

SEQUENCE LISTING

<110> Amicus Therapeutics, Inc.

<120> FORMULATIONS COMPRISING RECOMBINANT ACID ALPHA-GLUCOSIDASE

<130> AT-P8700-PCT

<160> 2

<170> PatentIn version 3.5

<210> 1

<211> 952

<212> PRT

<213> Homo sapiens

<400> 1

Met Gly Val Arg His Pro Pro Cys Ser His Arg Leu Leu Ala Val Cys
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Ala Leu Val Ser Leu Ala Thr Ala Ala Leu Leu Gly His Ile Leu Leu
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His Asp Phe Leu Leu Val Pro Arg Glu Leu Ser Gly Ser Ser Pro Val
35 40 45

Leu Glu Glu Thr His Pro Ala His Gln Gln Gly Ala Ser Arg Pro Gly
50 55 60

Pro Arg Asp Ala Gln Ala His Pro Gly Arg Pro Arg Ala Val Pro Thr
65 70 75 80

Gln Cys Asp Val Pro Pro Asn Ser Arg Phe Asp Cys Ala Pro Asp Lys
85 90 95

Ala Ile Thr Gln Glu Gln Cys Glu Ala Arg Gly Cys Cys Tyr Ile Pro
100 105 110

Ala Lys Gln Gly Leu Gln Gly Ala Gln Met Gly Gln Pro Trp Cys Phe
115 120 125

Phe Pro Pro Ser Tyr Pro Ser Tyr Lys Leu Glu Asn Leu Ser Ser Ser
130 135 140

Glu Met Gly Tyr Thr Ala Thr Leu Thr Arg Thr Thr Pro Thr Phe Phe
145 150 155 160

Pro Lys Asp Ile Leu Thr Leu Arg Leu Asp Val Met Met Glu Thr Glu
165 170 175

Asn Arg Leu His Phe Thr Ile Lys Asp Pro Ala Asn Arg Arg Tyr Glu
180 185 190

Val Pro Leu Glu Thr Pro Arg Val His Ser Arg Ala Pro Ser Pro Leu
195 200 205

Tyr Ser Val Glu Phe Ser Glu Glu Pro Phe Gly Val Ile Val His Arg
210 215 220

Gln Leu Asp Gly Arg Val Leu Leu Asn Thr Thr Val Ala Pro Leu Phe
225 230 235 240

Phe Ala Asp Gln Phe Leu Gln Leu Ser Thr Ser Leu Pro Ser Gln Tyr
245 250 255

Ile Thr Gly Leu Ala Glu His Leu Ser Pro Leu Met Leu Ser Thr Ser
260 265 270

Trp Thr Arg Ile Thr Leu Trp Asn Arg Asp Leu Ala Pro Thr Pro Gly
275 280 285

Ala Asn Leu Tyr Gly Ser His Pro Phe Tyr Leu Ala Leu Glu Asp Gly
290 295 300

Gly Ser Ala His Gly Val Phe Leu Leu Asn Ser Asn Ala Met Asp Val
305 310 315 320

Val Leu Gln Pro Ser Pro Ala Leu Ser Trp Arg Ser Thr Gly Gly Ile

325

330

335

Leu Asp Val Tyr Ile Phe Leu Gly Pro Glu Pro Lys Ser Val Val Gln
 340 345 350

Gln Tyr Leu Asp Val Val Gly Tyr Pro Phe Met Pro Pro Tyr Trp Gly
 355 360 365

Leu Gly Phe His Leu Cys Arg Trp Gly Tyr Ser Ser Thr Ala Ile Thr
 370 375 380

Arg Gln Val Val Glu Asn Met Thr Arg Ala His Phe Pro Leu Asp Val
 385 390 395 400

Gln Trp Asn Asp Leu Asp Tyr Met Asp Ser Arg Arg Asp Phe Thr Phe
 405 410 415

Asn Lys Asp Gly Phe Arg Asp Phe Pro Ala Met Val Gln Glu Leu His
 420 425 430

Gln Gly Gly Arg Arg Tyr Met Met Ile Val Asp Pro Ala Ile Ser Ser
 435 440 445

