

FIG. 1

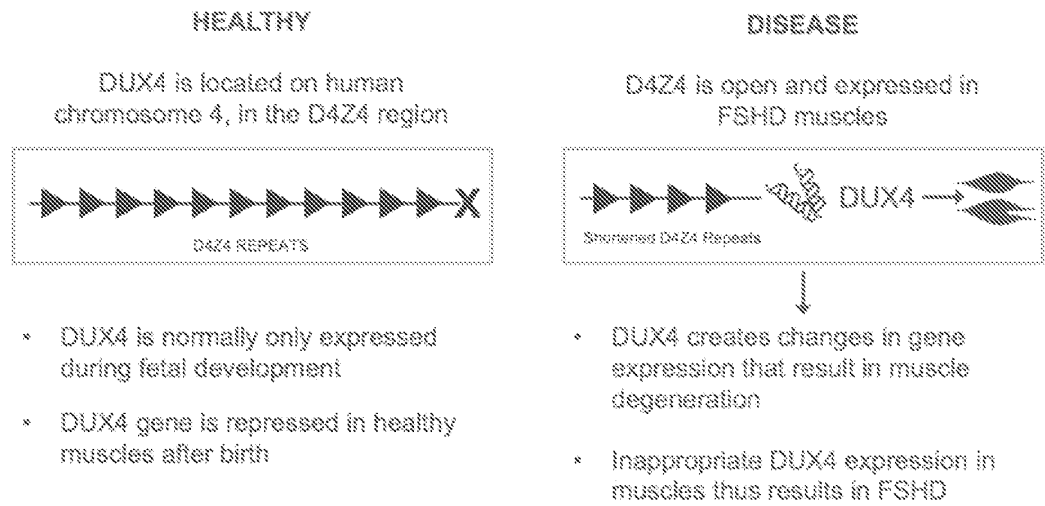


FIG. 2

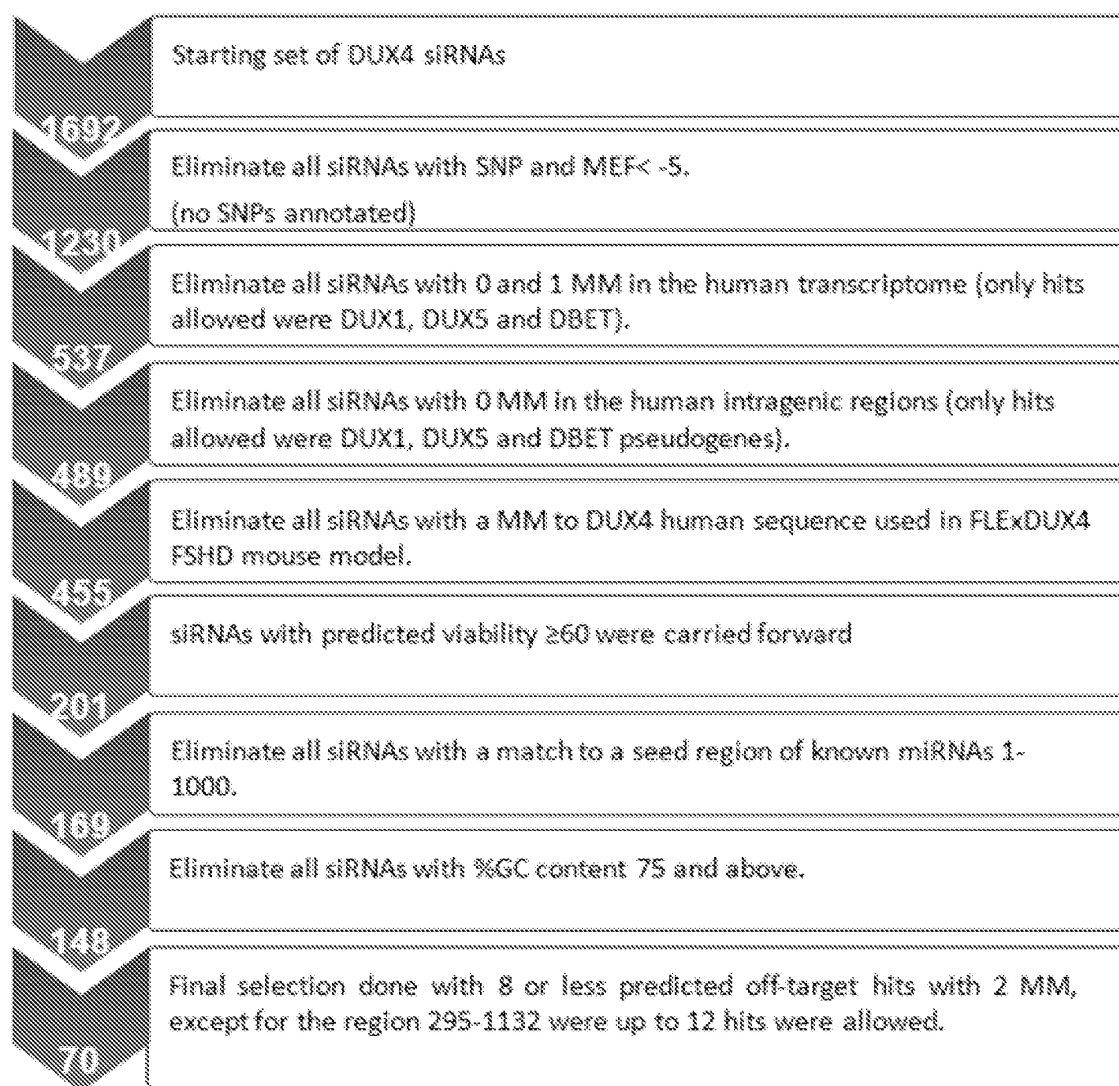


FIG. 3

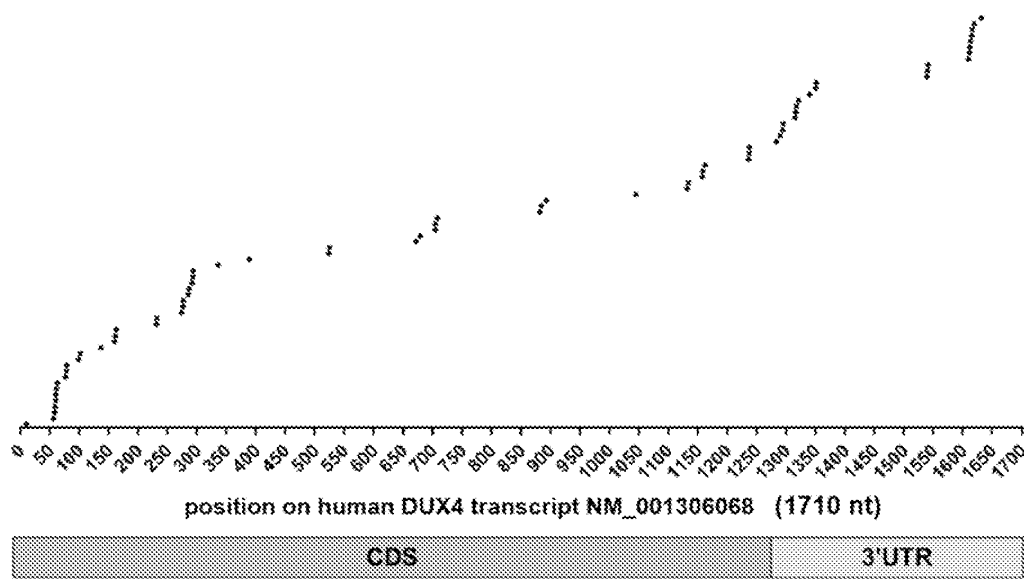


FIG. 4

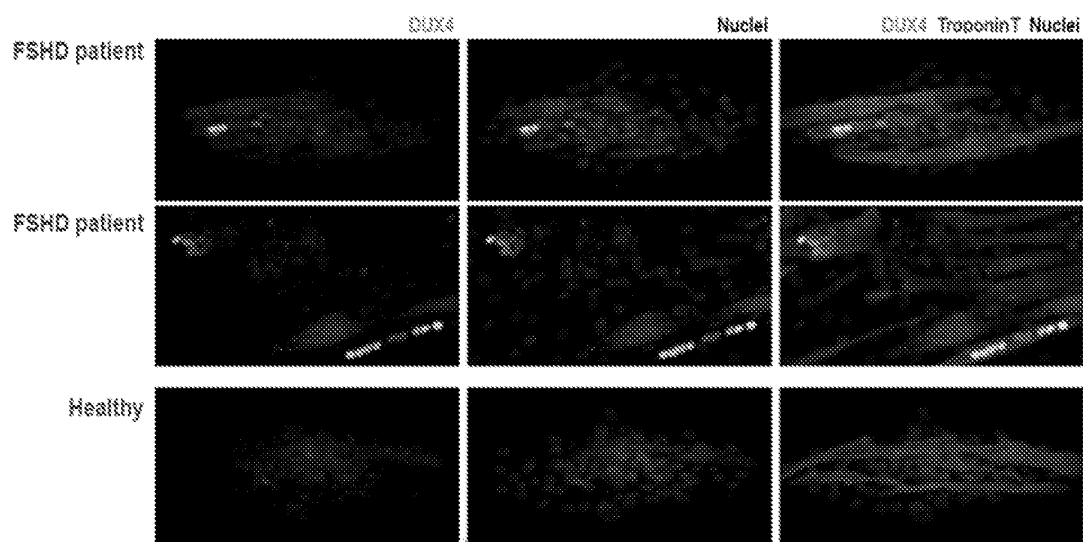
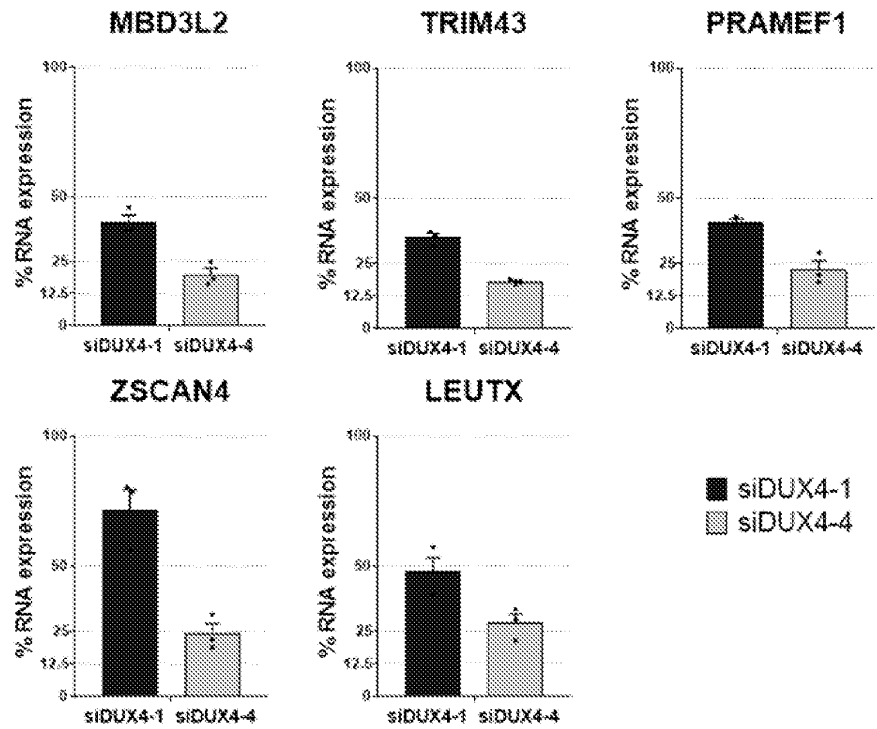


FIG. 5



