

Figure 1

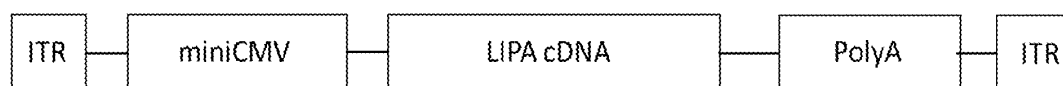


Figure 2

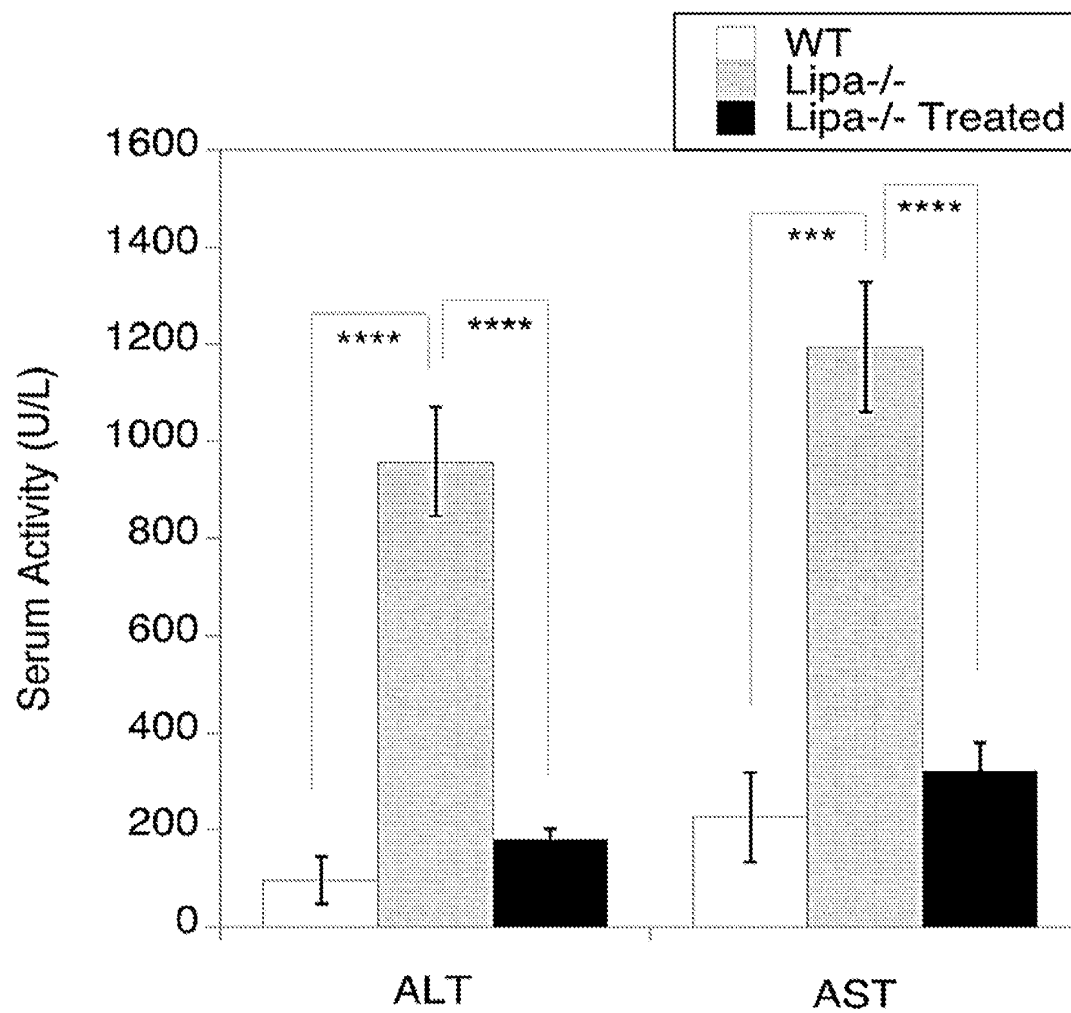


Figure 3

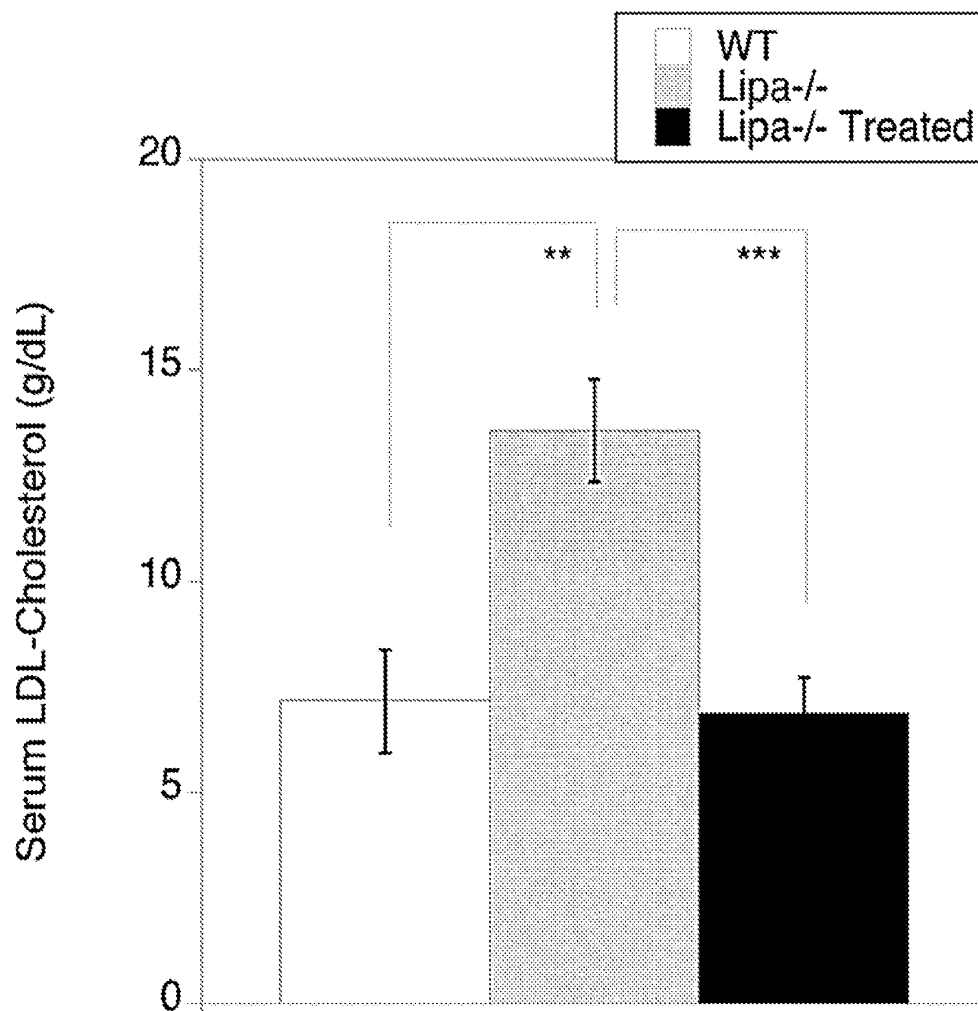


Figure 4

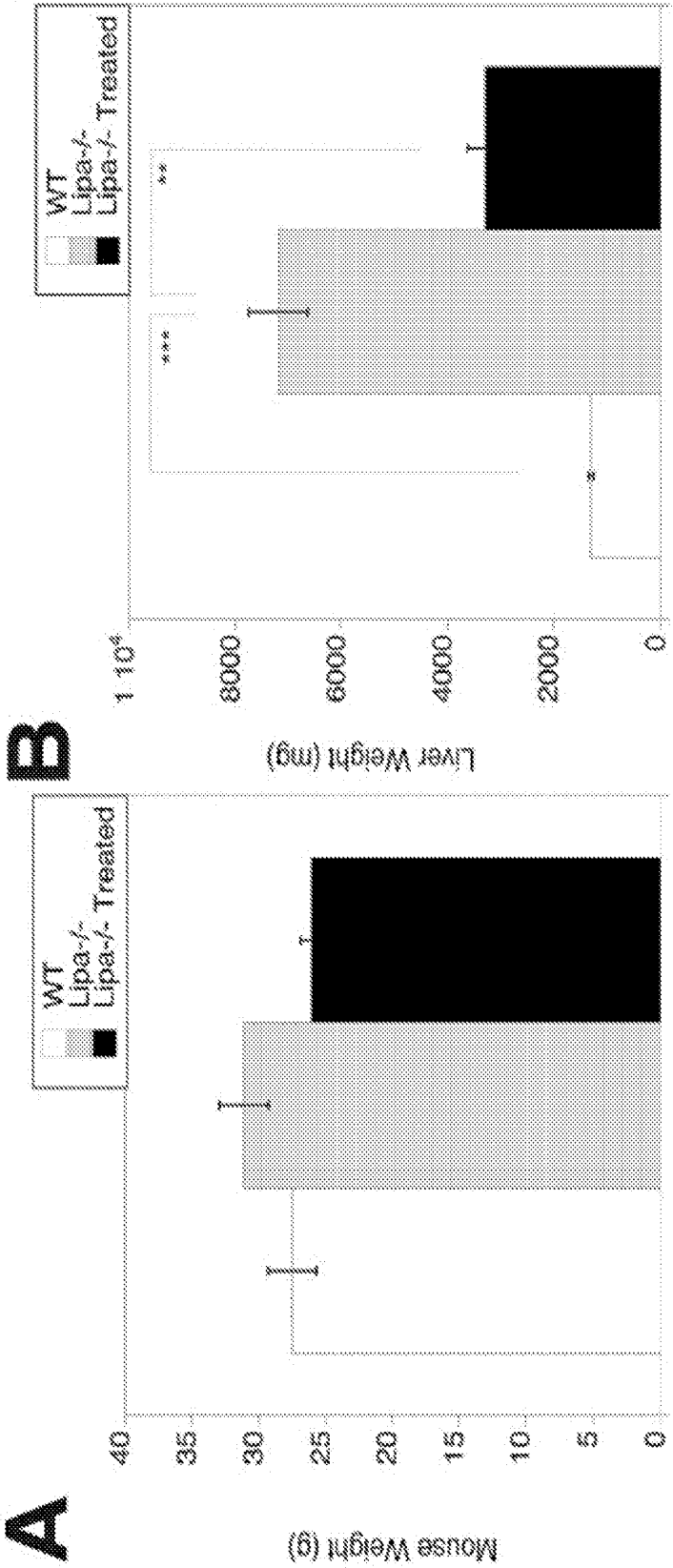


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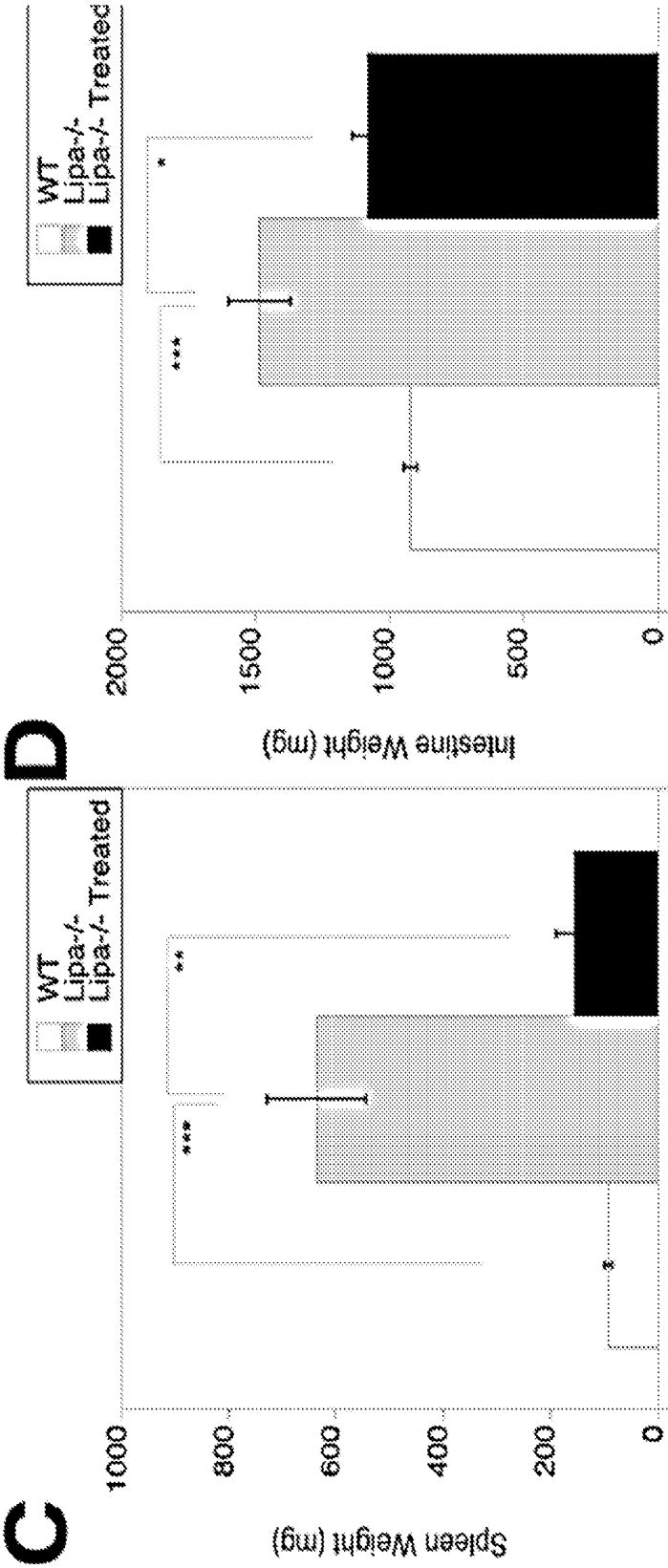
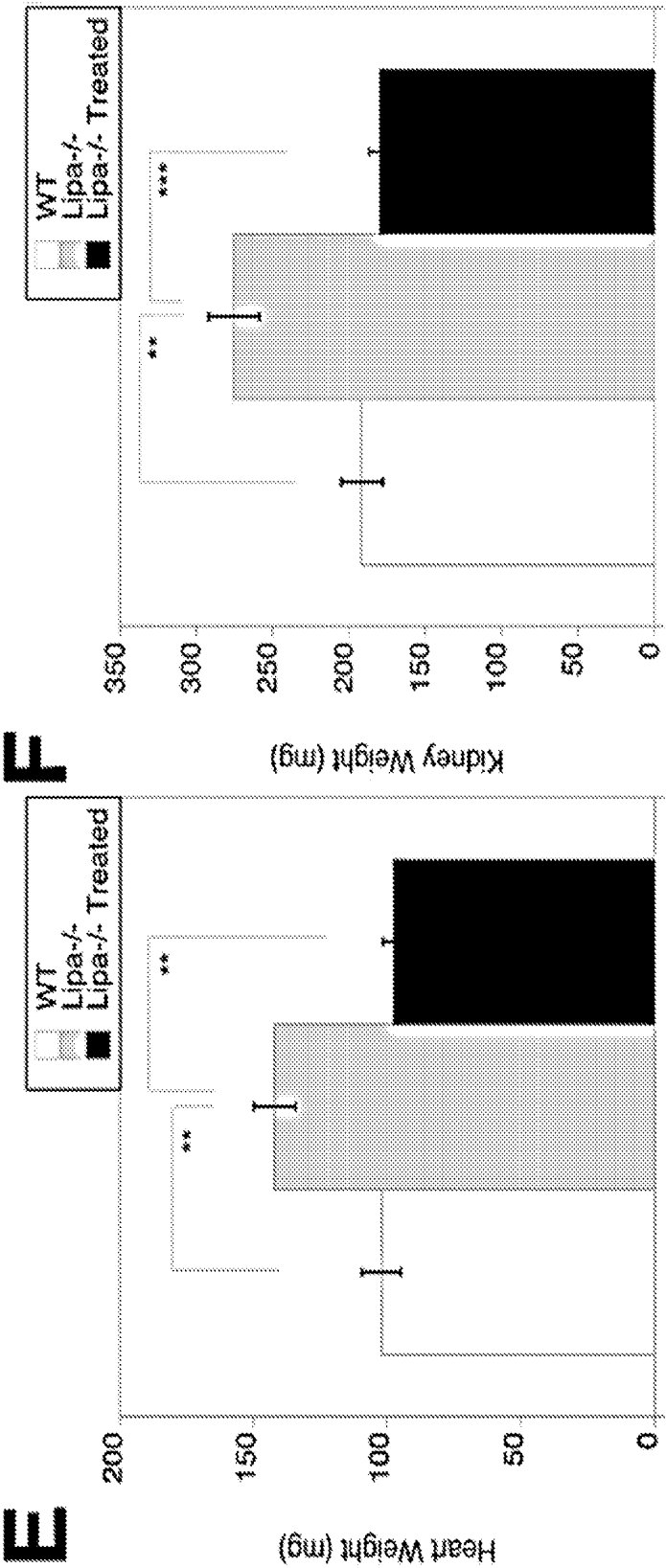


