What is claimed is:

1. Use of a human iduronate-2-sulfatase (hIDS) in the preparation of a composition for treating mucopolysaccharidosis II (MPS II) in a human subject, wherein:

a. the level of D2S6 present in a biological sample obtained from the human subject is measured;

b. the human subject is diagnosed with MPS II when the level of D2S6 in the biological sample is higher than a reference; and

c. the composition is suitable to be delivered to the central nervous system (CNS) of the human subject diagnosed with MPS II.

2. Use of a human iduronate-2-sulfatase (hIDS) in the preparation of a composition for treating mucopolysaccharidosis II (MPS II) in a human subject, wherein the composition is suitable to be delivered to the central nervous system (CNS) of the human subject diagnosed with MPS II, and wherein the level of D2S6 present in a biological sample obtained from the human subject is measured after the composition is delivered to the CNS.

3. The use of claim 1 or 2, wherein a ratio of D2S6 to total heparan sulfate disaccharides (HS) in the biological sample is calculated, wherein the total HS comprises disaccharides D2S6, D0A0, D0S0, and D0A6.

4. The use of claim 3, wherein the human subject is determined to be responsive to the hIDS if the D2S6 level is at least 20% of the total HS.

5. The use of any one of claims 1-4, wherein the composition is suitable to be delivered to the CNS via a medicament comprising an adeno-associated virus (AAV) vector comprising a nucleotide sequence encoding hIDS.

6. The use of any one of claims 1-4, wherein the composition is suitable to be delivered to the CNS via an enzyme replacement therapy with recombinant hIDS.

7. The use of any one of claims 1-4, wherein the composition is suitable to be delivered via a viral vector, liposome, a lipid-containing complex, a macromolecular complex, a synthetic modified mRNA, an unmodified mRNA, a small molecule, a non-biologically active molecule, a polymerized molecule, a naked DNA, a plasmid, a phage, a transposon, a cosmid, or an episome.

8. The use of any one of claims 1-7, wherein the composition is suitable to be delivered to the CNS via intrathecal, intracisternal (IC), intravenous, or intracerebroventricular (ICV) administration.

9. The use of any one of claims 1-7, wherein the composition is suitable to be delivered to the CNS via intracisternal (IC) or intracerebroventricular (ICV) injection.

10. The use of any one of claims 1-9, wherein the biological sample is obtained from the cerebrospinal fluid (CSF) of the human subject.

11. The use of any one of claims 1 and 3-10, wherein the reference is the level of D2S6 in a CSF sample obtained from a human subject with attenuated MPS II.

12. The use of any one of claims 1 and 3-10, wherein the reference is the level of D2S6 in a CSF sample obtained from a human subject not diagnosed with MPS II.

13. The use of any one of claims 1-12, wherein the MPS II is neuronopathic MPS II.

14. The use of any one of claims 1-13, wherein the human subject is:

a) an adult human subject;

b) younger than 18 years old;

c) 5 years old or older and younger than 18 years old; or

d) 4 months old or older and younger than 5 years old.

15. The use of any one of claims 1 and 3-14, wherein the level of D2S6 present in a biological sample obtained from the human subject is monitored after step c, wherein a decrease in the level of D2S6 compared to the level of D2S6 measured in step a is indicative of treatment.

16. The use of claim 15, wherein the biological sample obtained after step c is obtained from the human subject after about 1, 2, 3, 4, 5, 6, 7, 8, 10, 16, 20, 24, 30, 35, 40, 45, 48, 50, 52, or 56 weeks, or 1, 2, 3, 4, 5, or 6 months after step c.

17. The use of claim 15, wherein the decrease in the level of D2S6 after step c is about or at least about 5%, 10%, 15%, 20%, 25%, 30%, 35%, 40%, 45%, 50%, 55%, 60%, 65%, 70%, 75%, 80%, 85%, or 90%, as compared to the level of D2S6 measured in step a.

18. The use of any one of claims 1 and 3-14, wherein the level of D2S6 is higher by about or at least about 5%, 10%, 15%, 20%, 25%, 30%, 35%, 40%, 45%, 50%, 55%, 60%, 65%, 70%, 75%, 80%, 85%, 90%, or 95% than the reference.

19. The use of any one of claims 2, 3-10, 13, and 14, wherein the level of D2S6 is measured in a first biological sample obtained from the human subject before the composition is delivered to the human subject and the level of D2S6 present in a second biological sample obtained from the human subject is measured after the composition is delivered to the human subject, and wherein the first and second biological samples are obtained from the cerebrospinal fluid (CSF) of the human subject.

20. The use of any one of claims 1 and 3-18, wherein the reference is a predetermined value.