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FIG. 6A

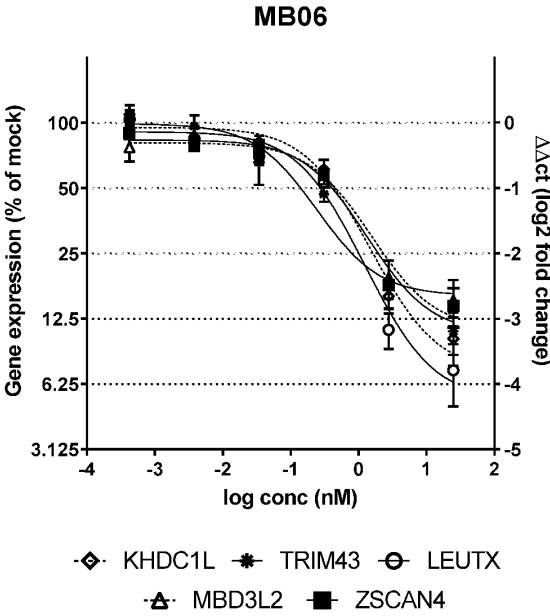
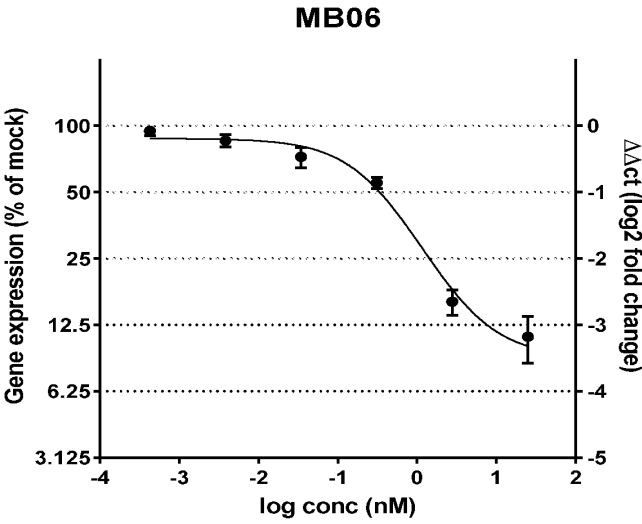


FIG. 6B



Calculations of the FSHD composite:
Dct = (Av. ct 5 DUX4 target genes) – (Av. ct 2 HKGs)
DDct = Dct (siDUX4) – Dct (mock)
Composite = $2^{-DDct} \times 100$ (%)

