

Medication Considerations on Next Page



Reassess the appropriateness of Care Guidelines as condition changes and 24 hrs after admission. This guideline is a tool to aid clinical decision making. It is not a standard of care. The physician should deviate from the guideline when clinical judgment so indicates.



Medication Considerations

Benzodiazepines: Recommend aggressive dosing to adequately control movements

- Clonazepam: 0.125mg PO/Gtube BID-TID, increase by 0.25 0.5mg/day q3 days. Max: 20 mg/day
- Diazepam (preferred due to rapid onset of action): 0.2mg/kg/dose IV q4hr PRN, increase up to 10mg q4h IV PRN or scheduled
- Midazolam:
 - Intranasal 0.2mg/kg/dose may increase to 0.8mg/kg/dose for benzodiazepine tolerate patients. Max 10mg/dose. May repeat x1 after 10 minutes.
- Lorazepam: 0.1mg/kg IV q2hr PRN. Max 4mg/dose.
- Flumazenil: available for rescue

Special considerations:

- These patients are often benzodiazepine tolerant and hence may require high doses to achieve therapeutic effect, this
 also limits side effects.
- Respiratory depression and sedation are side effects to monitor for. If decreasing respiratory rate and oxygen saturation consider monitoring of end-tidal CO2 to assess hypoventilation, particularly if giving supplemental oxygen. If concerns for respiratory symptomatology consider transfer to PICU for positive pressure ventilation therapy and possible intubation as indicated.
 - Avoid neuroleptics without discussion with movement team. If given add antimuscarinic.
 - o Consider prolactin level prior to administration
 - Titration of medication or progression to next step is based on need to abate movements either due to risk of patient harming themselves or others, or dystonic storm (as assessed by trending CK)
 - o If persistent concerns at maximum dosing of clonazepam/diazepam escalate to ICU for continuous infusions

For consideration in ICU (in order of recommendation):

- Midazolam: 0.03 0.2 mg/kg/hr IV continuous infusion
- Dexmedetomidine: 1.5 mcg/kg/hr IV (titrating to 2mcg/kg/hr)
- Ketamine: 0.5 2mg/kg/dose IV x1, then 5 60mcg/kg/min IV
- Propofol: 150 mcg/kg/min, titrating to 250 mcg/kg/min

Triggers to consider: pain, constipation, infection, headaches.

• May require dental evaluation, KUB, skeletal survey, etc.

References

- Allen, N. M., Lin, J. P., Lynch, T. & King, M. D. Status dystonicus: a practice guide. *Developmental Medicine & Child Neurology* 56, 105–112 (2014). (Level V)
- Goswami, J. N., Roy, S. & Patnaik, S. K. Pediatric Dystonic Storm. *Neurology: Clinical Practice* **11**, e645–e653 (2021). (Level IV)
- Kay, L. *et al.* Intranasal midazolam as first-line in hospital treatment for status epilepticus: a pharmaco-EEG cohort study. *Ann Clin Transl Neurol* 6, 2413–2425 (2019). (Level IV)
- Luc, Q. N. & Querubin, J. Clinical Management of Dystonia in Childhood. *Pediatric Drugs 2017 19:5* 19, 447–461 (2017). (Level V)
- Termsarasab, P. & Frucht, S. J. Dystonic storm: a practical clinical and video review. *Journal of Clinical Movement Disorders* **4**, 1–8 (2017). (Level V)