CLAIMS

1. A lyophilized formulation comprising;

a fusion protein including an anti-human transferrin receptor (hTfR) antibody and a human iduronate-2-sulfatase, as an active ingredient, and further

sodium chloride, sucrose, a nonionic surfactant, and a phosphate buffer,

wherein the nonionic surfactant is polysorbate or poloxamer, and

wherein the pH is 5.5 to 7.5 when the lyophilized formulation is dissolved in pure water.

2. The lyophilized formulation according to claim 1, wherein the nonionic surfactant is selected from the group consisting of polysorbate 20, polysorbate 80, and polyoxyethylene(160)polyoxypropylene(30)glycol.

3. The lyophilized formulation according to claim 1, wherein the nonionic surfactant is polyoxyethylene(160)polyoxypropylene(30)glycol.

4. The lyophilized formulation according to any one of claims 1 to 3, wherein the amounts of sodium chloride, sucrose, and the nonionic surfactant are 0.015 to 2.5 (w/w), 2.5 to 200 (w/w), and 0.005 to 6 (w/w), respectively, relative to the amount of the fusion protein.

5. The lyophilized formulation according to any one of claims 1 to 3, wherein the amounts of sodium chloride, sucrose, and the nonionic surfactant are 0.05 to 0.5 (w/w), 5 to 50 (w/w), and 0.02 to 0.2 (w/w), respectively, relative to the amount of the fusion protein.

6. The lyophilized formulation according to any one of claims 1 to

3, wherein the amounts of sodium chloride, sucrose, and the nonionic surfactant is 0.1 to 0.25 (w/w), 10 to 25 (w/w), and 0.04 to 0.1 (w/w), respectively, relative to the amount of the fusion protein.

7. The lyophilized formulation according to any one of claims 1 to 6, wherein the fusion protein is a fusion protein in which the human iduronate-2-sulfatase is linked to the light chain or the heavy chain of the antibody on the C-terminal side thereof by a peptide bond.

8. The lyophilized formulation according to any one of claims 1 to 6, wherein the fusion protein is a fusion protein in which the human iduronate-2-sulfatase is linked to the heavy chain of the antibody on the C-terminal side thereof by a peptide bond.

9. The lyophilized formulation according to any one of claims 1 to 6, wherein the fusion protein is a fusion protein in which the human iduronate-2-sulfatase is linked to the light chain or the heavy chain of the antibody on the C-terminal side thereof via a linker including one or more of amino acids.

10. The lyophilized formulation according to any one of claims 1 to 6, wherein the fusion protein is a fusion protein in which the human iduronate-2-sulfatase is linked to the heavy chain of the antibody on the C-terminal side thereof via a linker including one or more of amino acids.

11. The lyophilized formulation according to claim 9 or 10, wherein the linker includes the amino acid sequence of Gly-Ser.

12. The lyophilized formulation according to any one of claims 1 to 11, wherein the antibody is a human antibody or a humanized antibody.

13. The lyophilized formulation according to claim 12, wherein the antibody is a humanized anti-human transferrin receptor (hTfR) antibody.

14. The lyophilized formulation according to claim 12, wherein the antibody is a humanized anti-hTfR antibody, and wherein the fusion protein is selected from the group consisting of (a) to (c) below;

(a) the fusion protein, wherein the light chain of the humanized anti-hTfR antibody has the amino acid sequence set forth as SEQ ID NO:2, and wherein the heavy chain of the humanized anti-hTfR antibody having the amino acid sequence set forth as SEQ ID NO:8 is linked, on the C-terminal side thereof, to the human iduronate-2-sulfatase via a linker sequence,

(b) the fusion protein, wherein the light chain of the humanized anti-hTfR antibody has the amino acid sequence set forth as SEQ ID NO:4, and wherein the heavy chain of the humanized anti-hTfR antibody having the amino acid sequence set forth as SEQ ID NO:9 is linked, on the C-terminal side thereof, to the human iduronate-2-sulfatase via a linker sequence,

(c) the fusion protein, wherein the light chain of the humanized anti-hTfR antibody has the amino acid sequence set forth as SEQ ID NO:6, and wherein the heavy chain of the humanized anti-hTfR antibody having the amino acid sequence set forth as SEQ ID NO:10 is linked, on the C-terminal side thereof, to the human iduronate-2-sulfatase via a linker sequence.

15. The lyophilized formulation according to claim 12, wherein the antibody is a humanized anti-hTfR antibody, and wherein the fusion

protein is selected from the group consisting of (a) to (c) below;

(a) the fusion protein, wherein the light chain of the humanized anti-hTfR antibody has the amino acid sequence set forth as SEQ ID NO:2, and wherein the heavy chain of the humanized anti-hTfR antibody is linked, on the C-terminal side thereof and via a linker sequence of Gly-Ser, to the human iduronate-2-sulfatase, and the whole linked heavy chain has the amino acid sequence set forth as SEQ ID NO:13,

(b) the fusion protein, wherein the light chain of the humanized anti-hTfR antibody has the amino acid sequence set forth as SEQ ID NO:4, and wherein the heavy chain of the humanized anti-hTfR antibody is linked, on the C-terminal side thereof and via a linker sequence of Gly-Ser, to the human iduronate-2-sulfatase, and the whole linked heavy chain has the amino acid sequence set forth as SEQ ID NO:15, and

(c) the fusion protein, wherein the light chain of the humanized anti-hTfR antibody has the amino acid sequence set forth as SEQ ID NO:6, and wherein the heavy chain of the humanized anti-hTfR antibody is linked, on the C-terminal side thereof and via a linker sequence of Gly-Ser, to the human iduronate-2-sulfatase, and the whole linked heavy chain has the amino acid sequence set forth as SEQ ID NO:17.

16. The lyophilized formulation according to any one of claims 1 to 15, sealed in a container formed of a borosilicate glass or a hydrophobic resin.

17. The lyophilized formulation according to claim 16, wherein the

container is formed by a cycloolefin copolymer, a ring-opened polymer of cycloolefin, or a hydrogenated ring-opened polymer of cycloolefin.