

STATEMENT ON A NONPROPRIETARY NAME ADOPTED BY THE USAN COUNCIL

USAN (DE-132) RAVULIZUMAB

PRONUNCIATION rav" ue liz' ue mab

THERAPEUTIC CLAIM Treatment of paroxysmal nocturnal hemoglobinuria (PNH), atypical hemolytic uremic syndrome (aHUS)

CHEMICAL NAMES

1. Immunoglobulin G2/G4, anti-(human complement C5) (human-*Mus musculus* ALXN1210 heavy chain), disulfide with human-*Mus musculus* ALXN1210 κ -chain, dimer
2. Immunoglobulin G2/G4-kappa, anti-(human complement C5 (C3 and PZP-like alpha-2-macroglobulin domain-containing protein 4)); humanized mouse monoclonal antibody: $\gamma 2/\gamma 4$ heavy chain (1-447) [human VH (*Homo sapiens* IGHV1-46*01 (82%) –(IGHD)-IGHJ4*01 (93%)) [8.8.15] (1-122) -*Homo sapiens* IGHG2*01{CH1-H} (123-232) -*Homo sapiens* IGHG4*01{CH2 E³>P(235), L⁵>V(237), G⁶>A(238); CH3 M⁸⁸>L(429), N⁹⁴>S(435), K¹⁰⁷>del(448)} (233-447)], (136-214')-disulfide with kappa light chain (1'-214') [human V-KAPPA (*Homo sapiens* IGKV1-39*01 (84%) –IGKJ1*01) [6.3.9] (1'-107') -*Homo sapiens* IGKC*01 (108'-214')]; dimer (224-224":225-225":228-228":231-231")-tetrakisdisulfide

STRUCTURAL FORMULA

Heavy chain

QVQLVQSGAE	VKKPGASVKV	SCKASGHIFS	NYWIQWVRQA	PGQGLEWMGE	50
ILPGSGHTEY	TENFKDRVTM	TRDTSTSTVY	MELSSLRSED	TAVYVCARYF	100
FGSSPNWYFD	VWGQGLLVTV	SSASTKGPSV	FPLAPCSRST	SESTAALGCL	150
VKDYFPEPVT	VSWNSGALTS	GVHTFPAVLQ	SSGLYSLSSV	VTVPSSNFGT	200
QTYTCNVDHK	PSNTKVDKTV	ERKCCVECPP	CPAPPVAGPS	VFLFPPKPKD	250
TLMISRTPEV	TCVVVDVDSQE	DPEVQFNWYV	DGVEVHNAKT	KPREEQFNST	300
YRVVSVLTVL	HQDWLNGKEY	KCKVSNKGLP	SSIEKTISKA	KGQPREPQVY	350
TLPPSQEEMT	KNQVSLTCLV	KGFYPSDIAV	EWESNGQPEN	NYKTTTPVLD	400
SDGSFFLYSR	LTVDKSRWQE	GNVFSCSVLH	EALHSHYTKQ	SLLSLGL	447

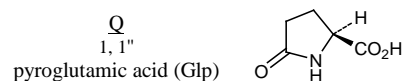
Light chain

DIQMTQSPSS	LSASVGDRVT	ITCGASENIY	GALNWFYQKP	GKAPKLLIYG	50'
ATNLADGVPS	RFGSGSGTD	FSLTISLQP	EDFATYYCQN	VLNTPPLTFGQ	100'
GTKVEIKRTV	AAPSVFIFPP	SDEQLKSGTA	SVVCLLNPFY	PREAKVQWKV	150'
DNALQSGNSQ	ESVTEQDSKD	STYLSLSTLT	LSKADYEKHK	VYACEVTHQG	200'
LSSPVTKSFN	RGEC				214'

Disulfide bridges location

22-96	22"-96"	23'-88'	23'"-88"	134'-194'	134'"-194"
136-214'	136"-214"	149-205	149'"-205"	224-224"	225-225"
228-228"	231-231"	262-322	262'"-322"	368-426	368'"-426"

Modified residues



Glycosylation sites (N)

Asn-298 Asn-298"

MOLECULAR FORMULA	C ₆₄₃₀ H ₉₈₈₈ N ₁₆₉₆ O ₂₀₂₈ S ₄₈ (unglycosylated)
MOLECULAR WEIGHT	144.9 kDa (unglycosylated)
TRADEMARK	None as yet
SPONSOR	Alexion Pharmaceuticals, Inc.
CODE DESIGNATIONS	ALXN1210
<u>CAS</u> REGISTRY NUMBER	1803171-55-2
UNII	C3VX249T6L
WHO NUMBER	10659

gbk