Lab Dept: Chemistry

Test Name: MUCOPOLYSACCHARIDOSIS (MPS) PANEL

**General Information**

**Lab Order Codes:** MPSP - whole panel, or individually, codes are listed below:
- ALID – Alpha-iduronidase
- ID2S – Iduronate-2-sulfatase
- HESU – Heparan N-sulfatase (type A)
- NAAG – N-acetyl alpha-glucosaminidase (type B)
- ACDA – Acetyl CoA: glucosamine N-acetyl transferase (type C)
- NAG6 – N-acetyl glucosamine-6-sulfatase (type D)
- NAGS – N-acetyl galactosamine-6-sulfase
- BGAL – Beta-galactosidase
- ARYB – Arylsulfatase B
- BGLU – Beta-glucuronidase

**Synonyms:**
- Alpha-iduronidase (Hurler’s Syndrome, MPS I); Iduronate-2-sulfatase (Hunter Syndrome, MPS II); Heparin Sulfamidase (Sanfilippo’s Syndrome Type A, MPS IIIA); N-acetyl-alpha-glucosaminidase (Sanfilippo syndrome Type B, MPS IIIB); Acetyl Coa:glucosaminidase N-acetytrasferase (Sanfilippo Syndrome Type C, MPS IIIC); N-acetyl glucosamine-6-sulfatase (Sanfilippo Syndrome Type D, MPS IIID); Galactosamine-6-sulfatase (Morquio Syndrome Type A, MPS IVA); Beta-galactosidase (Morquio Syndrome Type B, MPS IVB); Arylsulfatase B (Maroteaux Lamy’s Syndrome, MPS VI); Beta-glucuronidase (Sly Syndrome)

**CPT Codes:** 82657 x5 – Enzyme activity in blood cells, cultured cells, or tissue, not elsewhere specified; nonradioactive substrate, each specimen.

**Test Includes:** Ten quantitative enzyme levels. See Reference Range for listing.

**Logistics**

**Test Indications:** Monitoring patients with MPS who have had bone marrow transplants or are receiving enzyme therapy or screening for mucopolysaccharidosis (MPS).

**Lab Testing Sections:** Chemistry – Sendouts

**Referred to:** Greenwood Genetic Center

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

**Test Availability:** Monday - Thursday, 24 hours

**Turnaround Time:** 21 days
**Special Instructions:** Restricted draw times, collect **Mon-Thurs 1130-1430 ONLY.**

**Specimen**

**Specimen Type:** Whole Blood

**Container:** Green top (NaHep) tubes (2 Tubes)

**Draw Volume:** Minimum: 6 mL (3 mL blood in EACH tube)

**Processed Volume:** Same as Draw Volume

**Collection:** Routine venipuncture

**Special Processing:** Lab Staff: **Do Not** Centrifuge. Specimen should remain in the original collection containers. Store and ship at room temperature. Forward promptly. **Specimens must arrive at Greenwood Genetic Center within 24 hours of collection.**

**Patient Preparation:** None

**Sample Rejection:** Mislabeled and unlabeled specimens; specimens more than 24 hours old

**Interpretive**

**Reference Range:**

<table>
<thead>
<tr>
<th>Mucopolysaccharides, Quantitative:</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Alpha-Iduronidase</td>
<td>6 – 71.4 nmol substrate/hr/mg protein</td>
</tr>
<tr>
<td>Iduronate-2-sulfatase</td>
<td>155 - 1082 nmol substrate/hr/mL plasma</td>
</tr>
<tr>
<td>Heparan N-sulfatase (type A)</td>
<td>2.21 - 27.34 nmol substrate/17hr/mg protein</td>
</tr>
<tr>
<td>N-acetyl alpha-glucosaminidase (type B)</td>
<td>97 - 1064 nmol substrate/17hr/mL plasma</td>
</tr>
<tr>
<td>Acetyl CoA: glucosamine N-acetyl transferase (type C)</td>
<td>3.1 - 31.7 nmol substrate/17hr/mg protein</td>
</tr>
<tr>
<td>N-acetyl glucosamine-6-sulfatase (type D)</td>
<td>5.3 – 35.4 nmol substrate/24hr/mg protein</td>
</tr>
<tr>
<td>N-acetyl galactosamine-6-sulfatase</td>
<td>76.1 – 255.1 nmol substrate/17hr/mg protein</td>
</tr>
<tr>
<td>Beta-galactosidase</td>
<td>13.5 - 176 nmol substrate/hr/mg protein</td>
</tr>
<tr>
<td>Test</td>
<td>Reference Range</td>
</tr>
<tr>
<td>---------------------</td>
<td>------------------------------------------------------</td>
</tr>
<tr>
<td>Arylsulfatase B</td>
<td>31.8 – 215.9 nmol substrate/hr/mg protein</td>
</tr>
<tr>
<td>Beta-glucuronidase</td>
<td>31.4 - 224 nmol substrate/hr/mg protein</td>
</tr>
</tbody>
</table>

**Critical Values:** N/A

**Limitations:** N/A

**Methodology:** 4-methylumbelliferyl substrate

**References:** Greenwood Genetics November 2017

**Update:**
- 4/6/2011: Change in reference range for Arylsulfatase B.
- 4/11/2011: Change in reference range for N-acetyl alpha glucosaminidase (type B)
- 6/16/2011: Reference range updates
- 11/8/2017: Turnaround time update