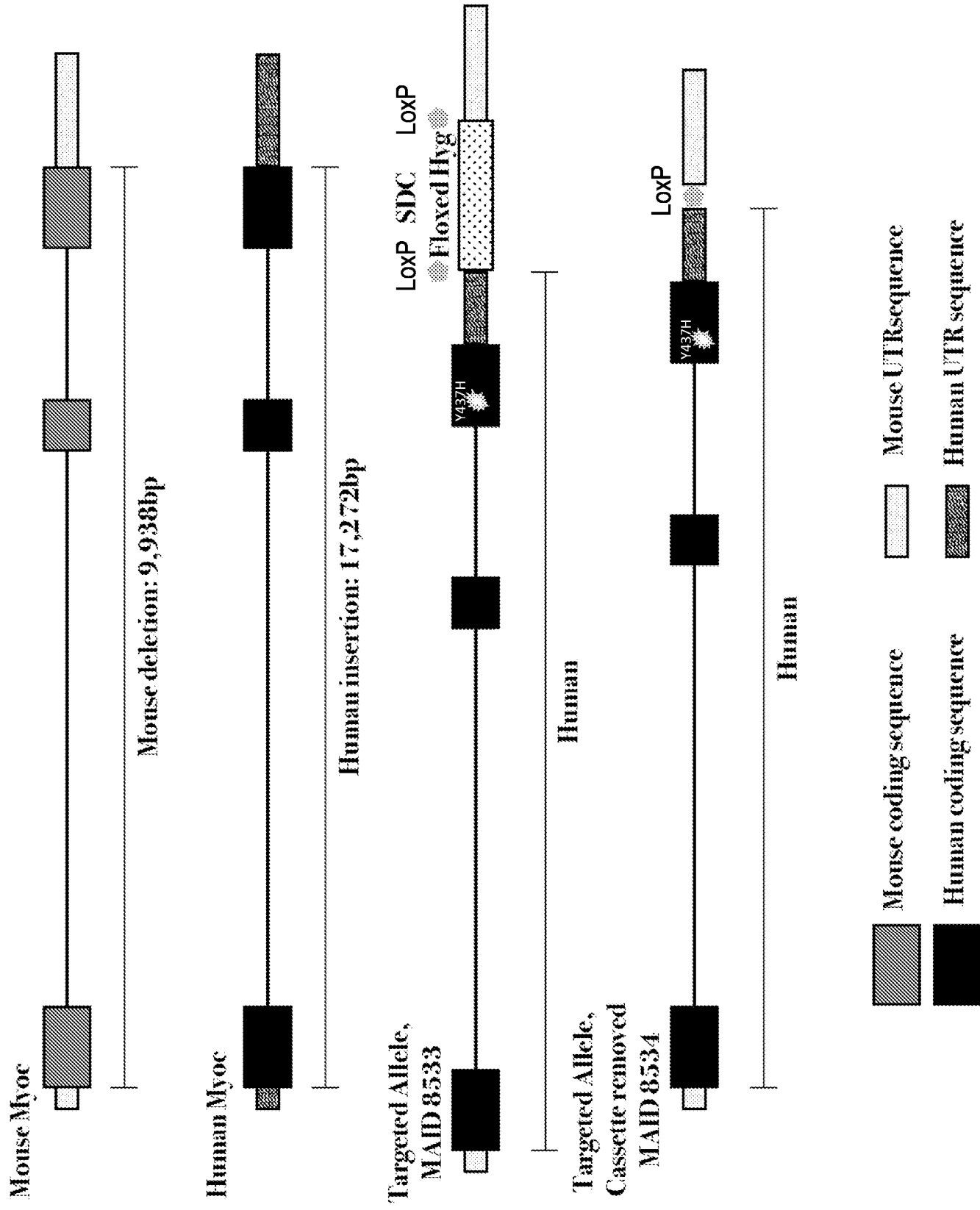


**FIG. 1**

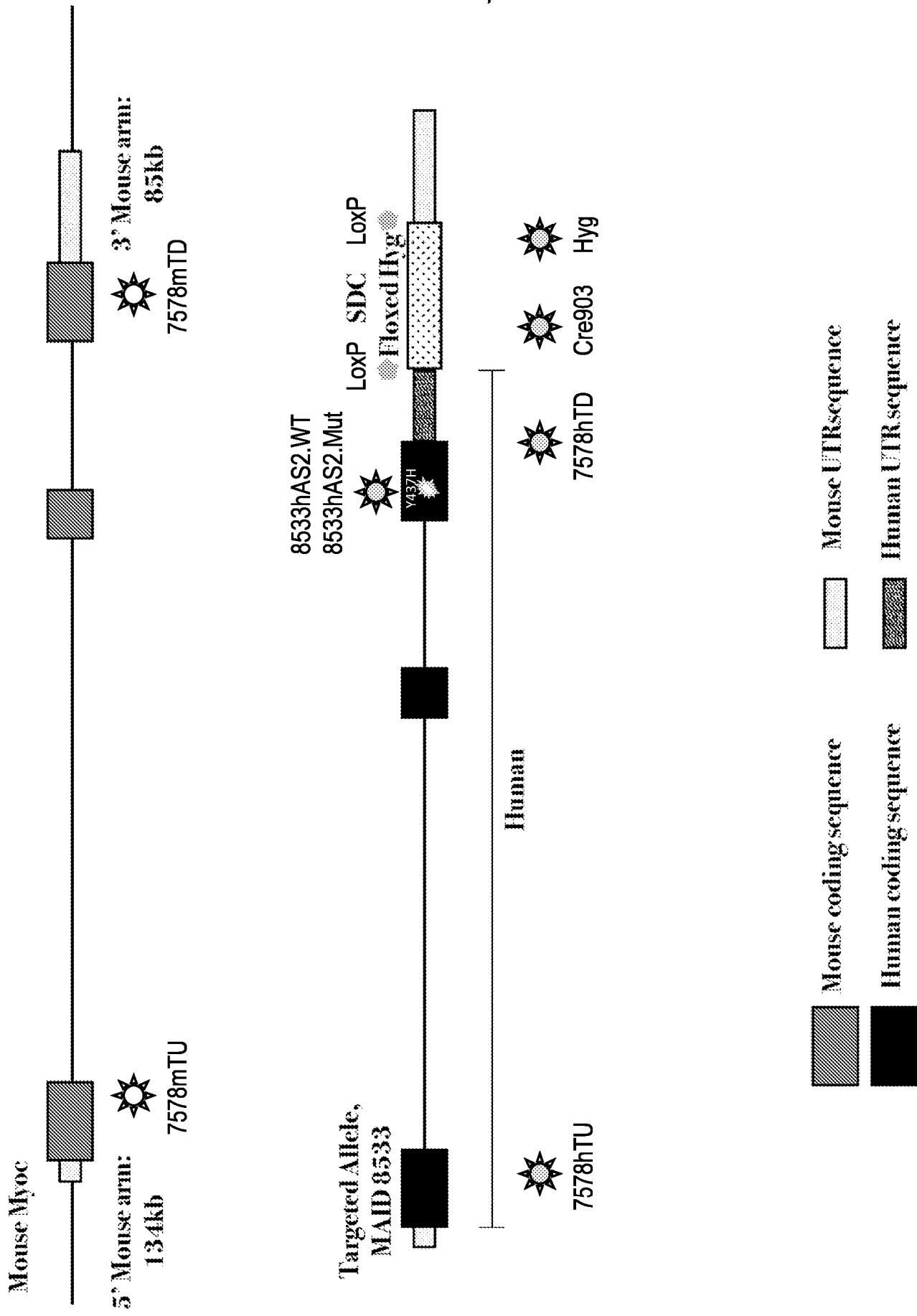


FIG. 2

Signal peptide

Human MYOC: MRFFCARCCSFGEPPAVQLLLLLACLVDVGAR
Mouse MYOC: MPALHLLFLACLVDGMGAR

Human MYOC: QRLDLEATKARLSLESLLHQLTLDQAARPQETQEGLRQLRRERDQLETQTRELETAYSNLLRDKSVLEEEKKRLR
Mouse MYOC: QHADLESTKARVRSLESLLHQMTLGRVTGTQEAQEGLRQLGALLRRERDQLETQTRDLEAAYNNLLRDKSALEEEKRQLE

Human MYOC: QENENLARRRLESSQEVARLRGQCQTRDTARAVPPGSRREVSTWNLDTLAFQELKSELTEVPASRIILKESPSGYLRSGE
Mouse MYOC: QENEDLARRRLESSSEEVTRLRRGQCPSTQYPSQDMLPGSRREVSWNLDTLAFQELKSELTEVPASQILKENPSGRPRSKE

N-terminal fragment | C-terminal fragment

CAPN2 cleavage

Human MYOC: GDTGCGELVWVGEPLTLRTAETITGKYGVWMRDPKPTYPYTQETTWRIDTVGTDVRQVFEYDILISQFMQGYPSKVHILPR
Mouse MYOC: GDKGCGALVWVGEPTLRTAETIAGKYGVWMRDPKPTHPYTQESTWRIDTVGTEIRQVFEYSQISQFEQGYPSKVHVILPR

Human MYOC: PLESTGAVVYSGSLYFQGAESRTVIRYELNTEETVKAKEIPGAGYHGQFPYSWGGYTDIDLAVDEAGLWVIYSTDEAKGA
Mouse MYOC: ALESTGAVVYAGSLYFQGAESRTVVRYELDTETVKAKEIPGAGYHGHPYAWGGYTDIDLAVDESGLWVIYSTEEAKGA

Human MYOC: IVLSKLNPENLELEQTWETNIRKQSVANAFIICGTLITVSSYTSADATVNFAYDTGTGTSKTLTIPFKNRYKYSSMIDYN
Mouse MYOC: IVLSKLNPANLELERTWETNIRKQSVANAFVICGILITVSSYSSAHATVNFAYDTKTGTSTLTIPFTNRYKYSSMIDYN

Y437

Human MYOC: PLEKKLFAWDNLNMVTYDIKLSKM (SEQ ID NO: 1)
Mouse MYOC: PLERKLFAWDNFMVTYDIKLLEM (SEQ ID NO: 6)