Figure 4- Continued



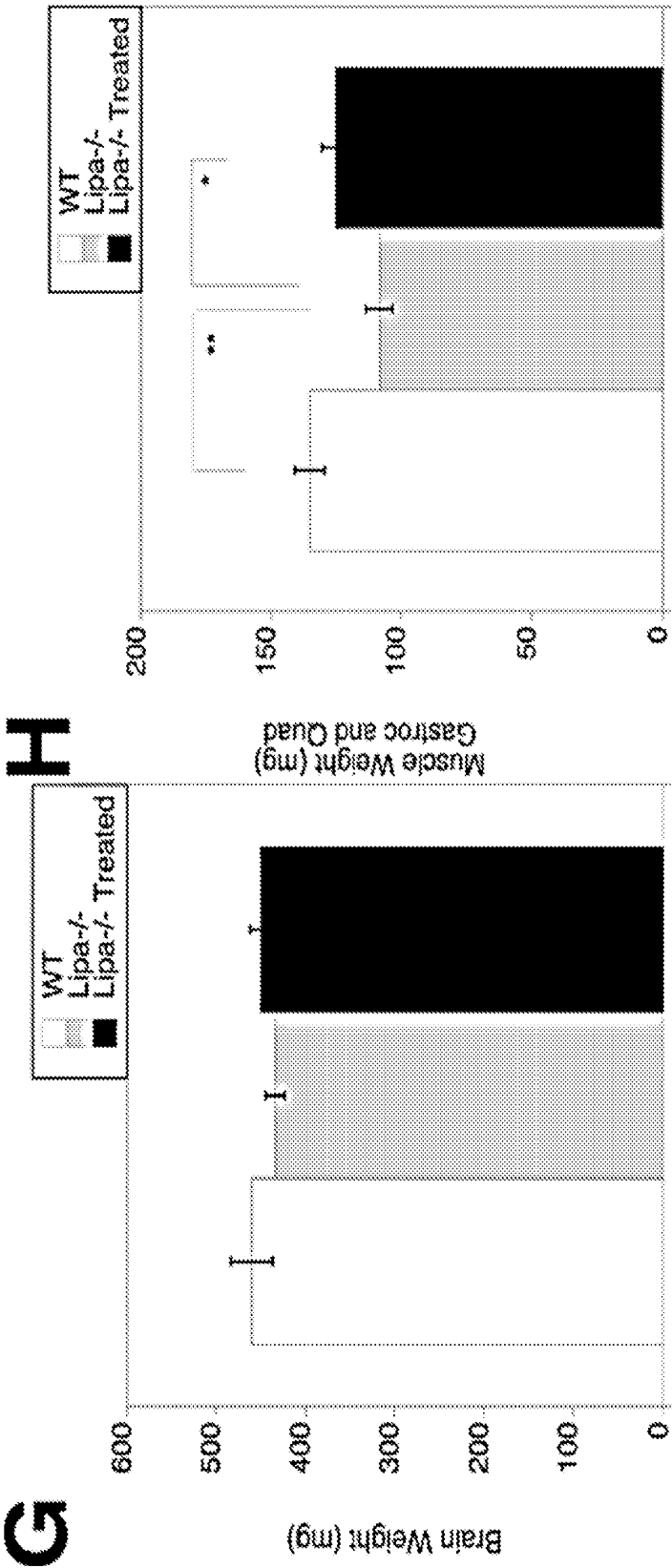
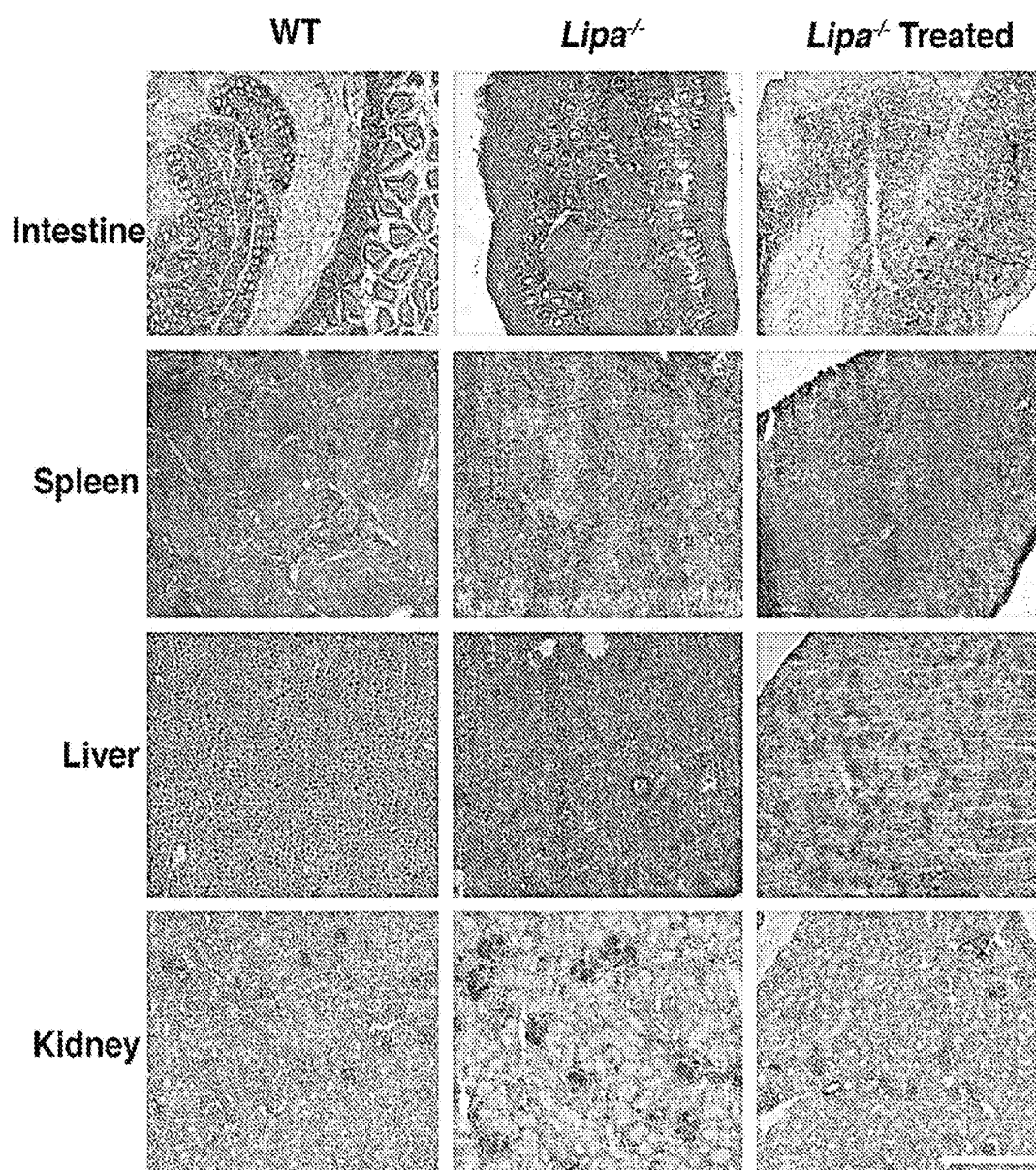


Figure 4- Continued

Figure 5



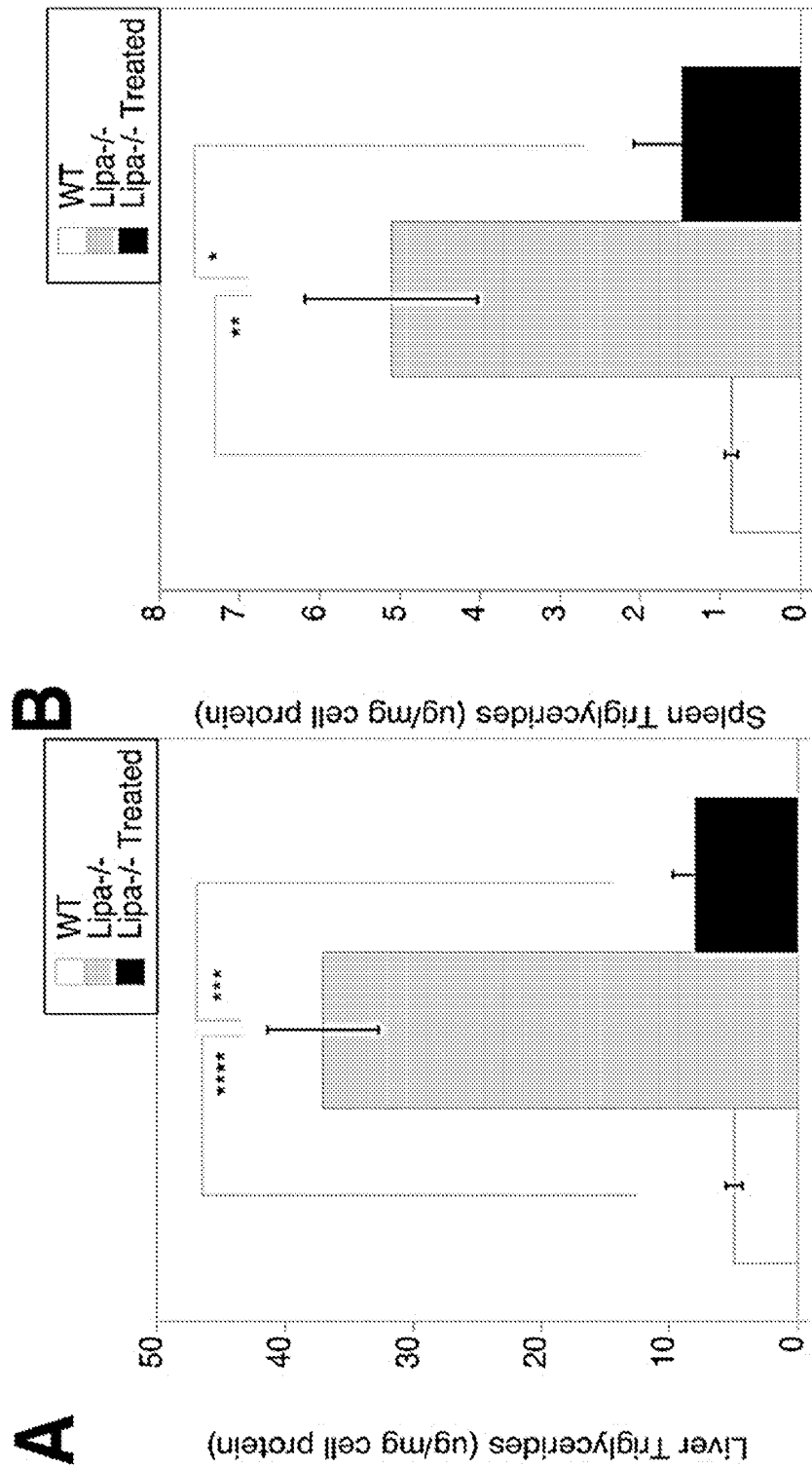


Figure 6

Figure 7

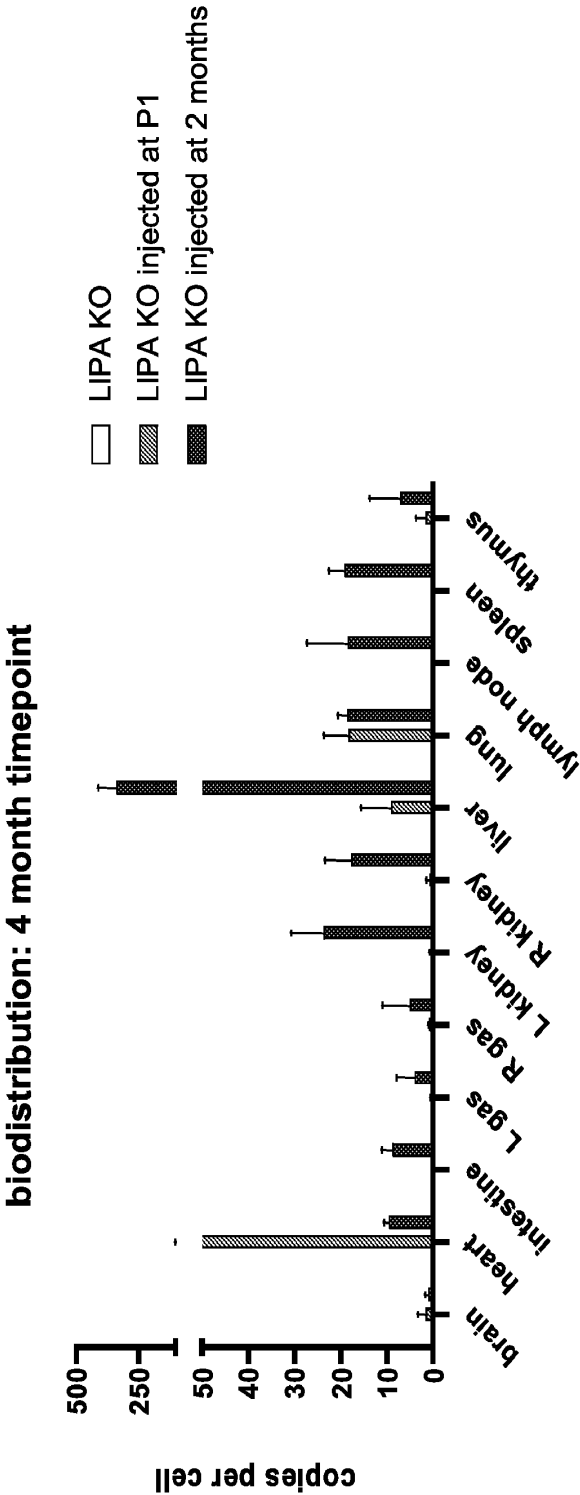


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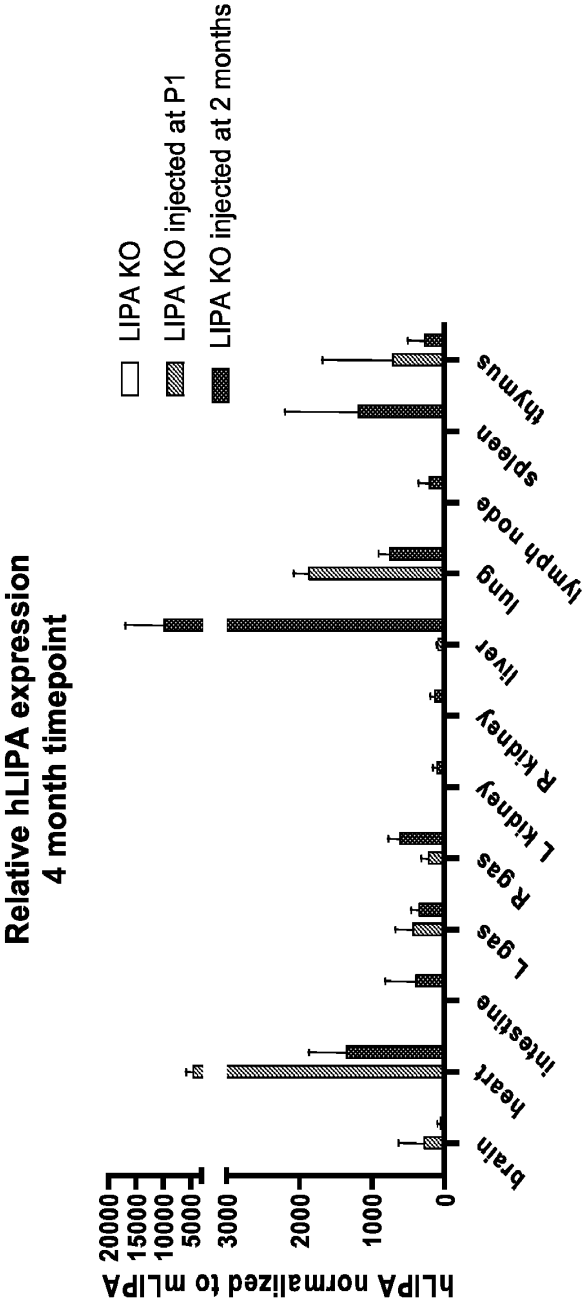


