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Hunter Syndrome (MPS II)

Emergency Quick Guide for First Responders

What is Hunter Syndrome?

Hunter syndrome (MPS II) is a rare genetic disorder where the body cannot properly break down certain sugars (glycosaminoglycans). These build up over time and affect multiple systems in the body.

Patients may appear stable but can have complex, hidden medical risks—especially in emergency situations.

How It May Present:

Individuals with Hunter syndrome may have a wide range of symptoms and abilities. First responders may notice:

- Facial features: Coarse facial features, enlarged tongue, or thickened lips
- Speech & communication differences: Some individuals may be nonverbal or have limited speech
- Behavioral presentation:
    - May appear anxious, overwhelmed, or resistant in unfamiliar or high-stress situations
    - Possible hyperactivity or impulsivity
    - Difficulty following directions or processing information quickly
- Cognitive differences: Ranging from typical cognition to developmental delay
- Hearing loss: May impact ability to respond to verbal commands
- Mobility limitations: Joint stiffness, difficulty walking, or reduced range of motion

Important Note: Behavioral responses are often a result of communication challenges, sensory sensitivities, or fear not noncompliance.

Key Emergency Considerations:

Airway – HIGH RISK

- Difficult airway is common
- Thickened tissues, large tongue, and narrowed airway
- Limited neck mobility and cervical spine concerns

*Intubation may be difficult—prepare for advanced airway support
Consider smaller tube size than expected

Cardiac Concerns

- Many patients have heart valve disease or cardiomyopathy
- May not tolerate stress or fluid shifts well

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*Monitor closely for cardiac instability

Neurological

- Some patients may have developmental delays or cognitive impairment
- Seizures may occur in some individuals

*Communication may be limited—caregivers are key

Spine & Mobility

- Cervical spine instability possible
- Joint stiffness and skeletal abnormalities

*Use caution with neck movement and positioning

Respiratory

- Chronic airway disease is common
- Higher risk of respiratory distress or obstruction

*Monitor oxygenation closely

Medications & Treatment:

- Some patients receive enzyme replacement therapy (ERT) or other treatments
- Ask caregivers about:
    - Current medications
    - Recent infusions or treatment

Caregiver Guidance is Critical:

- Parents/caregivers often know the patient's needs best
- Ask:
    - "What works best in an emergency?"
    - "Have there been airway or anesthesia challenges before?"

Transport Considerations:

- Transport to a facility familiar with complex or rare conditions if possible
- Early communication with receiving hospital is recommended

When in Doubt: Treat as a medically complex patient with high airway risk

Why This Matters: Hunter syndrome is rare, and many providers may not encounter it often. Quick recognition of these risks can prevent complications and save lives.