FIG. 3

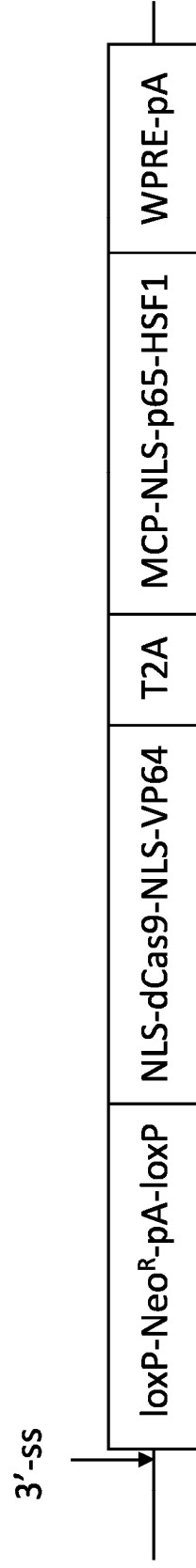


FIG. 4A

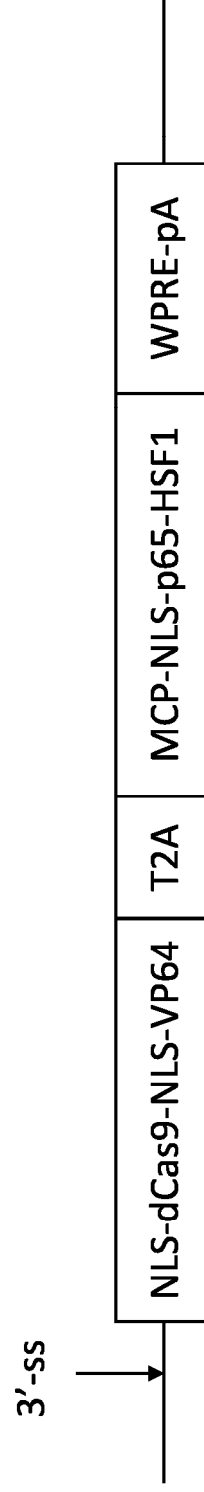


FIG. 4B

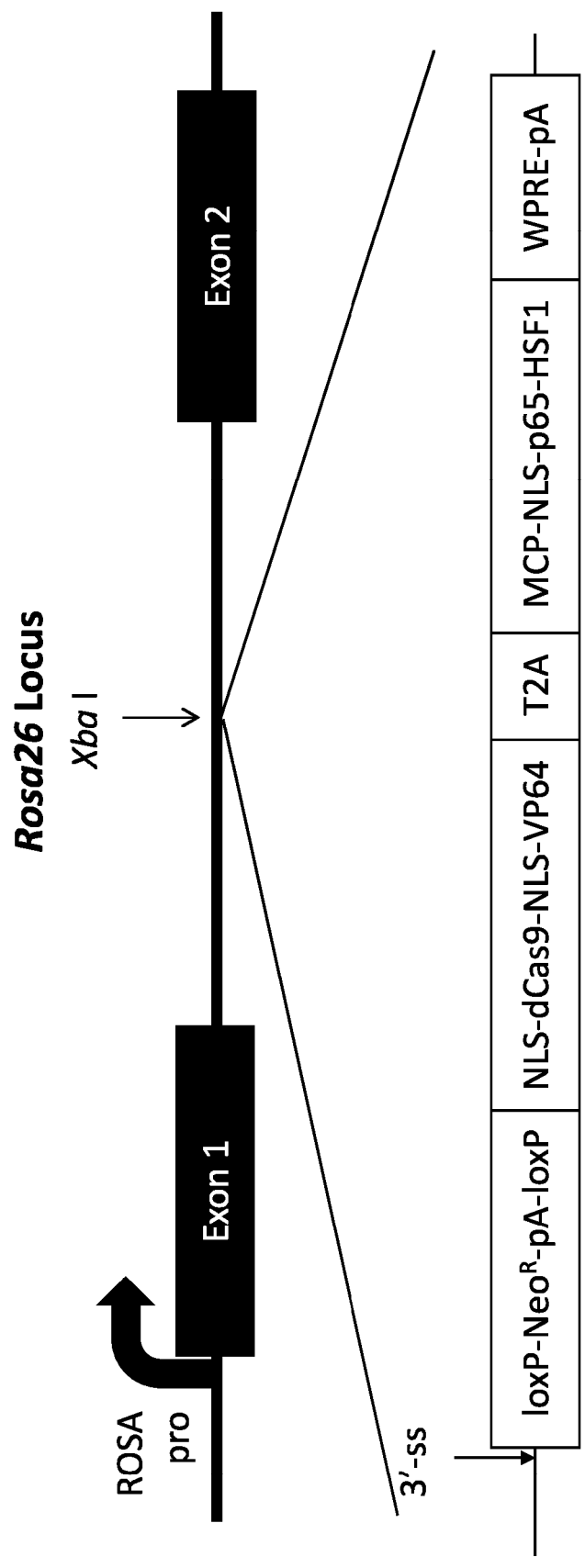


FIG. 5

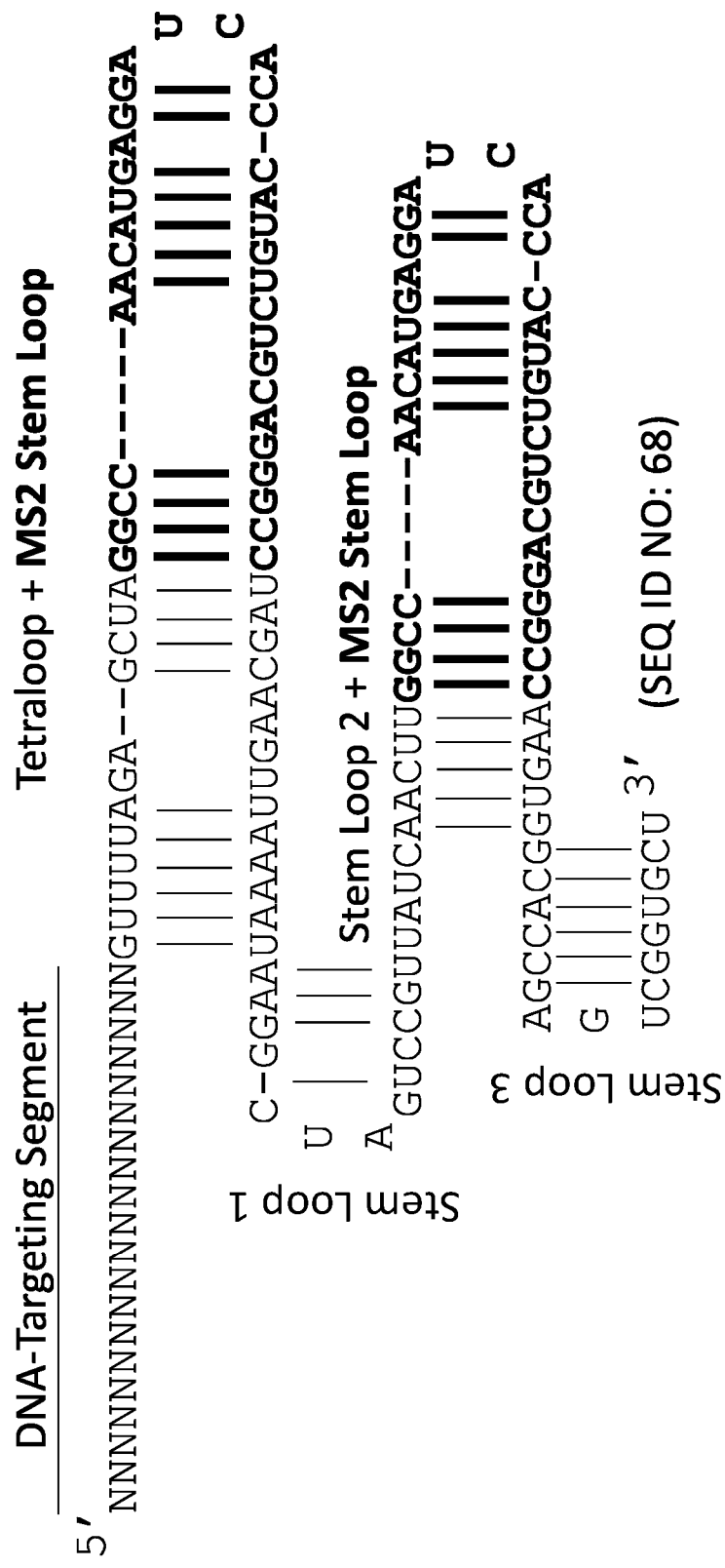


FIG. 6

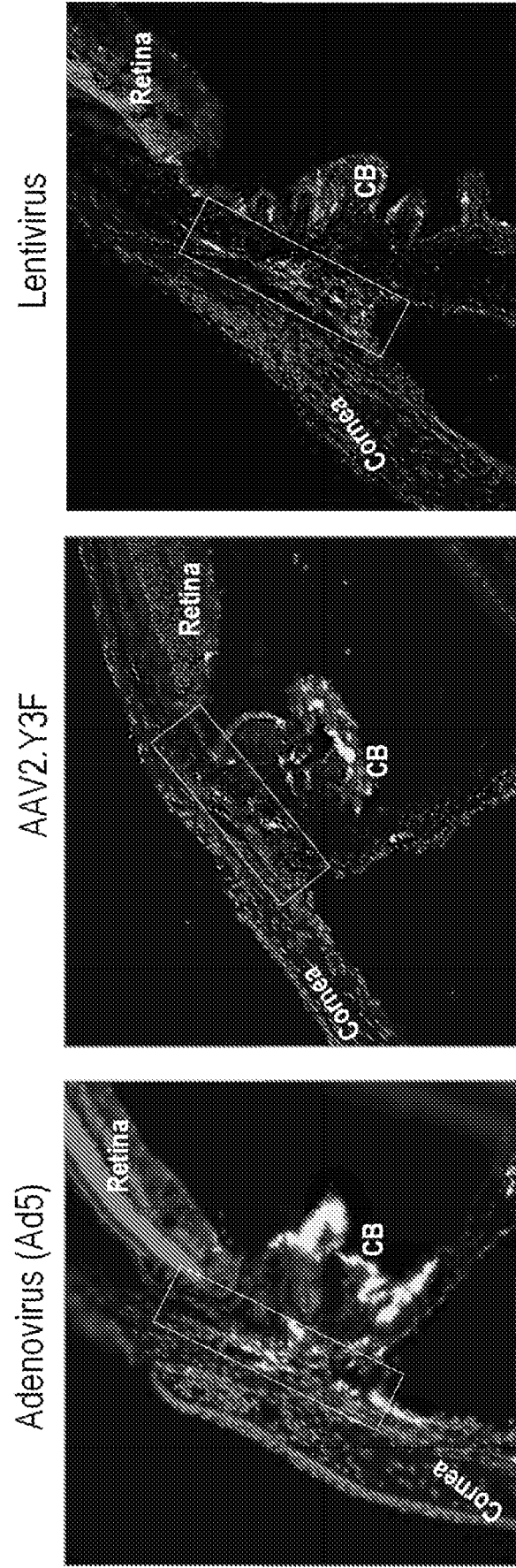


FIG. 7

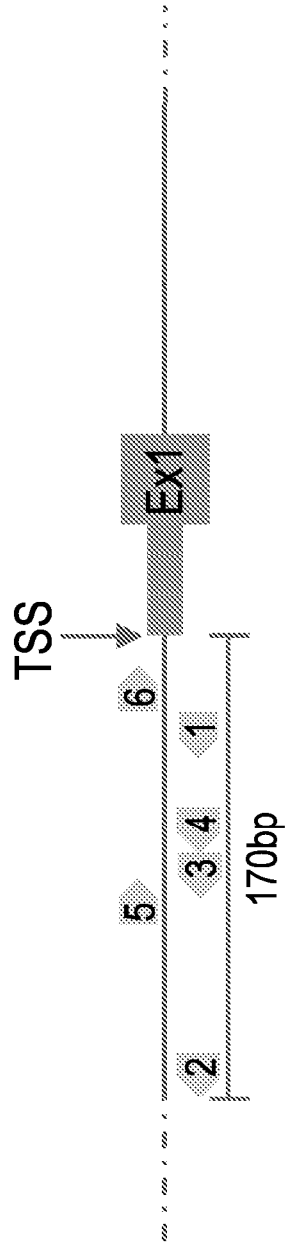


