CLAIMS

[Claim 1]. Use of a conjugate of an anti-human transferrin receptor antibody and a human acid α-glucosidase in the manufacture of a medicament for use in treating Pompe disease, wherein the human acid α-glucosidase has a physiological activity to be exerted in muscle.

[Claim 2]. The use according to claim 1, wherein the anti-human transferrin receptor antibody is a Fab antibody, a $F(ab')_2$ antibody, or a F(ab') antibody.

[Claim 3]. The use according to claim 1, wherein the anti-human transferrin receptor antibody is a single-chain antibody selected from the group consisting of scFab, scF(ab'), $scF(ab')_2$, and scFv.

[Claim 4]. The use according to claim 3, wherein in the single-chain antibody a light chain and a heavy chain of the anti-human transferrin receptor antibody are linked via a linker sequence.

[Claim 5]. The use according to claim 4, wherein the light chain and the heavy chain of the anti-human transferrin receptor antibody are linked on the C-terminal side of the light chain via the linker sequence.

[Claim 6]. The use according to claim 4, wherein the light chain and the heavy chain of the anti-human transferrin receptor antibody are linked on the C-terminal side of the heavy chain via the linker sequence.

[Claim 7]. The use according to any one of claims 4 to 6, wherein the linker sequence comprises 8 to 50 amino acid residues.

[Claim 8]. The use according to claim 7, wherein the linker sequence comprises an amino acid sequence selected from the group consisting of the amino acid sequence (Gly Ser), the amino acid sequence (Gly Gly Ser), the amino acid sequence (Gly Gly Gly), the amino acid sequence set forth as SEQ ID NO: 3, and the amino acid sequence set forth as SEQ ID NO: 4.

[Claim 9]. The use according to claim 8, wherein the linker sequence comprises 15 amino acid residues in which the amino acid sequence set forth as SEQ ID NO: 3 is consecutively repeated three times.

[Claim 10]. The use according to any one of claims 1 to 9, wherein the anti-human transferrin receptor antibody comprises the amino acid sequence set forth as SEQ ID NO: 22 in the variable region of the light chain and the amino acid sequence set forth as SEQ ID NO: 23 in the variable region of the heavy chain.

[Claim 11]. The use according to claim 10, wherein the anti-human transferrin receptor antibody comprises the light chain comprising an amino acid sequence set forth as SEQ ID NO: 24 and the heavy chain comprising an amino acid sequence set forth as SEQ ID NO: 25.

[Claim 12]. The use according to any one of claims 1 to 11, selected

from the group consisting of (1) to (4) below:

(1) a conjugate in which the protein is linked to the C-terminal side of the light chain of the anti-human transferrin receptor antibody by a peptide bond,

(2) a conjugate in which the protein is linked to the N-terminal side of the light chain of the anti-human transferrin receptor antibody by a peptide bond,

(3) a conjugate in which the protein is linked to the C-terminal side of the heavy chain of the anti-human transferrin receptor antibody by a peptide bond, and

(4) a conjugate in which the protein is linked to the N-terminal side of the heavy chain of the anti-human transferrin receptor antibody by a peptide bond.

[Claim 13]. The use according to claim 12, wherein the protein is linked to the anti-human transferrin receptor antibody via a linker sequence.

[Claim 14]. The use according to claim 13, wherein the linker sequence consists of 1 to 50 amino acid residues.

[Claim 15]. The use according to claim 14, wherein the linker sequence comprises an amino acid sequence selected from the group consisting of a single glycine, a single serine, the amino acid sequence

(Gly Ser), the amino acid sequence (Gly Gly Ser), the amino acid sequence set forth as SEQ ID NO:3, the amino acid sequence set forth as SEQ ID NO:4, and the amino acid sequence consisting of 1 to 10 amino acid sequences thereof that are linked consecutively.

[Claim 16]. The use according to any one of claims 1 to 15, wherein the anti-human transferrin receptor antibody is a humanized anti-human transferrin receptor antibody.

[Claim 17]. The use according to claim 1, wherein the light chain of the humanized anti-hTfR antibody comprises the amino acid sequence set forth as SEQ ID NO: 24, and wherein the heavy chain of the humanized anti-hTfR antibody is linked on its C-terminal side to the human acid α-glucosidase via a linker sequence (Gly Ser), and the whole linked heavy chain has the amino acid sequence set forth as SEQ ID NO: 27.

[Claim 18]. The use according to any one of claims 1 to 17, wherein the muscle is skeletal muscle, cardiac muscle, or smooth muscle.