



PROJECT/**ALIVE**

ANNUAL HEALTH  
MAINTENANCE FOR  
NEURONOPATHIC MPS II FAMILIES

Hunter Syndrome  
Research and Advocacy

March 2021

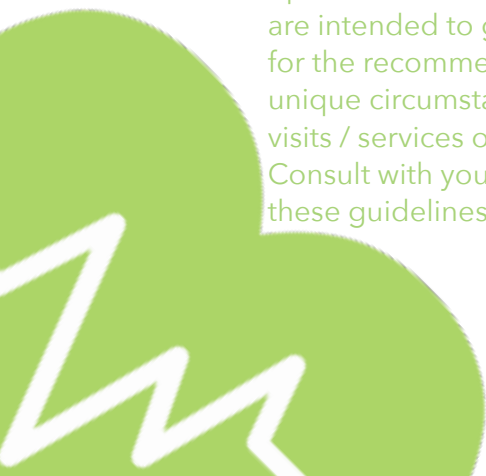
## RECURRING LAB TESTS:

- ☐ Urine GAGs - periodic, based on recommendation of MPS specialist
- ☐ I2S antibody testing - periodic, based on recommendation of MPS specialist (*start after 3 months / 12 infusions of Elaprase ERT*)
- ☐ I2S leukocyte enzyme level - only for children who underwent HSCT, based on recommendation of bone marrow transplant specialist

## RECURRING RADIOLOGY TESTS / PROCEDURES / STUDIES:

- ☐ MRI brain / spine - every 1-3 years, *per MPS specialist or neurosurgeon recommendation*
  - Monitor for hydrocephalus, spinal cord compression
- ☐ Cervical spine flexion / extension x-rays - *per MPS specialist recommendation*
  - Monitor for atlantoaxial (*cervical spine*) instability
- ☐ Orthopedic x-rays including kyphosis series, hip series, etc - *per orthopedist recommendation*
  - Monitor for kyphosis, hip dysplasia, other orthopedic conditions
- ☐ ECG and echocardiogram - every 1-3 years, *per cardiologist recommendation*
  - Evaluate heart valves and function
- ☐ EMG / Nerve conduction studies - every 1-3 years, *per MPS specialist or neurologist recommendation*
  - Evaluate carpal tunnel syndrome, possible tarsal tunnel syndrome
- ☐ Audiology exam - every 6-12 months, *depending on concerns*
  - Assess hearing, adjust hearing aids (*if applicable*)
- ☐ Sleep Study - every 1-3 years
  - Evaluate for obstructive sleep apnea (OSA)
- ☐ Pulmonary function testing - every 1-3 years (*after age 5-6yrs, if child is able to participate*)
- ☐ EEG - as needed, *per neurologist recommendation*
  - Evaluate for seizures

DISCLAIMER: These guidelines are based upon the evidence-based consensus opinions of several MPS II medical experts and experienced MPS II parents. They are intended to guide families as a point of reference, but they do not substitute for the recommendations of your own medical providers. Depending on the unique circumstances of each child or care team, your child may not require all visits / services outlined above, or your child may require others that are not listed. Consult with your child's MPS specialist or primary medical team to personalize these guidelines.



# RECURRING MEDICAL SPECIALTY VISITS:

## **Audiologist** - every 6-12 months depending on concerns

- ☐ Consider ABR test under anesthesia if unable to cooperate with behavioral audiometry testing
- ☐ Adjust hearing aids (if applicable)

## **Bone Marrow Transplant Specialist** - only for children who underwent HSCT; frequency determined by specialist

- ☐ Order & interpret bone marrow follow-up labs, monitor for post-transplant complications

## **Cardiologist** - every 1-3 years based on symptoms / previous results

- ☐ Interpret periodic ECG and Echocardiogram

## **Developmental-Behavioral Pediatrician** - as needed or referred by MPS specialist

- ☐ Evaluate and treat behavioral and sleep issues

## **Endocrinologist** - consider consultation @ age 8-9 years (pre-puberty)

- ☐ Assess growth trajectory, monitor bone age, consider growth hormone therapy

## **Gastroenterologist** - as needed or routinely if specific issues are identified

- ☐ Evaluate and treat gastroesophageal reflux
- ☐ Consider nutrition evaluation and G-tube (feeding tube) placement (if needed)

## **Hand Surgeon** - as needed based on EMG / nerve conduction testing results

- ☐ Evaluate carpal tunnel syndrome to consider surgical intervention

## **MPS Specialist / Geneticist** - every 6-12 months

- ☐ Prescribe ERT (if applicable)
- ☐ Review periodic lab, radiology, & other diagnostic testing
- ☐ Discuss changes in symptoms / status, new treatment options, or clinical trials