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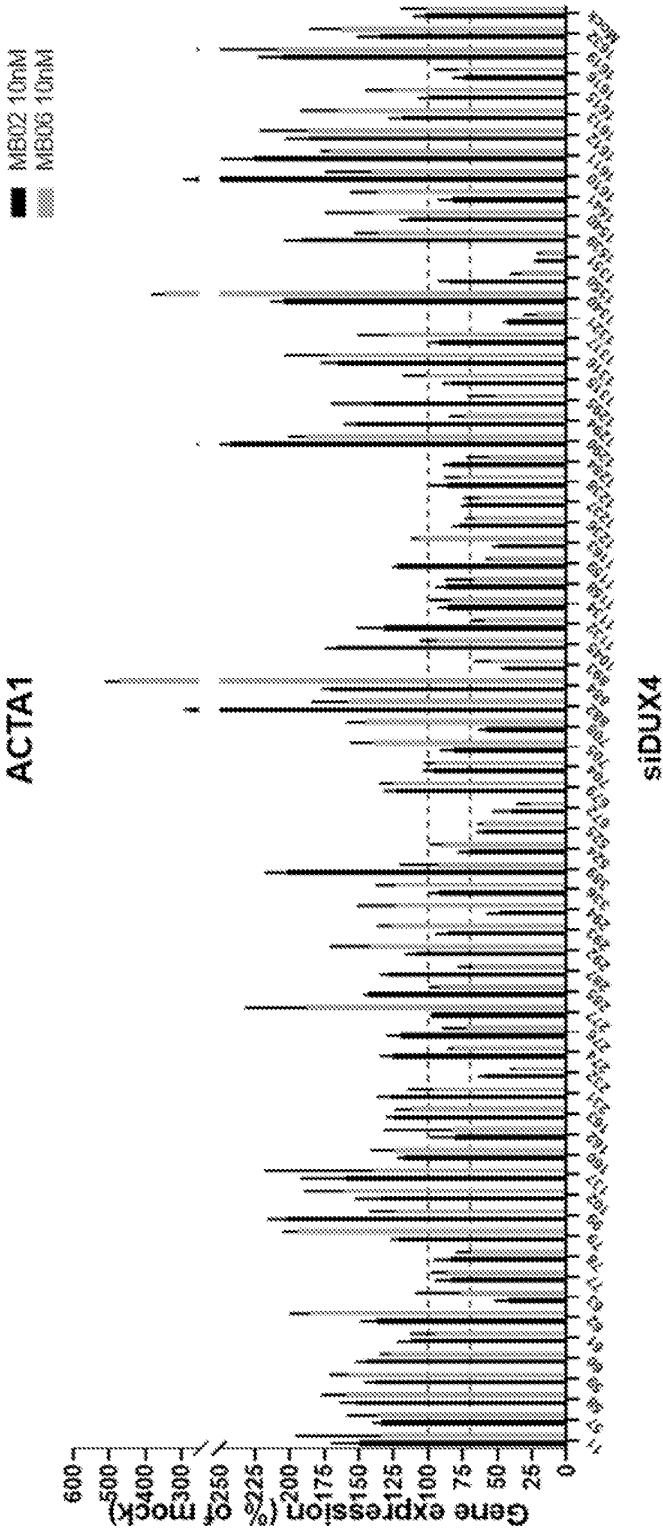


FIG. 7

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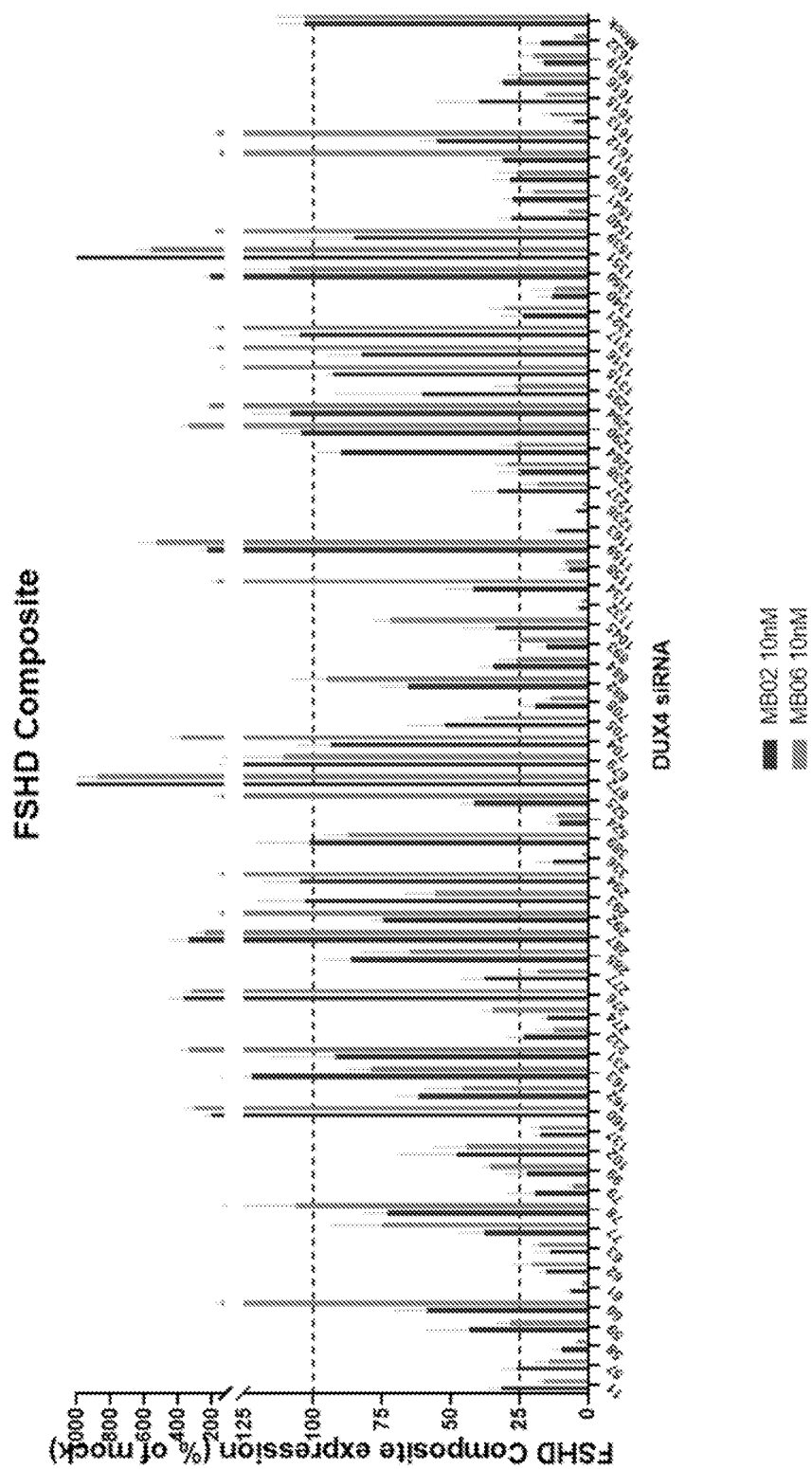


