Paroxysmal Sympathetic Hyperactivity

Clinical Practice Guideline

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Clinical features consistent with PSH and PSH-AM ≥8? (see Appendix A)

no → Alternative diagnoses ruled out? (see Appendix B)

yes → Nursing to document CFS scores Q-shift and with acute episodes

no → Address alternative diagnosis

yes → Alternative diagnoses ruled out? (see Appendix B)

no → Address alternative diagnosis

yes → PSH-AM ≥ 8?

yes →

Non-pharmacologic interventions:

- Reduce stimulation level in patient room
- Check skin for wounds
- Reposition patient for comfort
- Reposition medical devices
- Rule out urinary retention and constipation

Other Measures:

- Nursing to document CFS scores Qshift and with acute episodes
- Primary team to document PSH-AM scores in daily note with smartphrase ".PSHSCORE"
- Consider checking CK level with prolonged posturing
- Consider expert consultation
  - New dx: PM&R or Neurology
  - Chronic dx: Primary outpatient managing team (PM&R, Complex Care, Palliative Care)

PSH symptoms continue?

Consider targeted pharmacotherapy towards primary symptom. (see Appendix C for dosing)

Targeted Pharmacotherapy

Primary Symptoms	Increased tone/posturing	Vital sign abnormalities	Pain control
1st line medication:	Baclofen	Propranolol	Gabapentin
2nd line medication:	Diazepam (PO/IV)	Clonidine (enteral or cutaneous patch)	Opioids (oxycodone enteral or morphine IV)
3rd line medication:	Gabapentin	Diazepam (PO/IV)	----

- Refractory patients may require consideration for dexmedetomidine bolus or infusion (ICU only).
- If chronic PSH patient, consider escalation of existing medications first.
- Ensure that PRN orders specify what symptom the PRN is to be used for.

If PSH symptoms not improved despite 2 pharmacologic measures and/or 48h duration (whichever comes first), consider expert consultation if not already involved

Requiring re-assessment or PSH therapy > Q2H and on floor?

Yes → **Consider ICU RRT**

Prior to Discharge:

- Needs outpatient follow-up with PM&R if new patient, or with primary outpatient managing team (PM&R, complex care, palliative care) if chronic patient.
- Update chart (problem list) with medications (particularly PRNs) that were effective in managing patient's PSH during admission, for future references.
- Add diagnosis "Sympathetic Storming" (ICD-10 code G90.8) to problem list if not already present.

Appendix A – PSH-AM Scoring Criteria

Clinical Feature Scale (CFS):

	0	1	2	3
Heart Rate	1-4 years: < 100 5-15 years: < 100 16+ years: < 100	1-4 years: 110-124 5-15 years: 100-119 16+ years: 100-119	1-4 years: 125-139 5-15 years: 120-139 16+ years: 120-139	1-4 years: ≥ 140 5-15 years: ≥ 140 16+ years: ≥ 140
Respiratory Rate	1-4 years: < 30 5-15 years: < 25 16+ years: < 18	1-4 years: 30-34 5-15 years: 25-29 16+ years: 18-23	1-4 years: 35-39 5-15 years: 30-34 16+ years: 24-29	1-4 years: ≥ 40 5-15 years: ≥ 35 16+ years: ≥ 30
Systolic Blood Pressure	1-4 years: < 100 5-15 years: < 120 16+ years: < 140	1-4 years: 100-109 5-15 years: 120-129 16+ years: 140-159	1-4 years: 110-119 5-15 years: 130-139 16+ years: 160-179	1-4 years: ≥ 120 5-15 years: ≥ 180 16+ years: ≥ 140
Temperature	< 37 C	37-37.9 C	38-38.9 C	≥ 39C
Diaphoresis	Normal	Mild	Moderate	Severe
Posturing During Episodes	Absent	Mild	Moderate	Severe

Diaphoresis:

Mild – moist or glistening skin
Moderate – beads of sweat
Severe – profuse sweating

Posturing:

Mild – hypertonicity increases but the tone is easily overcome
Moderate – hypertonicity and the tone is hard to overcome
Severe – inescapable hypertonicity

Diagnosis Likelihood Tool (DLT): one point per feature present

- Antecedent acquired brain injury
- Clinical features occur simultaneously
- Episodes are paroxysmal in nature
- Sympathetic over-reactivity to normally non-noxious stimuli
- Absence of parasympathetic features during episodes
- Features persist for > 3 consecutive days
- Features persist for > 2 weeks post brain injury
- 2 or more episodes daily
- Absence of other presumed causes of features
- Features persist despite treatment of alternative differential diagnoses
- Medication administered to decrease sympathetic features

CFS scoring is intended to be completed by nursing staff and recorded in flowsheet, while DLT and overall assessment are to be completed by the physician.

PSH-AM = CFS subtotal + DLT subtotal

PSH-AM score:

< 8 = PSH unlikely, 8-16 = PSH possible, > 16 = PSH probable

- **Appendix B – Alternative Diagnoses:**

- Manifestations of paroxysmal sympathetic hyperactivity have significant overlap with other clinical diagnoses, so alternative or concurrent conditions must be considered, and providers should use clinical judgment in performing appropriate diagnostic tests based on the individual patient. One should also consider that the extent of exclusion is less necessary in a patient with known history of paroxysmal sympathetic hyperactivity.