## **Neurologist** - every 12 months

- ☐ Interpret periodic EMG / nerve conduction studies to evaluate for carpal tunnel syndrome
- ☐ Diagnose / manage hydrocephalus (if applicable)
- ☐ Diagnose / manage seizures (if applicable)

## **Neurosurgeon** - as needed or referred by MPS specialist

- ☐ Monitor and/or consider surgery for spinal cord compression or hydrocephalus

## **Neuropsychologist** - every 12 months or per MPS specialist recommendation

- ☐ Perform / interpret neurocognitive testing

## **Ophthalmologist** - every 12 months

- ☐ Perform visual acuity and dilated eye exam (assess for MPS-associated retinal changes)

## **Orthopedist** - every 12 months or as needed for specific conditions

- ☐ Order / interpret hip, knee or spine x-rays as needed based on symptoms
- ☐ Consider surgery as needed for specific conditions such as genu valgum or hip dysplasia

## **Otolaryngologist (ear-nose-throat specialist, ENT)** - every 12 months

- ☐ Check ears, tonsils, adenoids, airway
- ☐ Consider surgery (including T&A, ear tubes) as needed based on symptoms

## **Pediatrician** - routine care & vaccines (every 6-12 months depending on age)

- ☐ Monitor head growth, hepatosplenomegaly, inguinal or umbilical hernias

## **Pediatric Dentist** - routine care (every 6 months)

- ☐ Routine dental care (common findings = delayed tooth eruption and poor enamel)

## **Physiatrist / Pediatric Rehabilitation Medicine Specialist** as needed or referred by MPS specialist or PT/OT providers

- ☐ Evaluate for adaptive equipment (including stroller or wheelchair), orthotics
- ☐ Modify therapy prescriptions / recommendations

## **Pulmonologist** - every 12 months or based on symptoms

- ☐ Interpret periodic sleep studies and pulmonary function testing
- ☐ Consider bronchoscopy as needed to evaluate pulmonary function or airways



## RECURRING THERAPIES:

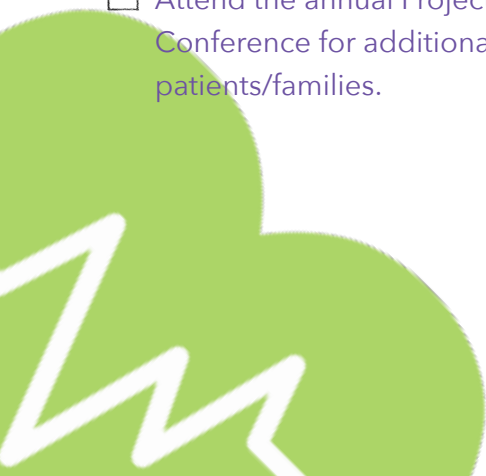
- ☐ Speech Therapy - *often 1-2 times weekly*
  - Consider augmentative communication evaluation (*multidisciplinary assessment with speech, OT, education team*)
- ☐ Occupational Therapy (OT) - *often 1-2 times weekly*
  - Consider augmentative communication evaluation - see above
- ☐ Physical Therapy (PT) - *often 1-2 times weekly*
- ☐ Behavioral Therapy (such as ABA) - *per MPS specialist or neuropsychologist recommendation*
- ☐ Other potential therapies to consider:
  - Hippotherapy
  - Aqua therapy
  - Art therapy
  - Music therapy

## EDUCATIONAL / DEVELOPMENTAL SERVICES:

- ☐ Early childhood intervention services (*0-3yrs*)
- ☐ School-based services (*>3yrs or school-aged*) - *individualized education plan (IEP) or Section 504 plan*
- ☐ Other local agencies / programs servicing special needs (*medical, educational, developmental*)

## ADDITIONAL CONSIDERATIONS:

- ☐ Identify palliative care / hospice care resources in your community
- ☐ Identify and apply for state-based health insurance waiver program (*aka Medicaid, Katie Beckett, HCBS*)
- ☐ Identify local home healthcare resources / agencies
- ☐ Update & maintain your Backpack Health profile
- ☐ Follow [clinicaltrials.gov](https://clinicaltrials.gov) for up-to-date information on new clinical trials in MPSII
- ☐ Attend the annual Project Alive Hunter Syndrome Community Conference or MPS Society Family Conference for additional opportunities to engage with MPS experts, researchers, and other MPS II patients/families.





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