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**Lab Dept:** Chemistry

**Test Name:** MUCOPOLYSACCHARIDOSIS (MPS) PANEL

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**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-acetyltransferase (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.

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**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**.

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***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

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***Interpretive***

**Reference Range:**

<b>Mucopolysaccharides, Quantitative:</b>	
Alpha-Iduronidase	6 – 71.4 nmol substrate/hr/mg protein
Iduronate-2-sulfatase	155 - 1082 nmol substrate/hr/mL plasma
Heparan N-sulfatase (type A)	2.21 - 27.34 nmol substrate/17hr/mg protein
N-acetyl alpha-glucosaminidase (type B)	97 - 1064 nmol substrate/17hr/mL plasma
Acetyl CoA: glucosamine N-acetyl transferase (type C)	3.1 - 31.7 nmol substrate/17hr/mg protein
N-acetyl glucosamine-6-sulfatase (type D)	5.3 – 35.4 nmol substrate/24hr/mg protein
N-acetyl galactosamine-6-sulfatase	76.1 – 255.1 nmol substrate/17hr/mg protein
Beta-galactosidase	13.5 - 176 nmol substrate/hr/mg protein

Arylsulfatase B	31.8 – 215.9 nmol substrate/hr/mg protein
Beta-glucuronidase	31.4 - 224 nmol substrate/hr/mg protein

**Critical Values:** N/A

**Limitations:** N/A

**Methodology:** 4-methylbumbelliferyl 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