- Including but not limited to:
  - Infectious process (bacteremia, meningitis, etc.)
  - Non-PSH neurological deterioration (increased ICP, intracranial hemorrhage, intracranial edema, seizures, encephalitis)
  - Pulmonary embolism
  - Thyrotoxicosis
  - Alcohol or drug withdrawal
  - Neuroleptic malignant syndrome
  - Serotonin syndrome
  - Malignant hyperthermia

Appendix C - Dosing Information for Pharmacologic Agents:

Baclofen (enteral)

- Starting maintenance dose: 2.5 mg/dose TID (< 8 yo), 5 mg/dose TID (> 8 yo)
- PRN regimen: 2.5-5 mg Q6H PRN (within bounds of max daily dose)
- Target total daily dose (combined maintenance and PRN): 10-20 mg/day (< 2 yo), 20-40 mg/day (> 2 yo)
- Daily maximum dose (combined maintenance and PRN): 40 mg/day (< 2 yo), 60 mg/day (> 2 yo)
- Side effects to monitor: drowsiness, confusion, hypotonia, nausea/vomiting, hypotension, seizure
- Lab monitoring needed: none
- Other notes: If maintenance therapy is started and then later stopped, this medication should be tapered off over time rather than abruptly discontinued to avoid withdrawal symptoms (including tachycardia, pruritus, hypertonia, seizures).

Diazepam (enteral/IV have same dosing)

- Starting maintenance dose: 0.1-0.2 mg/kg/dose every 6-8 hours up to a maximum dose of 10 mg per dose.
- PRN regimen: 0.1-0.2 mg/kg/dose Q12H PRN (maximum single dose 10 mg, stay within bounds of max daily dose).
- Target total daily dose (combined maintenance and PRN): N/A, titrate to clinical improvement
- Daily maximum dose (combined maintenance and PRN): 0.8 mg/kg/day or 40 mg/day
- Side effects to monitor: drowsiness, confusion, hypotension, bradycardia (at higher doses)
- Lab monitoring needed: none during short term inpatient use
- Other notes: If maintenance therapy is started and then later stopped, this medication should be tapered off over time rather than abruptly discontinued to avoid withdrawal.

Gabapentin (enteral)

- Starting maintenance dose: 5 mg/kg/dose TID (< 12 yo), 300 mg/dose TID (> 12 yo)
- PRN regimen: 5 mg/kg/dose Q12H PRN (< 12 yo, within bounds of max daily dose), 300 mg/dose Q12H PRN (> 12 yo, within bounds of max daily dose)
- Target total daily dose (combined maintenance and PRN): Titrate to clinical improvement, but well tolerated regimens are 30-40 mg/kg/day (< 12 yo), 900-1800 mg/day (> 12 yo)
- Daily maximum dose (combined maintenance and PRN): 50 mg/kg/day (< 12 yo), 3600 mg/day (> 12 yo)
- Side effects to monitor: drowsiness, dizziness, ataxia, peripheral edema (adolescents)
- Lab monitoring needed: none
- Other notes: Dosing needs to be adjusted with renal impairment. Although withdrawal symptoms are mild, they can occur with abrupt cessation of long term therapy.

Propranolol (enteral)

- Starting maintenance dose: 1 mg/kg/day divided TID
- PRN regimen: Not to be used PRN, consider other option such as clonidine
- Daily maximum dose: 4 mg/kg/day
- Side effects to monitor: respiratory infections, constipation/diarrhea, agitation, drowsiness, bradycardia, hypotension
- Lab monitoring needed: none
- Other notes: Use with caution in patients with compromised cardiac function.

Clonidine (enteral, immediate release)

- Starting maintenance dose: 5-10 mcg/kg/day divided Q8H to Q12H
- PRN regimen: 2-5 mcg/kg/dose Q6H to Q8H PRN (within bounds of max daily dose)
- Target total daily dose (combined maintenance and PRN): 5-25 mcg/kg/day
- Daily maximum dose (combined maintenance and PRN): 0.9 mg/day
- Side effects to monitor: drowsiness, dizziness, headache, hypotension, bradycardia, prolonged QT, dry mouth
- Lab monitoring needed: none
- Other notes: Use with caution in patients with compromised cardiac function. Bradycardia, sedation, and hypotension may also be more likely in patients with renal failure.

Clonidine (patch)

- Transdermal dose/24 hours approximately equivalent to the total enteral daily dose (above) may be used, exchange patch every 5-7 days.

Oxycodone (enteral)

- Starting maintenance and PRN doses: 0.025-0.05 mg/kg/dose Q4H to Q6H (< 6 mo), 0.1-0.2 mg/kg/dose Q4H to Q6H (> 6 mo and < 50 kg), 5-10 mg Q4H to Q6H (> 50 kg). This regimen can be used scheduled or PRN.
- Daily maximum dose (combined maintenance and PRN): N/A, discuss with pharmacy/expert consultants if uptitration beyond starting dose is required.
- Side effects to monitor: drowsiness, constipation, headache, nausea, dependency with chronic use
- Lab monitoring needed: none
- Other notes: If maintenance therapy is started and then later stopped (duration > 5 days), this medication should be tapered off over time rather than abruptly discontinued to avoid withdrawal.

Morphine (IV)

- Starting PRN dose: 0.025-0.05 mg/kg every 2-4 hours PRN (up to 2-4 mg/dose every 2-4 hours) Use within bounds of max daily dose.
- Target total daily dose (combined maintenance and PRN): N/A, titrate to clinical improvement
- Daily maximum dose: 0.1 mg/kg every 2-4 hours (up to 8 mg every 2 to 4 hours)
- Side effects to monitor: drowsiness, constipation, headache, nausea, dependency with chronic use
- Lab monitoring needed: none
- Other notes: If maintenance therapy is started and then later stopped (duration > 5 days), this medication should be tapered off over time rather than abruptly discontinued to avoid withdrawal.