FIG. 8

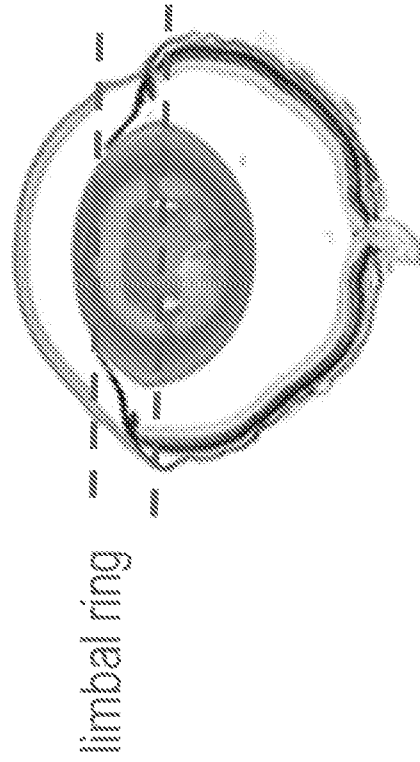


FIG. 9

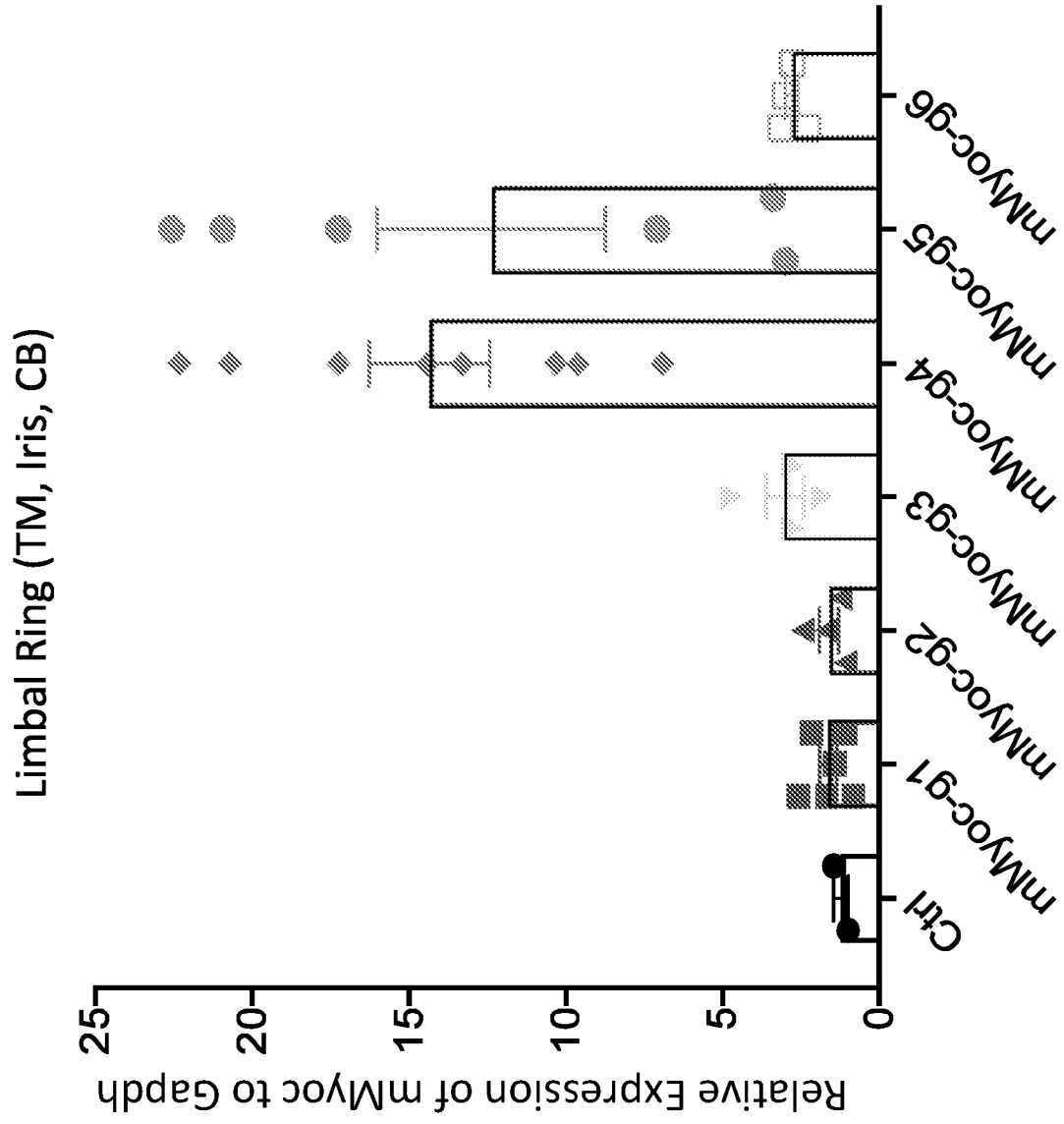


FIG. 10A

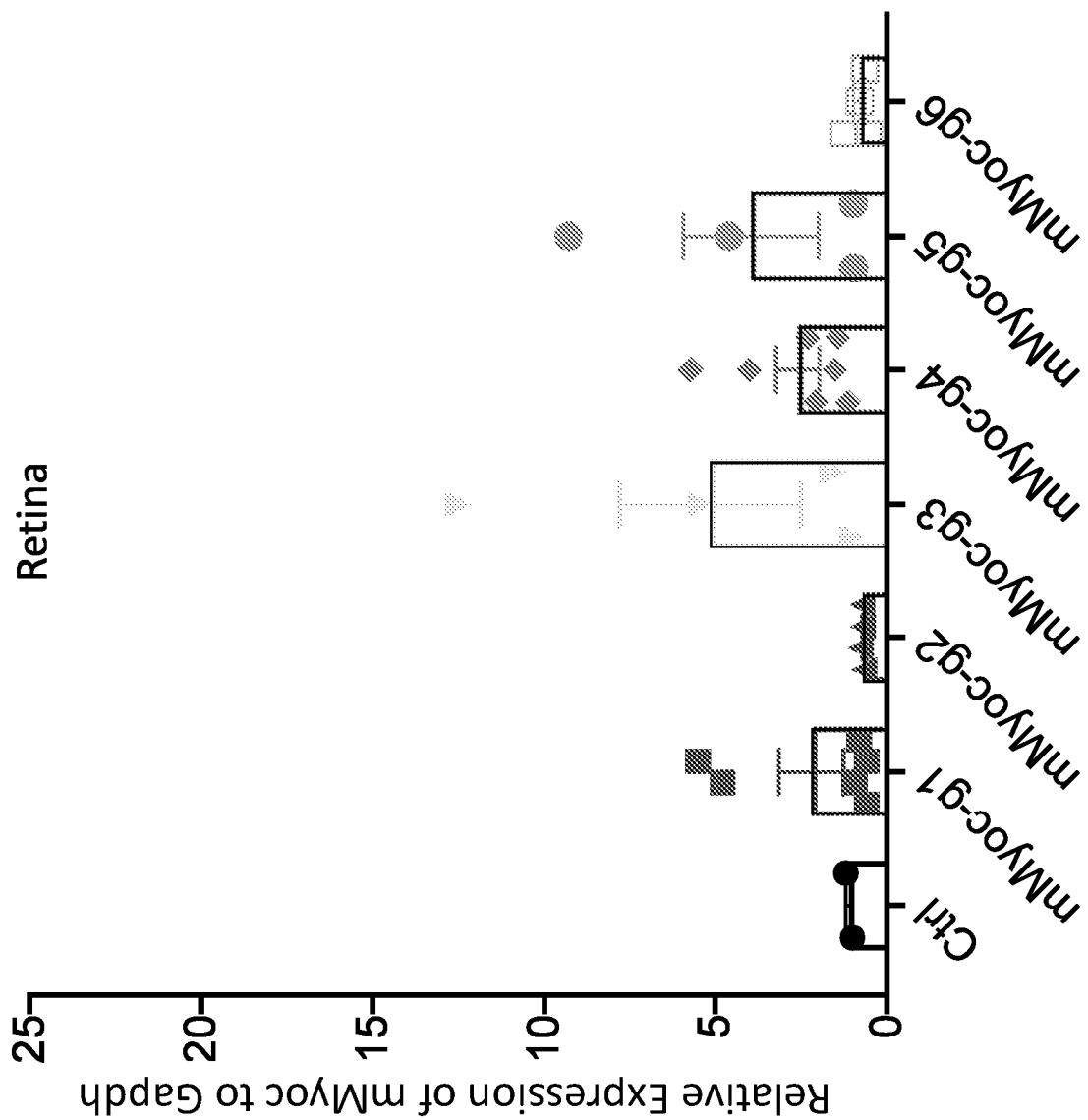


FIG. 10B

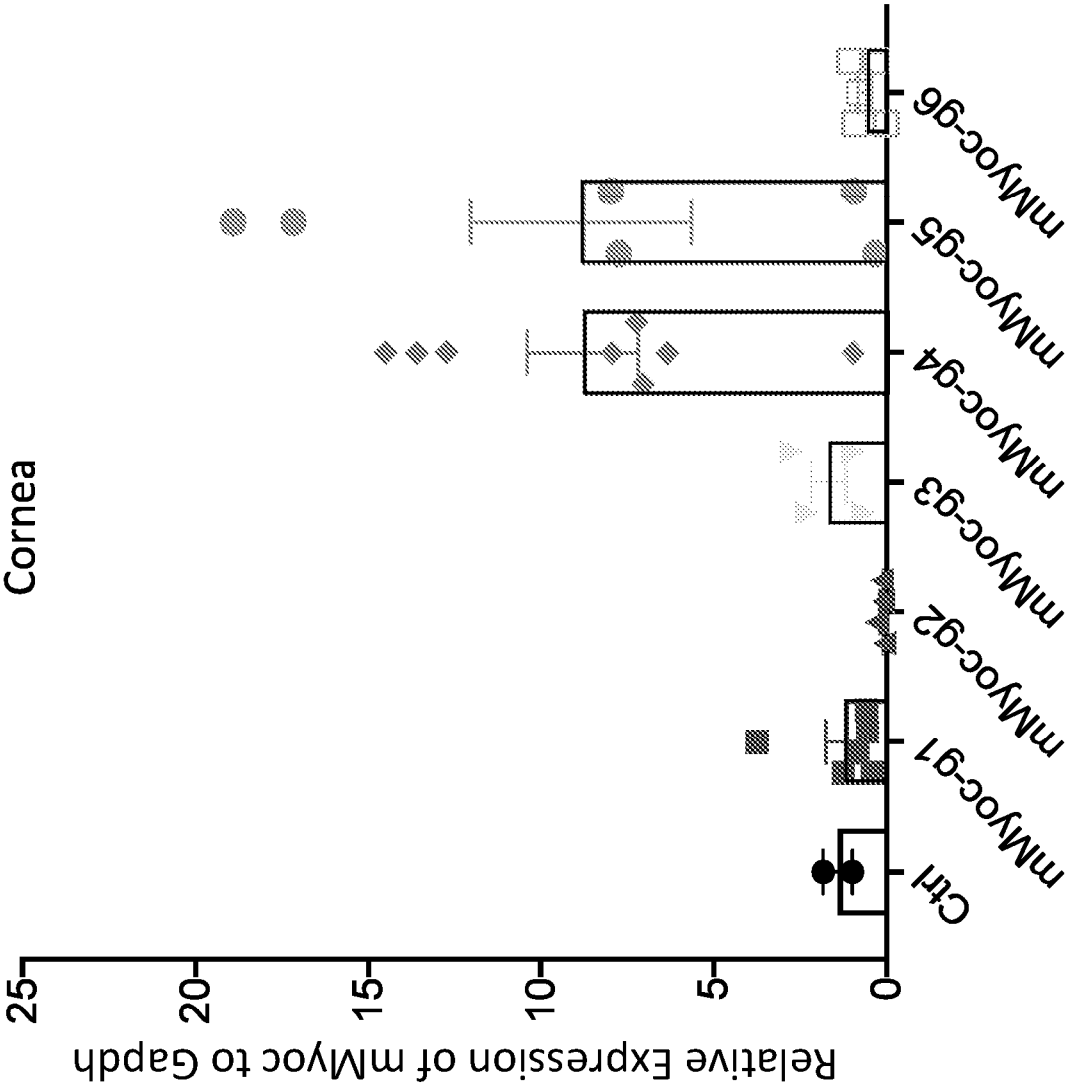


FIG. 10C

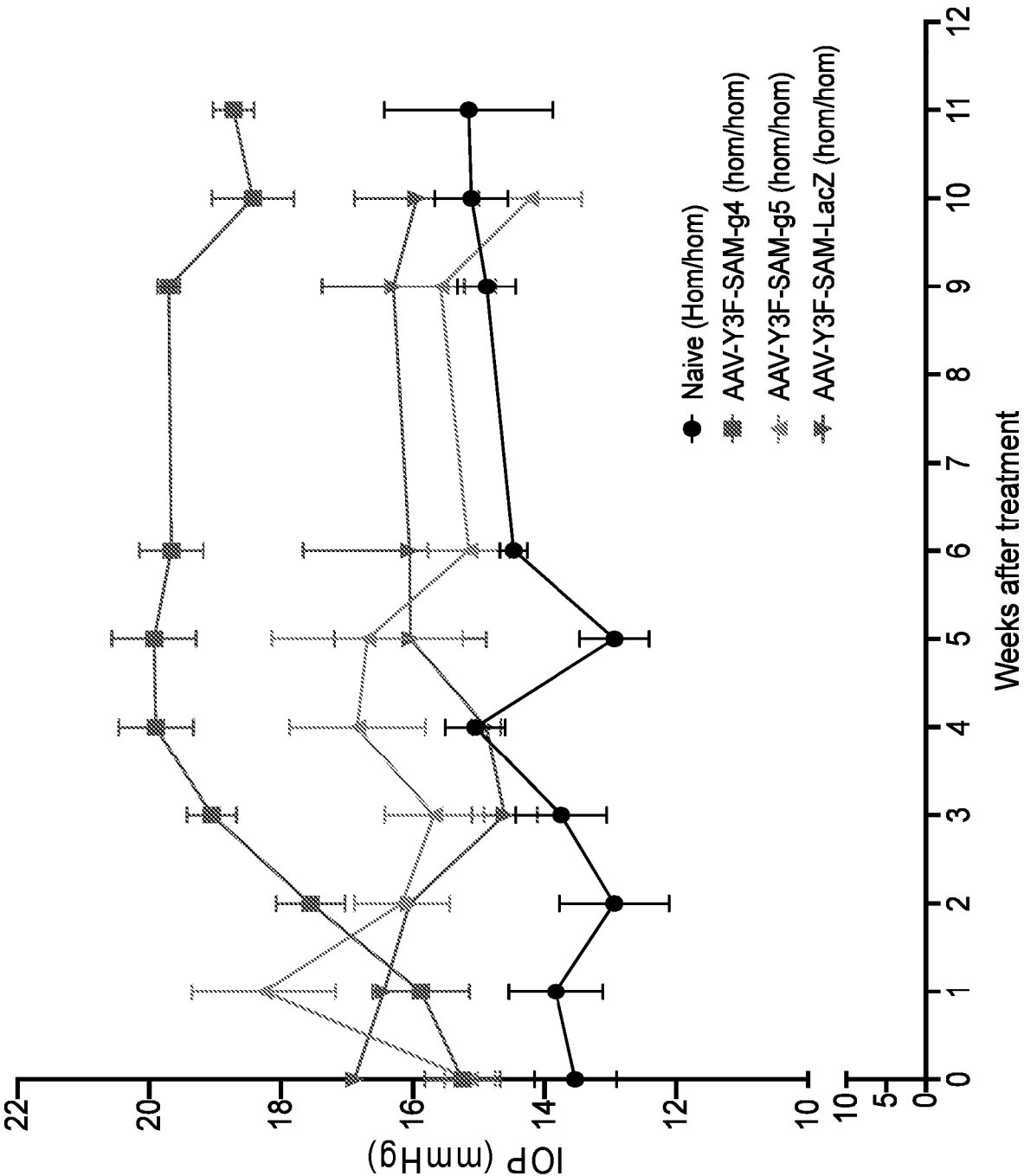


FIG. 11

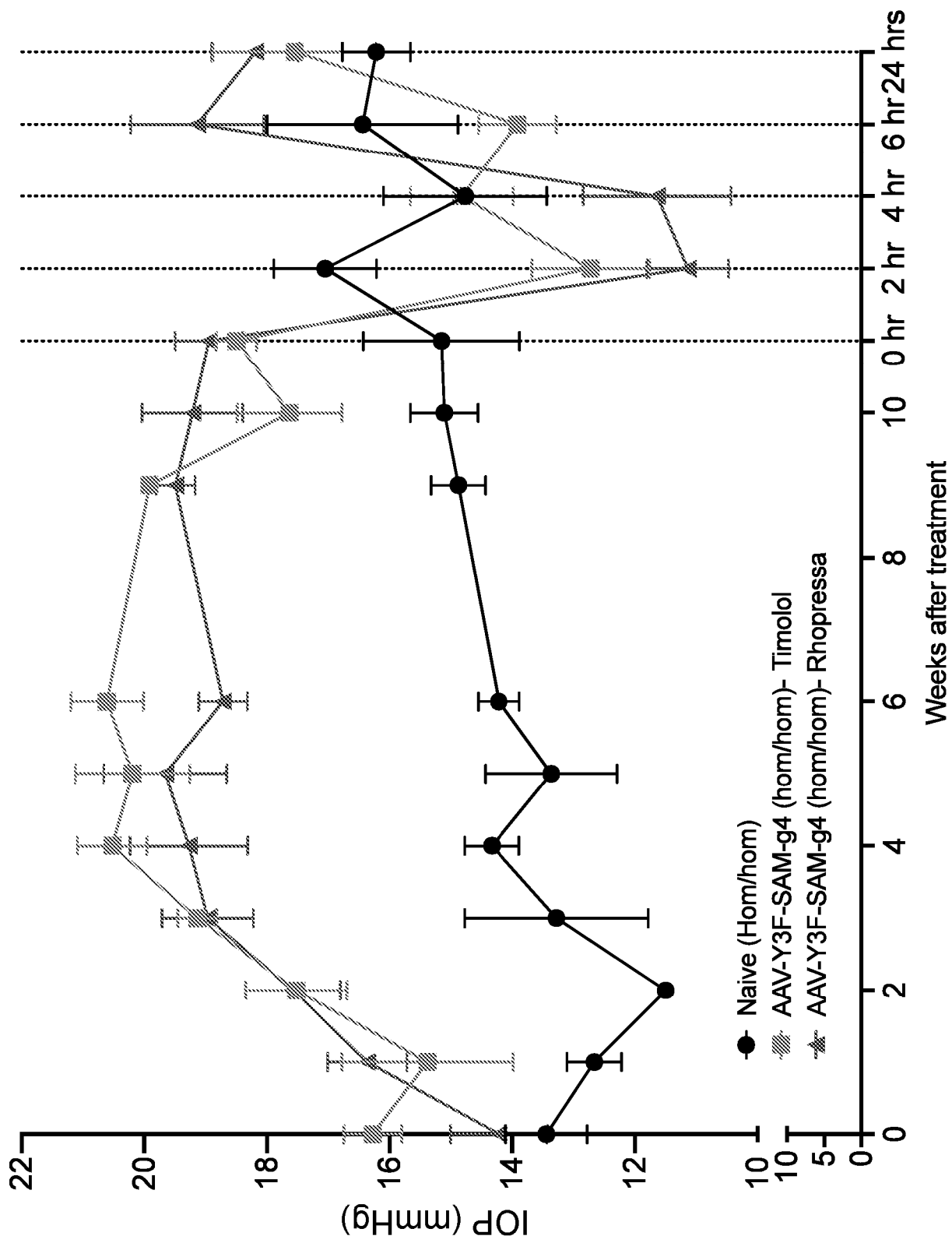


