

October 26, 2016

## STATEMENT ON A NONPROPRIETARY NAME ADOPTED BY THE USAN COUNCIL

USAN (EF-03)	EMICIZUMAB
PRONUNCIATION	em" i siz' ue mab
THERAPEUTIC CLAIM	Preventing bleeding in adults and children with hemophilia A

## CHEMICAL NAMES

1. Immunoglobulin G4, anti-(human blood-coagulation factor IXa/blood-coagulation factor IX) (human-Rattus norvegicus monoclonal Q499-z121/J327-z119/L404-k heavy chain), disulfide with human-Mus musculus-Rattus norvegicus monoclonal Q499-z121/J327-z119/L404-k light chain, dimer with immunoglobulin G4, anti-(human blood-coagulation factor Xa/blood-coagulation factor X) (human-Mus musculus monoclonal Q499-z121/J327-z119/L404-k heavy chain), disulfide with human-Mus musculus-Rattus norvegicus monoclonal Q499-z121/J327-z119/L404-k light chain
2. Immunoglobulin G4-kappa, bispecific, anti-[Homo sapiens F9a (activated coagulation factor F9, activated coagulation factor IX) and anti-[Homo sapiens F10 (coagulation factor 10, coagulation factor X)], humanized monoclonal antibody; gamma4 heavy chain (1-448) [humanized VH (Homo sapiensIGHV3-23\*04 (87.80%) -(IGHD)-IGHJ4\*01 (1- 123)), IGHG4\*01 (CH1 K100>Q (202) (124-221), hinge S10>P (231) (222-233), CH2 F84.3>Y (299) (234-343), CH3 E12>K (359), R88>K (412), H115>R (438), L125>P (448) (344-448), CHS>del) (124-448)], (137-214')-disulfide with kappa light chain (1'-214') [humanized V-KAPPA (Homo sapiens IGKV1-39\*01 (80.00%) -IGKJ4\*01 [6.3.9] (1'-107') -Homo sapiens IGKC\*01, Km3 (108'-214')]; gamma4 heavy chain (1-444) [VH (Homo sapiens IGHV1- 2\*02 (75.50%) -(IGHD)-IGHJ6\*03 Q120>E (111"), T123>L (114") (1"-119")), IGHG4\*01 (CH1 A100>Q (198") (120"- 217"), hinge S10>P (227") (218"-229"), CH2 F84.3>Y (295"), (230"-339"), CH3 R88>K (408), K119>E (438), L125>P (444) (340"-444"), CHS>del) (120"-444")], (133"- 214"")-disulfide with kappa light chain (1""-214"" [humanized V-KAPPA (Homo sapiens IGKV1-39\*01 (80.00%) -IGKJ4\*01 [6.3.9] (1""-107'") -Homo sapiens IGKC\*01, Km3 (108""-214'")]; dimer (229-225":232-228")- bisdisulfide

## STRUCTURAL FORMULA

Heavy chain anti-FIXa

QVQLVESGGG	LVQPGGSLRL	SCAASGFTFS	YYDIQWVRQA	PGKGLEWVSS	50
ISPSGQSTYY	RREVKGRFTI	SRDNSKNTLY	LQMNSLRAED	TAVVYCARRT	100

GREYGGGWYF	DYWGQGLTVT	VSSASTKGPS	VFPLAPCSRS	TSESTAALGC	150
LVKDYFPEPV	TVSWNSGALT	SGVHTFPAVL	QSSGLYSLSS	VVTVPSSSLG	200
TQTYTCNVDH	KPSNTKVDKR	VESKYGPCCP	PCPAPEFLGG	PSVFLFPPKP	250
KDTLMI SRTP	EVTCTVVVDVS	QEDPEVQFNW	YVDGVEVHNA	KTKPREEQYN	300
STYRVVSVLT	VLHQDWLNGK	EYKCKVSNKG	LPSSIEKTIS	KAKGQPREPQ	350
VYTLPPSQKE	MTKNQVSLTC	LVKGFYPSDI	AVEWESNGQP	ENNYKTTTPPV	400
LDS DGSFFLY	SKLTVDKSRW	QEGNVFSCSV	MHEALHNRYT	QKSLSLSP	448

## Heavy chain anti-FX

QVQLVQSGSE	LKKPGASVKV	SCKASGYTFT	DNNMDWVRQA	PGQGLEWMDG	50
INTRSGGSIY	NEEFQDRVIM	TVDKSTDTAY	MELSSLRSED	TATYHCARRK	100
SYGYLDEWEG	EGTLVTVSSA	STKGPSVFPPL	APCSRSTSES	TAALGCLVKD	150
YFPEPVTVSW	NSGALTSQVH	TFPAVLQSSG	LYSLSSVTVV	PSSSLGTQTY	200
TCNVDPKPSN	TKVDKRVESK	YGPPCPPCPA	PEFLGGPSVF	LFPPKPKDTL	250
MISRTPEVTC	VVVDVSEQEDP	EVQFNWYVDG	VEVHNAKTKP	REEQYNSTYR	300
VVSVLTVLHQ	DWLNGKEYKC	KVSNKGLPSS	IEKTISKAKG	QPREPQVYTL	350
PPSQEEMTKN	QVSLTCLVKG	FYPSDIAVEW	ESNGQPENNY	KTTPPVLDSD	400
GSFFLYSKLT	VDKSRWQEGN	VFSCSVMHEA	LHNHYTQESL	SLSP	444

## Light chain

DIQMTQSPSS	LSASVGDRVT	ITCKASRNIE	RQLAWYQQKP	GQAPELLIYQ	50'
ASRKESGVPD	RFSGSRVYTD	FLLTISSLQP	EDIATYQCQQ	YSDPPLTFGG	100'
GTKVEIKRTV	AAPSVFIFPP	SDEQLKSGTA	SVVCLLNNFY	PREAKVQWKV	150'
DNALQSGNSQ	ESVTEQDSKD	STYLSLSTLT	LSKADYEKHK	VYACEVTHQG	200'
LSSPVTKSFN	RGEC				214'

## Disulfide bridges

Light chain, light chain	23-88	134-194		
Heavy chain anti-FX	22-96	146-202	260-320	366-424
Heavy chain anti-FIXa	22-96	150-206	264-324	370-428
Interchain bridges	L(C214)-H anti-FX(C133)	L(C214)-H anti-FIXa(C137)	H anti-FX(C225)-H anti-FIXa(C229)	H anti-FX(C228)-H anti-FIXa(C232)

## Glycosylation sites

Heavy chain anti-FIXa	300
Heavy chain anti-FX	296

MOLECULAR FORMULA  $C_{6434}H_{9940}N_{1724}O_{2047}S_{45}$

MOLECULAR WEIGHT 145.6 kDa

TRADEMARK None as yet

SPONSOR Chugai Pharma USA

CODE DESIGNATIONS ACE910

CAS REGISTRY NUMBER 1610943-06-0

UNII 7NL2E3F6K3

WHO NUMBER 10115

gbk