FIG. 8

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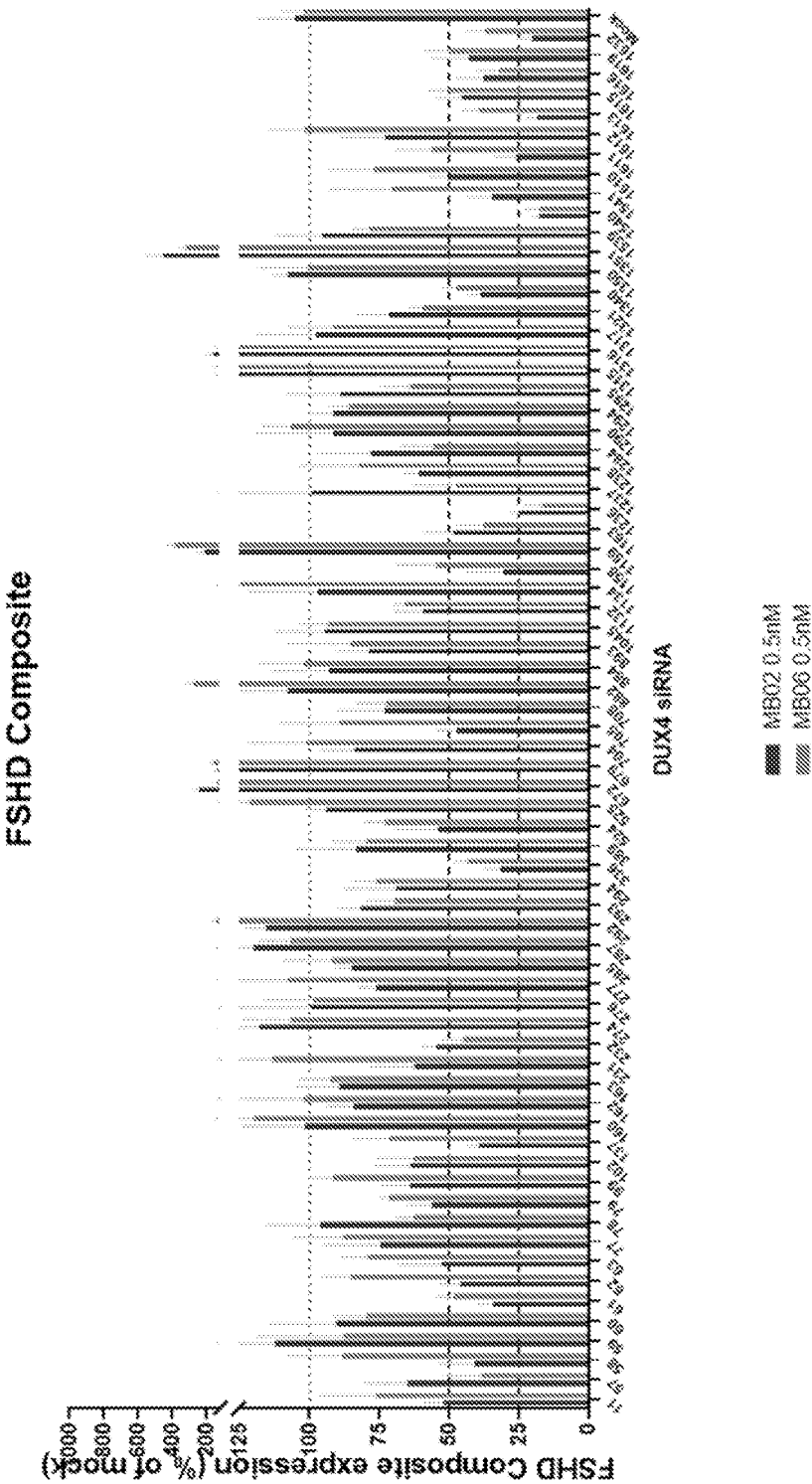
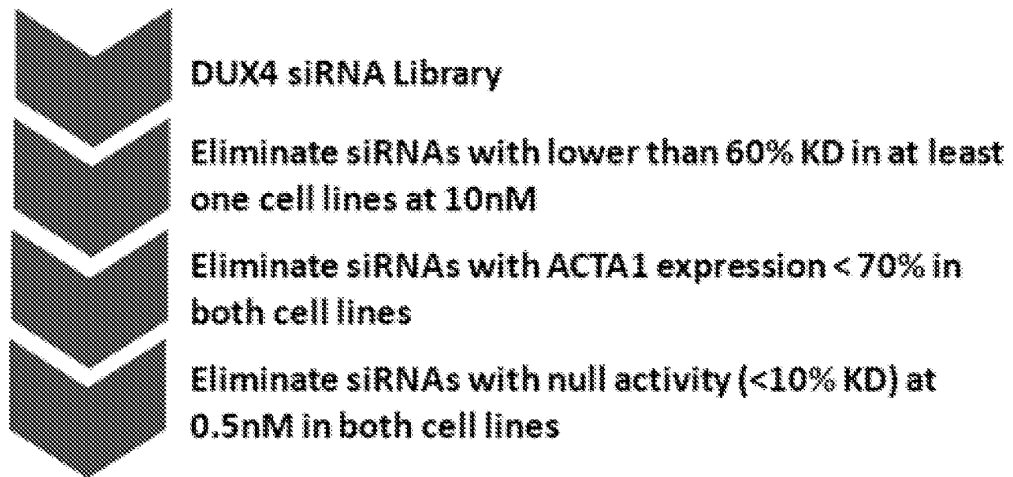