Figure 9

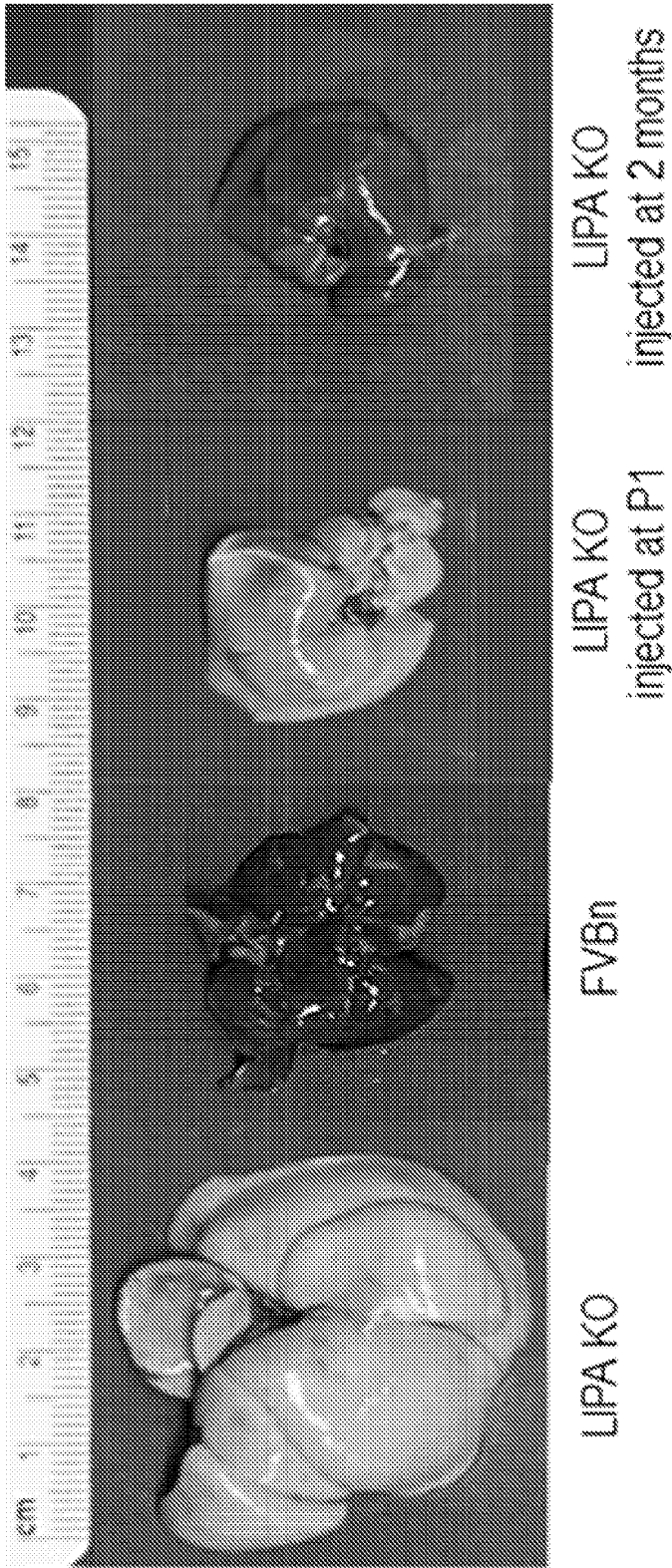


Figure 10

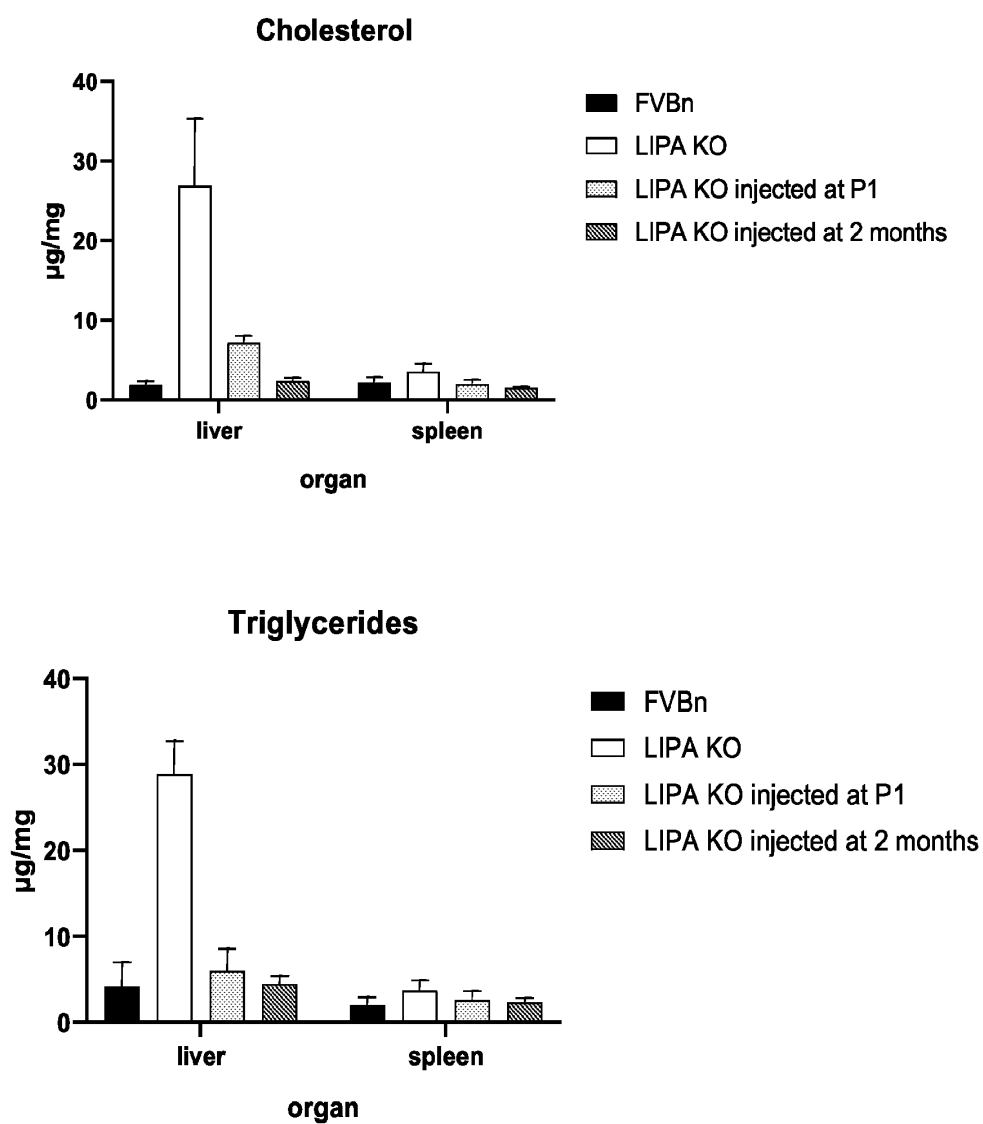


Figure 11

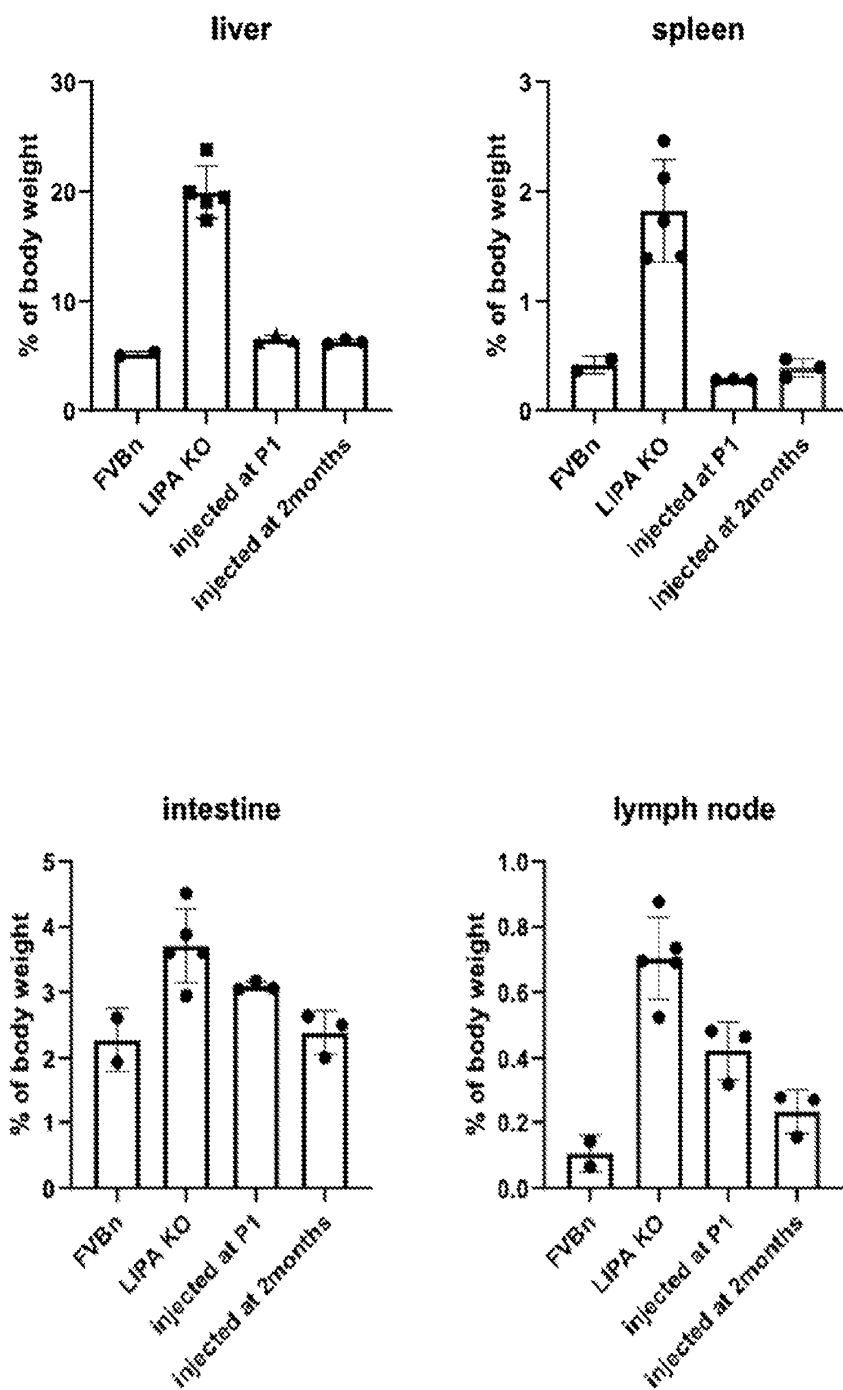


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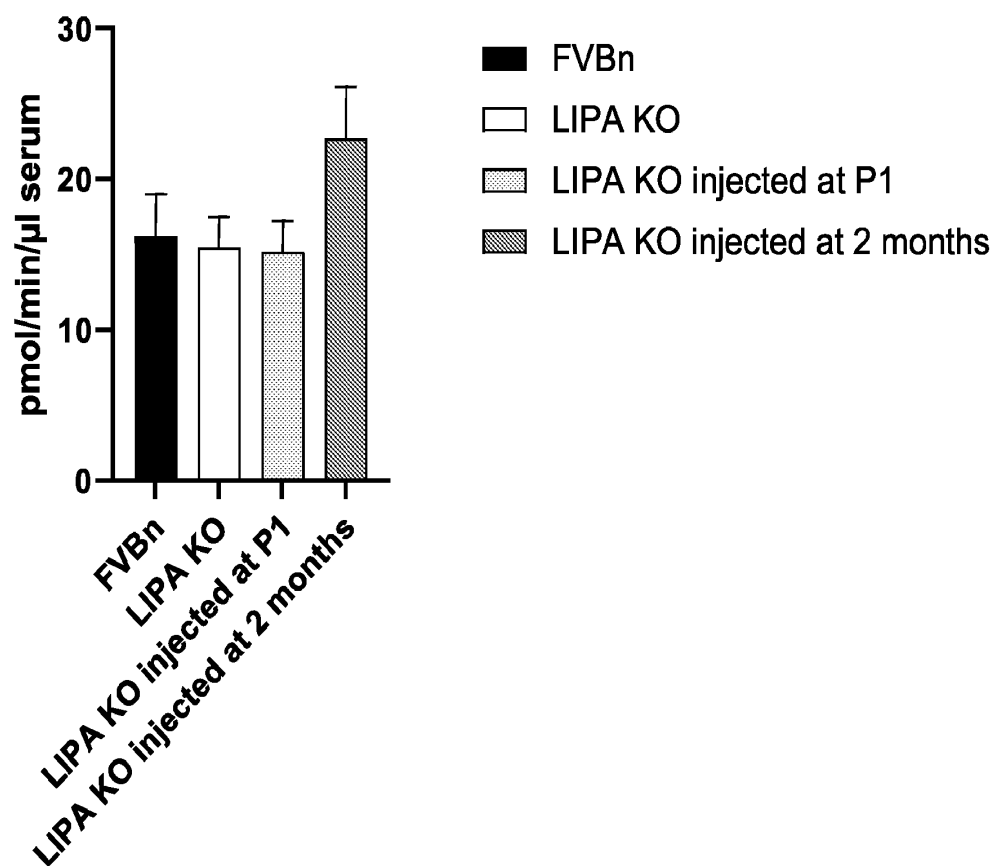
**LAL enzyme activity - serum
4 month timepoint**

