



MPS EMERGENCY: ACT IMMEDIATELY



INITIAL ACTIONS:

Immediate Emergency Response

- Check Airway, Breathing, Circulation (ABC)
- Monitor oxygen, pulse, blood pressure
- Provide immediate support as needed

Breathing Difficulties

- Sit upright
- Clear secretions
- Oxygen or inhalers
- Prepare airway support

Seizures

- Protect from injury
- Do not restrain
- Nothing in mouth
- Give prescribed rescue medication
- Monitor vital signs

Cardiac Issues

- Check vital signs immediately
- Monitor for cyanosis, swelling, breathlessness
- Follow advanced life support if unstable

KEY CLINICAL REMINDERS:

- Check Health Passport / MPS Toolkit
- Review anaesthetic history
- Anticipate difficult airway
- Involve specialist teams early

HIGH RISK WARNINGS:

Airway/Anesthesia Risk

- Narrow airway / short neck / limited neck movement
- Enlarged tonsils/adenoids
- Tracheal narrowing
- High risk of obstruction with sedation

Airway Management

- Avoid blind intubation
- Early anaesthetic involvement required
- Use advanced airway equipment (fiberoptic / LMA)
- Previous airway records are essential
- Maintain oxygenation at all times

Infusion Therapy (ERT)

- Maintain scheduled ERT where possible
- Do not abruptly interrupt
- Watch for: fever, rash, hypotension, respiratory distress
- Document last dose + date

Systemic Risks

- Cervical spine instability / scoliosis
- Joint stiffness → positioning risk
- Valvular disease / cardiomyopathy
- Respiratory infections may deteriorate rapidly
- Handle carefully during transfers