## *Recommended schedule of assesments for patients with MPS VI*[1]

| *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 | ● | | ● | |
| Fundoscopic 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.

Abbreviations: MPS VI, mucopolysaccharidosis VI; MRI, magnetic resonance imaging; uGAG, urinary glycosaminoglycan.

[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 Enzyme replacement therapy (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.

**Reference: 1.** Giugliani R, Harmatz P, Wraith JE. Management guidelines for mucopolysaccharidosis VI. *Pediatrics*. 2007;120:405-418. doi:10.1542/peds.2006-2184.