Figure 13

LAL enzyme activity - liver 4 month timepoint

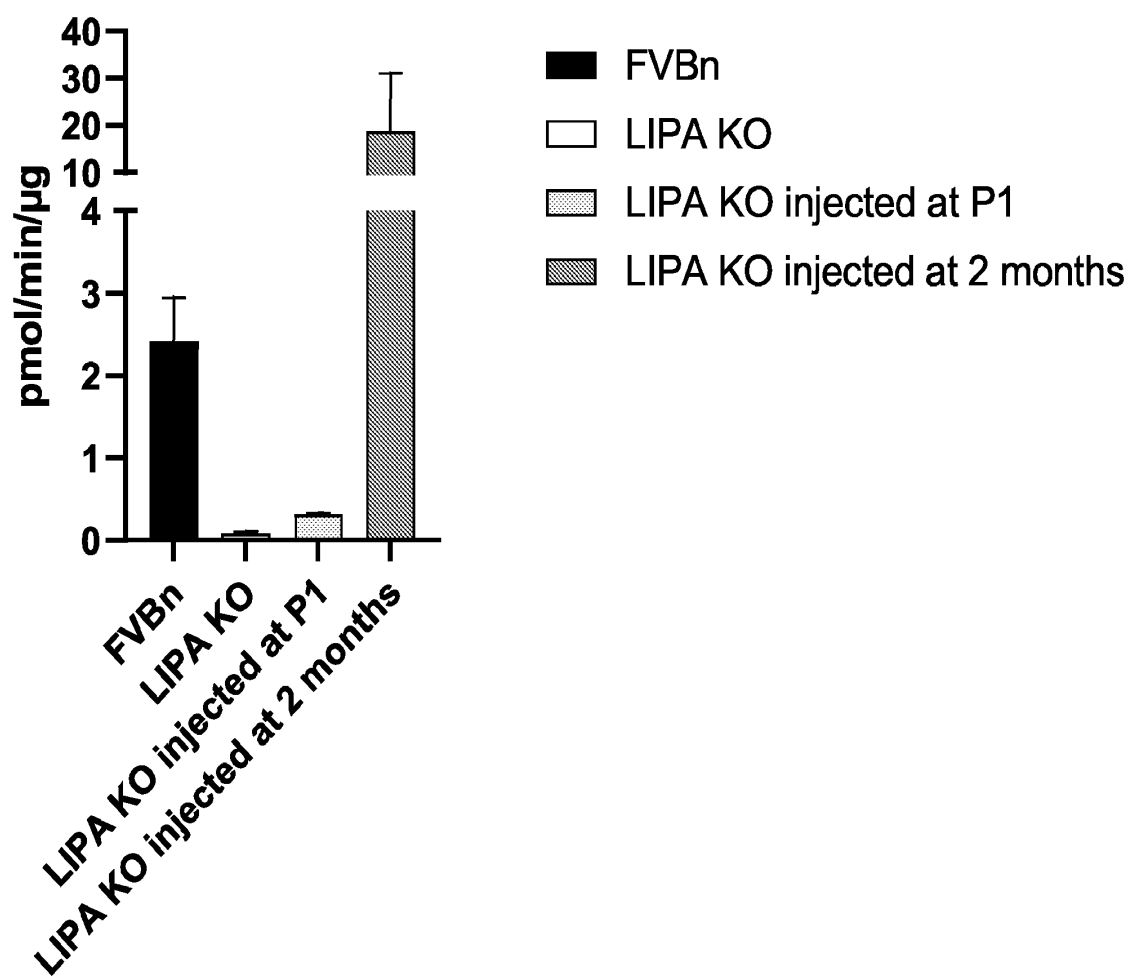


Figure 14

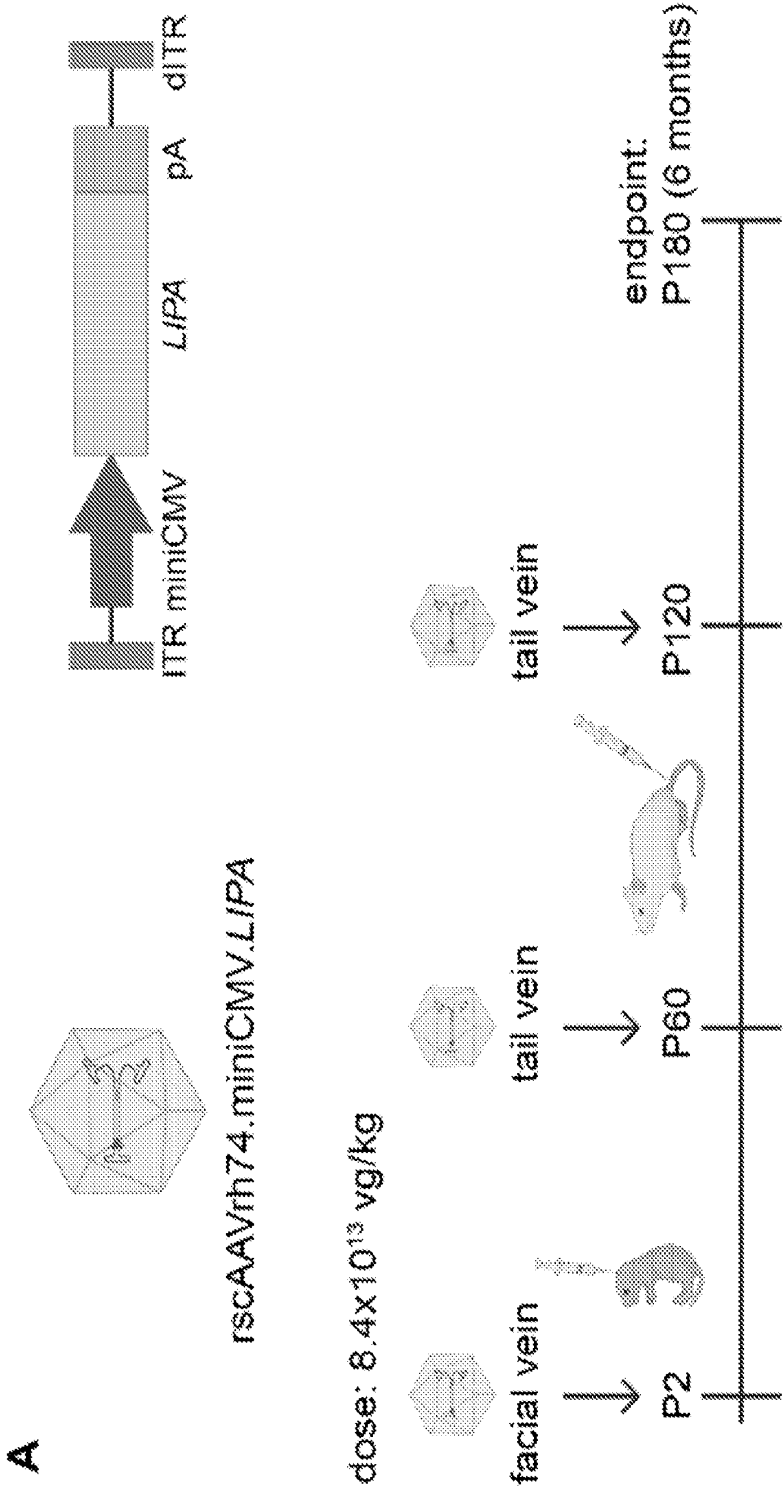


Figure 14 Continued

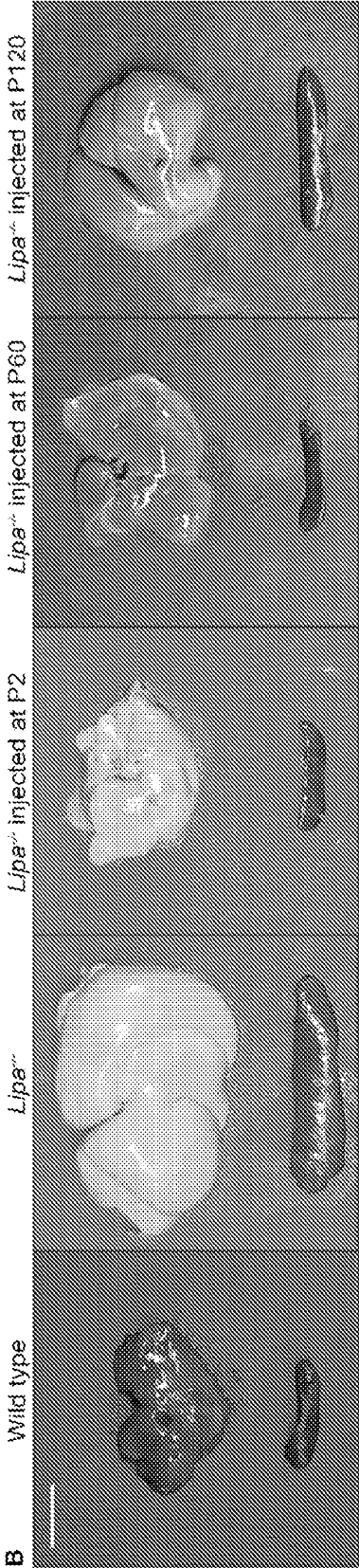


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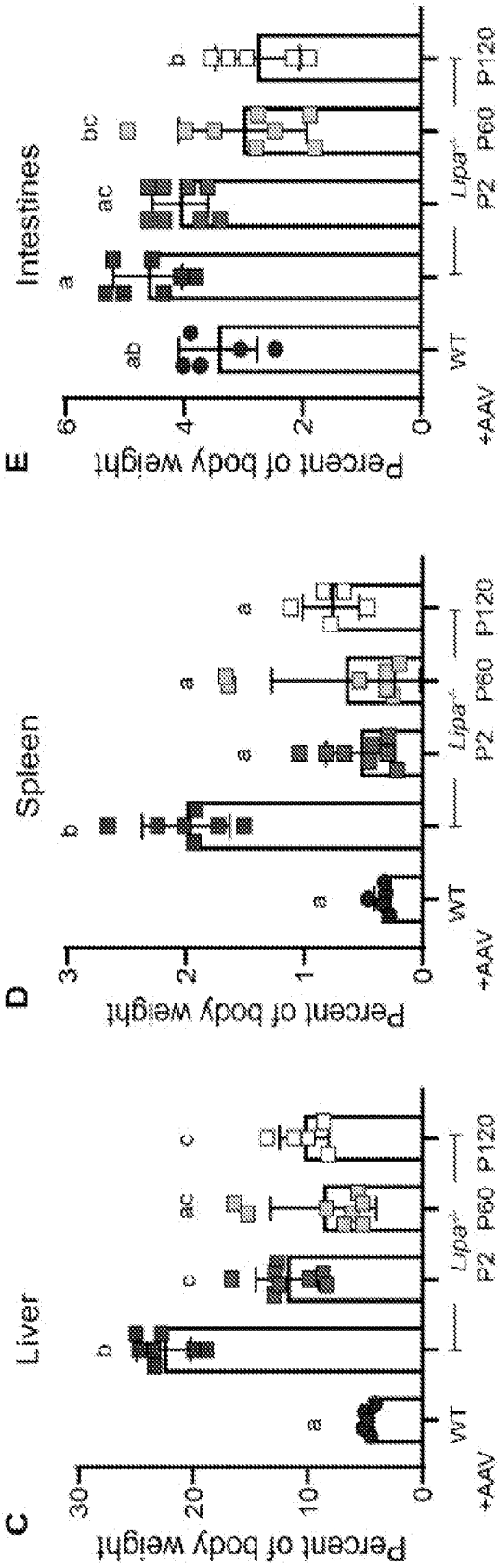


Figure 14 Continued

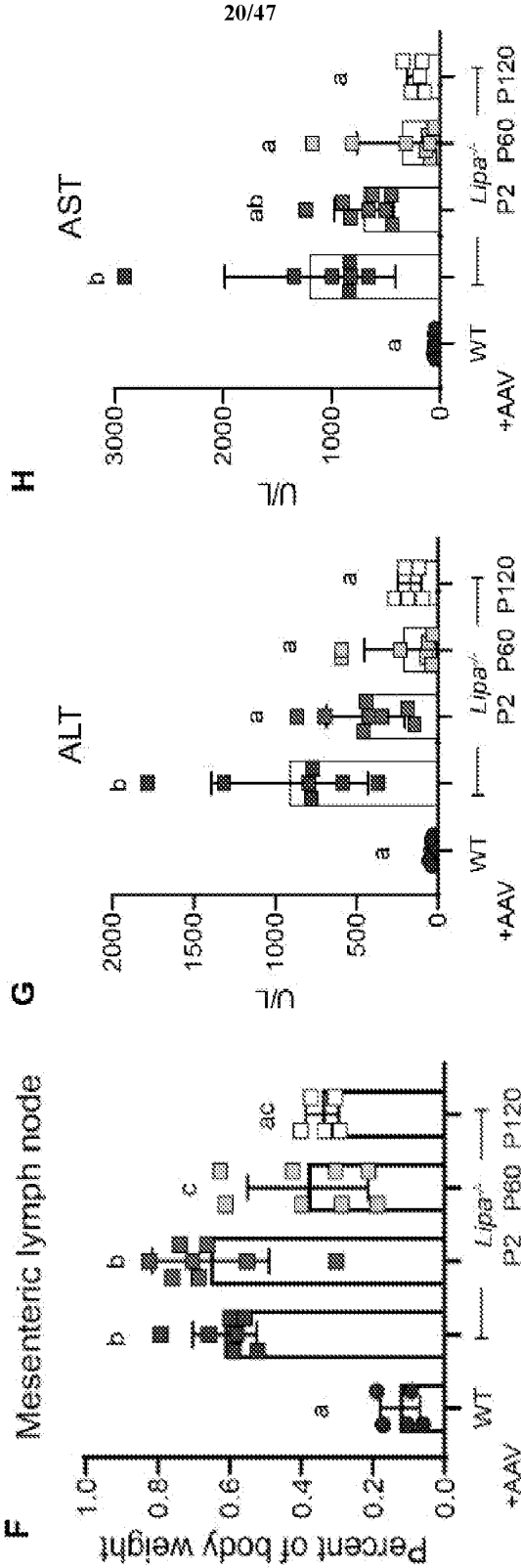


Figure 15

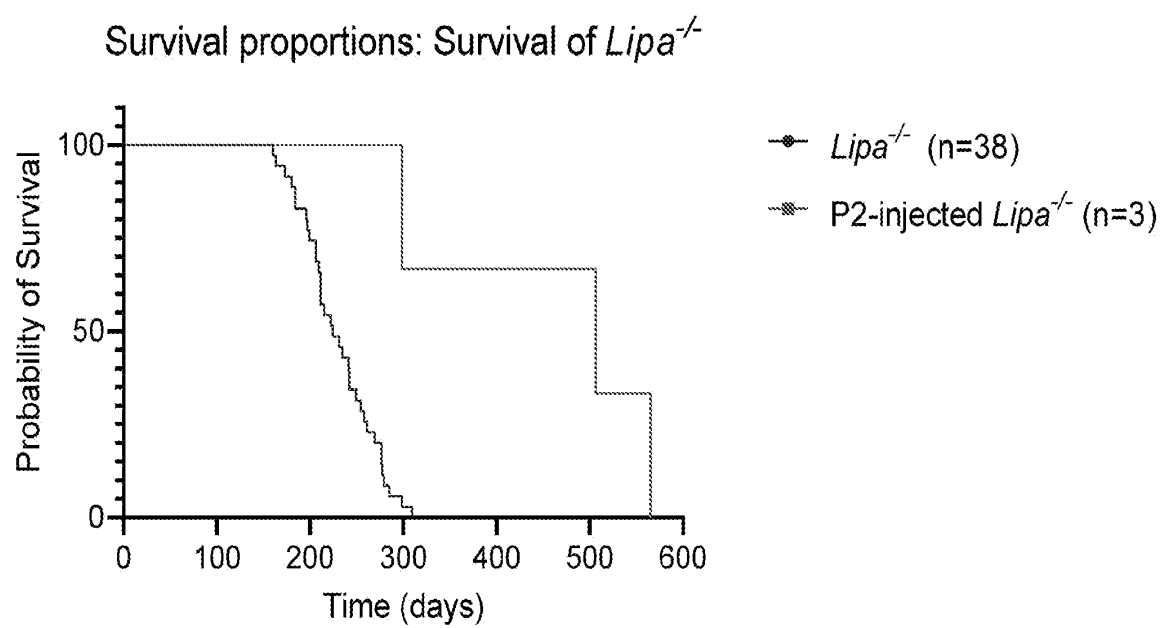


Figure 16

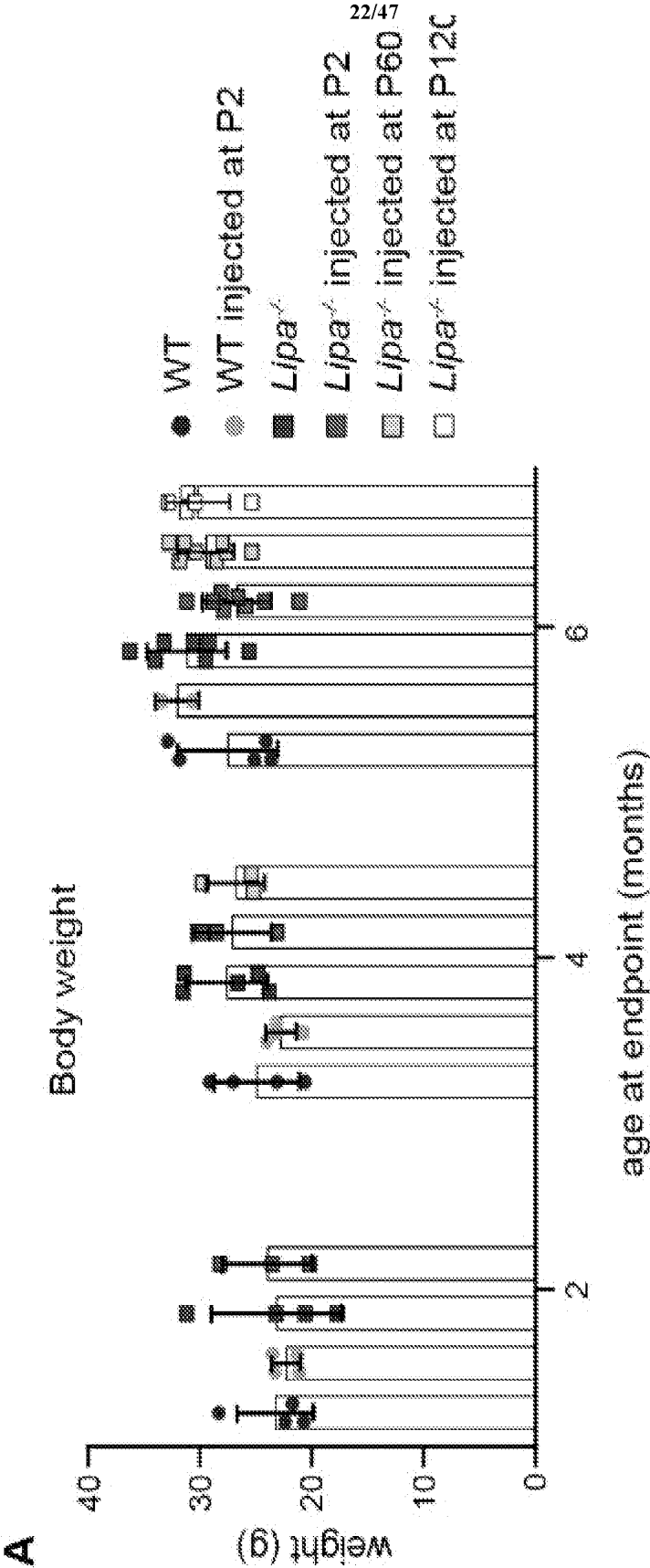


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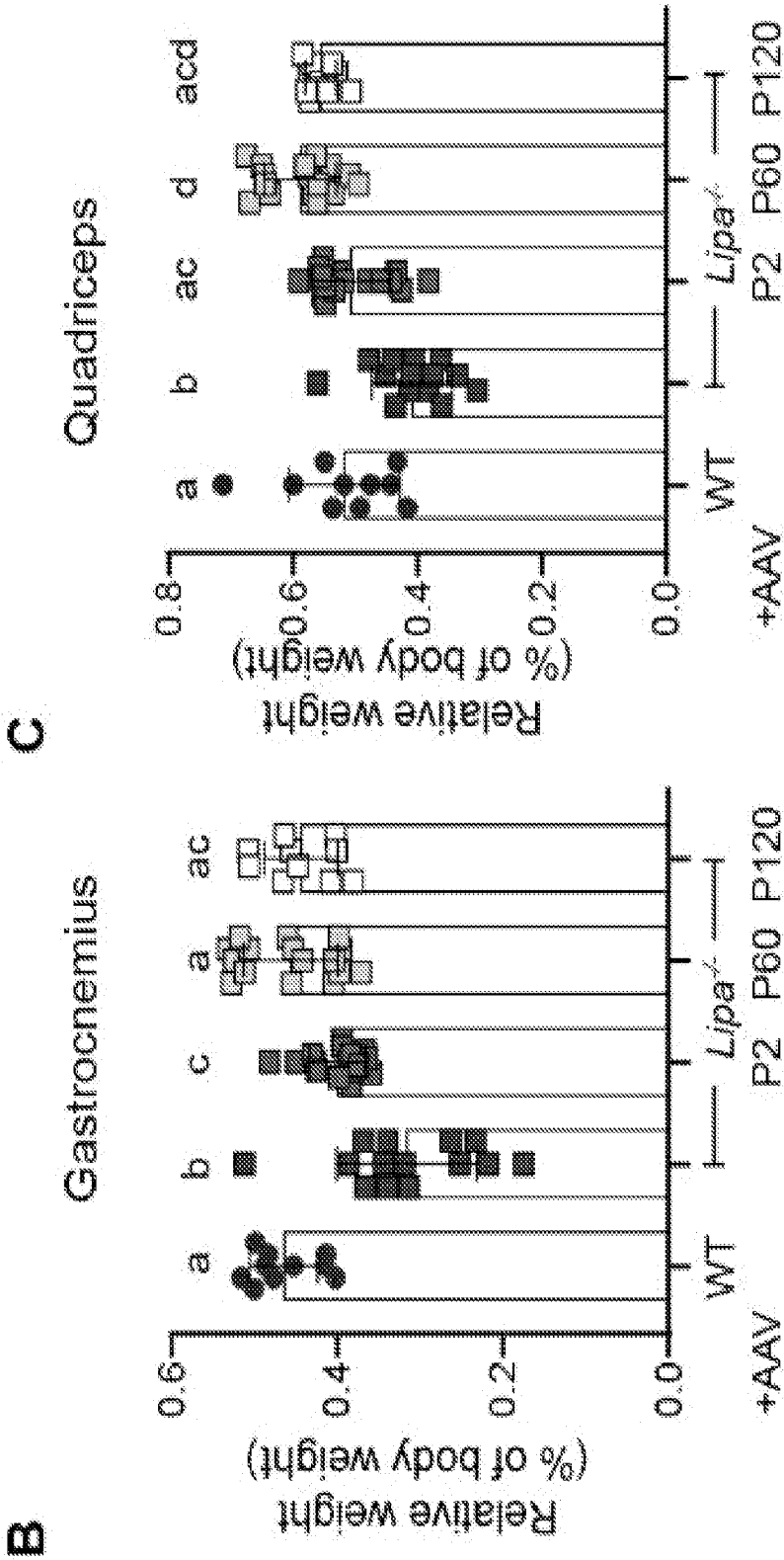


