**Lab Dept:** Chemistry  
**Test Name:** MUCOPOLYSACCHARIDOSIS (MPS) PANEL

### General Information

**Lab Order Codes:** MPSP - whole panel, or individually, codes are listed below:
- ALID – Alpha-IIduronidase
- 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-acetyltrasferase (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 (Morqio 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**

<table>
<thead>
<tr>
<th>Reference Range:</th>
<th>Mucopolysaccharides, Quantitative:</th>
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<tbody>
<tr>
<td></td>
<td>Alpha-Iduronidase</td>
</tr>
<tr>
<td></td>
<td>Iduronate-2-sulfatase</td>
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<tr>
<td></td>
<td>Heparan N-sulfatase (type A)</td>
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<td></td>
<td>N-acetyl alpha-glucosaminidase (type B)</td>
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<tr>
<td></td>
<td>Acetyl CoA: glucosamine N-acetyl transferase (type C)</td>
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<td></td>
<td>N-acetyl glucosamine-6-sulfatase (type D)</td>
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<td></td>
<td>N-acetyl galactosamine-6-sulfatase</td>
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<tr>
<td></td>
<td>Beta-galactosidase</td>
</tr>
<tr>
<td>Arylsulfatase B</td>
<td>31.8 – 215.9 nmol substrate/hr/mg protein</td>
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<tr>
<td>Beta-glucuronidase</td>
<td>31.4 - 224 nmol substrate/hr/mg protein</td>
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</tbody>
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**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