Ser Gly Pro Ala Gly Ser Tyr Arg Pro Tyr Asp Glu Gly Leu Arg Arg
 450 455 460

Gly Val Phe Ile Thr Asn Glu Thr Gly Gln Pro Leu Ile Gly Lys Val
 465 470 475 480

Trp Pro Gly Ser Thr Ala Phe Pro Asp Phe Thr Asn Pro Thr Ala Leu
 485 490 495

Ala Trp Trp Glu Asp Met Val Ala Glu Phe His Asp Gln Val Pro Phe
 500 505 510

Asp Gly Met Trp Ile Asp Met Asn Glu Pro Ser Asn Phe Ile Arg Gly
 515 520 525

Ser Glu Asp Gly Cys Pro Asn Asn Glu Leu Glu Asn Pro Pro Tyr Val
530 535 540

Pro Gly Val Val Gly Gly Thr Leu Gln Ala Ala Thr Ile Cys Ala Ser
545 550 555 560

Ser His Gln Phe Leu Ser Thr His Tyr Asn Leu His Asn Leu Tyr Gly
565 570 575

Leu Thr Glu Ala Ile Ala Ser His Arg Ala Leu Val Lys Ala Arg Gly
580 585 590

Thr Arg Pro Phe Val Ile Ser Arg Ser Thr Phe Ala Gly His Gly Arg
595 600 605

Tyr Ala Gly His Trp Thr Gly Asp Val Trp Ser Ser Trp Glu Gln Leu
610 615 620

Ala Ser Ser Val Pro Glu Ile Leu Gln Phe Asn Leu Leu Gly Val Pro
625 630 635 640

Leu Val Gly Ala Asp Val Cys Gly Phe Leu Gly Asn Thr Ser Glu Glu
645 650 655

Leu Cys Val Arg Trp Thr Gln Leu Gly Ala Phe Tyr Pro Phe Met Arg
660 665 670

Asn His Asn Ser Leu Leu Ser Leu Pro Gln Glu Pro Tyr Ser Phe Ser
675 680 685

Glu Pro Ala Gln Gln Ala Met Arg Lys Ala Leu Thr Leu Arg Tyr Ala
690 695 700

Leu Leu Pro His Leu Tyr Thr Leu Phe His Gln Ala His Val Ala Gly
705 710 715 720

Glu Thr Val Ala Arg Pro Leu Phe Leu Glu Phe Pro Lys Asp Ser Ser

725

730

735

Thr Trp Thr Val Asp His Gln Leu Leu Trp Gly Glu Ala Leu Leu Ile
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Thr Pro Val Leu Gln Ala Gly Lys Ala Glu Val Thr Gly Tyr Phe Pro
 755 760 765

Leu Gly Thr Trp Tyr Asp Leu Gln Thr Val Pro Ile Glu Ala Leu Gly
 770 775 780

Ser Leu Pro Pro Pro Pro Ala Ala Pro Arg Glu Pro Ala Ile His Ser
 785 790 795 800

Glu Gly Gln Trp Val Thr Leu Pro Ala Pro Leu Asp Thr Ile Asn Val
 805 810 815

His Leu Arg Ala Gly Tyr Ile Ile Pro Leu Gln Gly Pro Gly Leu Thr
 820 825 830

Thr Thr Glu Ser Arg Gln Gln Pro Met Ala Leu Ala Val Ala Leu Thr
 835 840 845

Lys Gly Gly Glu Ala Arg Gly Glu Leu Phe Trp Asp Asp Gly Glu Ser
 850 855 860

Leu Glu Val Leu Glu Arg Gly Ala Tyr Thr Gln Val Ile Phe Leu Ala
 865 870 875 880

Arg Asn Asn Thr Ile Val Asn Glu Leu Val Arg Val Thr Ser Glu Gly
 885 890 895

Ala Gly Leu Gln Leu Gln Lys Val Thr Val Leu Gly Val Ala Thr Ala
 900 905 910

Pro Gln Gln Val Leu Ser Asn Gly Val Pro Val Ser Asn Phe Thr Tyr
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Ser Pro Asp Thr Lys Val Leu Asp Ile Cys Val Ser Leu Leu Met Gly
930 935 940