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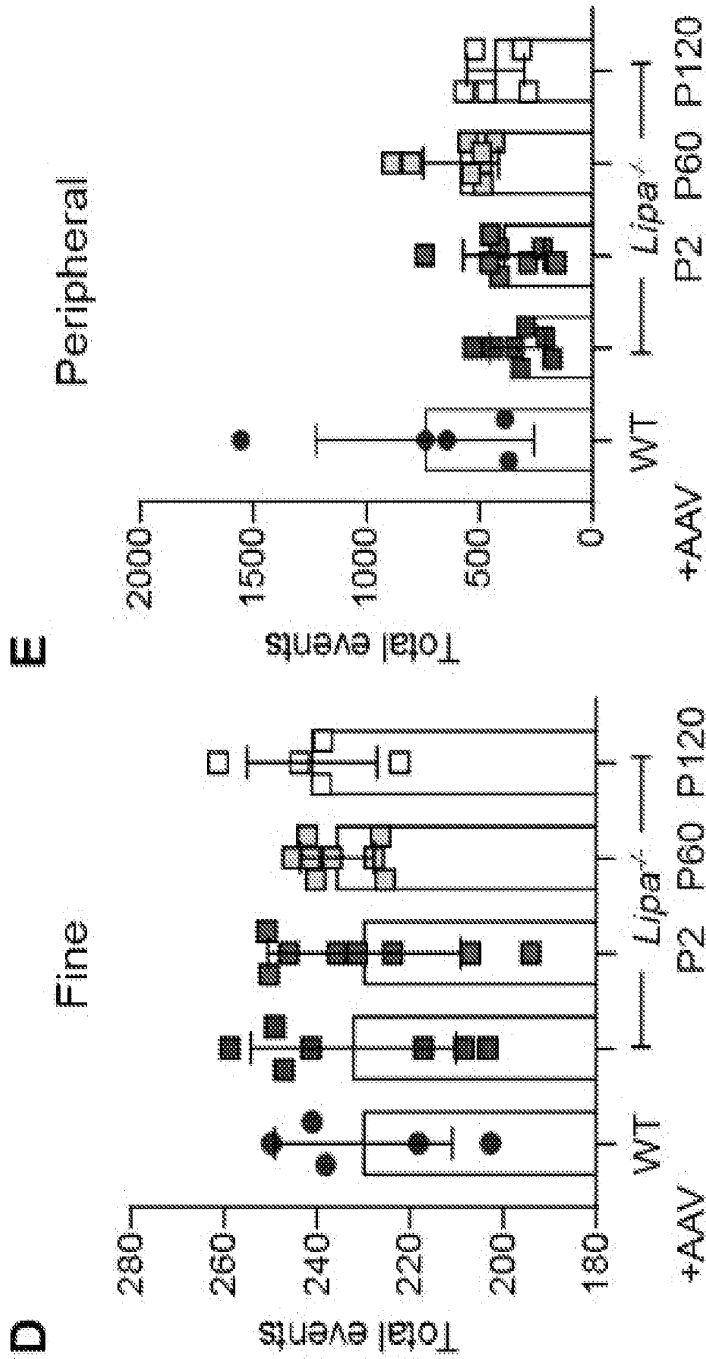