FIG. 12

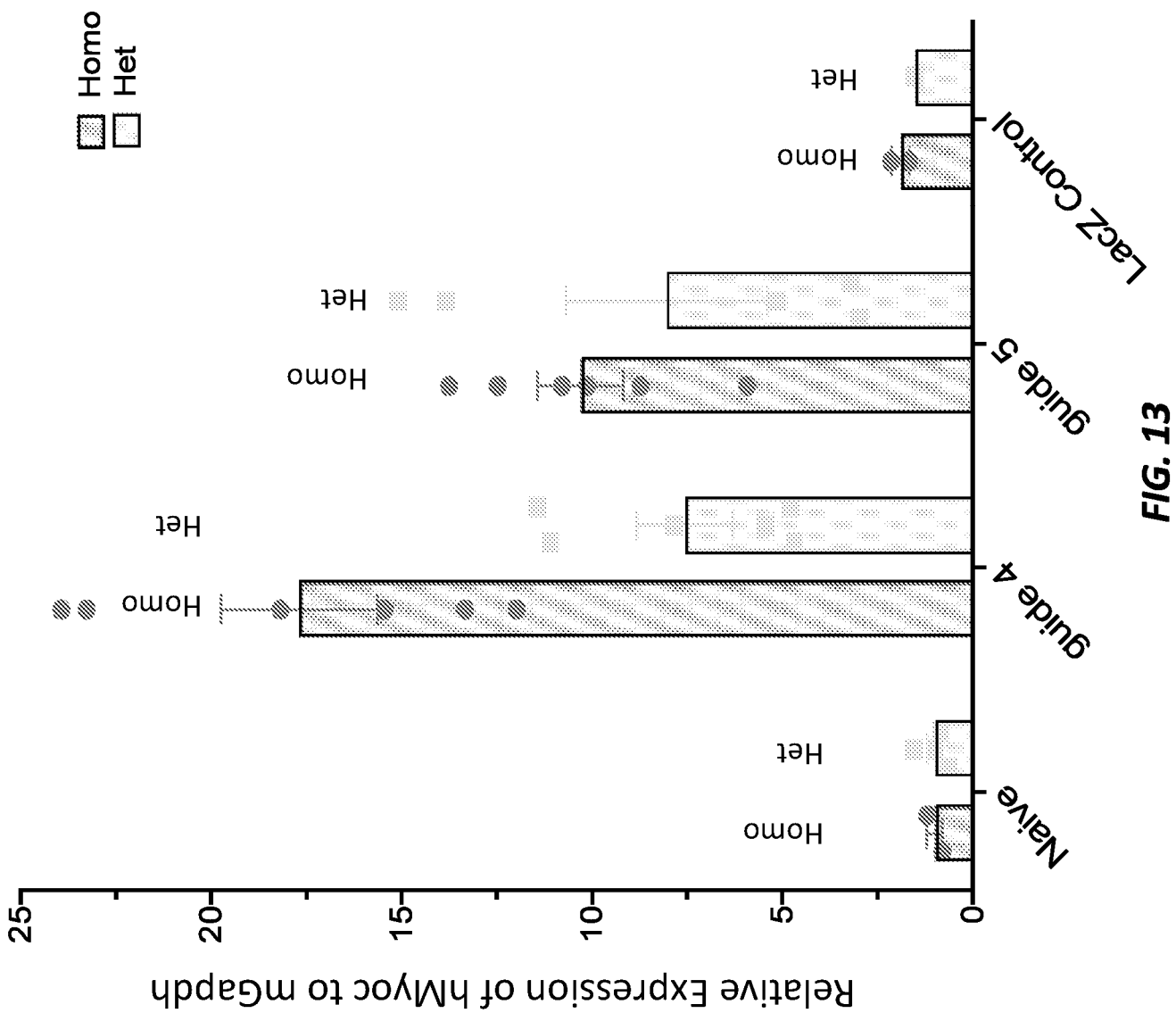
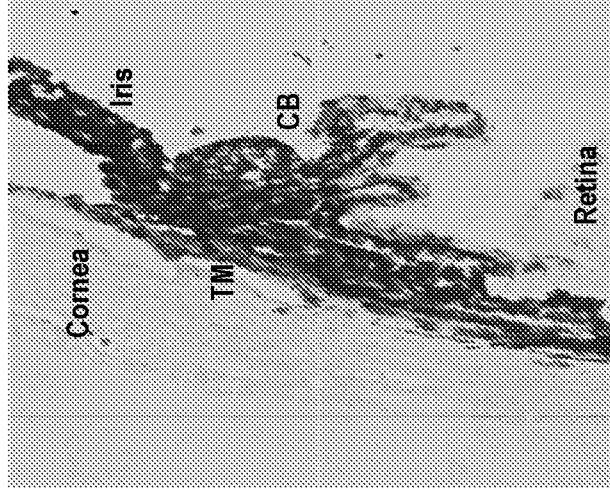
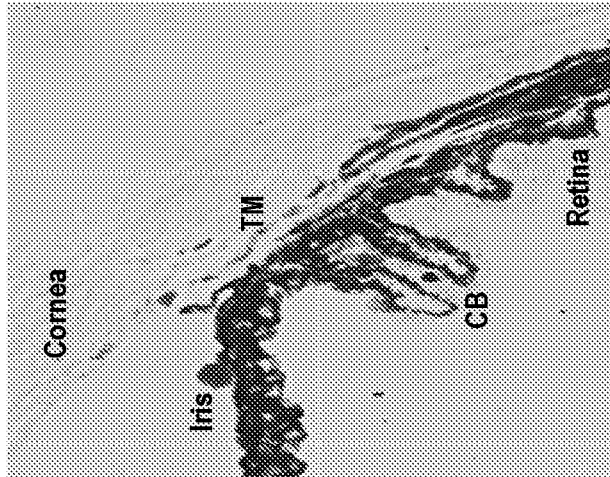


FIG. 13

Y437H Homo



Y437H Het



Naïve

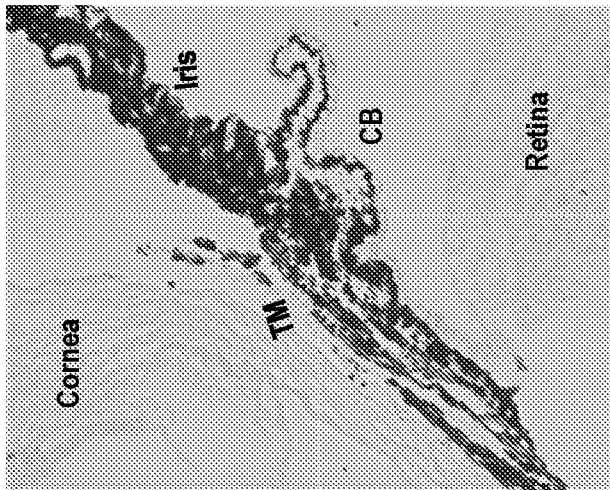


FIG. 14

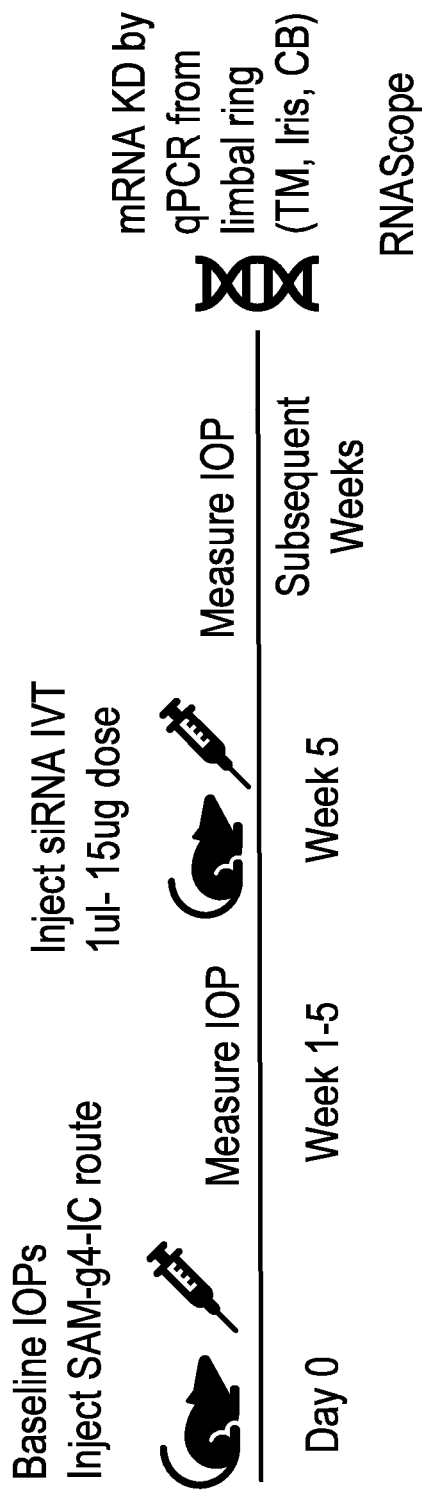
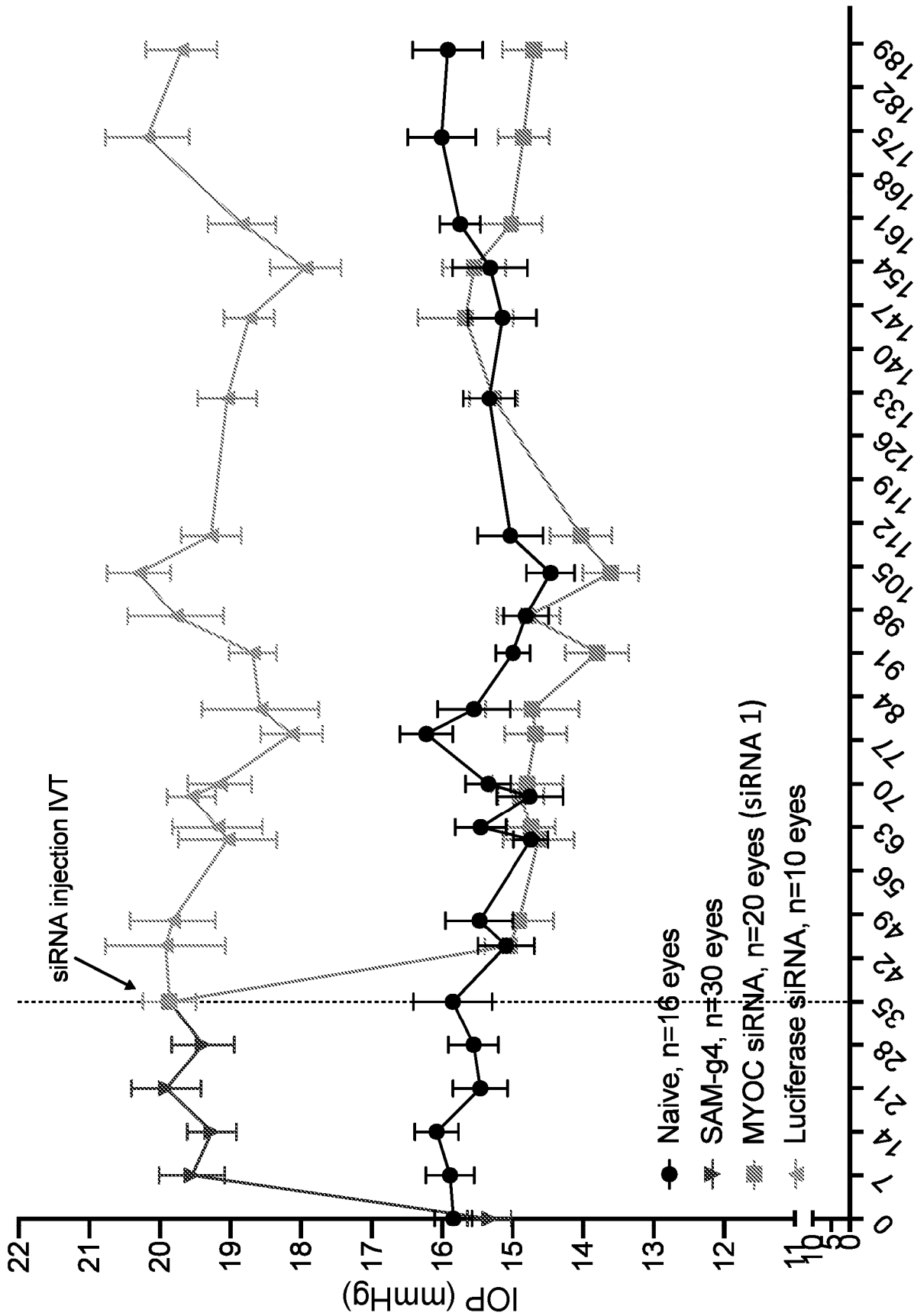