Glu Gln Phe Leu Val Ser Trp Cys
945 950

<210> 2
<211> 896
<212> PRT
<213> Homo sapiens

<400> 2

Gln Gln Gly Ala Ser Arg Pro Gly Pro Arg Asp Ala Gln Ala His Pro
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Gly Arg Pro Arg Ala Val Pro Thr Gln Cys Asp Val Pro Pro Asn Ser
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Arg Phe Asp Cys Ala Pro Asp Lys Ala Ile Thr Gln Glu Gln Cys Glu
35 40 45

Ala Arg Gly Cys Cys Tyr Ile Pro Ala Lys Gln Gly Leu Gln Gly Ala
50 55 60

Gln Met Gly Gln Pro Trp Cys Phe Phe Pro Pro Ser Tyr Pro Ser Tyr
65 70 75 80

Lys Leu Glu Asn Leu Ser Ser Ser Glu Met Gly Tyr Thr Ala Thr Leu
85 90 95

Thr Arg Thr Thr Pro Thr Phe Phe Pro Lys Asp Ile Leu Thr Leu Arg
100 105 110

Leu Asp Val Met Met Glu Thr Glu Asn Arg Leu His Phe Thr Ile Lys
115 120 125

Asp Pro Ala Asn Arg Arg Tyr Glu Val Pro Leu Glu Thr Pro Arg Val
130 135 140

His Ser Arg Ala Pro Ser Pro Leu Tyr Ser Val Glu Phe Ser Glu Glu
145 150 155 160

Pro Phe Gly Val Ile Val His Arg Gln Leu Asp Gly Arg Val Leu Leu
165 170 175

Asn Thr Thr Val Ala Pro Leu Phe Phe Ala Asp Gln Phe Leu Gln Leu
180 185 190

Ser Thr Ser Leu Pro Ser Gln Tyr Ile Thr Gly Leu Ala Glu His Leu
195 200 205

Ser Pro Leu Met Leu Ser Thr Ser Trp Thr Arg Ile Thr Leu Trp Asn
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Arg Asp Leu Ala Pro Thr Pro Gly Ala Asn Leu Tyr Gly Ser His Pro
225 230 235 240

Phe Tyr Leu Ala Leu Glu Asp Gly Gly Ser Ala His Gly Val Phe Leu
245 250 255

Leu Asn Ser Asn Ala Met Asp Val Val Leu Gln Pro Ser Pro Ala Leu
260 265 270

Ser Trp Arg Ser Thr Gly Gly Ile Leu Asp Val Tyr Ile Phe Leu Gly
275 280 285

Pro Glu Pro Lys Ser Val Val Gln Gln Tyr Leu Asp Val Val Gly Tyr
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Pro Phe Met Pro Pro Tyr Trp Gly Leu Gly Phe His Leu Cys Arg Trp
305 310 315 320

Gly Tyr Ser Ser Thr Ala Ile Thr Arg Gln Val Val Glu Asn Met Thr
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Arg Ala His Phe Pro Leu Asp Val Gln Trp Asn Asp Leu Asp Tyr Met
340 345 350