Figure 16 Continued

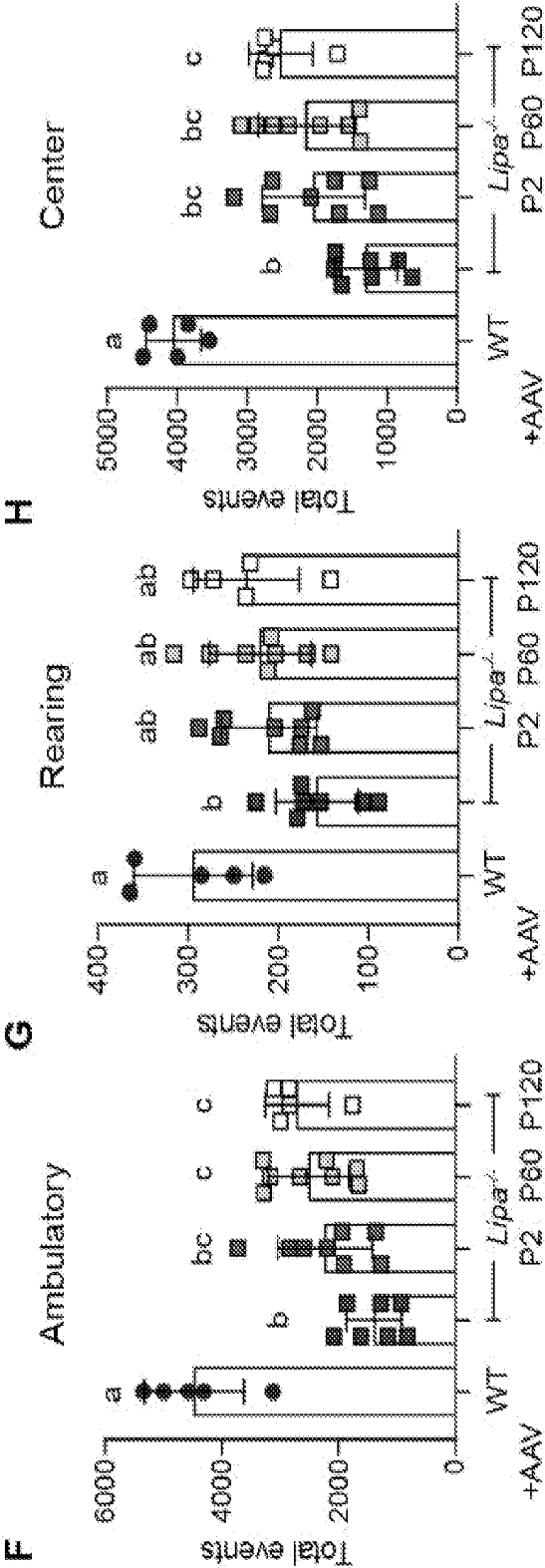


Figure 17

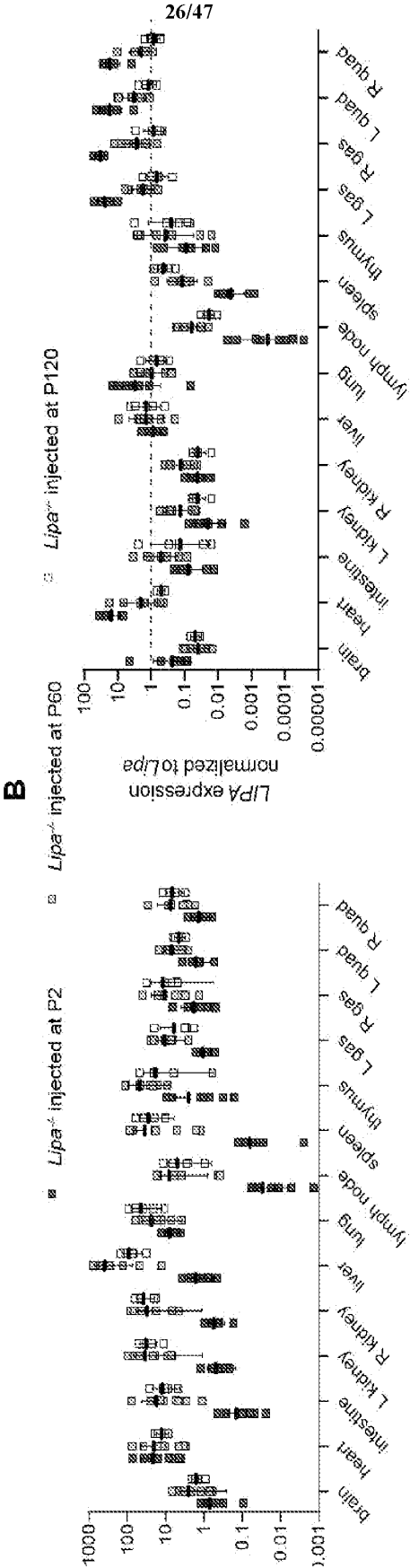


Figure 17 Continued

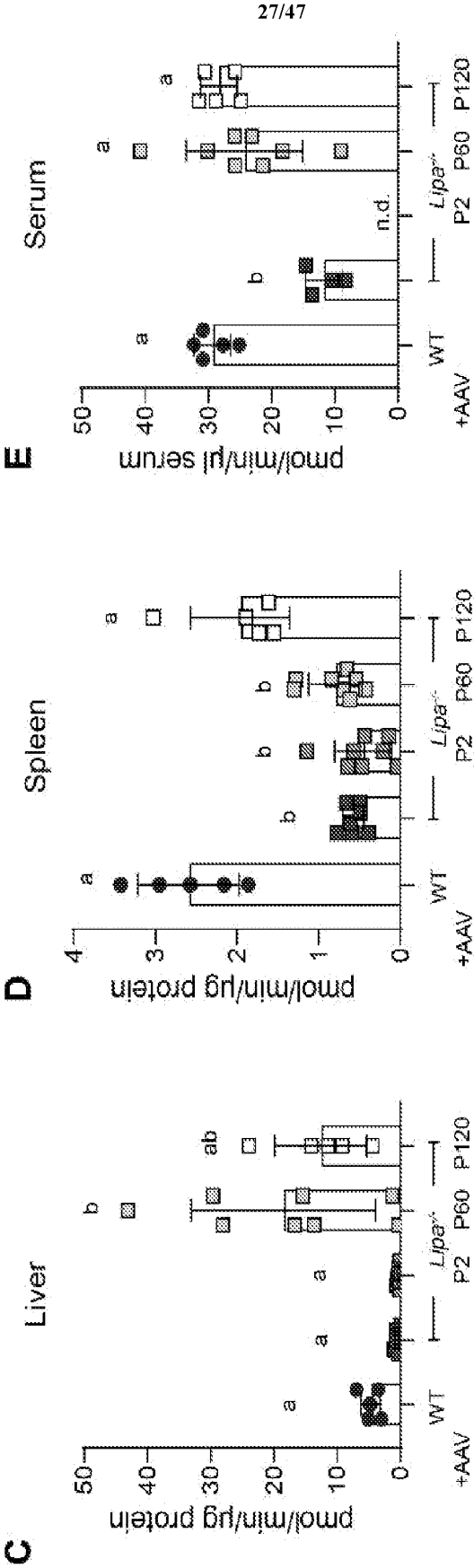


Figure 18

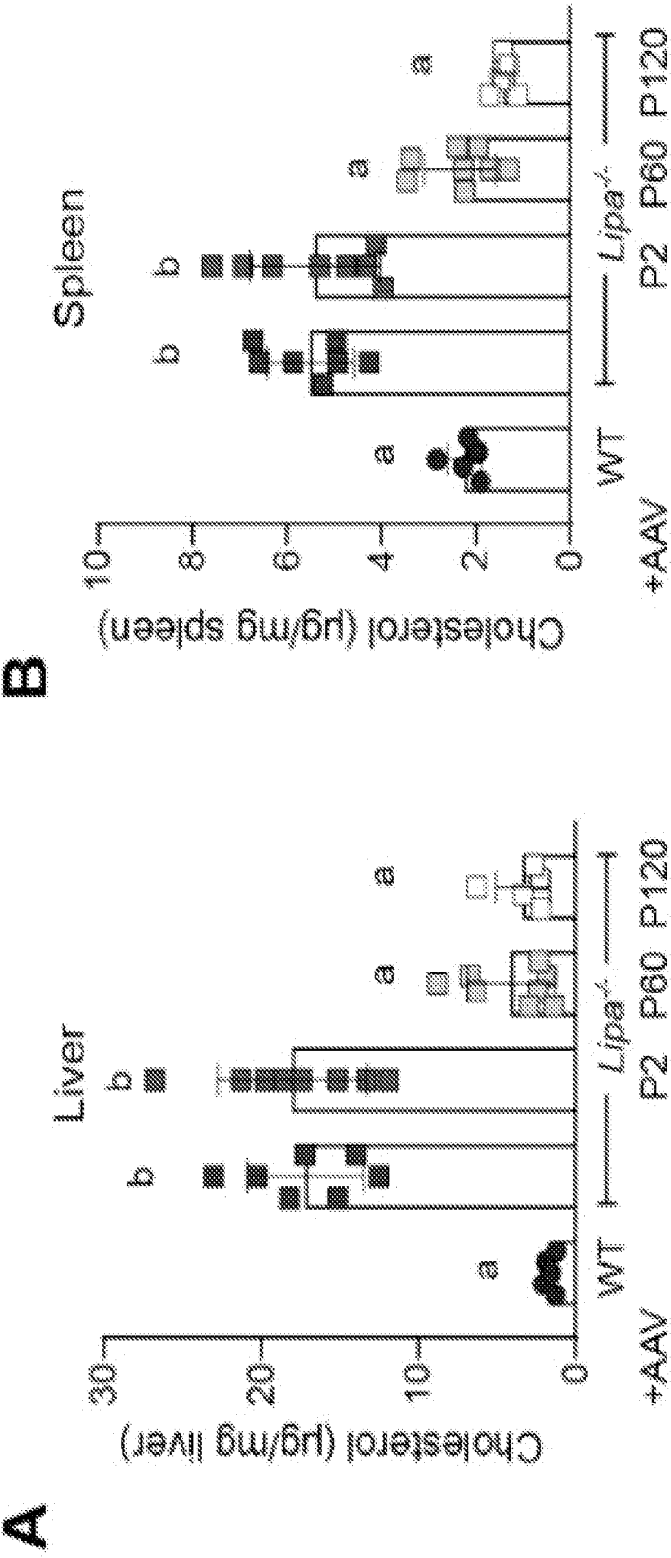


Figure 18 Continued

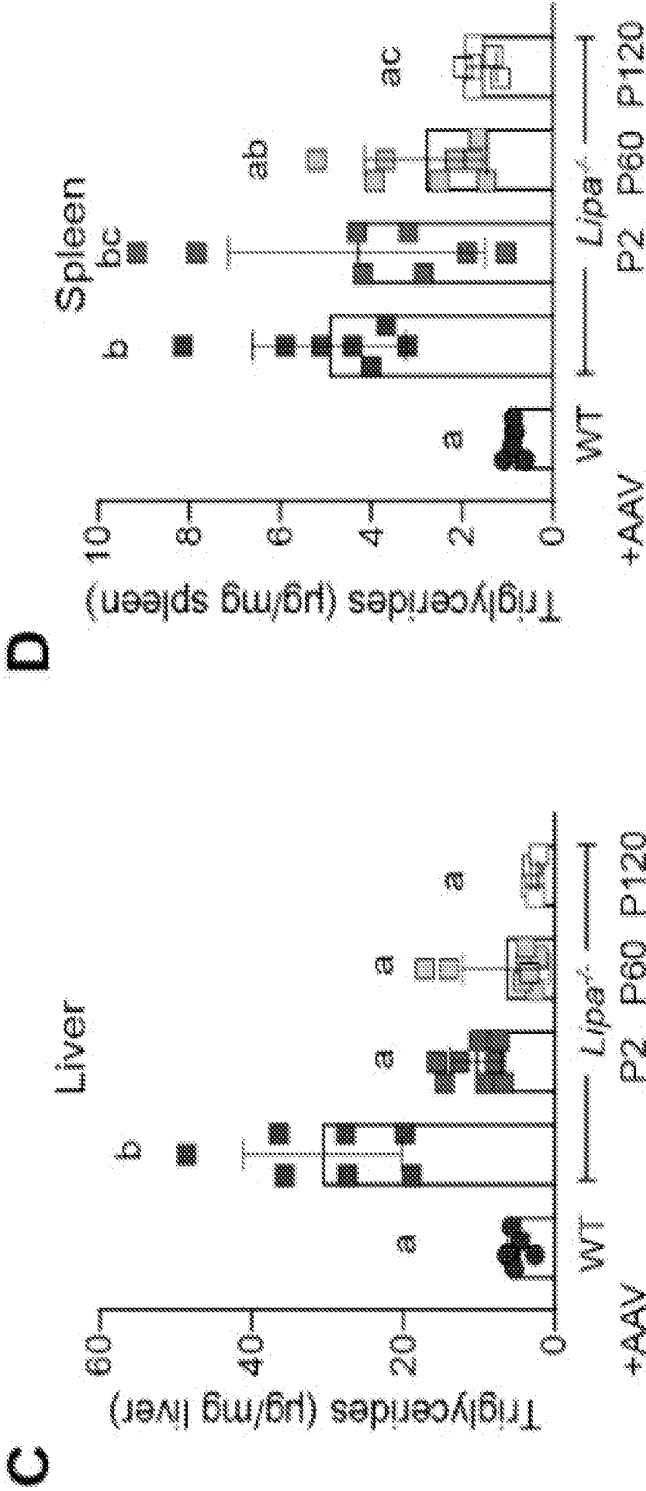


Figure 18 Continued

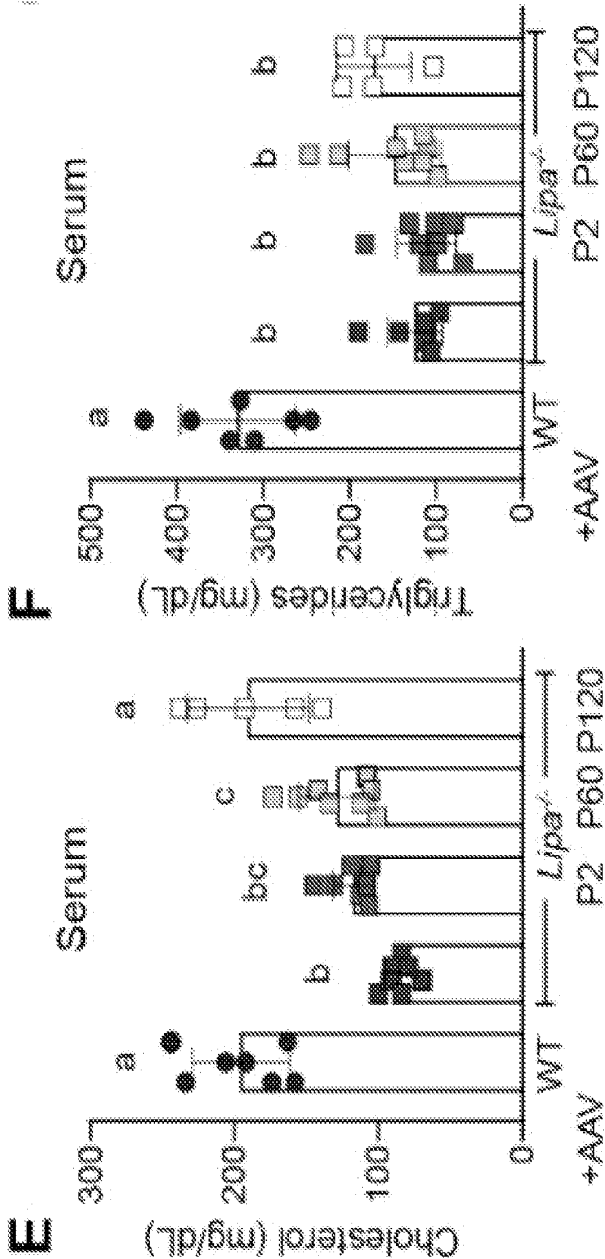


Figure 18 Continued

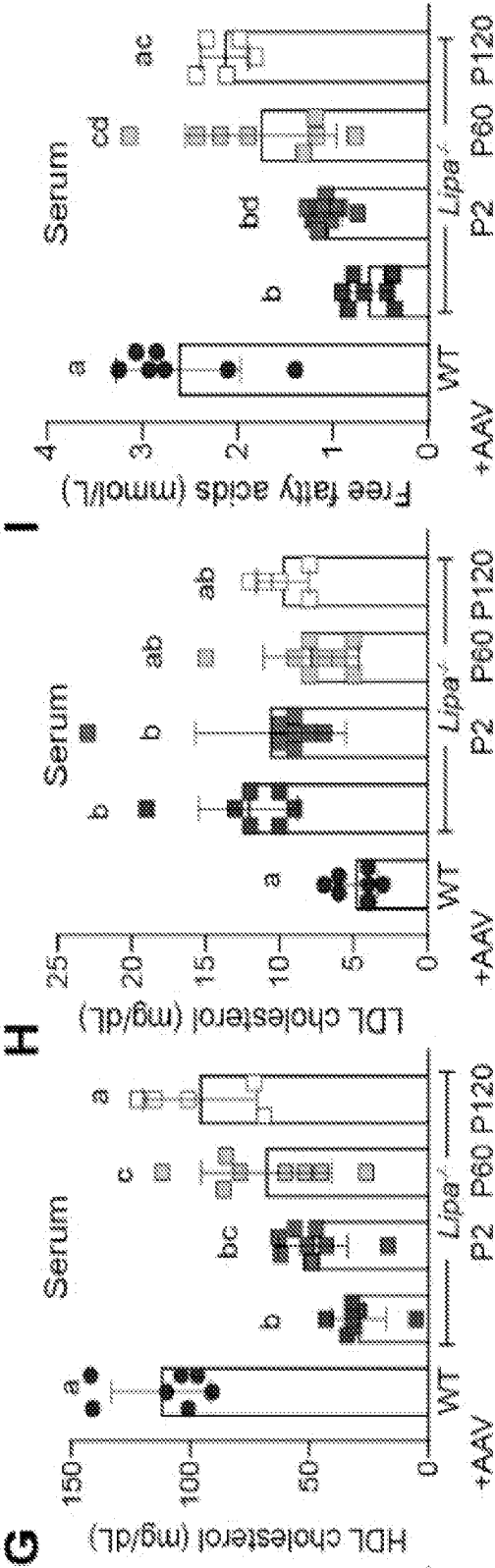


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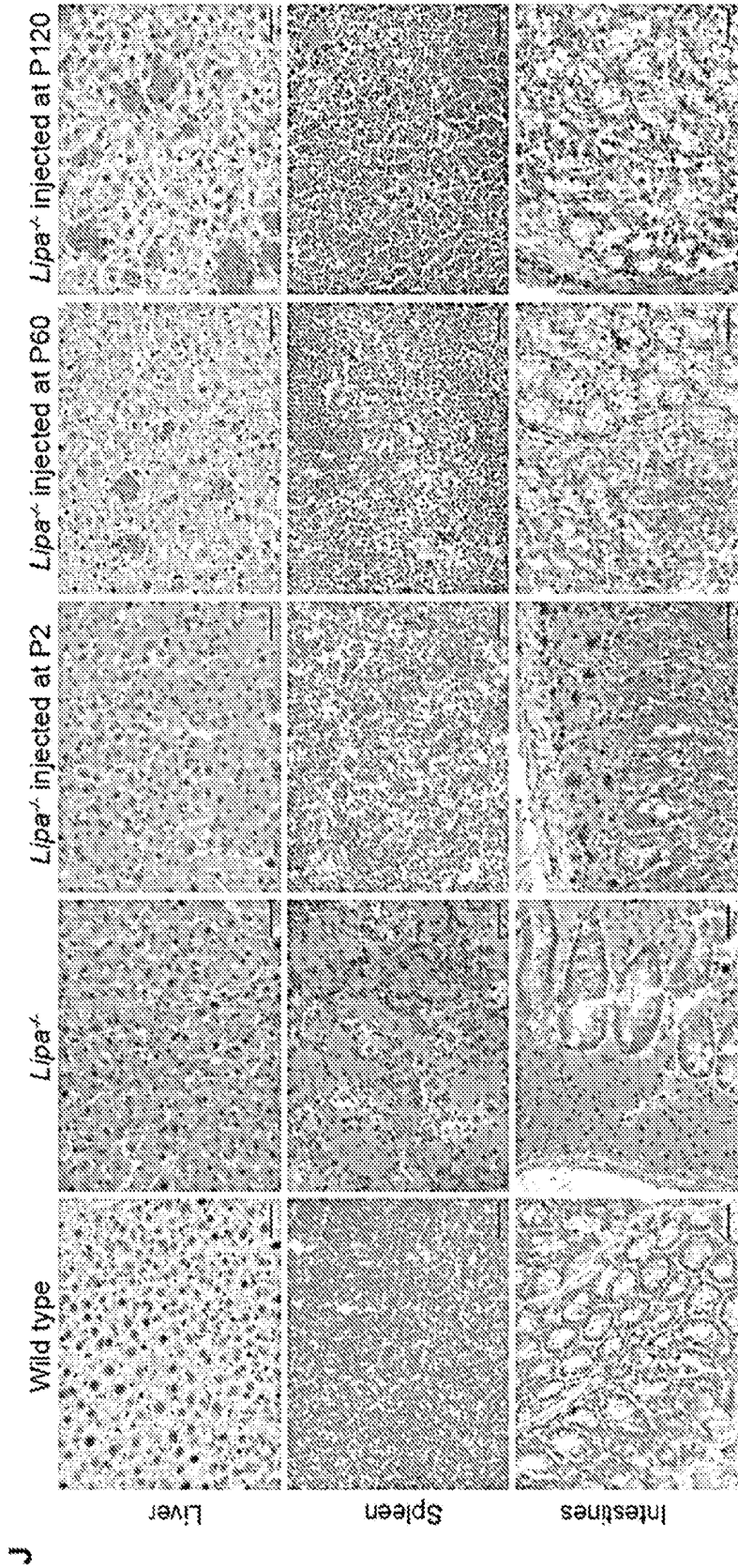


Figure 19

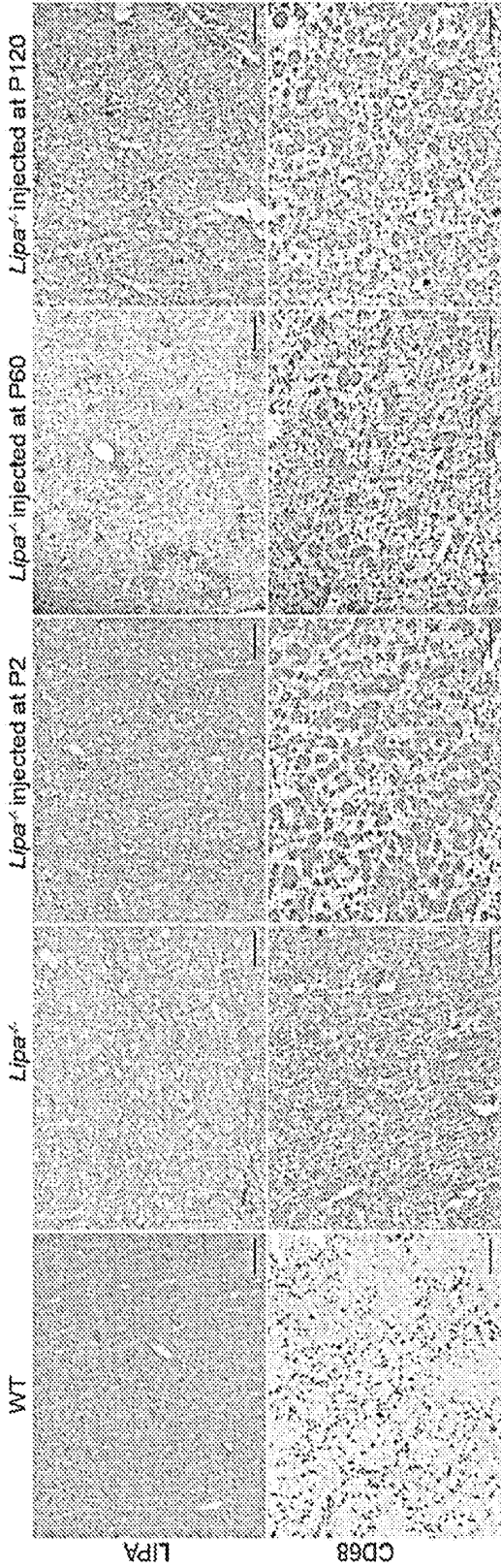


Figure 20

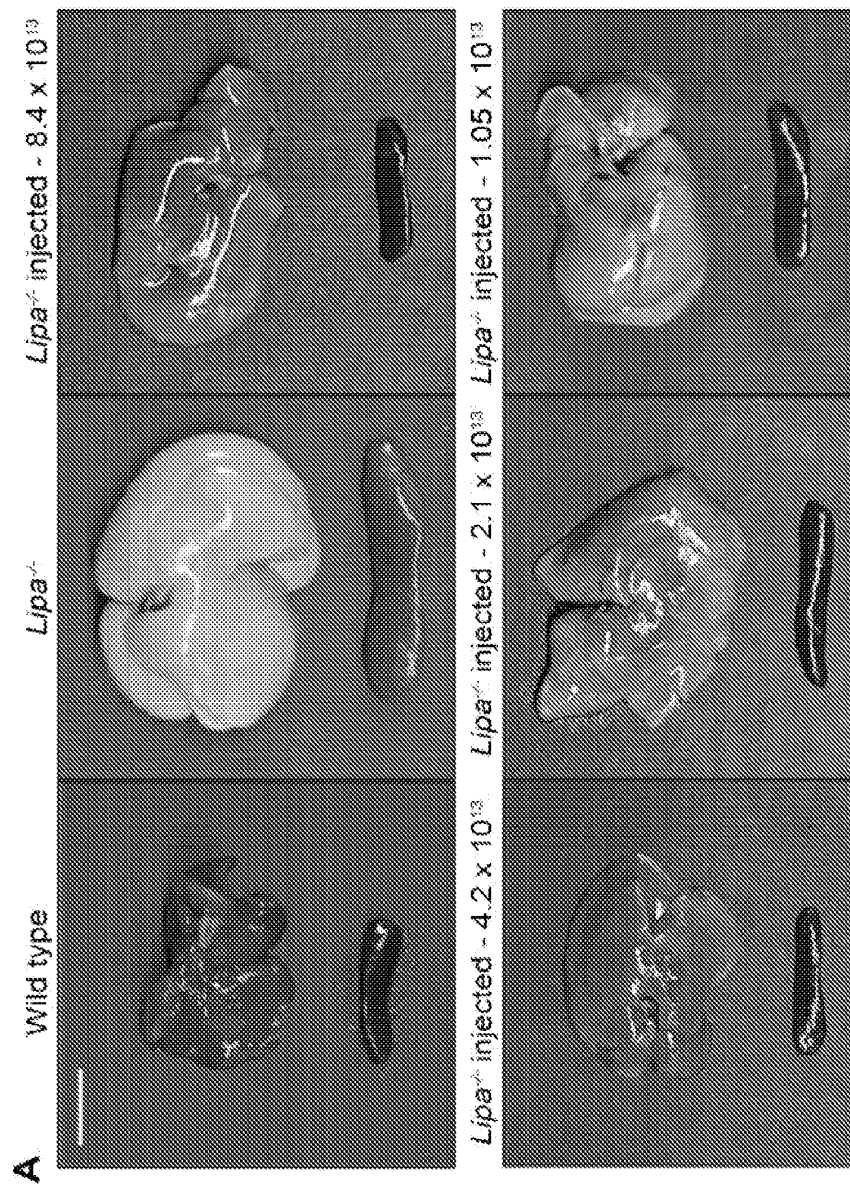


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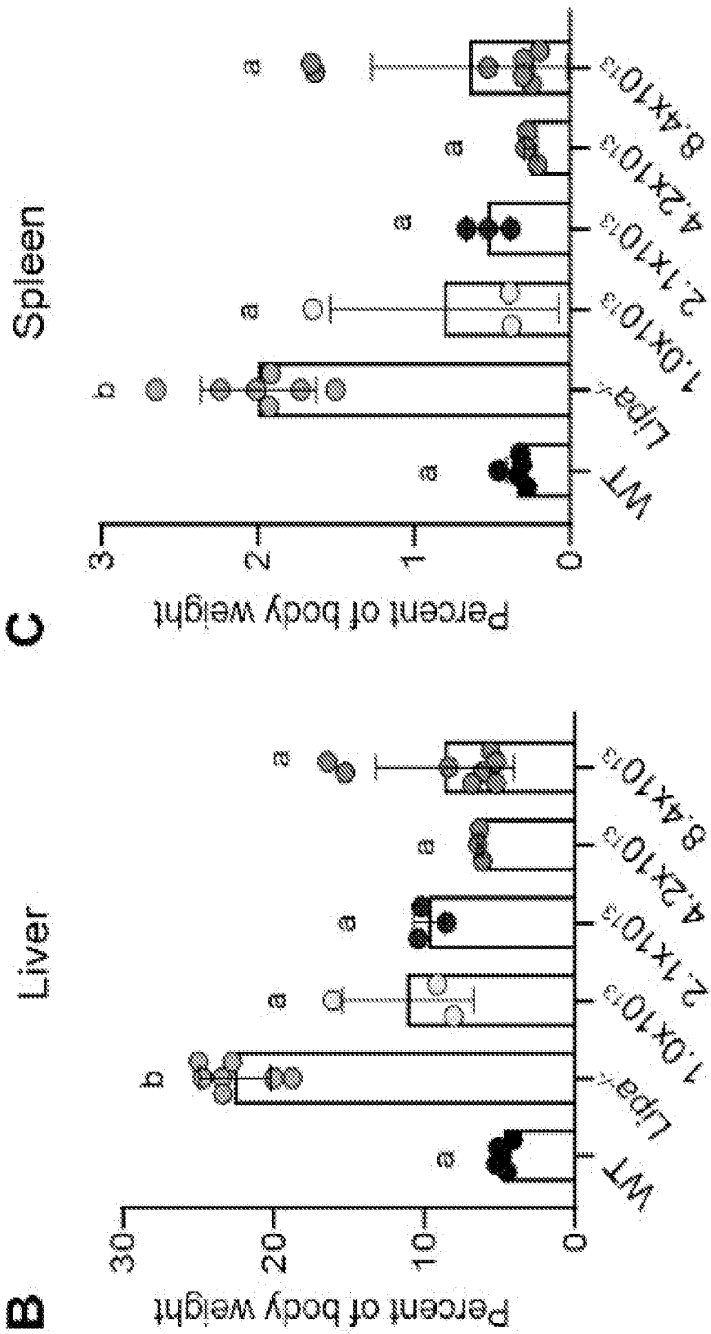
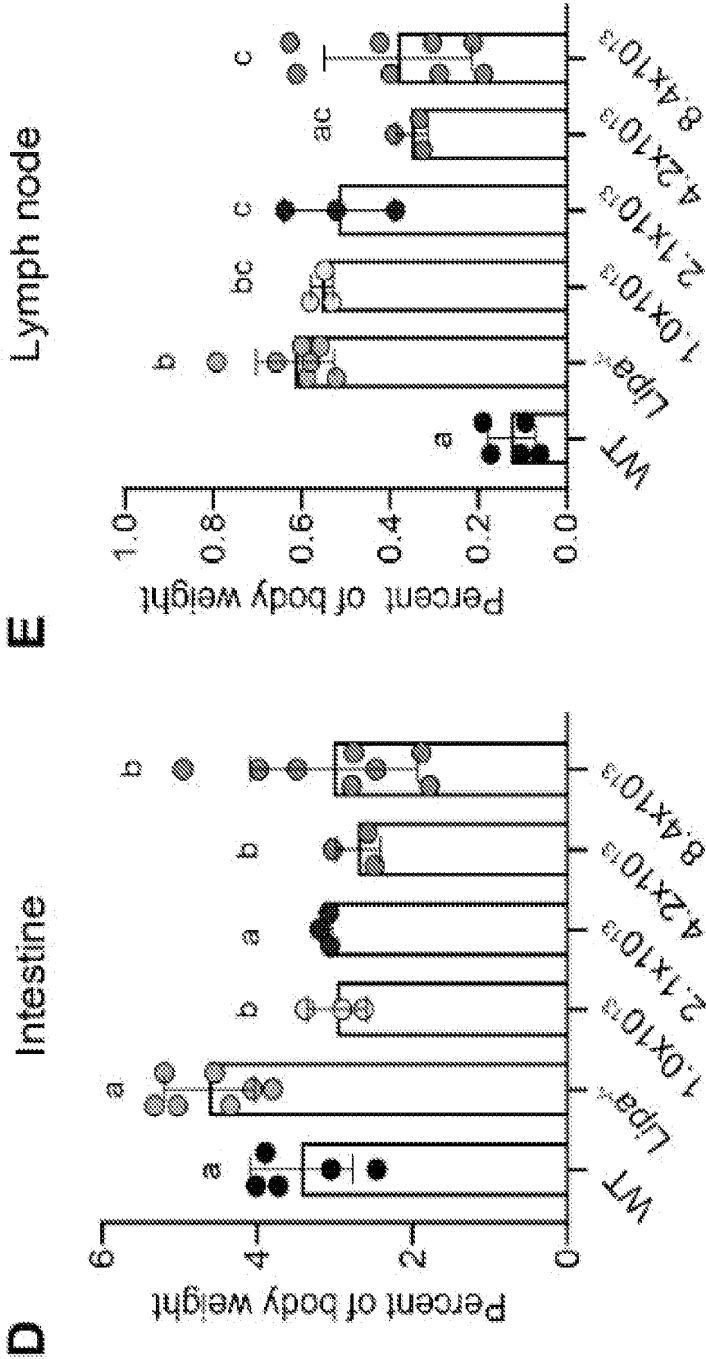


