

What is claimed is:

1. A pharmaceutical formulation comprising:
 - (d) a recombinant acid α -glucosidase, wherein the recombinant acid α -glucosidase is expressed in Chinese hamster ovary (CHO) cells and comprises an increased content of N-glycan units bearing one or two mannose-6-phosphate residues when compared to a content of N-glycan units bearing one or two mannose-6-phosphate residues of α -glucosidase alfa;
 - (e) at least one buffer selected from the group consisting of a citrate, a phosphate and combinations thereof; and
 - (f) at least one excipient selected from the group consisting of mannitol, polysorbate 80, and combinations thereof,wherein the formulation has a pH of from about 5.0 to about 7.0.
2. The pharmaceutical formulation of claim 1, wherein the recombinant acid α -glucosidase is present in a concentration of about 5 to about 50 mg/mL.
3. The pharmaceutical formulation of claim 1 or 2, wherein the recombinant acid α -glucosidase is present in a concentration of about 15 mg/mL.
4. The pharmaceutical formulation of any of claims 1-3, wherein the formulation has a pH of from about 5.5 to about 7.0.
5. The pharmaceutical formulation of any of claims 1-4, wherein the formulation has a pH of about 6.0.
6. The pharmaceutical formulation of any of claims 1-5, wherein the at least one buffer comprises citrate.
7. The pharmaceutical formulation of any of claims 1-6, wherein the at least one buffer comprises a potassium, sodium or ammonium salt.
8. The pharmaceutical formulation of any of claims 1-7, wherein the at least one buffer comprises sodium citrate.
9. The pharmaceutical formulation of any of claims 1-8, wherein the at least one buffer is present in a concentration of about 10 to about 100 mM.

10. The pharmaceutical formulation of any of claims 1-9, wherein the at least one buffer is present in a concentration of about 25 mM.
11. The pharmaceutical formulation of any of claims 1-10, wherein trehalose, sucrose, glycine or combinations thereof is excluded.
- 5 12. The pharmaceutical formulation of any of claims 1-11, wherein the at least one excipient is mannitol present in a concentration of about 10 to about 50 mg/mL.
13. The pharmaceutical formulation of any of claims 1-12, wherein the at least one excipient is polysorbate 80 present in a concentration of about 0.2 to about 0.5 mg/mL.
- 10 14. The pharmaceutical formulation of any of claims 1-13, wherein the mannitol is present at a concentration of about 20 mg/mL and the polysorbate 80 is present at a concentration of about 0.5 mg/mL.
15. The pharmaceutical formulation of any of claims 1-14, further comprising:
- (d) water.
16. The pharmaceutical formulation of any of claims 1-15, further comprising:
- 15 (e) an alkalizing agent; and/or
- (f) an acidifying agent,
- wherein the alkalizing agent and acidifying agent are present in amounts to maintain the pharmaceutical formulation at a pH of from about 5.0 to about 6.0.
- 20 17. The pharmaceutical formulation of any of claims 1-16, wherein at least 30% of molecules of the recombinant human acid α -glucosidase comprise one or more N-glycan units bearing one or two mannose-6-phosphate residues.
18. The pharmaceutical formulation of any of claims 1-17, wherein the recombinant human acid α -glucosidase comprises on average from 0.5 to 7.0 moles of N-glycan units bearing one or two mannose-6-phosphate residues per mole of recombinant human acid
- 25 α -glucosidase.
19. The pharmaceutical formulation of any of claims 1-18, wherein the recombinant human acid α -glucosidase comprises on average at least 3 moles of mannose-6-phosphate

residues per mole of recombinant human acid α -glucosidase and at least 4 moles of sialic acid residues per mole of recombinant human acid α -glucosidase.

20. The pharmaceutical formulation of any of claims 1-19, wherein the recombinant human acid α -glucosidase comprises seven potential N-glycosylation sites, at least 50% of molecules of the recombinant human acid α -glucosidase comprise an N-glycan unit bearing two mannose-6-phosphate residues at the first site, at least 30% of molecules of the recombinant human acid α -glucosidase comprise an N-glycan unit bearing one mannose-6-phosphate residue at the second site, at least 30% of molecules of the recombinant human acid α -glucosidase comprise an N-glycan unit bearing two mannose-6-phosphate residue at the fourth site, and at least 20% of molecules of the recombinant human acid α -glucosidase comprise an N-glycan unit bearing one mannose-6-phosphate residue at the fourth site.
21. The pharmaceutical formulation of any of claims 1-20, wherein the pharmaceutical formulation consists essentially of:
- (a) the recombinant acid α -glucosidase;
 - (b1) sodium citrate;
 - (b2) citric acid monohydrate;
 - (c1) mannitol;
 - (c2) polysorbate 80;
 - (d) water;
 - (e) optionally, an acidifying agent; and
 - (f) optionally, an alkalizing agent,
- wherein the formulation has a pH of from about 5.0 to about 6.0.
22. The pharmaceutical formulation of any of claims 1-21, wherein the pharmaceutical formulation consists essentially of:
- (a) the recombinant acid α -glucosidase, present at a concentration of about 15 mg/mL;
 - (b) sodium citrate buffer, present at a concentration of about 25 mM;

- (c1) mannitol, present at a concentration of about 20 mg/mL;
(c2) polysorbate 80, present at a concentration of about 0.5 mg/mL; and
(d) water;
(e) optionally, an acidifying agent; and
5 (f) optionally, an alkalizing agent,
wherein the formulation has a pH of from about 5.0 to about 6.0.
23. A pharmaceutical composition comprising the formulation of any of claims 1-22 after lyophilization.
24. A pharmaceutical composition comprising a lyophilized mixture comprising:
- 10 (a) a recombinant acid α -glucosidase, wherein the recombinant acid α -glucosidase is expressed in Chinese hamster ovary (CHO) cells and comprises an increased content of N-glycan units bearing one or two mannose-6-phosphate residues when compared to a content of N-glycan units bearing one or two mannose-6-phosphate residues of alglucosidase alfa;
- 15 (b) a buffer selected from the group consisting of a citrate, a phosphate and combinations thereof; and
(c) at least one excipient selected from the group consisting of trehalose, mannitol, polysorbate 80, and combinations thereof.
25. Use of the pharmaceutical formulation of claims 1-22 for treating Pompe disease.
- 20 26. The use of claim 25, further wherein the pharmaceutical formulation is diluted prior to administration to the patient.
27. Use of the pharmaceutical composition of claim 23 or 24 for treating Pompe disease, wherein the pharmaceutical composition is reconstituted and the reconstituted pharmaceutical composition is administered to a patient in need thereof.
- 25 28. A method of preparing the pharmaceutical formulation of any of claims 1-22, the method comprising:
adding the at least one buffer, at least one excipient and recombinant acid α -glucosidase to water to provide a solution;

optionally adjusting the pH of the solution; and
optionally adding additional water to the solution.

29. The method of claim 28, further comprising filtering the solution.

30. The method of claim 28 or 29, further comprising storing the solution.