Lab Dept: Urine/Stool

Test Name: MUCOPOLYSACCHARIDE SCREEN, (MPS), URINE

**General Information**

Lab Order Codes: MSUS

Synonyms: Acid Mucopolysaccharides; AMPS; Berry Spot; Chondroitin Sulfate; Dermatan Sulfate; GAGS; Glycoaminoglycans; Heparan Sulfate; Hunter's Syndrome; Hurler's Syndrome; Hunter Syndrome; Keratan Sulfate; Mucopolysaccharides;

CPT Codes: 83864 – Mucopolysaccharides, acid, quantitative
82542 – Mucopolysaccharide screen (column chromatography, includes mass spectrometry, if performed, non-drug analyte(s) not elsewhere specified, qualitative or quantitative, each specimen

Test Includes: Urine mucopolysaccharides quantitative with interpretive report and reported in mg/mmol Cr. Includes Heparin Sulfate, Chondroitin-6 Sulfate and Keratan Sulfate

**Logistics**

Test Indications: Supporting the biochemical diagnosis of one of the mucopolysaccharidoses: Types I, II, III, IV, VI, or VII. This test is not appropriate for the diagnosis of a specific mucopolysaccharidosis (MPS). Follow-up enzymatic testing must be performed to confirm a diagnosis of an MPS.

Lab Testing Sections: Urine/Stool – Sendouts

Referred to: Mayo Medical Laboratories (MML Test: MPSQU)

Phone Numbers: MIN Lab: 612-813-6280
STP Lab: 651-220-6550

Test Availability: Daily, 24 hours

Turnaround Time: 10 – 14 days

Special Instructions: Early morning specimen preferred

**Specimen**

Specimen Type: Urine, random

Container: Plastic leakproof container (No preservatives)
Draw Volume: 5 mL random urine

Processed Volume: Same as Draw Volume

Collection: A random urine sample may be obtained by voiding into a urine cup and is often performed at the laboratory. Bring the refrigerated container to the lab. Make sure all specimens submitted to the laboratory are properly labeled with the patient's name, medical record number and date of birth.

Special Processing: Lab Staff: Mix random sample. Aliquot 5 mL (Minimum: 1-2 mL) urine into a urine tube. Store and ship at refrigerated temperatures.

Patient Preparation: Morning specimen preferred. Do not administer low-molecular weight heparin prior to collection.

Sample Rejection: Warm specimens; mislabeled and unlabeled specimens

**Interpretive**

<table>
<thead>
<tr>
<th>Reference Range:</th>
<th>Mucopolysaccharide</th>
<th>Range (mg/mmol creatinine):</th>
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<tbody>
<tr>
<td></td>
<td>Dermatan Sulfate</td>
<td>&lt; or = 1.00</td>
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<tr>
<td></td>
<td>Heparan Sulfate</td>
<td>&lt; or = 4 yrs: &lt; or = 0.50</td>
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<td></td>
<td></td>
<td>&gt; or = 5 yrs: &lt; or = 0.25</td>
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<tr>
<td></td>
<td>Chondroitan Sulfate</td>
<td>&lt; or = 24 months: &lt; or = 10.00</td>
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<td></td>
<td></td>
<td>25 months – 10 yrs: &lt; or = 2.50</td>
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<tr>
<td></td>
<td></td>
<td>&gt; or =11 yrs: &lt; or = 1.50</td>
</tr>
<tr>
<td></td>
<td>Keratan Sulfate</td>
<td>&lt; or = 12 months: &lt; or = 2.00</td>
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<tr>
<td></td>
<td></td>
<td>13 – 24 months: &lt; or = 1.50</td>
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<tr>
<td></td>
<td></td>
<td>25 months – 4 yrs: &lt; or = 1.00</td>
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<td>5 – 18 yrs: &lt; or = 0.50</td>
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<td>&gt; or = 19 yrs: &lt; or = 0.30</td>
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An interpretation will be provided

**Critical Values:** N/A
Limitations: This test alone is not appropriate for the diagnosis of a specific mucopolysaccharidosis. Follow-up enzymatic testing must be performed to confirm a diagnosis of an MPS.

Methodology: Liquid Chromatography – Tandem Mass Spectrometry (LC-MS/MS)

References: Mayo Clinical Laboratories Web Page February 2020

Updates: 1/28/2016: CPT update
2/7/2020: Method and reference range update