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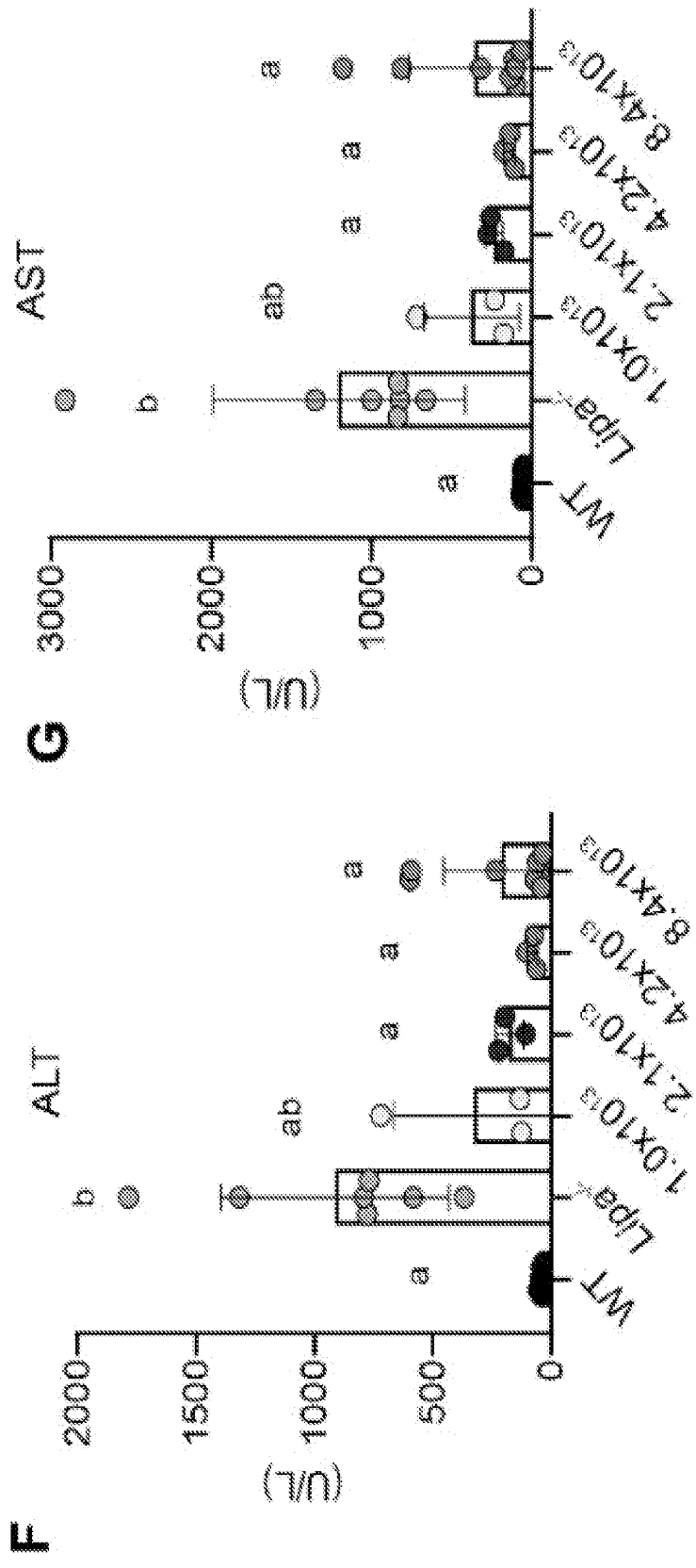
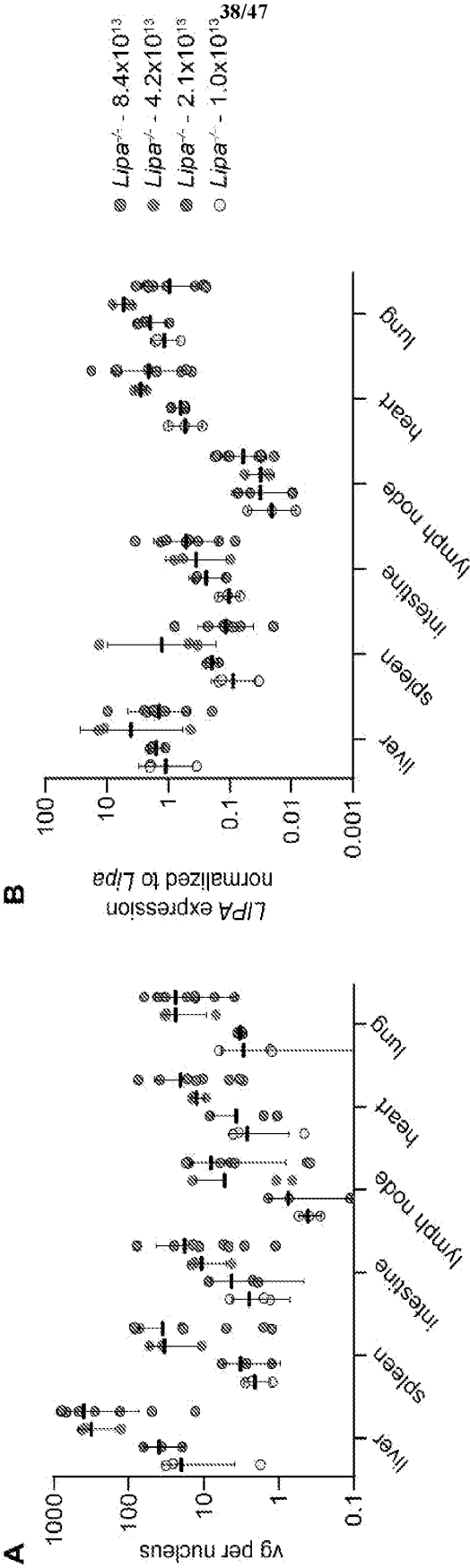


Figure 20 Continued

Figure 21



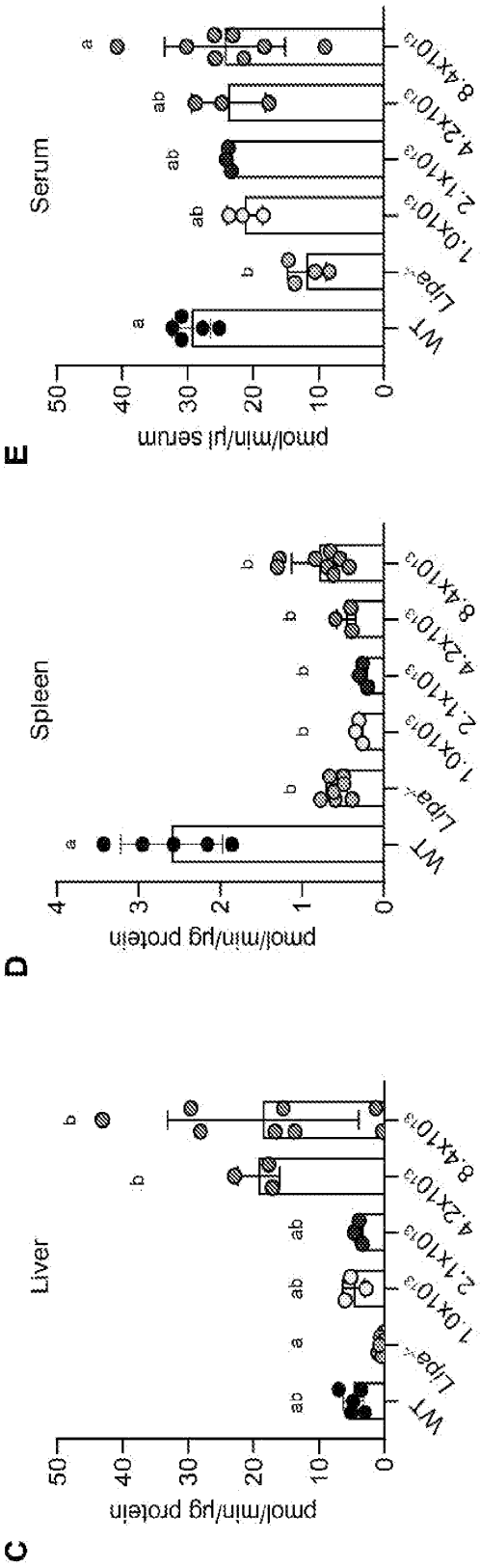


Figure 21 Continued

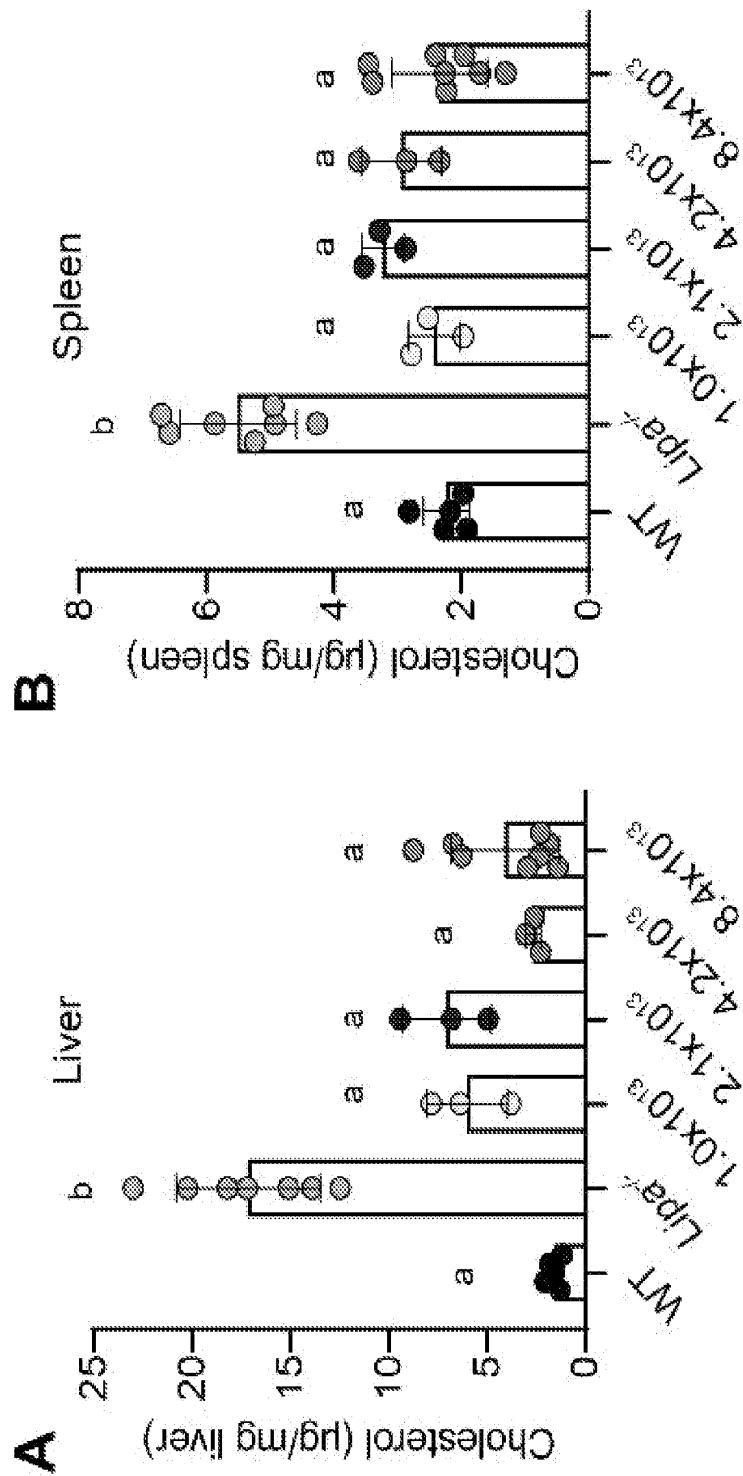
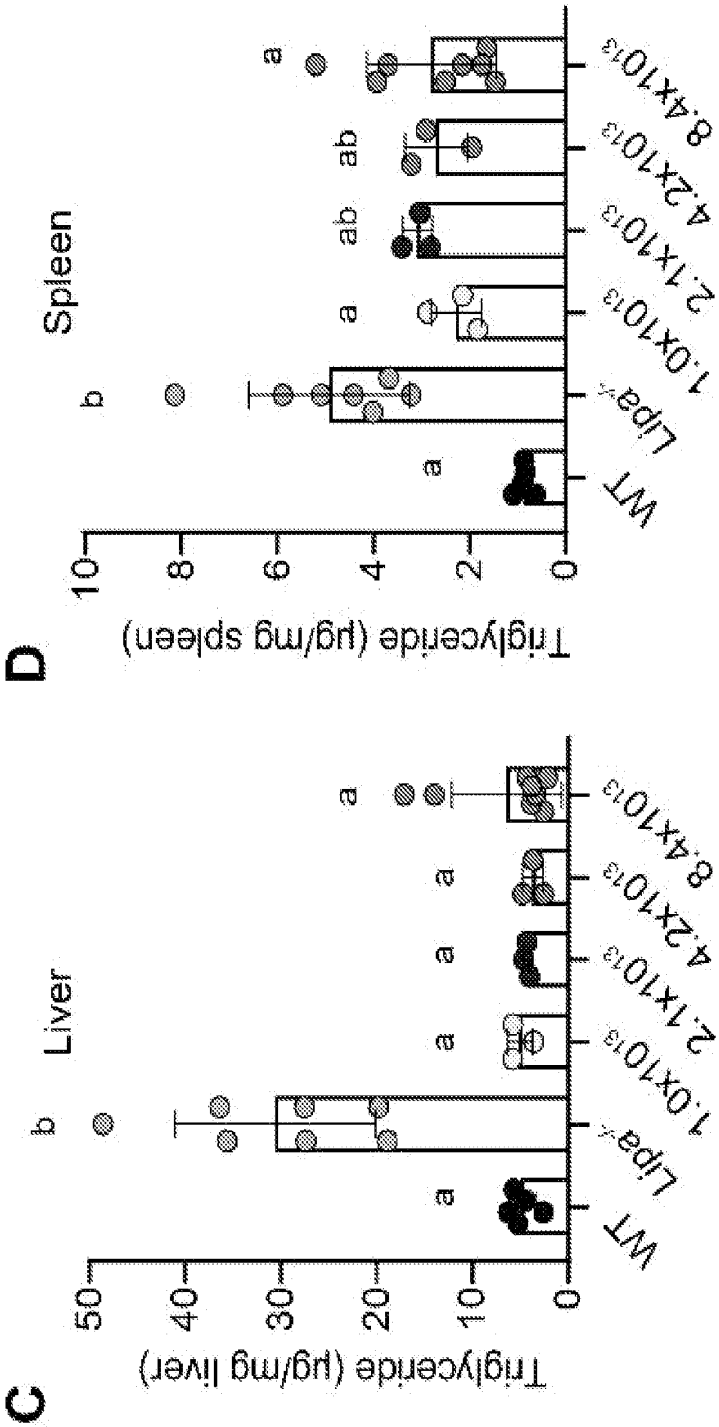


Figure 22

Figure 22 Continued



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20 40 60 80 100

GAATACTCATACTCTTCCTTTTTCAATATTATTGAAGCATTTATCAGGGTTATTGTCTCATGAGCGGATACATATTTGAATGTATTTAGAAAAATAACAAATAGGG
CTTATGAGTATGAGAAGGAAAAAGTTATAATAACTTCGTAAATAGTCCCAATAACAGAGTACTCGCCTATGTATAAACTTACATAAATCTTTTATTTGTTATCCC
120 140 160 180 200

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CAAGGCGCGTGTAAGGGGCTTTTACGGTGGACTGCAGATTCTTTGGTAATAAGTACTGTAATTGGATATTTTATCCGCATAGTGCTCCGGGAAGCAGAGCG
220 240 260 280 300 320

GCCTTTCGGTGATACGGTGAAAACCTCTGACACATGCAGCTCCCGGAGACGGTCACAGCTTGTCTGTAAGCGGATGCCGGGAGCAGACAAGCCCGTCAGGGCGCGT
CGCAAAGCCACTACTGCCACTTTTGGAGACTGTGTACGTGAGGGCCTCTGCCAGTGTGCAACAGACATTGCGCTACGGCCCTCGTCTGTTCCGGCAGTCCCGCGCA
340 360 380 400 420

CAGCGtGTGTTGGCGGGTGTGGGGCTGGCTTAACATATGCGGCATCAGAGCAGATTGTAAGAGTGCACCATATGCGGTGTGAAATACCGCACAGATGCGTAAGG
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440 460 480 500 520

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540 560 580 600 620 640

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660 680 700 720 740

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760 780 800 820 840

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860 880 900 920 940 960

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980 1,000 1,020 1,040 1,060

Figure 23

SUBSTITUTE SHEET (RULE 26)

