient presents to ED with questionable intrathecal baclofen (ITB) OVERDOSE

   Does patient present with DECREASED tone along with decreased alertness, and/or nausea/vomiting?

   Secure ABCs

   Recent ITB pump refill, dose/concentration change within past week?

   Yes

   Call rehab for further instructions
   • Monitor respiratory status and maintain SPO2 less than 92%
   • **Consider admission if** patient has:
     • Cardiorespiratory instability
     • Altered mental status without improvement for more than 6 hours

   No

   ITB overdose unlikely

   Other signs/symptoms of illness?

   Yes

   Work up for infectious etiology

   No

   History of CSF shunt placement?

   Yes

   If patient presents with HA, AMS, and/or vomiting, evaluate for shunt malfunction:
   • Consider MRI shunt series & XR shunt series
   • Consider Neurosurgery consult

   No

   History of seizure disorder?

   Yes

   If presentation is concerning for seizure, evaluate and consider Neurology consult.

   No

   Consider contacting Rehab for evaluation

   ITB overdose unlikely if patient does not have decreased tone
Algorithm 2. Patient presents to ED with questionable intrathecal baclofen (ITB) WITHDRAWAL

Patient presents to the ED with questionable intrathecal baclofen (ITB) WITHDRAWAL

- Does patient present with increased tone, fever, pruritis and/or agitation?
  - Yes: Other signs/symptoms of illness, change in skin integrity, constipation, or discomfort/pain?
    - Yes: Call Rehab for further recommendations on diagnostics and management
    - No: Not likely ITB related. Work-up for presenting problem.
  - No: Inclusion Criteria

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 dept other than the ED/UC

- Noxious stimuli will increase tone.
- Treat underlying problem.
- Patient may take 10 to 20 mg of enteral baclofen every 6 hours as needed for comfort until resolved.

- Criteria for admission:
  - Cardiorespiratory instability
  - Uncontrolled tone despite rescue baclofen doses
  - Need for IV tone management
  - Concern for serious infection
  - Clear indication of withdrawl after diagnostics and discussion with Rehab provider

- Has the patient/family heard the ITB alarm?
  - Yes: Call Rebab for ITB pump interrogation. Pump either needs medication refill or is malfunctioning.
  - No: Has the family given any supplemental baclofen or diazepam?

- improves with supplemental baclofen?
  - Yes: Continue supplemental baclofen every 6 hours
  - No: Administer 10 to 20 mg of enteral baclofen and watch for improvement (30 mins to 1 hr)

- Admit for further diagnostic studies and IV tone management medications including diazepam and/or midazolam. Draw CK. Maintain adequate hydration due to risk for rhabdomyolysis. May use diphenhydramine or cyproheptadine for pruritus. Call Rehab for ITB pump evaluation.

- Is patient stable for discharge?
  - Yes: Discharge to home with instructions to use 10 to 20 mg of supplemental baclofen every 6 hours prn. Schedule URGENT Rehab follow-up appointment.
  - No: Continue supplemental baclofen every 6 hours
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TARGET POPULATION

Inclusion Criteria

- Patients receiving intrathecal baclofen

Exclusion Criteria

- Patients with indwelling pumps delivering any medication other than baclofen
- Patients receiving only oral baclofen

TRIAGE CONSIDERATIONS

- 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 recently?
- When was the patient’s last bowel movement?

INITIAL CLINICAL MANAGEMENT

**All Patients** with Potential Intrathecal Baclofen (ITB) Complications

Monitoring

- Place on cardiorespiratory monitors
- Vital signs Q2H
- Pain assessment/re-assessment per local pain assessment and management policy
Fluids, Electrolytes, Nutrition

- Diet: NPO until patient has returned to baseline mentation and is able to tolerate oral or gastrostomy tube intake.
- Maintenance IV fluids are recommended while patient is NPO, and if there is concern for rhabdomyolysis [in acute withdrawal]

INITIAL CLINICAL MANAGEMENT OF POTENTIAL INTRATHECAL BACLOFEN OVERDOSE (ALGORITHM 1)

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 include:

- 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

Laboratory | Radiology

Patients presenting with signs/symptoms of acute illness, such as fever, should be worked up for an infectious etiology per clinician’s history and exam.

Consults

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

INITIAL CLINICAL MANAGEMENT OF POTENTIAL INTRATHECAL BACLOFEN WITHDRAWAL (ALGORITHM 2)

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\(^1\):

- Increased spasticity\(^4\), dystonia, clonus or spasms
- Fever leading to hyperthermia
- Tachycardia
- Pruritus
- Paresthesias
- Seizures
- Agitation, anxiety, irritability
- History of withdrawal symptoms close to scheduled refill date
- 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 include\(^1\):

- Autonomic dysreflexia (bradycardia with hypertension, lack of increased spasticity)
- Malignant hyperthermia (after anesthesia, familial disorder)
- Serotoninergic 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 (In addition to monitoring mentioned on page 1)**

- Bladder scan Q4H and straight catheterization for bladder volume calculated to be greater than or equal to \([(patient’s age + 2) \times 30 \text{ mL}] \) 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.*

**Consults**

Contact the Rehabilitation Medicine provider on call for any questions or concerns of diagnostics and management of intrathecal baclofen withdrawal.

**THERAPEUTICS**

- Oral baclofen 10 to 20 mg q6h may be administered as “rescue doses”; monitor for effect.
- Diazepam via PO (0.3 to 0.2 mg/kg/dose q6h) or IV (0.04 to 0.3 mg/kg/dose every 2 to 4 hours, Max 0.6 mg/kg 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.*

Radiology

- Patients presenting with signs/symptoms of acute illness, such as fever, should be worked up for an infectious etiology.

- 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 XR 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


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