

Appendix 4

Pharmacological therapy

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Drug	Mechanism	Dose	Clinical Effect	Side Effects
Morfine	Opioid agonist	<u>Bolus (EV)</u> – 2 to 8 mg	<u>Abortive</u> Improvement of most symptoms.	- Respiratory depression - Sedation - Hypotension - Ileus - Emesis - Tolerance
Propofol	Agonist GABA-A	<u>Bolus (EV)</u> – 10 to 20 mg <u>Continuous perfusion (EV)</u> ≤80 mcg/kg/min	<u>Abortive</u> - Bolus <u>Preventive</u> - Continuous perfusion Improvement of most symptoms.	- Deep sedation - Propofol infusion syndrome
Benzodiazepines	Agonist GABA-A	<u>Bolus (EV):</u> - Diazepam 5 to 10 mg - Lorazepam 1 to 4 mg - Midazolam 2 to 5 mg <u>Enteric route:</u> - Clonazepam 0,5 to 2 mg every 8 hours	<u>Abortive</u> - Diazepam - Lorazepam - Midazolam <u>Preventive</u> - Clonazepam Diazepam → Useful in hypertonicity Improves most symptoms. Less effective than morphine.	- Sedation - Respiratory depression - Tolerance
Dexmedetomidine	Alpha-2 adrenergic receptor agonist	<u>IV perfusion</u> - 0.2 to 1.5 mcg/kg/h	<u>Abortive and preventive</u> Improves mostly hypertension and tachycardia.	- Bradycardia - Hypotension - Sedation
Clonidine	Alpha-2 adrenergic receptor agonist	Enteric route - 0.1 to 0.3 mg every 6/8 hours	<u>Abortive and preventive</u> Improves mostly hypertension and tachycardia.	- Bradycardia - Hypotension - Sedation
Propranolol	Beta blocker non-cardioselective	Enteric route - 20 to 80 mg every 6/8 hours	<u>Preventive</u> Improves tachycardia, hypertension and diaphoresis. Possible improvement in dystonia. Less effect on hyperthermia	- Bradycardia - Hypotension - Sleep disorders

Gabapentin	Interaction with the alpha-2-delta subunit of voltage dependent calcium channels in the dorsal horn of the spinal cord	Enteric route - 100 to 300 mg every 8 hours (Maximum daily dose - 3600 to 4800 mg)	<u>Preventive</u> Most symptoms improved	- Light sedation
Bromocriptine	D2 dopamine receptor agonist	Enteric route - 1.25 mg every 12 hours (Maximum daily dose - 20 to 40 mg)	<u>Preventive</u> Modest and delayed effects	- Confusion - Agitation - Dyskinesias - Nausea and emesis - Orthostatic hypotension - Decreased seizure threshold
Baclofen	GABA-B agonist	Enteric route - 5 mg every 8 hours (Maximum daily dose - 80 mg. Intrathecal route possible)	<u>Preventive</u> Improving hypertonicity and dystonia	- Sedation - Muscle weakness - Risk of deprivation
Dantrolene	Ryanodine receptor interaction in myocytes	- IV - 0.5 to 2 mg/kg every 6-12 hours (Maximum daily dose - 10 mg/kg)	<u>Abortive</u> Improving hypertonicity and dystonia	- Hepatotoxicity - Respiratory depression - Muscle weakness