
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-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.

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: 7 - 10 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:

Mucopolysaccharides, Quantitative:	
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](#) June 2011

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