Paroxysmal Autonomic Instability with Dystonia
PAID Syndrome

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Paroxysmal Autonomic Instability with Dystonia
PAID Syndrome

Synonyms:
- Autonomic Crisis (Storm)
- Cerebrally Mediated Autonomic Dysfunction
- Paroxysmal Dysautonomia
- Paroxysmal sympathetic hyperactivity
- Mesencephalic Seizures
- Midbrain dysregulatory syndrome

• PAID: Blackman et al in Arch Neurol, 61, March 2004
Definition

- Syndrome of Paroxysmal autonomic hyperactivity
  - Increased heart rate, respiratory rate, BP, temperature and sweating
  - Dystonia
- Results from a brain lesion
  - Trauma, infection, hemorrhage, infarction, brain tumor, cerebral hypoxia, or degeneration
- Different entity from Autonomic Dysreflexia in SCI
PAID Syndrome

- Hypertension that is constant or fluctuating
- Fever that may range from low grade to high
- Tachycardia and tachypnea
- Pupillary dilation
- Diaphoresis, Agitation
- Dystonia or extensor posturing
- Reported in 10-30% of Brain injuries during acute phase
- Commonly seen after severe injury (low GCS, longer coma duration), brain stem injury or DAI
Additional Symptoms

- Electrocardiographic alterations including arrhythmias
  - Plasma catecholamines elevated significantly
- Increased ICP
- Hypohidrosis
- Subnormal temperature in flaccid limbs
- Neurogenic lung disease

All part of autonomic dysregulation
Diagnostic Criteria

- Blackman et al
- Severe Brain Injury, Rancho Los Amigos level $\leq 4$
- Temperature of at least 38.5°C
- Pulse at least 130/mt
- Respiratory rate at least 140/mt
- Agitation
- Diaphoresis
- Dystonia
- At least 1 cycle per day for at least 3 days
- Other conditions should be ruled out
Pathophysiology

• Loss of central inhibitory regulation of control centers in the medulla.
  • Dysfunction of autonomic centers in diencephalon
  • Loss of connection to cortical, subcortical or brainstem loci that mediate autonomic function
• Thermoregulatory dysfunction due to hypothalamic dysfunction. In part, hypermetabolic state with sustained muscle contraction contributes
• Rigidity and decerebrate posturing due to lesions in midbrain. Nuclei become tonically active
Essentials of Assessment

- Symptom episodes may be unprovoked. Individual episodes may vary in intensity. Episodes may persist for weeks but frequency, duration, and intensity decrease.
- Normal resting vital signs between episodes.
- Resting energy expenditure can be 3 times of norm.
- In Parkinson’s Disease carries worse prognosis.
- Rule out Malignant Neuroleptic syndrome, Serotonin syndrome, Malignant hyperthermia & Thyroid Storm.
- If symptoms are persistent without paroxysmal episodes then dysautonomia unlikely.
Outcomes

- Shorter the episodes, better the outcome
- Response to empirical treatment – better the outcome
- Higher Memory impairment, longer PTA
- No increase in length of ICU stay but longer Rehabilitation stay
- Lower GOS and FIM scores
- Average duration of dysautonomia is about 75 days
Medical management

<table>
<thead>
<tr>
<th>Medication</th>
<th>Classification</th>
<th>Common use</th>
<th>Adverse effect/contraindications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morphine sulfate, fentanyl, oxycodone</td>
<td>Opiate agonist</td>
<td>Analgesia</td>
<td>Sedation</td>
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<tr>
<td>Bromocriptine</td>
<td>Dopamine agonist</td>
<td>Lactation suppression, infertility, prolactin-secreting pituitary tumors</td>
<td>Seizures, use caution in patients with renal or hepatic disease</td>
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<tr>
<td>Carbidopa/levodopa</td>
<td></td>
<td>Parkinson disease</td>
<td></td>
</tr>
<tr>
<td>Propranolol</td>
<td>Nonselective β-adrenergic antagonist</td>
<td>Hypertension</td>
<td>Hypotension, use caution in patients with asthma or bronchial disease</td>
</tr>
<tr>
<td>Clonidine</td>
<td>α₂-adrenergic agonist</td>
<td>Hypertension</td>
<td></td>
</tr>
<tr>
<td>Labetalol</td>
<td>Nonselective β agonist, Selective α₁ adrenergic antagonist</td>
<td>Hypertension</td>
<td></td>
</tr>
<tr>
<td>Midazolam, diazepam, clonazepam</td>
<td>GABA-A agonist</td>
<td>Sedation</td>
<td>Sedation</td>
</tr>
<tr>
<td>Baclofen (oral/intrathecal)</td>
<td>GABA-B agonist</td>
<td>Spasms</td>
<td>Sedation</td>
</tr>
<tr>
<td>Chlorpromazine</td>
<td>Dopamine antagonist</td>
<td>Malignant hyperthermia</td>
<td>Sedation, lowers seizure threshold, extrapyramidal side effects</td>
</tr>
<tr>
<td>Phenytoin, carbamazepine</td>
<td>Anticonvulsant</td>
<td>Seizures</td>
<td>Sedation</td>
</tr>
<tr>
<td>Dantrolene</td>
<td>Other</td>
<td>Muscle relaxation</td>
<td>Hepatic disease</td>
</tr>
</tbody>
</table>

Abbreviation: GABA, γ-aminobutyric acid.
Management and Interventions

• For Bruxism and Opisthhotonus, Botulinum toxin injection into key muscles such as masseters, temporalis and paraspinals is indicated
• Dental consultation for mouth-piece
• Resting splints. PROM. Frequent turning
• Prevent noxious stimuli
• Intrathecal Baclofen. Continuous infusion via external pump