

Recommended schedule of assessments for patients with MPS VI⁴

Imaging study	Initial assessments	Every 3 months	Every 12 months	As clinically indicated ^a
Confirmation of MPS VI	●			
Medical history^b	●		●	
Physical examination	●		●	
Neurologic examination	●		●	
Height, weight	●		●	
Head circumference^c	●		●	
Tanner stage^d	●		●	
Photographs	●			●
Endurance^e				
12-minute walk test	●		●	
3-minute stair climb	●		●	
Ophthalmology				
Visual acuity	●		●	
Corneal examination	●		●	
Fundoscopy examination	●		●	
Intraocular pressure	●		●	
Refraction	●		●	
Audiometry	●		●	

Cardiology				
Echocardiogram	●		●	
Electrocardiogram	●		●	
Blood pressure	●		●	
Electrophysiology				
Nerve conduction ^f	●			●
Pulmonary function				
Forced vital capacity, forced expiratory volume in 1 second, maximum voluntary ventilation ^g	●		●	
Sleep study	●			●
Imaging studies				
Hip films ^h	●			●
Skeletal survey	●			●
Flex/ext radiograph of cervical spine	●			●
MRI of brain and spine ⁱ	●			●
Laboratory assessments				
uGAG levels	●		●	
Supplemental assessments for patients on ERT^j				
Total anti-ASB antibody ^k	●	●	Yearly after 24 months	

Adapted from Giugliani, *Pediatrics*, 2007.

Abbreviation: MRI, magnetic resonance imaging.

^a "As clinically indicated" generally means every 2 to 3 years depending on the rate of disease progression and clinical symptoms.

^b For infants, more frequent examinations are necessary.

^c Monitored until head growth has stopped.

^d Continue assessments until pubertal maturation is completed.

^e Endurance-testing paradigm before and after ERT: distance walked in 12 minutes (or 6-minute walk test per American Thoracic Society guidelines, but preferably same minute length as completed in previous test); number of stairs climbed in 3 minutes.

^f Median nerve conduction measured to evaluate carpal tunnel syndrome.

^g Pulmonary function tests are to include forced vital capacity, forced expiratory volume in 1 second, and maximum voluntary ventilation.

^h Anteroposterior and "frog-leg" lateral views of pelvis.

ⁱ MRI of brain and spinal cord may require sedation or general anesthesia depending on patient age and cooperation. General anesthesia carries substantial risk for patients with MPS VI.

^j For patients on ERT, results should be obtained at baseline, then at months 3, 6, 12, 18, and 24, and then yearly.

^k Anti-ASB antibody testing is only available for US patients enrolled in the clinical surveillance program.