FIG. 15A



Days after treatment SAM-g4; Days after siRNA inj

FIG. 15B

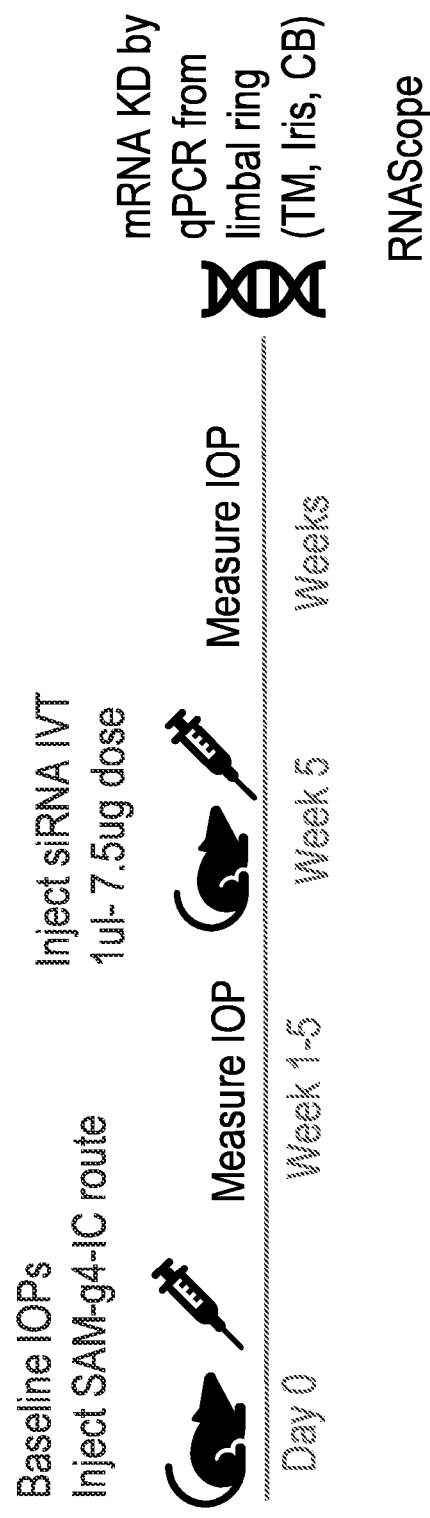
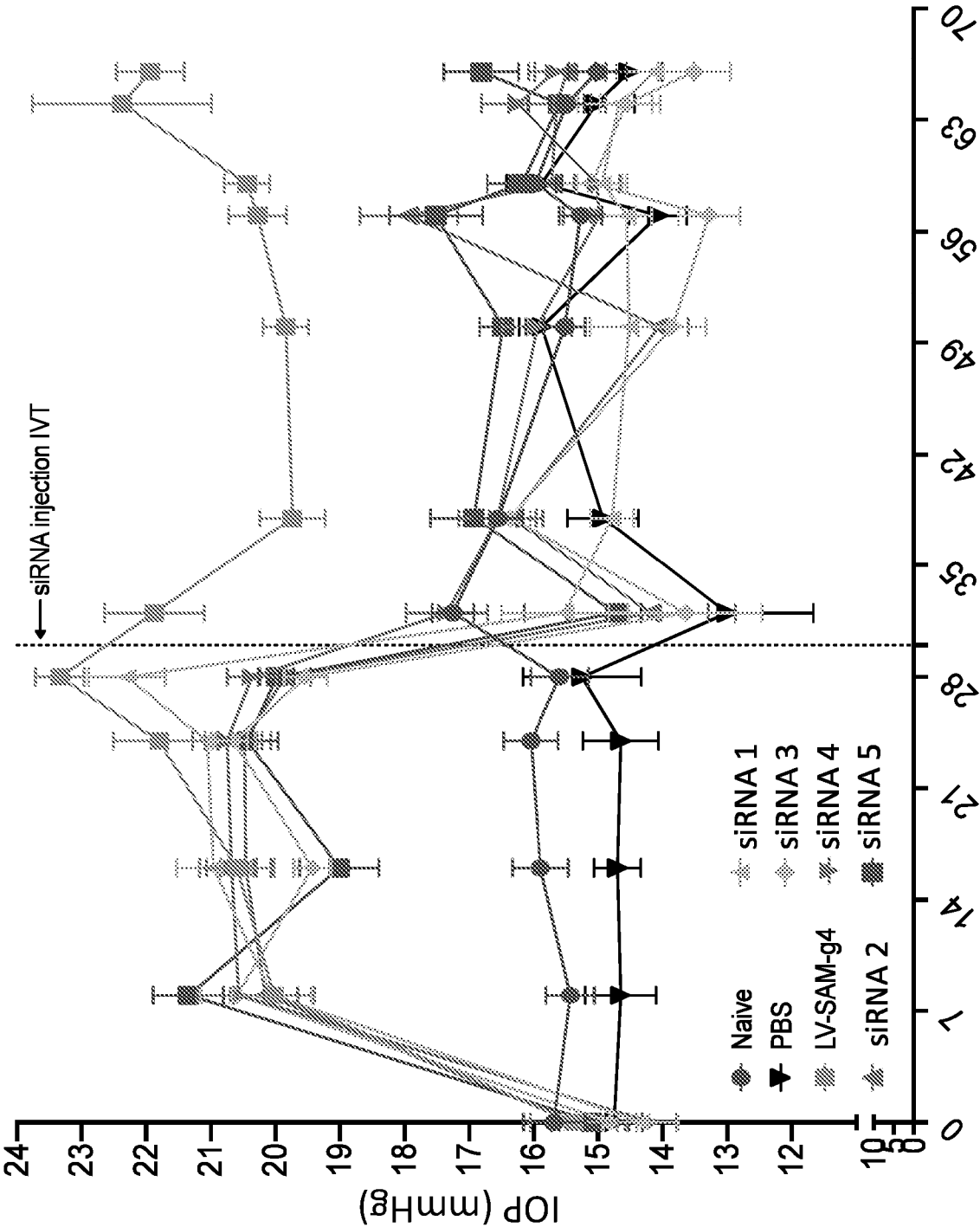
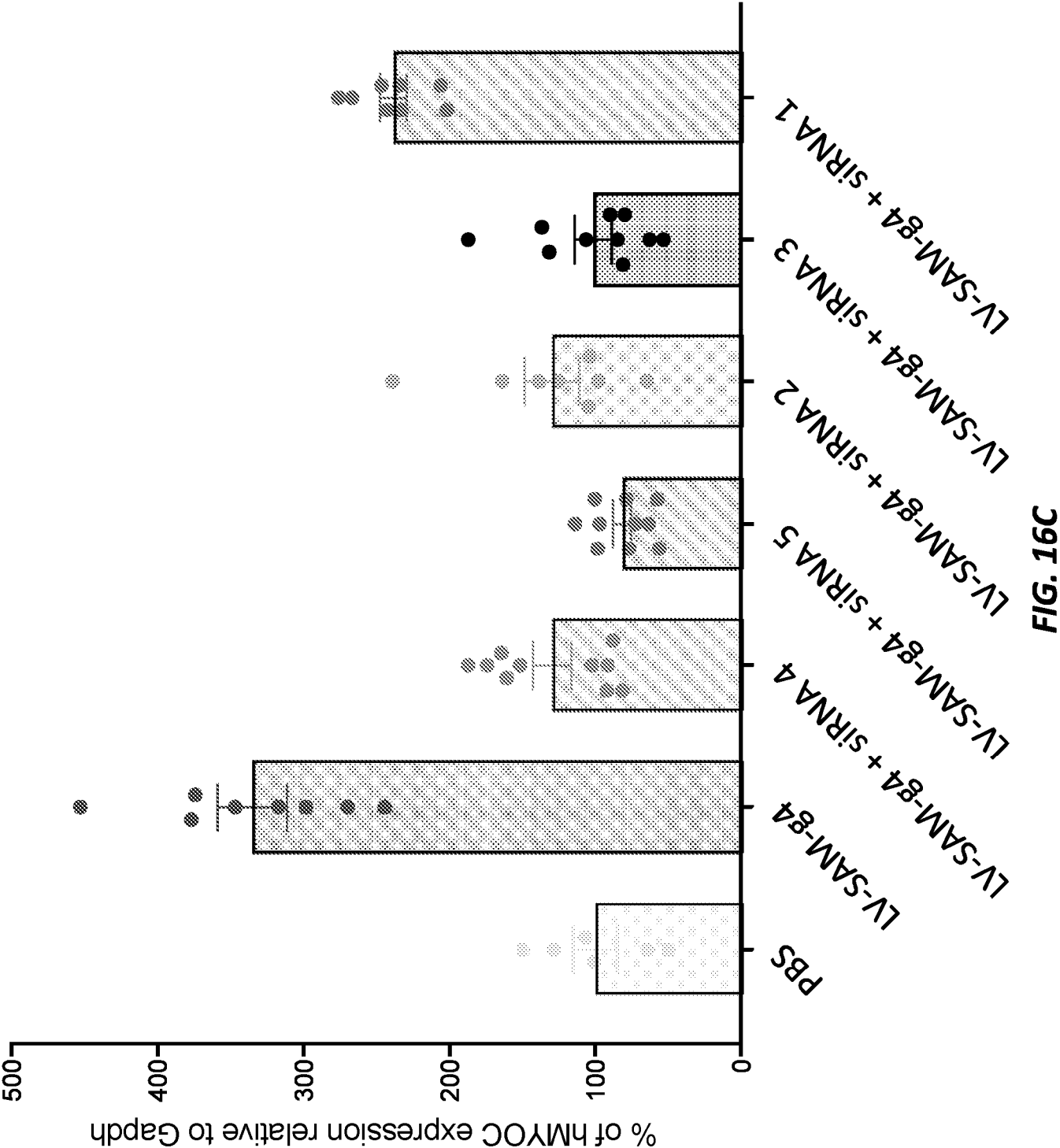