FIG. 9

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FIG. 10



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FIG. 11A

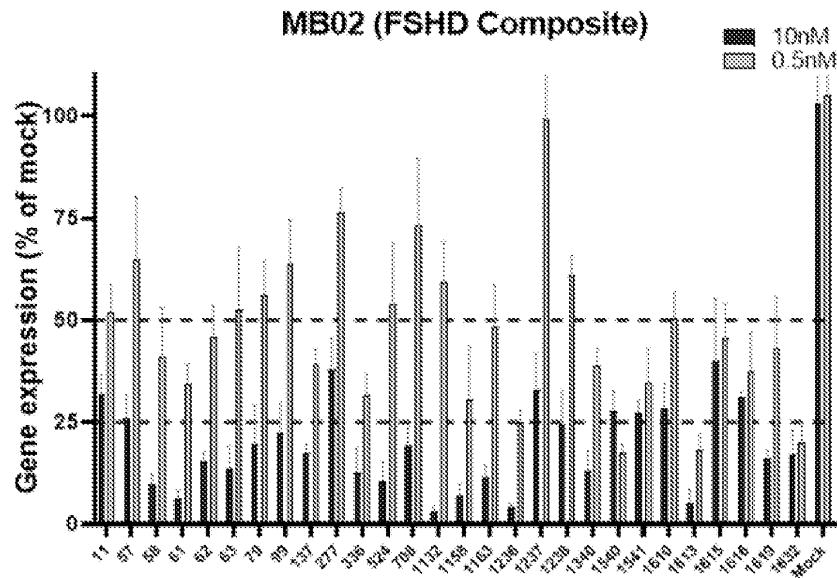
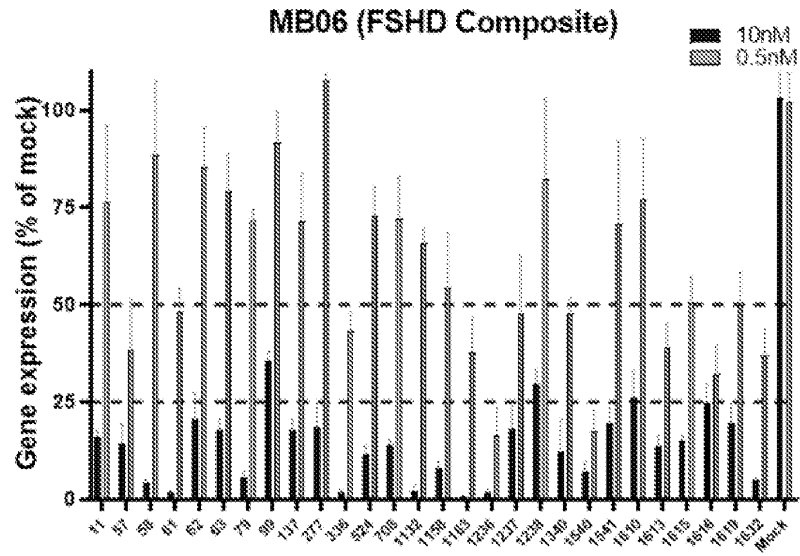
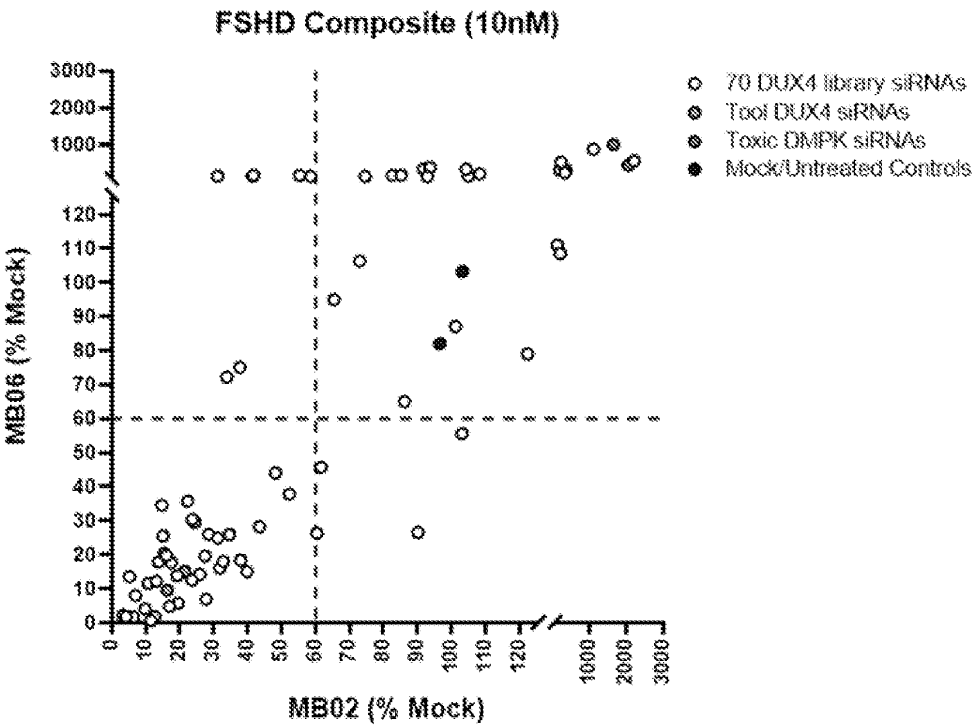


