UHealth
UNIVERSITY OF MIAMI HEALTH SYSTEM

MILLER
SCHOOL OF MEDICINE
UNIVERSITY OF MIAMI

EMERGENCY REGIMEN

Patient label

Primary Geneticist: _____

On call UM Geneticist Tel: 305-331-3023

Protocol seen and approved: _____

Mucopolysaccharidsosis Type I

The above named patient has Mucopolysaccharidosis Type I (MPS-I). MPS I is a multi-organ progressive lysosomal storage disorder where complex carbohydrates accumulate in lysosomes throughout the body. Main symptoms include hepatosplenomegaly, dystosis multiplex, joint contractures, hypoplasia of the second vertebrae (dens axis), spinal stenosis and others. Some patients are treated with enzyme replacement therapy and/or early bone marrow transplantation.

If a patient with MPS I presents with severe illness, please immediately notify the on-call UM Geneticist at 305-331-3023. This is a 24/7 service.

Affected patients with MPS I are at greatest risk for severe (irreversible) spinal cord injury during neck extension due to a potential hypoplasia of the dens axis. These patients are also very difficult to intubate when indicated requiring fiber optic intubation. Intubated patients with MPS I should be extubated as quickly as medically justifiable.

Disclaimer: the above recommendations cannot replace an individual medical evaluation by a board certified physician. The UM Geneticist on-call should always be informed. UM is not responsible in case the protocol has not been followed.

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