FIG. 16A



Days after treatment SAM-g4; Days after siRNA inj

FIG. 16B



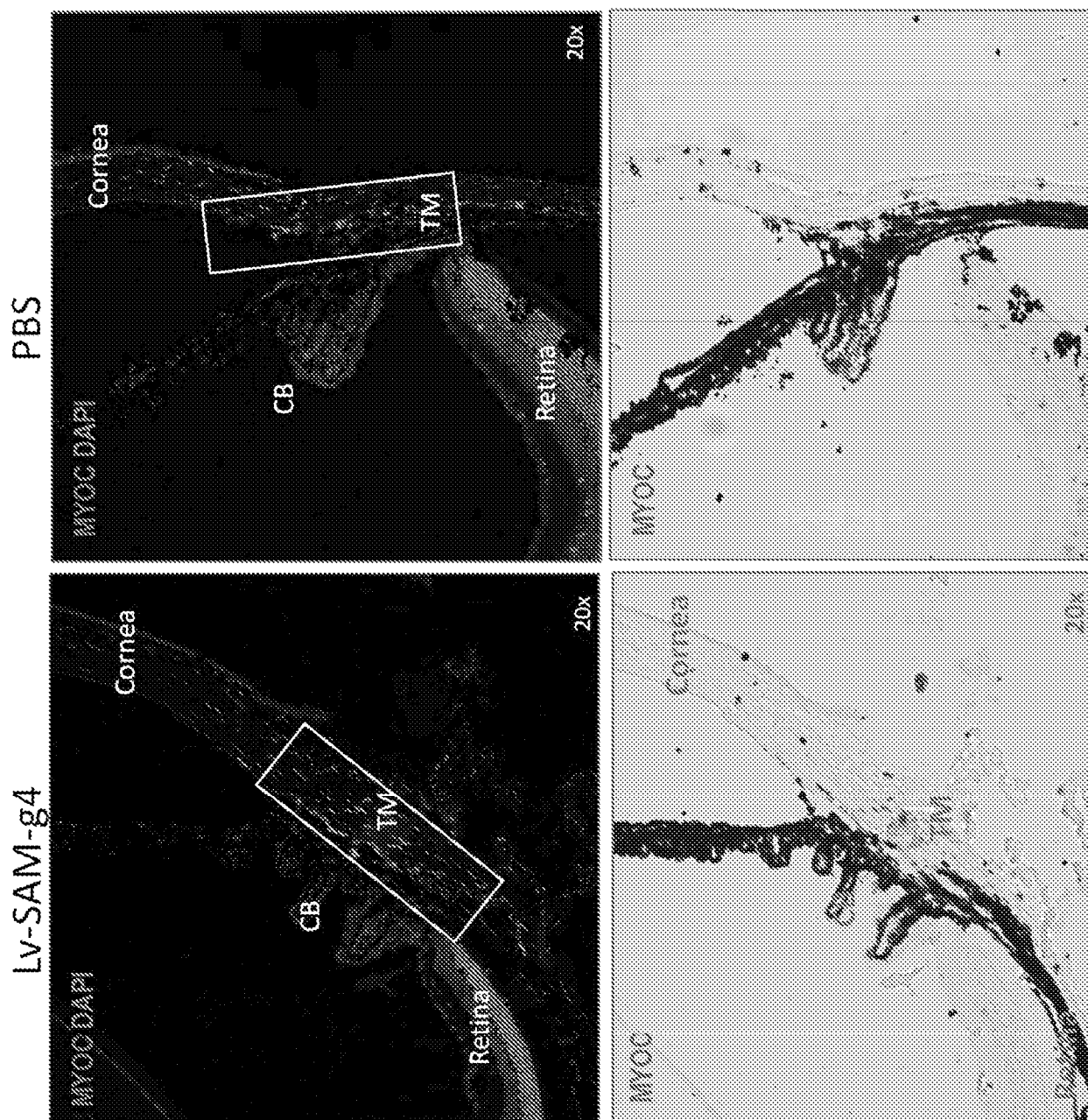


FIG. 16D

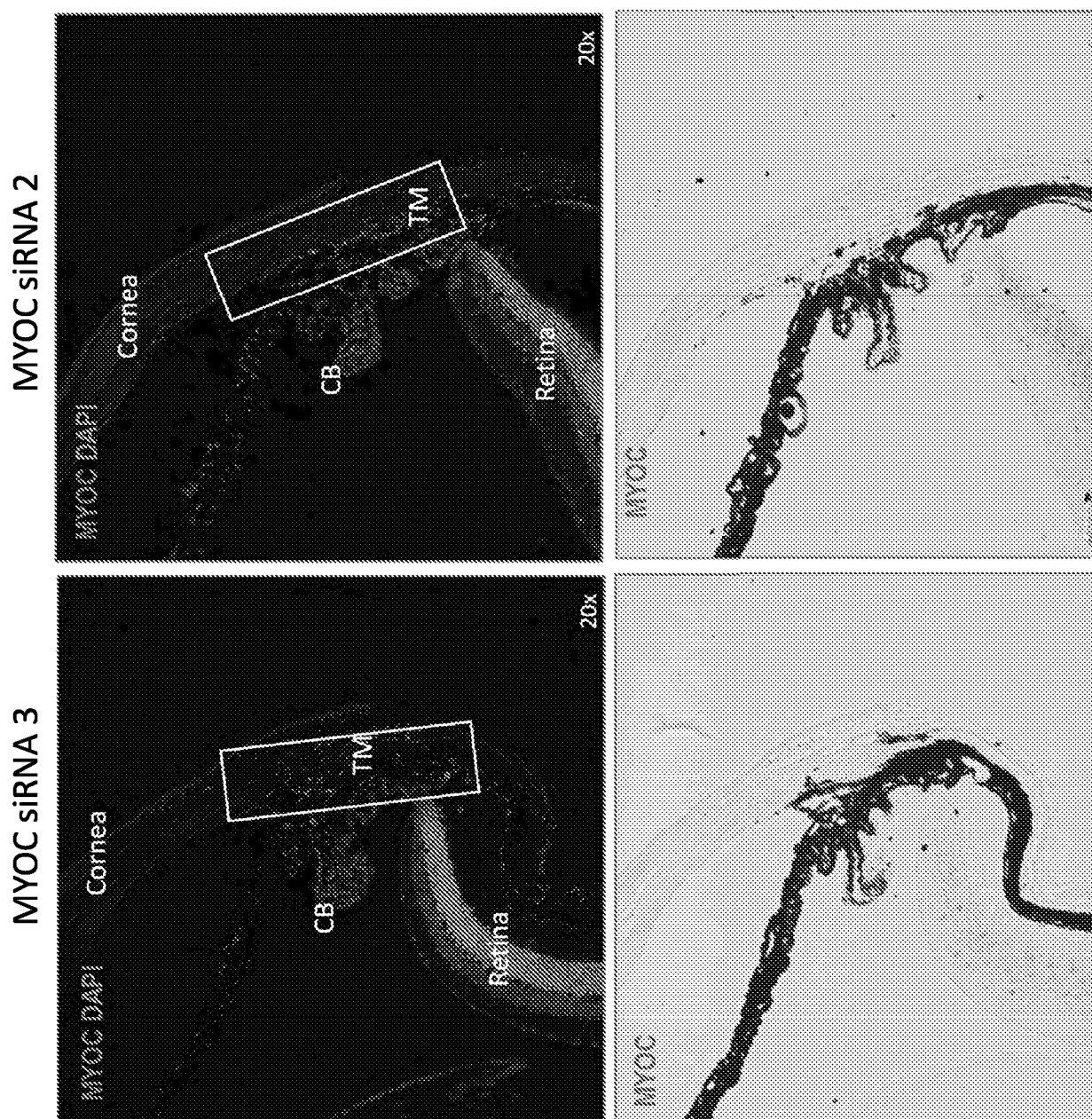


FIG. 16D continued

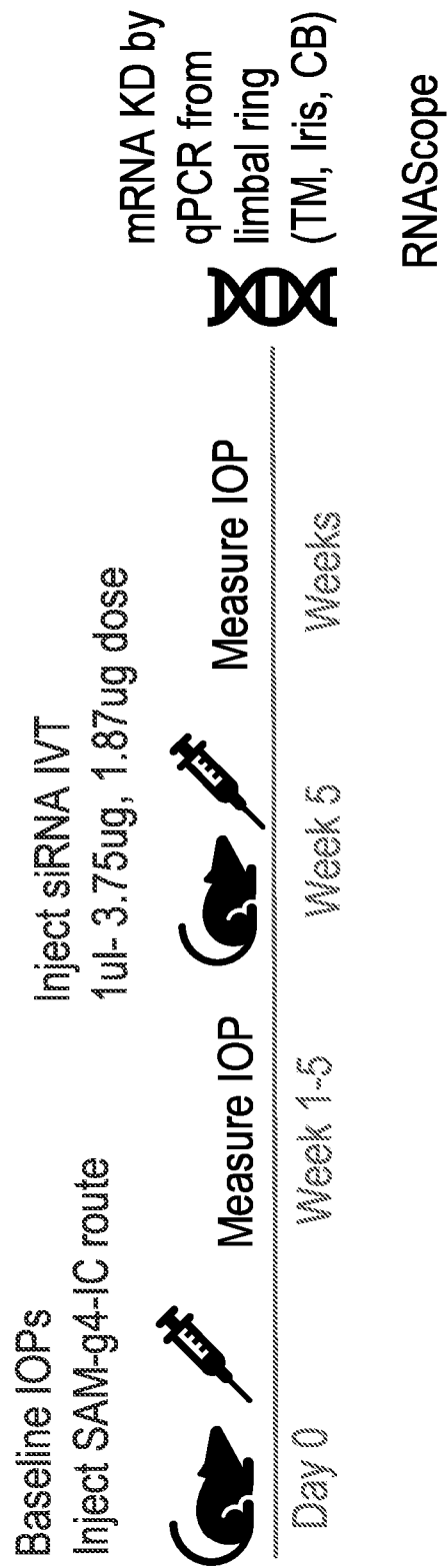


FIG. 17A

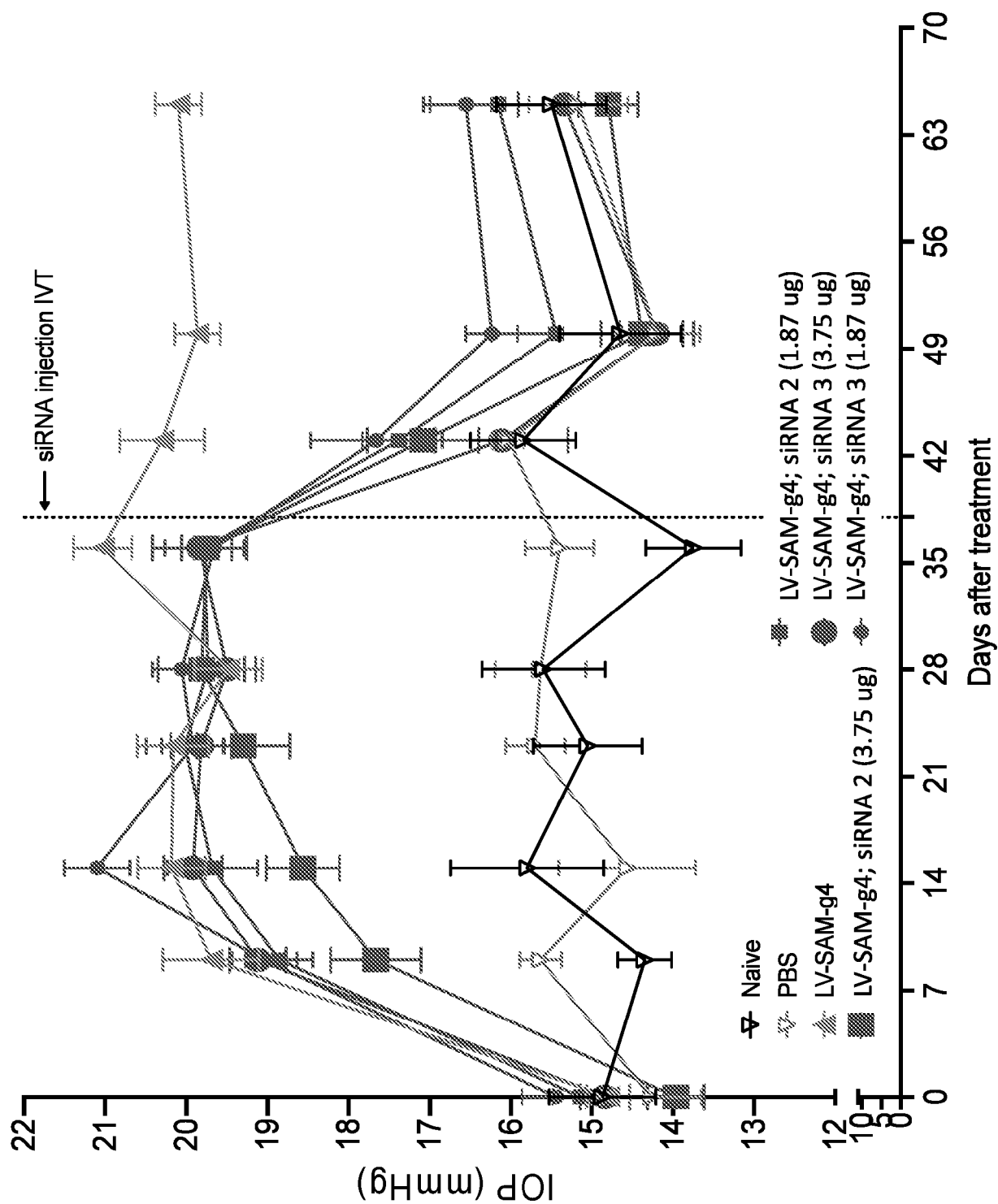
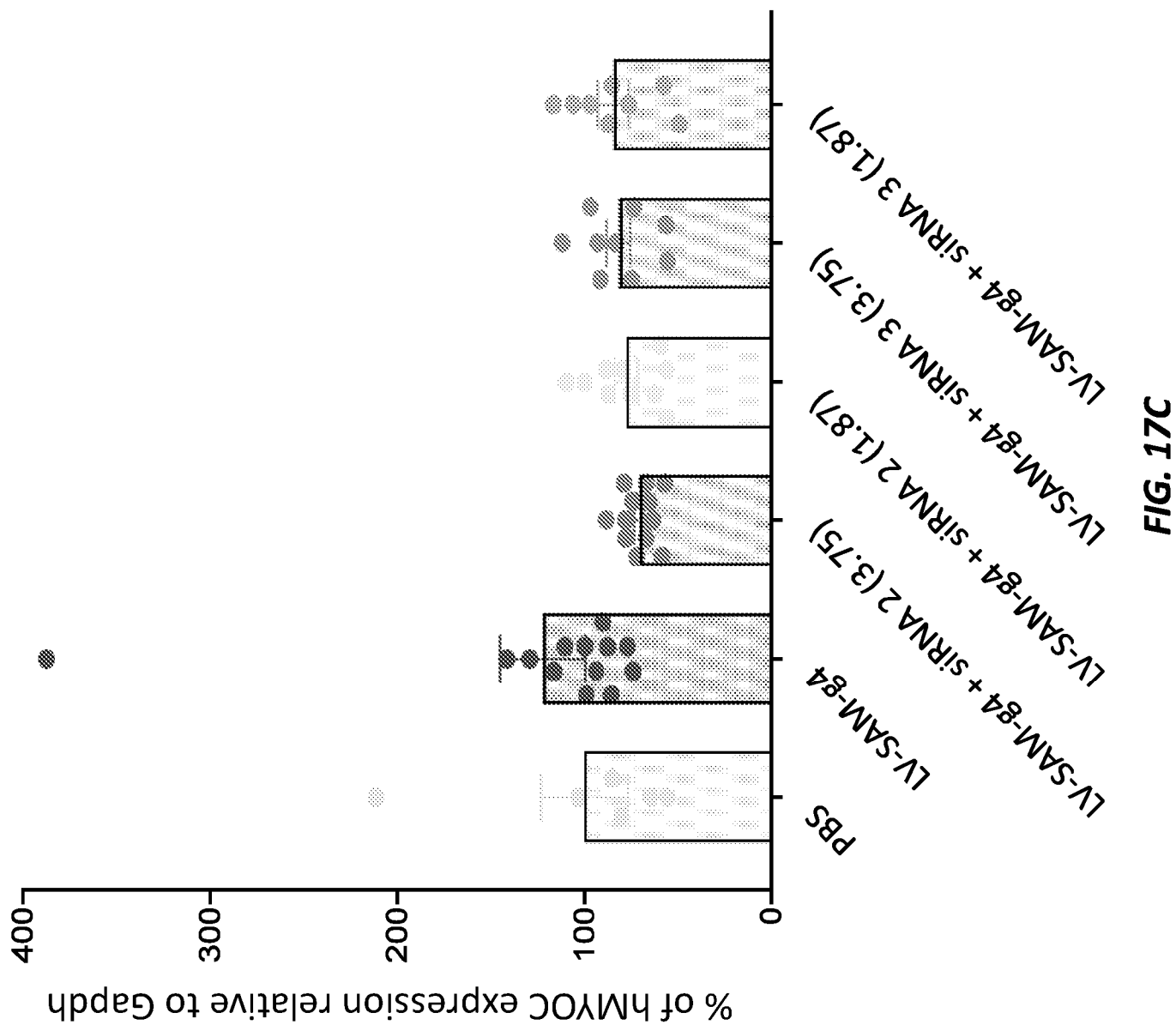