FIG. 11B



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FIG. 12



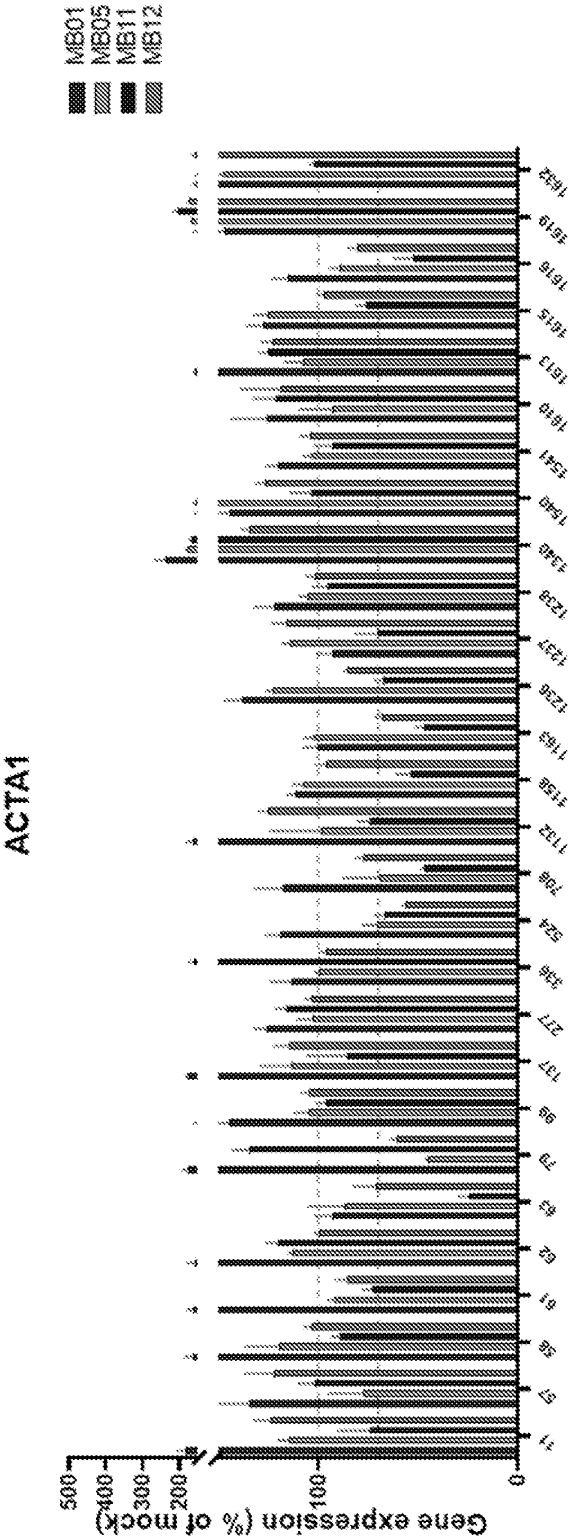


FIG. 13

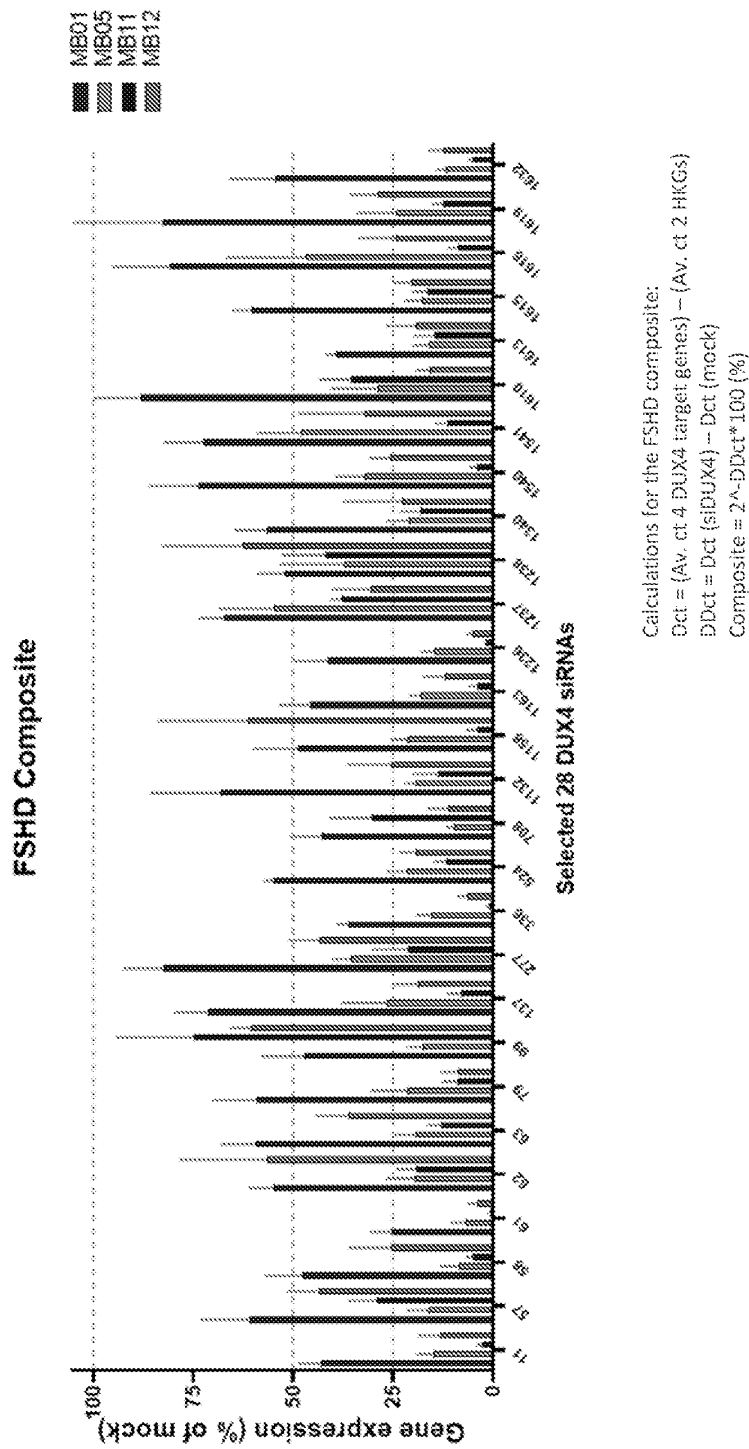


FIG. 14

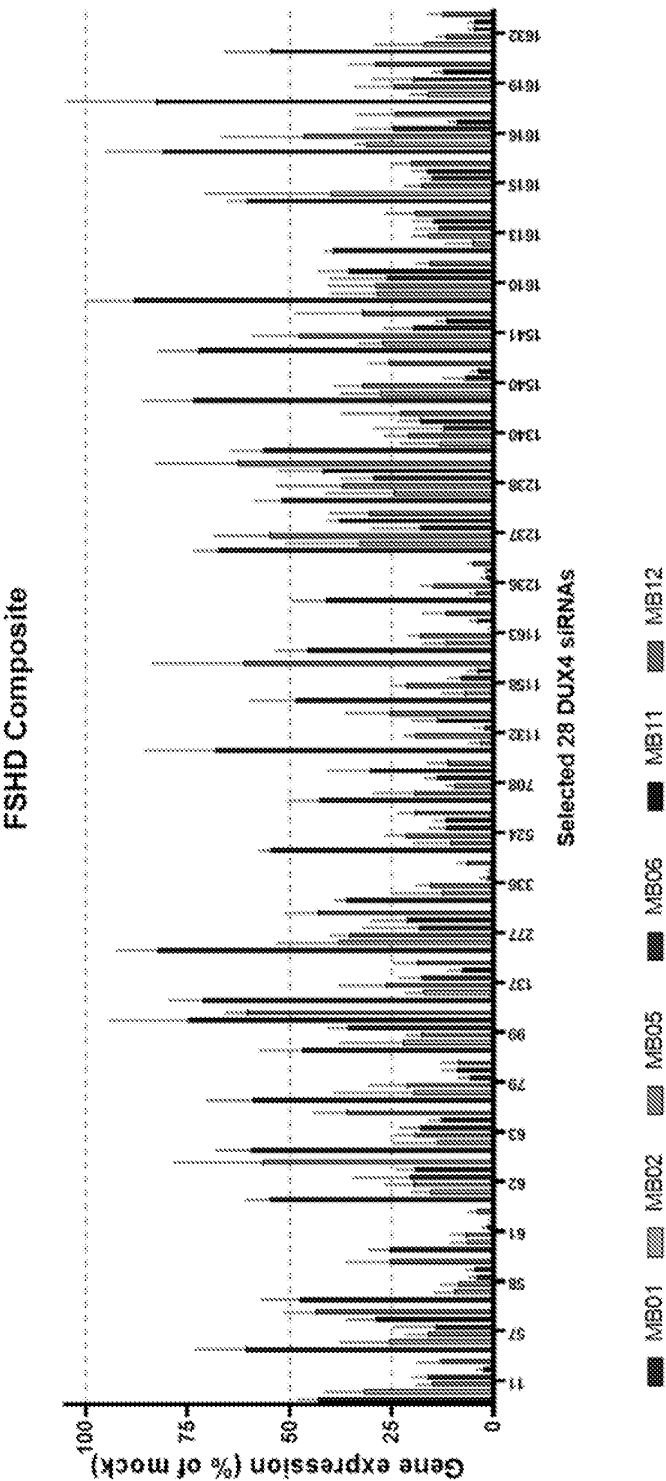


FIG. 15

FIG. 16A

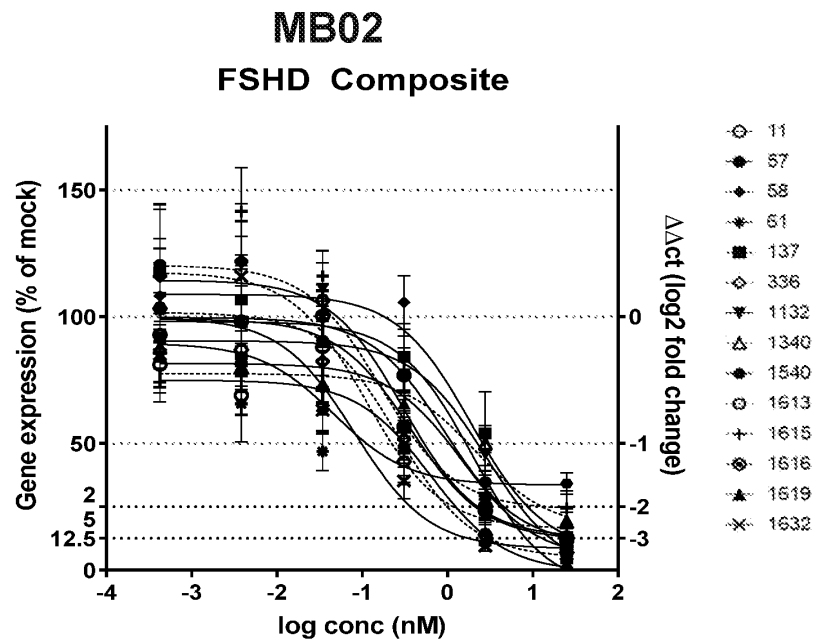
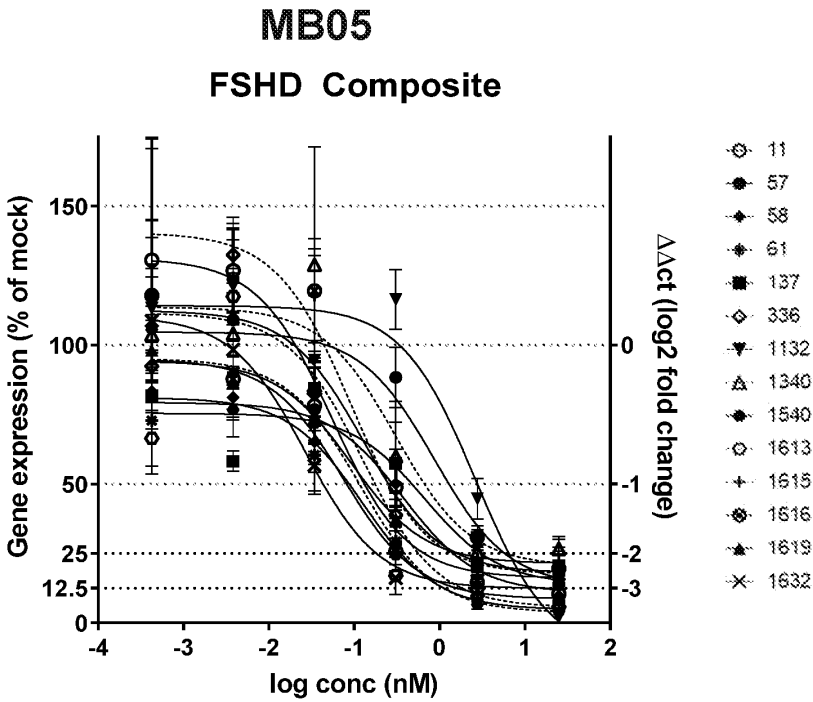