Asp Ser Arg Arg Asp Phe Thr Phe Asn Lys Asp Gly Phe Arg Asp Phe
355 360 365

Pro Ala Met Val Gln Glu Leu His Gln Gly Gly Arg Arg Tyr Met Met
370 375 380

Ile Val Asp Pro Ala Ile Ser Ser Ser Gly Pro Ala Gly Ser Tyr Arg
385 390 395 400

Pro Tyr Asp Glu Gly Leu Arg Arg Gly Val Phe Ile Thr Asn Glu Thr
405 410 415

Gly Gln Pro Leu Ile Gly Lys Val Trp Pro Gly Ser Thr Ala Phe Pro
420 425 430

Asp Phe Thr Asn Pro Thr Ala Leu Ala Trp Trp Glu Asp Met Val Ala
435 440 445

Glu Phe His Asp Gln Val Pro Phe Asp Gly Met Trp Ile Asp Met Asn
450 455 460

Glu Pro Ser Asn Phe Ile Arg Gly Ser Glu Asp Gly Cys Pro Asn Asn
465 470 475 480

Glu Leu Glu Asn Pro Pro Tyr Val Pro Gly Val Val Gly Gly Thr Leu
485 490 495

Gln Ala Ala Thr Ile Cys Ala Ser Ser His Gln Phe Leu Ser Thr His
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Tyr Asn Leu His Asn Leu Tyr Gly Leu Thr Glu Ala Ile Ala Ser His
515 520 525

Arg Ala Leu Val Lys Ala Arg Gly Thr Arg Pro Phe Val Ile Ser Arg
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Ser Thr Phe Ala Gly His Gly Arg Tyr Ala Gly His Trp Thr Gly Asp
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Val Trp Ser Ser Trp Glu Gln Leu Ala Ser Ser Val Pro Glu Ile Leu
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Gln Phe Asn Leu Leu Gly Val Pro Leu Val Gly Ala Asp Val Cys Gly
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Phe Leu Gly Asn Thr Ser Glu Glu Leu Cys Val Arg Trp Thr Gln Leu
595 600 605

Gly Ala Phe Tyr Pro Phe Met Arg Asn His Asn Ser Leu Leu Ser Leu
610 615 620

Pro Gln Glu Pro Tyr Ser Phe Ser Glu Pro Ala Gln Gln Ala Met Arg
625 630 635 640

Lys Ala Leu Thr Leu Arg Tyr Ala Leu Leu Pro His Leu Tyr Thr Leu
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Phe His Gln Ala His Val Ala Gly Glu Thr Val Ala Arg Pro Leu Phe
660 665 670

Leu Glu Phe Pro Lys Asp Ser Ser Thr Trp Thr Val Asp His Gln Leu
675 680 685

Leu Trp Gly Glu Ala Leu Leu Ile Thr Pro Val Leu Gln Ala Gly Lys
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Ala Glu Val Thr Gly Tyr Phe Pro Leu Gly Thr Trp Tyr Asp Leu Gln
705 710 715 720

Thr Val Pro Ile Glu Ala Leu Gly Ser Leu Pro Pro Pro Pro Ala Ala
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Pro Arg Glu Pro Ala Ile His Ser Glu Gly Gln Trp Val Thr Leu Pro
740 745 750

Ala Pro Leu Asp Thr Ile Asn Val His Leu Arg Ala Gly Tyr Ile Ile
755 760 765

Pro Leu Gln Gly Pro Gly Leu Thr Thr Thr Glu Ser Arg Gln Gln Pro
770 775 780

Met Ala Leu Ala Val Ala Leu Thr Lys Gly Gly Glu Ala Arg Gly Glu
785 790 795 800

Leu Phe Trp Asp Asp Gly Glu Ser Leu Glu Val Leu Glu Arg Gly Ala
805 810 815

Tyr Thr Gln Val Ile Phe Leu Ala Arg Asn Asn Thr Ile Val Asn Glu
820 825 830

Leu Val Arg Val Thr Ser Glu Gly Ala Gly Leu Gln Leu Gln Lys Val
835 840 845

Thr Val Leu Gly Val Ala Thr Ala Pro Gln Gln Val Leu Ser Asn Gly
850 855 860

Val Pro Val Ser Asn Phe Thr Tyr Ser Pro Asp Thr Lys Val Leu Asp
865 870 875 880

Ile Cys Val Ser Leu Leu Met Gly Glu Gln Phe Leu Val Ser Trp Cys
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