FIG. 17B



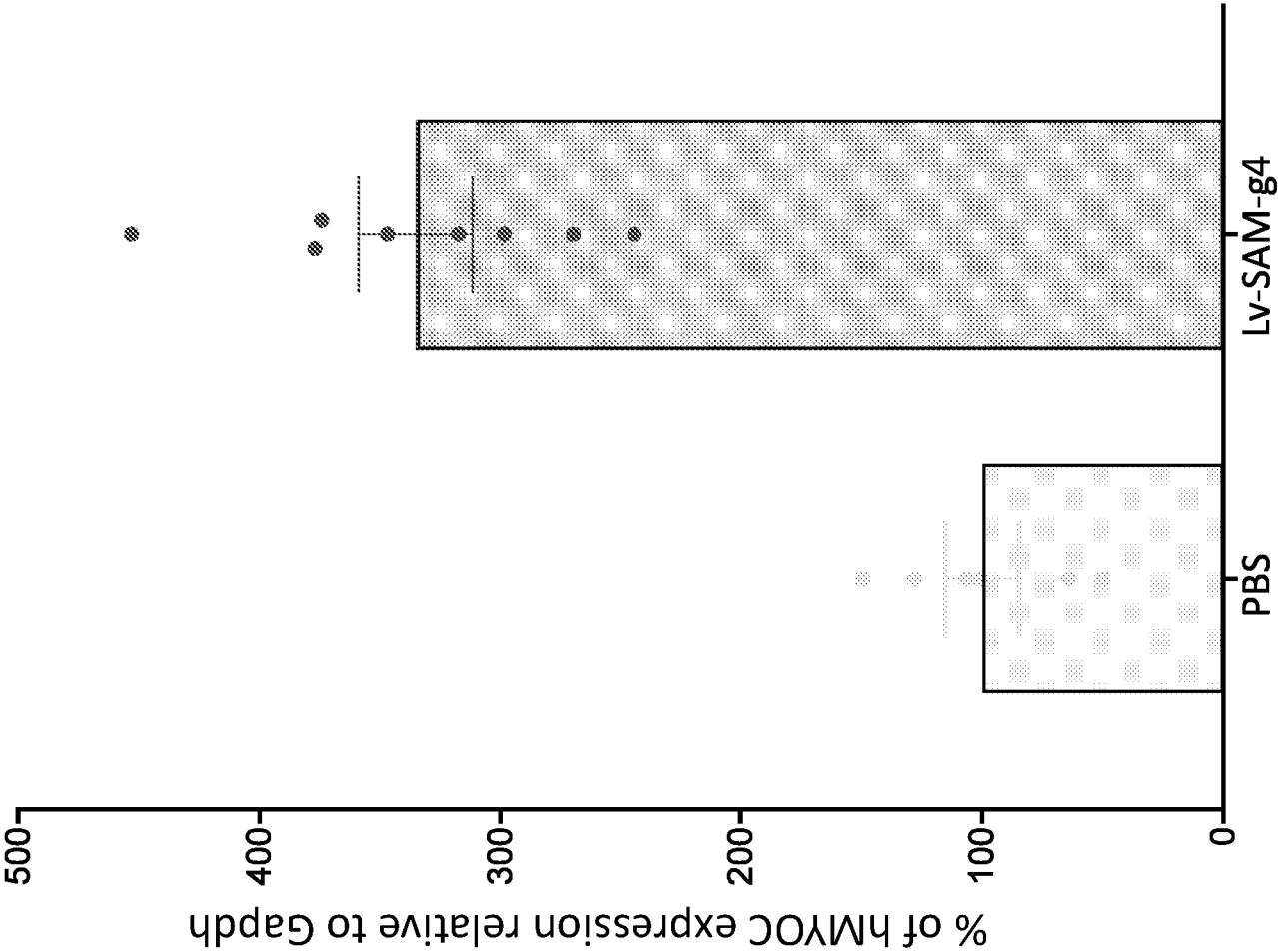


FIG. 18A

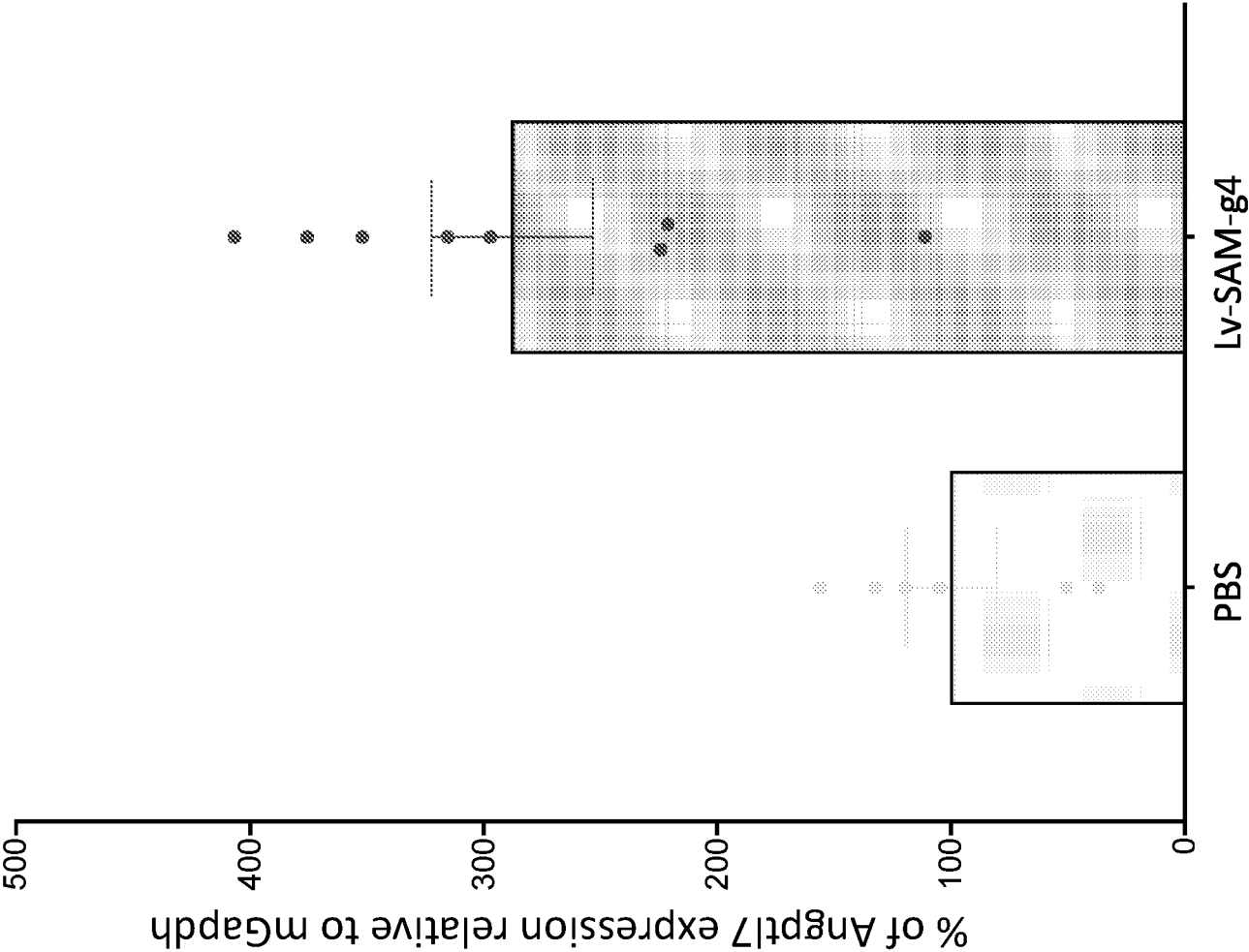


FIG. 18B

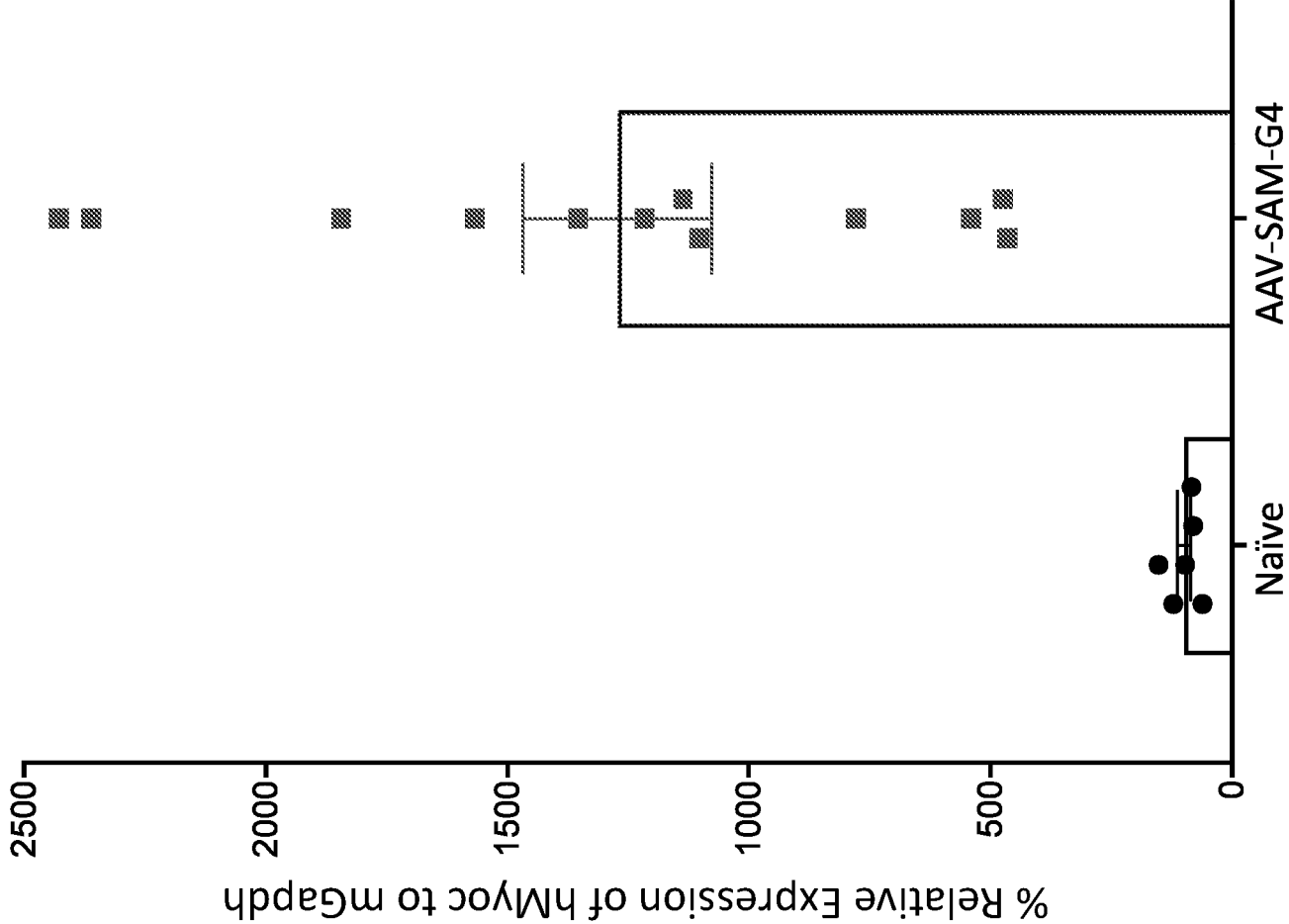


FIG. 18C

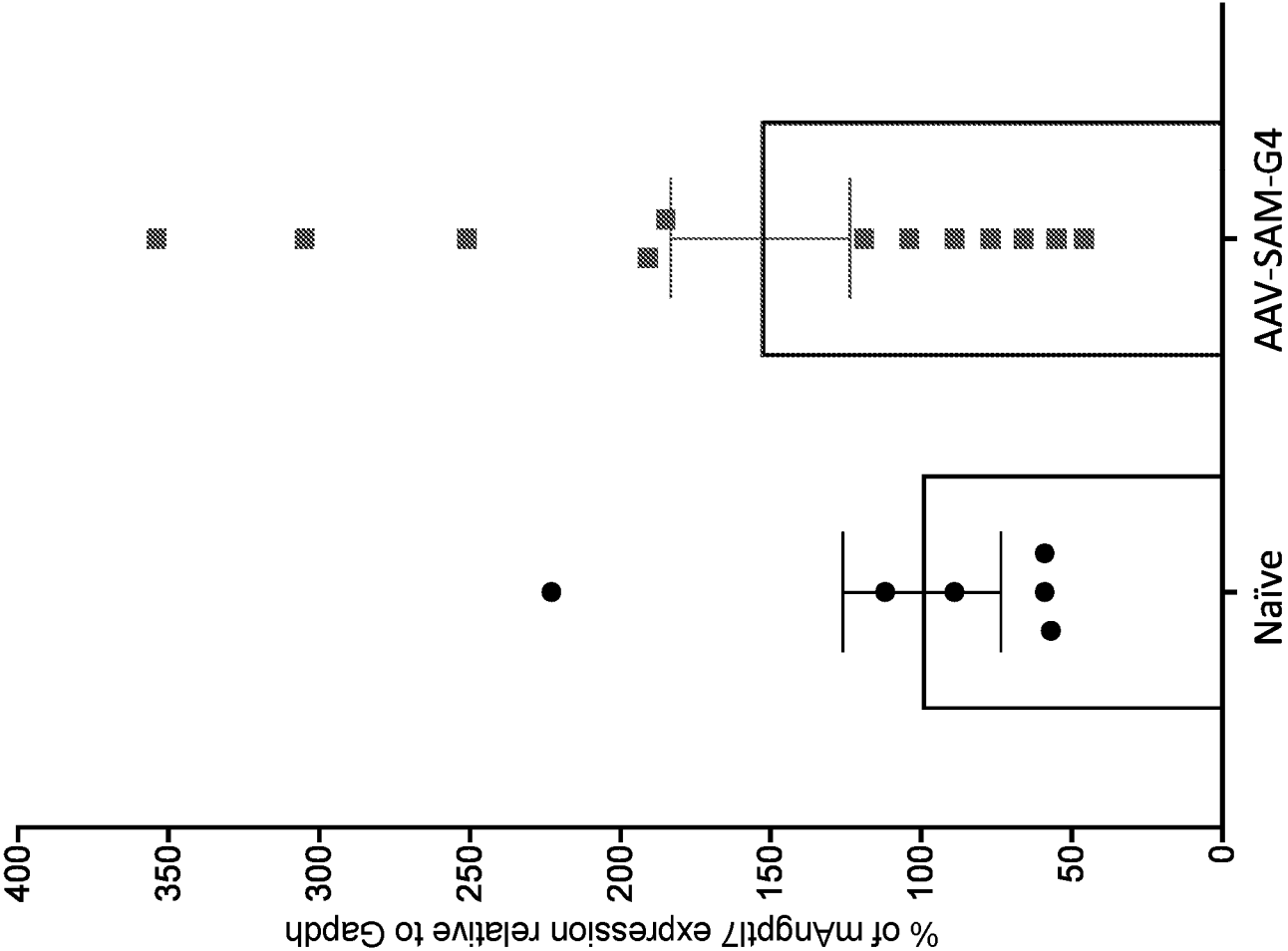


FIG. 18D

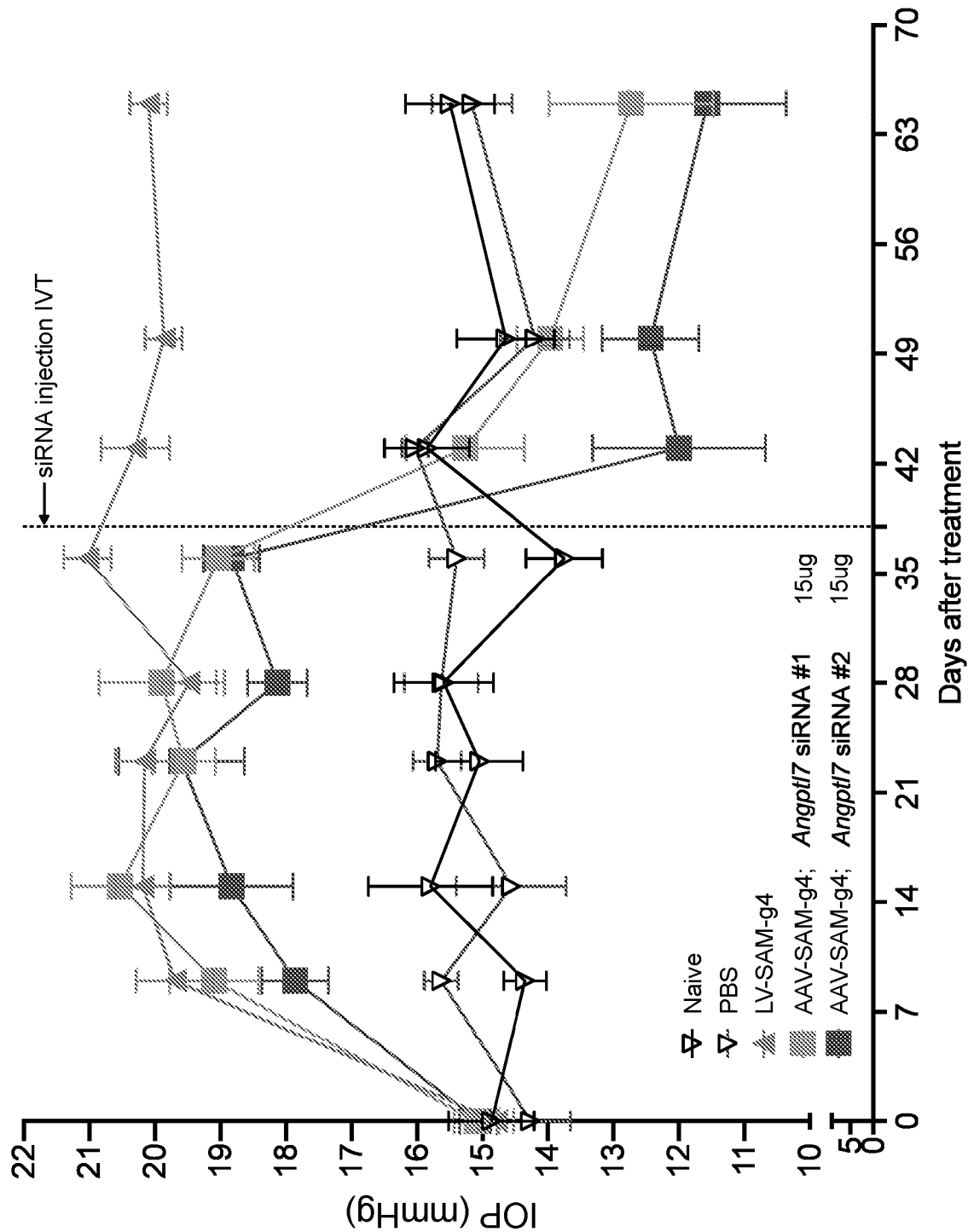