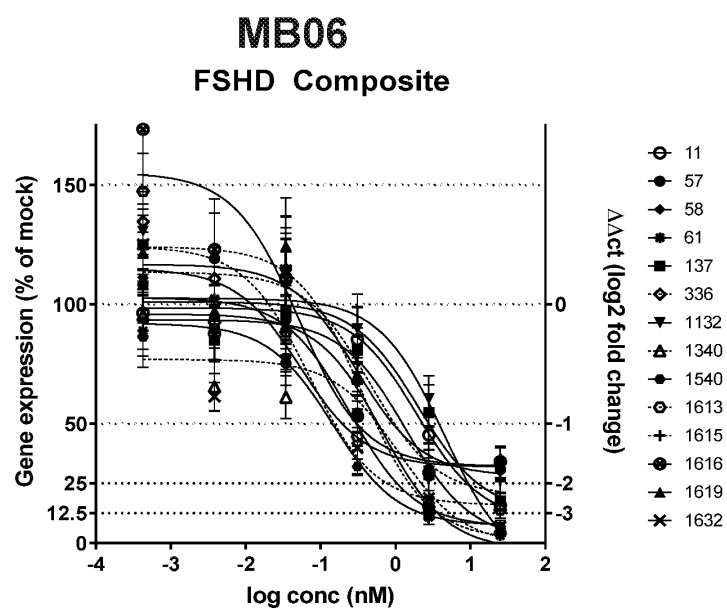


FIG. 16B



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FIG. 16C



Calculations for the FSHD composite:

Dct = (Av. ct 4 DUX4 target genes) – (Av. ct 2 HKGs)

DDct = Dct (siDUX4) – Dct (mock)

Composite = $2^{-DDct} \times 100$ (%)