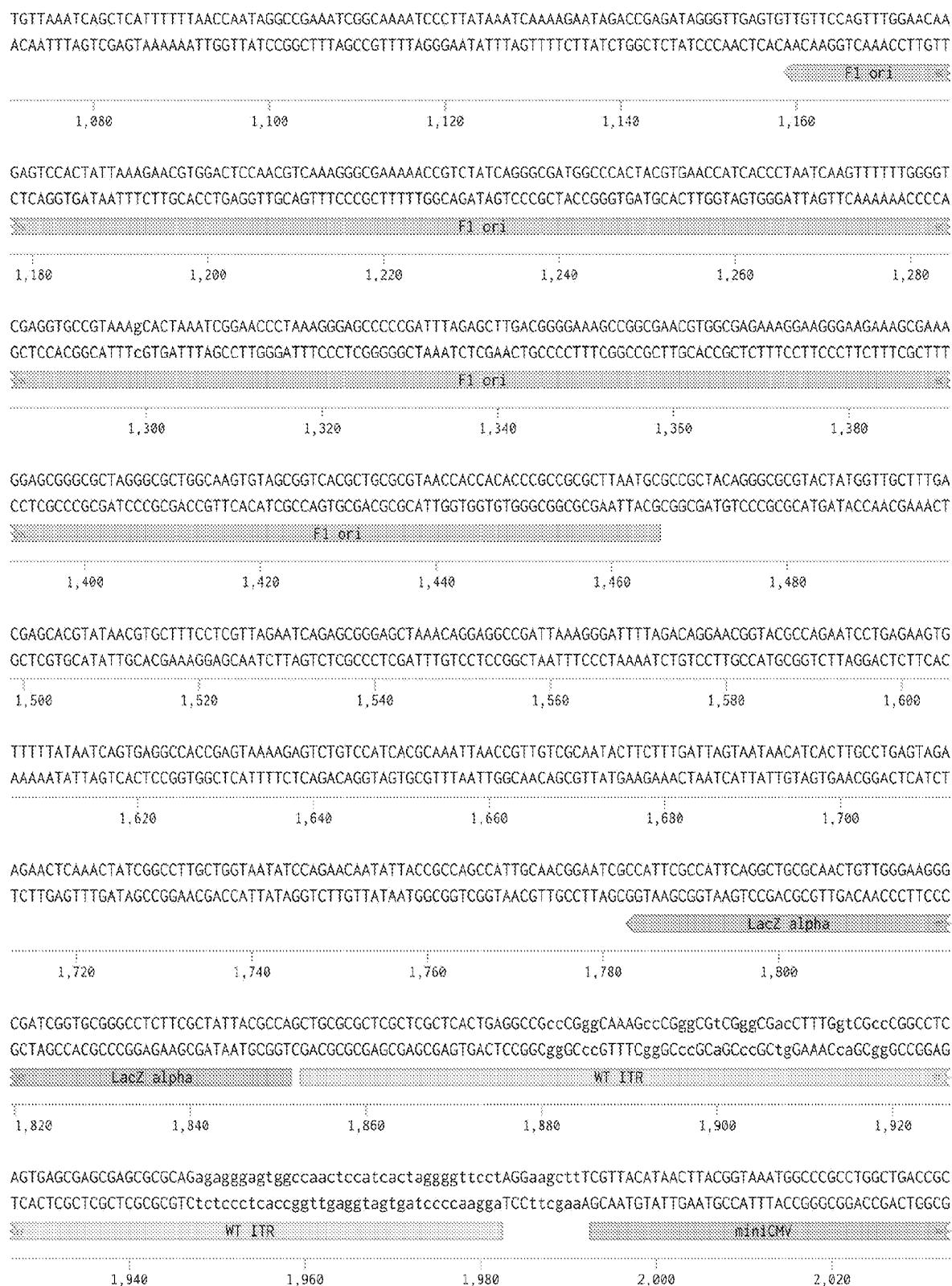


Figure 23 - Continued

SUBSTITUTE SHEET (RULE 26)

44/47

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GGTTGCTGGGGCCTGAGTGCCCTAAAGGTTCAAGGTGGGGTAAGTGCAGTTACCTCAAACAAAACCGTGGTTTTAGTTGCCCTGAAAGGTTTTACAGCATTGT

miniCMV
2,040 2,060 2,080 2,100 2,120 2,140

ACTCCGCCCCATTGACGCAAATGGGCGGTAGGCGTGTACGGTGGGAGGTCTATATAAGCAGAGCTCGTTTAGTAACCGGctgaCCGGgtaccggcgccgctagcc
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miniCMV
2,160 2,180 2,200 2,220 2,240

gccaccATGAAATGCGGTTCTTGGGGTGGTGGTCTGTTTGGTCTCTGGACCTGCATTCTGAGGGGTCTGGAGGAACTGACAGCTGTGGATCCTGAAACAAA
cgggtgTACTTTTACGCCAAGAACCCCAACCACGACAAACCAAGAGACCTGGGACGTAAGACTCCCAGACCTCCCTTTGACTGTGACACCTAGGACTTTGTTT

M K M R F L G L V V C L V L W T L H S E G S G G K L T A V D P E T N

LIPA transcript variant 1
2,260 2,280 2,300 2,320 2,340

CATGAATGTGAGTGAAATTATCTCTTACTGGGGATTCCCTAGTGAGGAATACCTAGTTGAGACAGAAGATGGATATATTCTGTGCCTTAACCGAATTCCTCATGGGA
GTACTTACACTCACTTTAATAGAGAATGACCCCTAAGGGATCACTCCTTATGGATCAACTCTGTCTTCTACCTATATAAGACACGGAATTGGCTTAAGGAGTACCTT

M N Y S E I I S Y W G F P S E E Y L V E T E D G Y I L C L N R I P H G

LIPA transcript variant 1
2,360 2,380 2,400 2,420 2,440 2,460

GGAAGAACCATTCTGACAAAGGTCCCAAACAGTTGTCTTCTGCAACATGGCTTGCTGGCAGATTCTAGTAAGTGGGTCACAAACCTTGCCAAACAGCAGCCTGGGC
CCTTCTTGTAAGACTGTTTCCAGGGTTTGGTCAACAGAAGGACGTTGTACCGAACGACCGTCTAAGATCATTGACCCAGTGTTTGAACGGTTGTCGTCGGACCCG

R K N H S D K G P K P Y V F L Q H G L L A D S S N W V T N L A N S S L G

LIPA transcript variant 1
2,480 2,500 2,520 2,540 2,560

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AAGTAAGAACGACTACGACCAAACTGCACACCTACCCGTTGTCGTCTCCTTTATGGACCAGAGCCTTTGTATTCTGTGAGAGTCAAAGAGTCTACTTAAGACCCG

F I L A D A G F D V W M G N S R G N T W S R K H K T L S V S Q D E F W A

LIPA transcript variant 1
2,580 2,600 2,620 2,640 2,660

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F S Y D E M A K Y D L P A S I N F I L N K T G Q E Q V Y Y V G H S Q G

LIPA transcript variant 1
2,680 2,700 2,720 2,740 2,760 2,780

Figure 23 - Continued

SUBSTITUTE SHEET (RULE 26)

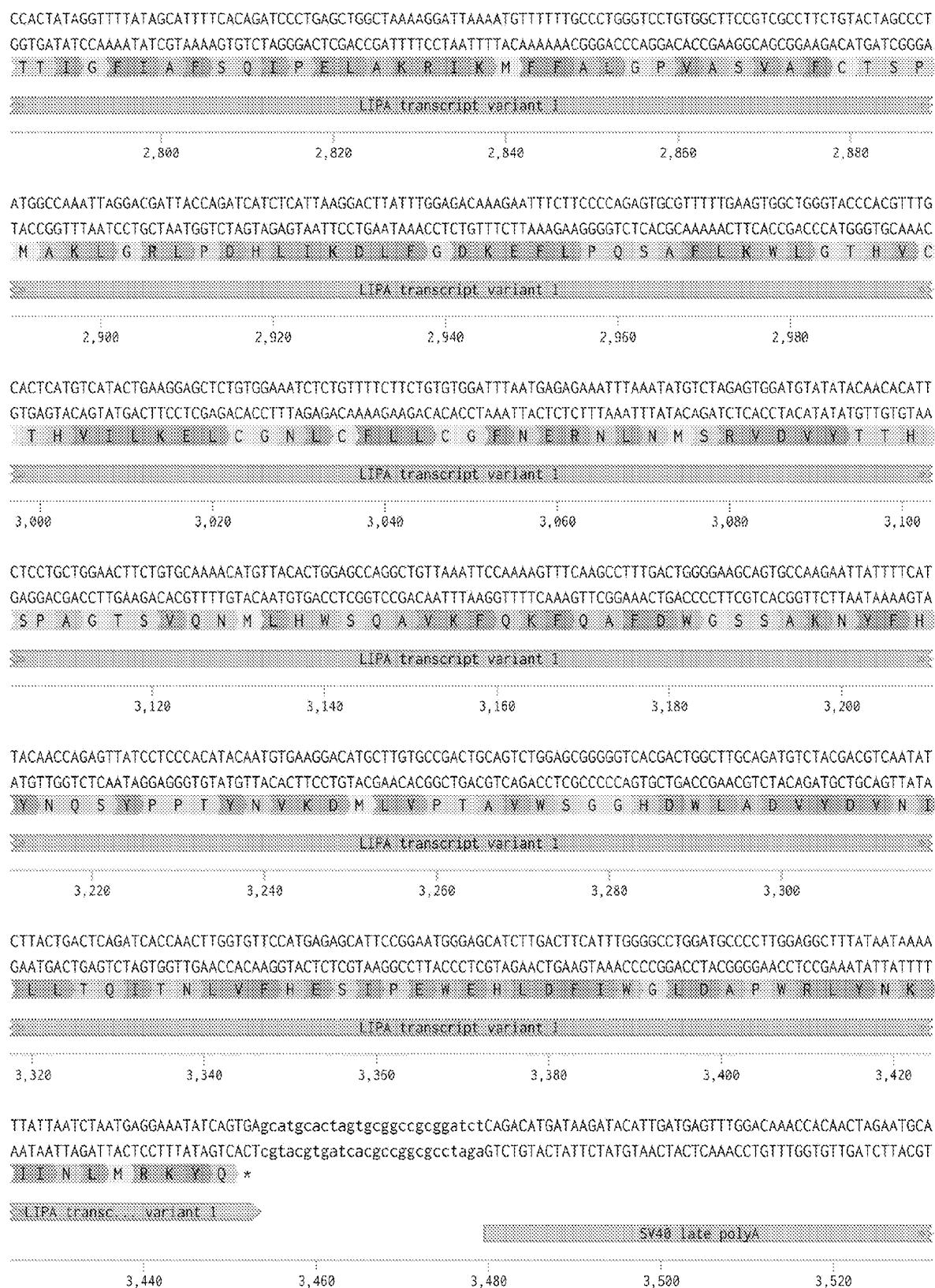


Figure 23 - Continued

SUBSTITUTE SHEET (RULE 26)

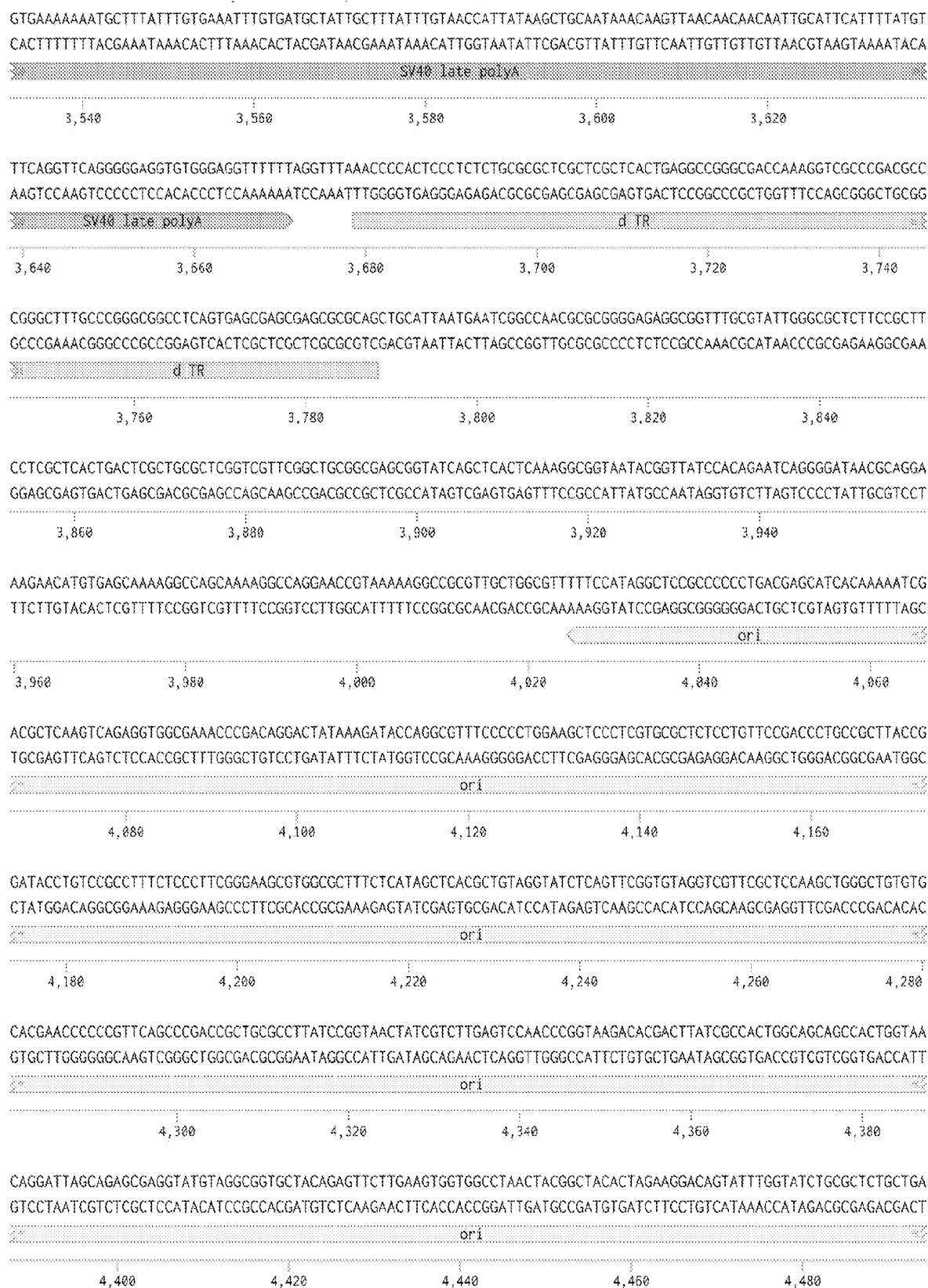


Figure 23 - Continued
SUBSTITUTE SHEET (RULE 26)

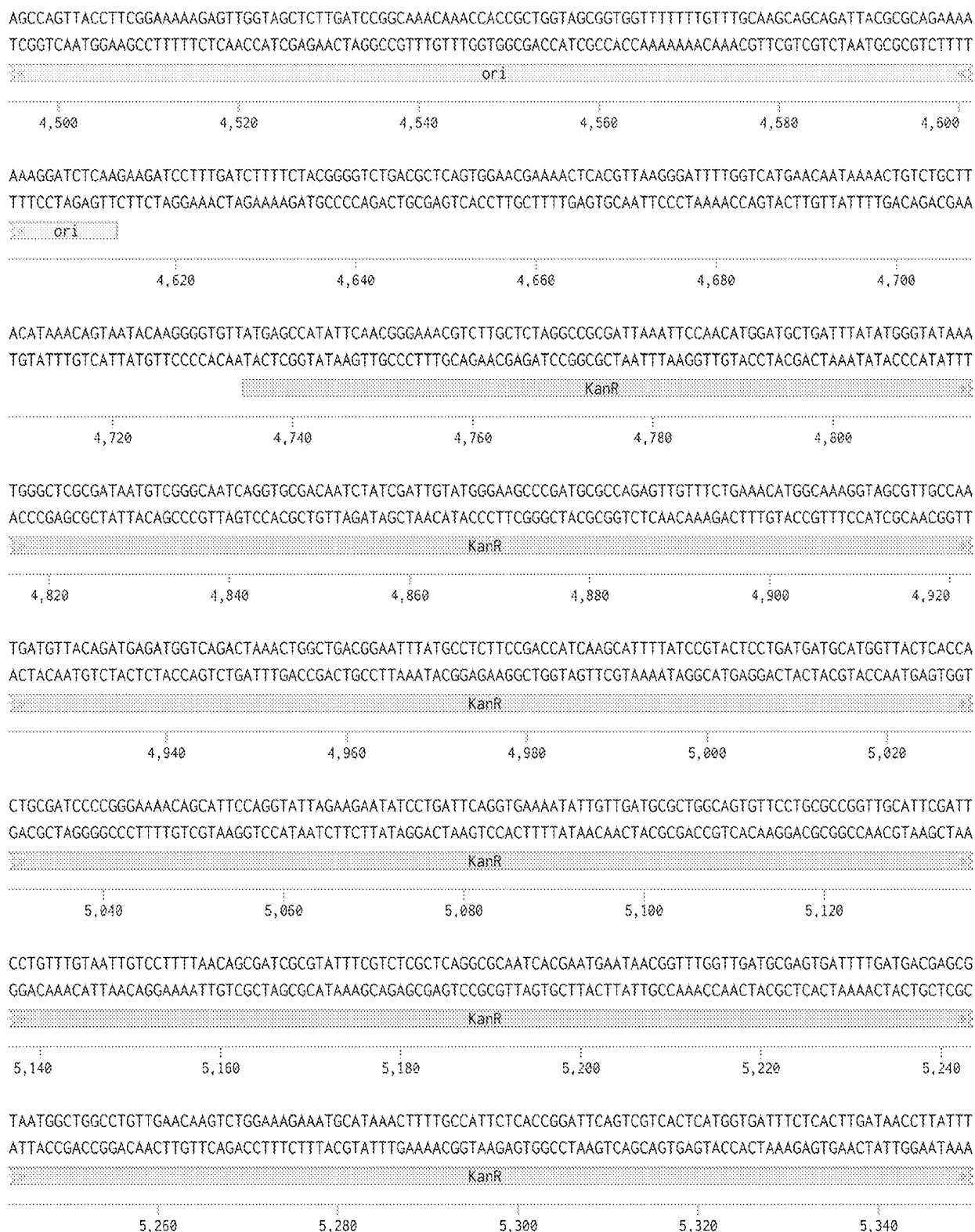


Figure 23 - Continued

SUBSTITUTE SHEET (RULE 26)