FIG. 19A

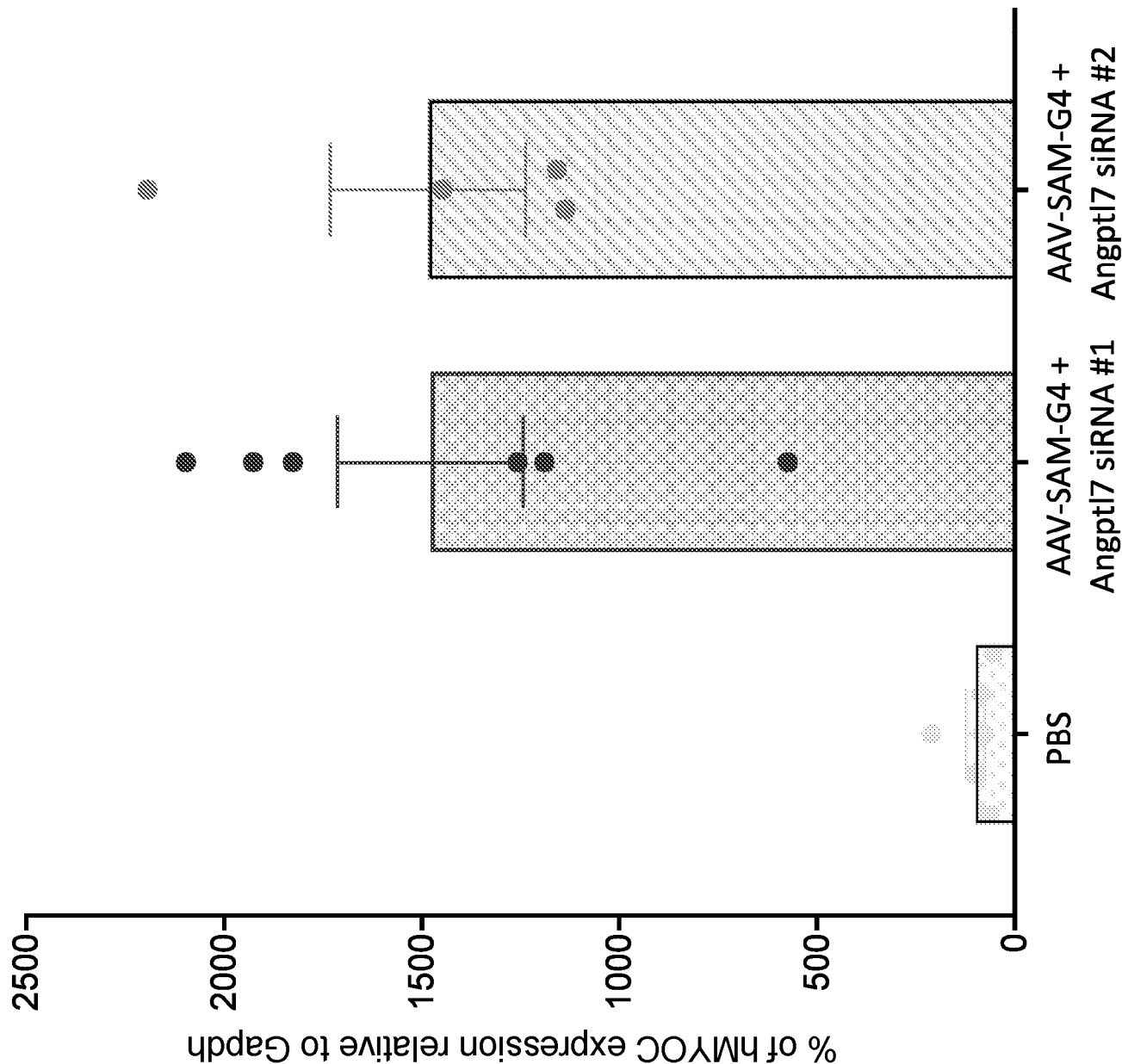


FIG. 19B

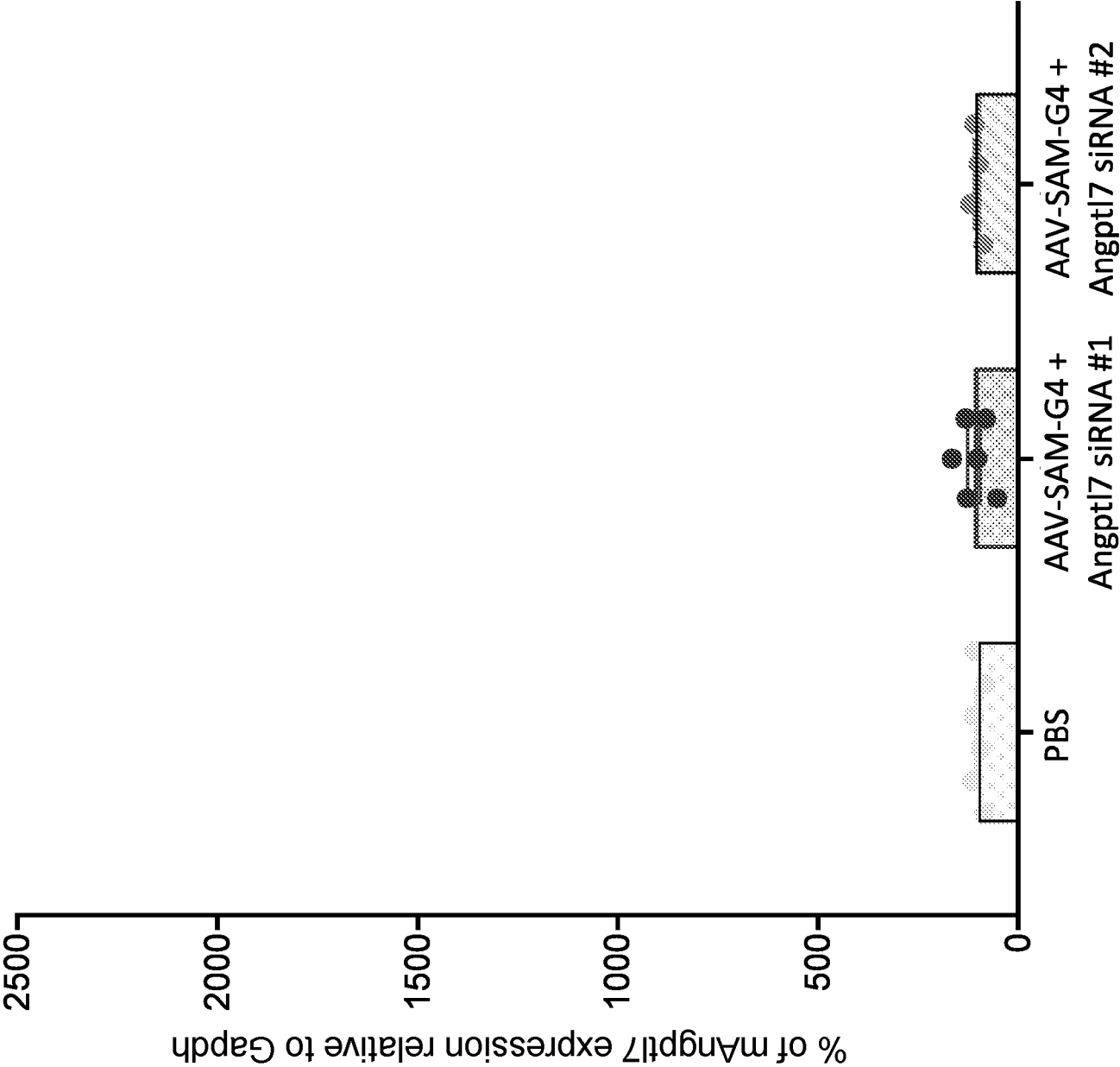


FIG. 19C

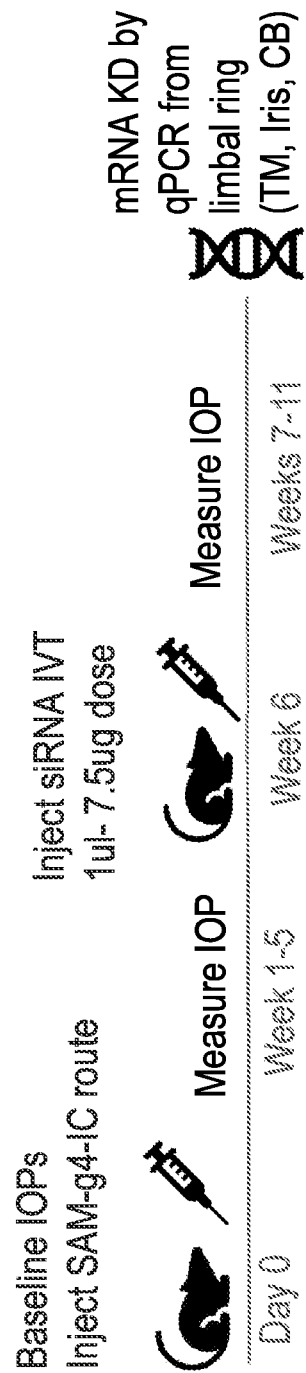


FIG. 20A

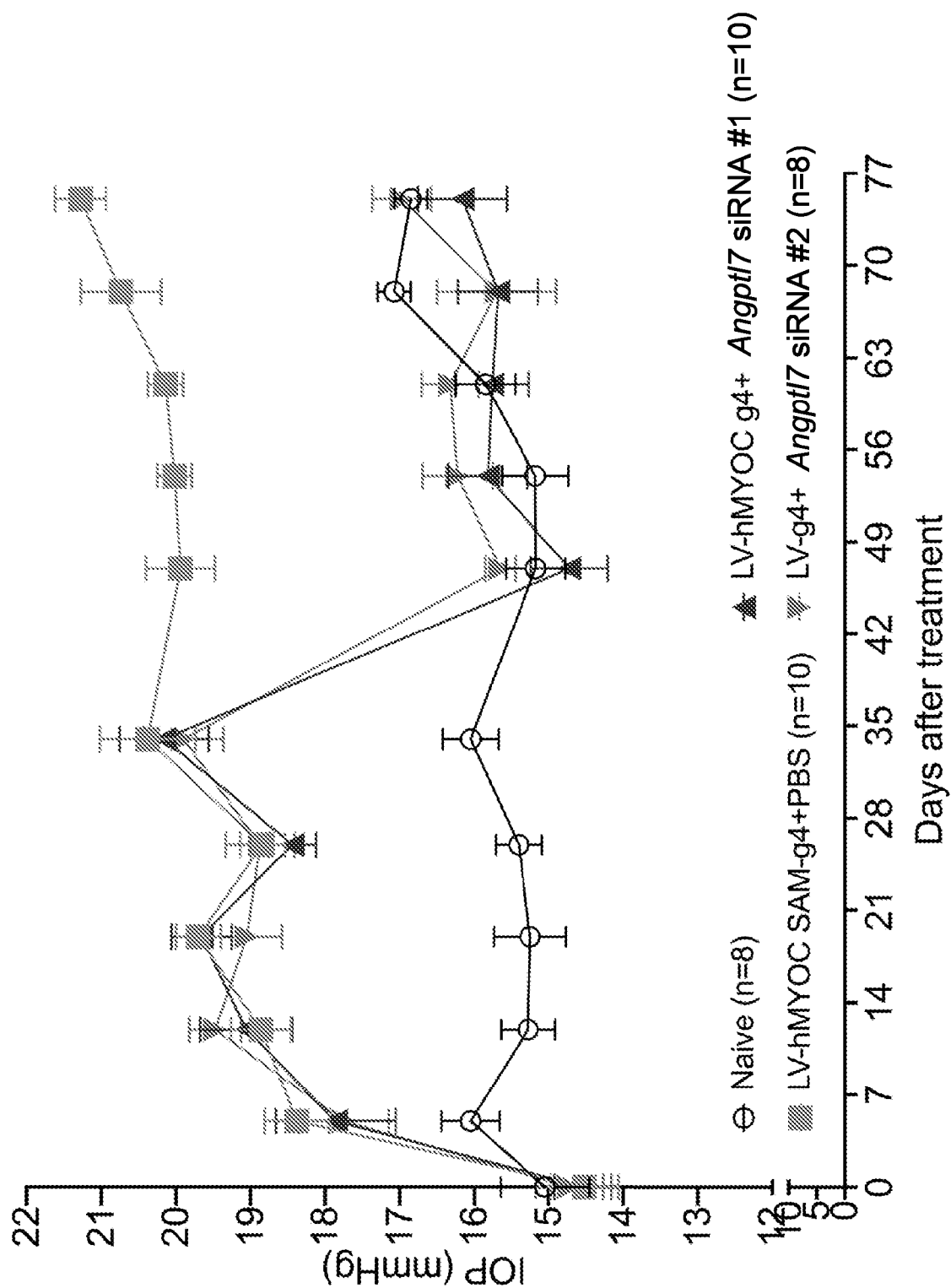


FIG. 20B

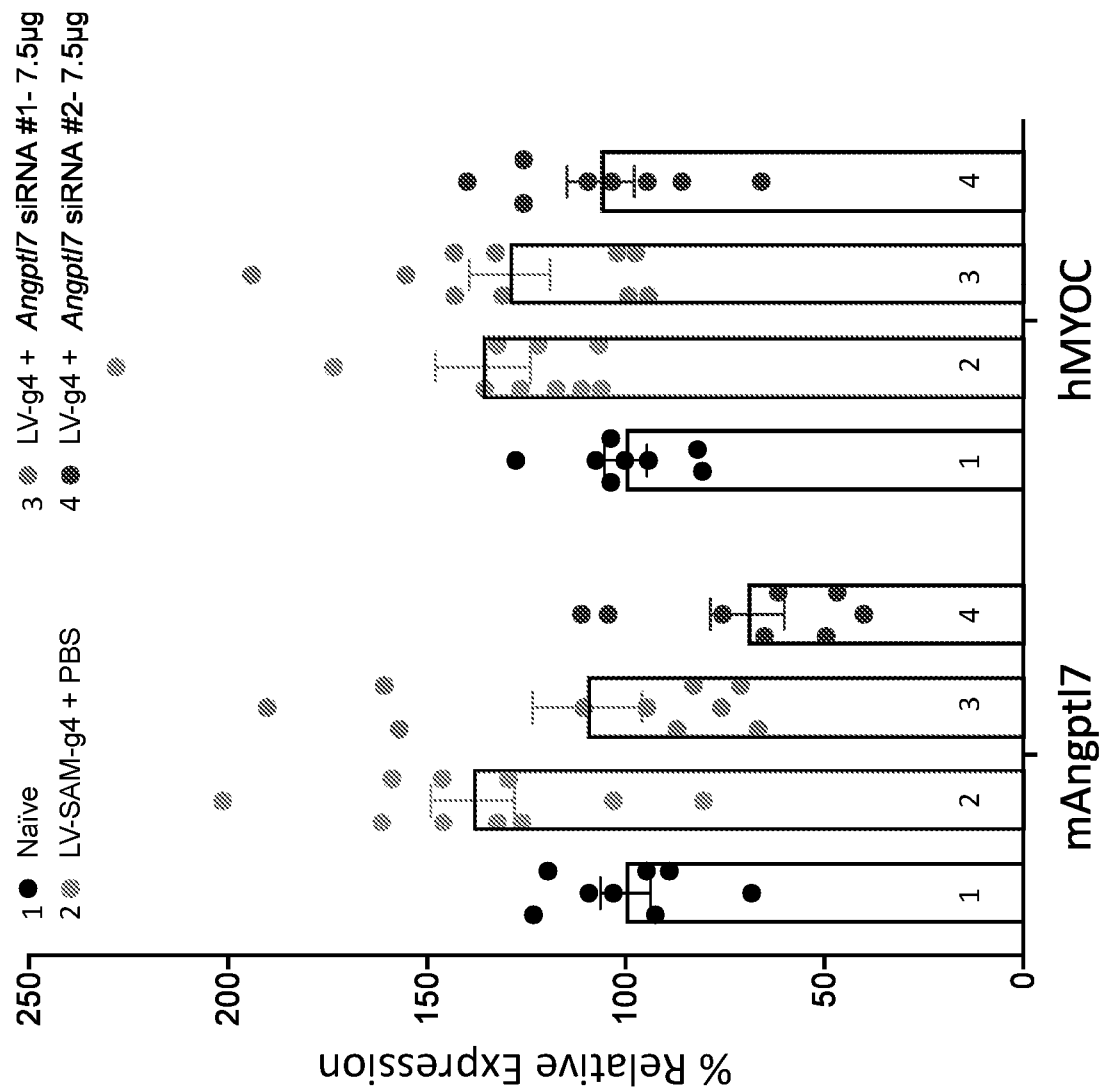


FIG. 20C