FIG. 17A

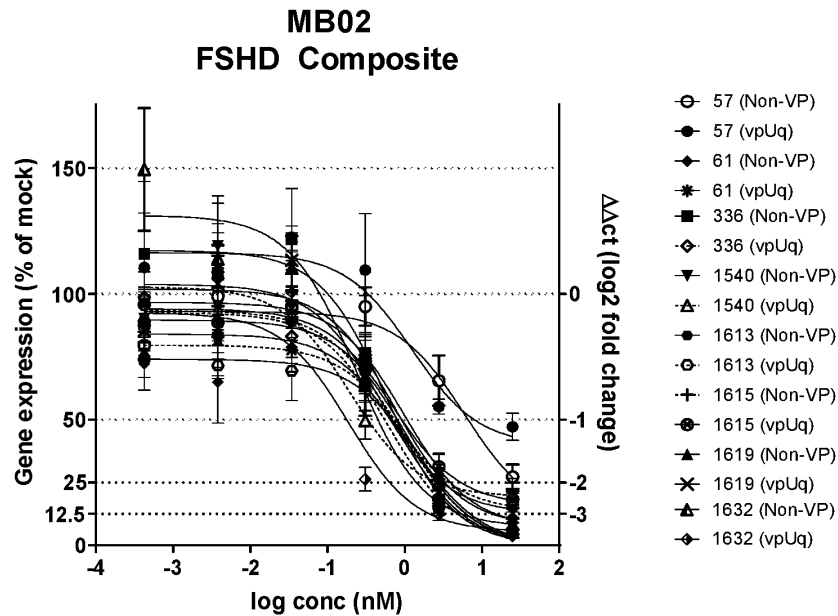


FIG. 17B

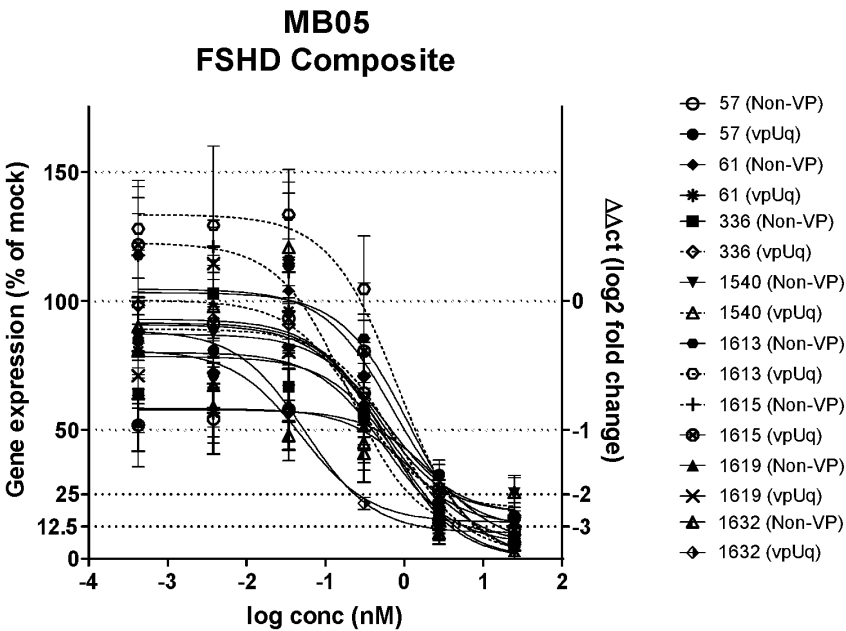


FIG. 17C

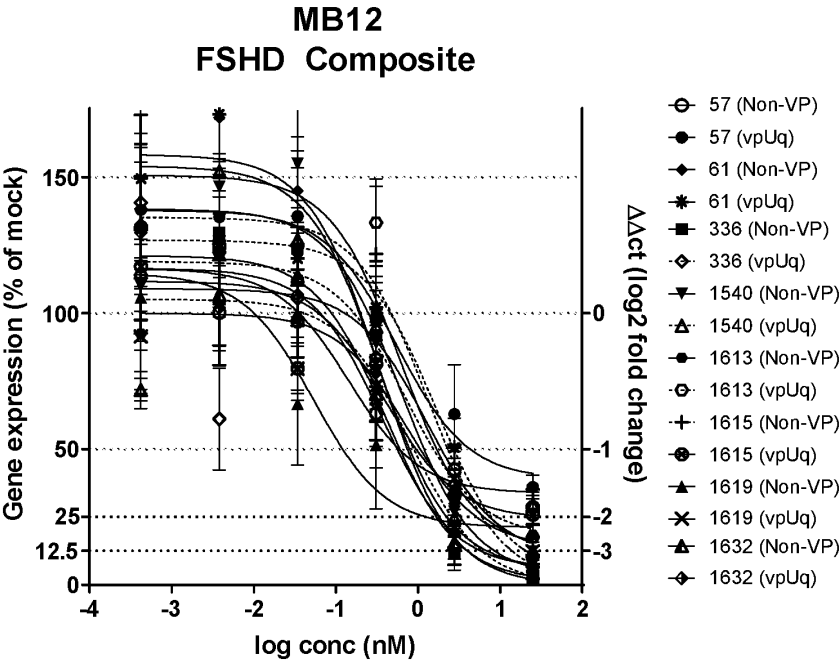


FIG. 18A

In Vitro Activities of DUX4-AOCs With
DUX4-siRNA With Vinylphosphonate

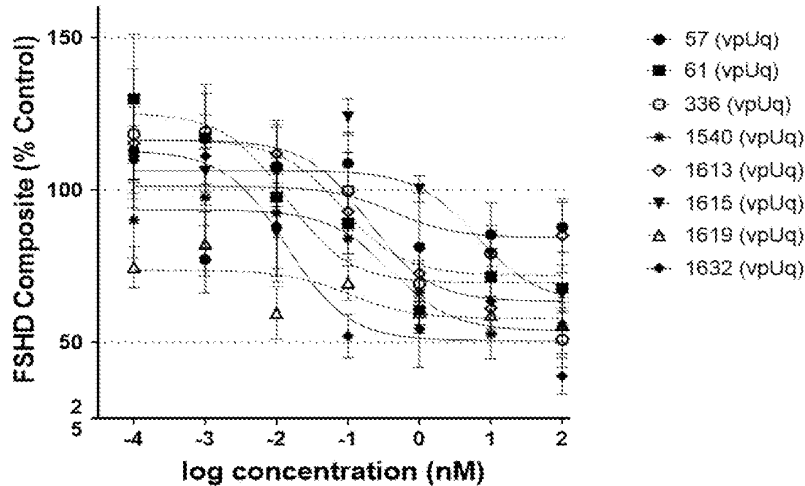
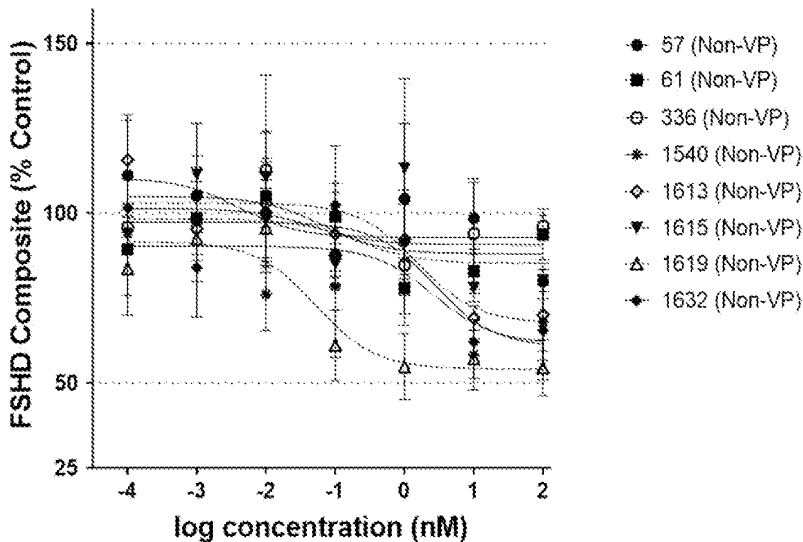


FIG. 18B

In Vitro Activities of DUX4-AOCs with
DUX-4siRNA Without Vinylphosphonate



In Vivo Reduction of Nuclear Localized Lnc-RNA Malat1 in Skeletal Muscle
in Mice by siMalat1-AOC

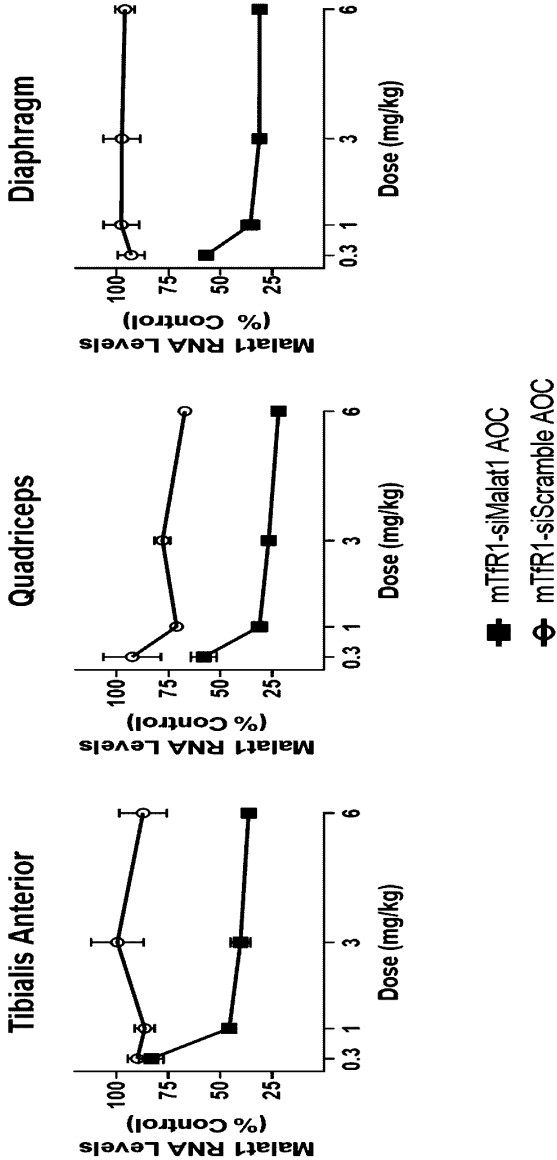


FIG. 19

FIG. 20

In Vivo Reduction of SSB mRNA in Skeletal Muscle in Mice by
siSSB-AOCs

