

Major musculoskeletal manifestations of MPS by type of disorder¹

OMIM	Disease	Deficient enzyme	Main storage materials
607014 607015 607016	MPS I (Hurler, Scheie, Hurler/Scheie)	α -L-iduronidase	Dermatan sulfate, heparan sulfate
309900	MPS II (Hunter)	Iduronate 2-sulfatase	Dermatan sulfate, heparan sulfate
252900	MPS IIIA (Sanfilippo A)	Heparan <i>N</i> -sulfatase	Heparan sulfate
252920	MPS IIIB (Sanfilippo B)	α - <i>N</i> -acetylglucosaminidase	Heparan sulfate
252930	MPS IIIC (Sanfilippo C)	Acetyl CoA: α -glucosaminidase <i>N</i> -acetyltransferase	Heparan sulfate
252940	MPS IIID (Sanfilippo D)	<i>N</i> -acetylglucosamine-6-sulfatase	Keratan sulfate, chondroitin 6-sulfate
253000	MPS IVA (Morquio A)	<i>N</i> -acetylgalactosamine-6-sulfatase	Keratan sulfate, chondroitin 6-sulfate
253010	MPS IVB (Morquio B)	β -galactosidase	Keratan sulfate
253200	MPS VI (Maroteaux–Lamy)	<i>N</i> -acetyl galactosamine 4-sulfatase (arylsulfatase B)	Dermatan sulfate
253220	MPS VII (Sly)	β -glucuronidase	Dermatan sulfate, heparan sulfate, chondroitin 6-sulfate
601492	MPS IX (Natowicz)	Hyaluronidase	Hyaluronan

Adapted from Filocamo, *Hum Genomics*, 2011.

Abbreviation: MPS, mucopolysaccharidosis.

Reference: 1. Filocamo M, Morrone A. Lysosomal storage disorders: molecular basis and laboratory testing. *Hum Genomics*. 2011;5(3):156-169. doi:10.1186/1479-7364-5-3-156.