

Figure 4–figure supplement 2. Amino Acid Sequence of FBN-1.

**FBN-1 isoform A**

1 **MSTRSRGSCSSPWFITPLVLLALCRGSHQIAKNFVNSSTPDLRPTFVVNF**  
51 **DTSTVICQHSSDPTDLHIHNMSSLCDGKQDCFVNPAMHDEVFPYCEHKCQ**  
↓*ns67(P116L)* ↓*ns283*  
101 **STCSGKGA** **CLYDGAKPQ** **CYCDSGFSGSA** **CELQDKNE** **CLEHPC** **HMMAQC** **QN**  
**(C148Y)**  
151 **TLGSYECR** **CLPGYEGNGHECT** **DID** **ECSDKLTSR** **CPEHSK** **CINLPGTY** **CN**  
201 **CTQGFTPKGNQGSGLDK** **CADINE** **CETGAHN** **CADEI** **CENSIGSF** **KCVNKC**  
251 **SPGYELIDGKCE** **DVNE** **CGSEK** **LHKCDV** **RADC** **VNTIGGYE** **CECE** **EEGFEGDG**  
301 **KN** **CQPKS** **SCRKNSA** **ICDR** **HASCH** **IVLDI** **CDCKT** **GYTGDG** **ITCH** **DINE** **CDA**  
351 **KDTP** **CSDGGR** **CLNLD** **GGYV** **CCKNG** **QDDAT** **CI** **KDQGA** **FC** **SGGC** **GDNA** **ICSN**  
401 **AT** **CACID** **GRGD** **PHKK** **CV** **DINE** **CVEND** **SV** **CGGV** **GDRC** **VN** **LFGG** **FKCC** **QH** **G**  
451 **STEA** **ECT** **DQAF** **SSDS** **STISS** **HGAD** **FTTT** **GEQI** **IEG** **SGSI** **QTSS** **GGSI** **TVT**  
501 **RGLIP** **KDVEL** **TTSGRL** **ACTSY** **CPP** **NSECV** **GGYCE** **CV** **SGYGG** **NALV** **GC** **EDI**  
551 **DECITEI** **CN** **IEAN** **EWCV** **NLIG** **GFVCC** **NPTN** **ATHDD** **CIEIT** **KEK** **GLHV** **IGG**  
601 **NEED** **TVVAT** **RSNH** **STSD** **QLIT** **QVVQ** **QSRN** **FSTG** **QIIL** **TRGK** **VSSG** **EAVT** **Q**  
651 **TTDA** **DEFGL** **EISA** **ADLAG** **SGSG** **ITLPT** **TLEPK** **IEG** **SGK** **KASG** **GVWTE** **EDE**  
↓*tm290(deletion)*  
701 **GEDE** **DLME** **EGSG** **SWST** **TING** **TGIT** **GS** **PR** **SEGT** **IR** **VRIT** **TLG** **EDGE** **PETAT**  
751 **KPGI** **SAPDK** **TGEG** **SKTES** **DGEE** **KLTVE** **KDG** **KEA** **QSSG** **SATSS** **GKK** **SEAT**  
801 **SGSS** **SSSA** **KSGT** **GSEAS** **GSSG** **ASSSS** **SGSV** **SGES** **GSSV** **STES** **GS** **FGT** **SSS**  
851 **GSV** **GSEAT** **GSTG** **VDG** **SESG** **KPSK** **SSTEE** **KLP** **FTK** **NGEK** **SPI** **SGSD** **TTG** **KE**  
901 **SSE** **ETTS** **RKPI** **EGSD** **SLTE** **GS** **SGE** **WFET** **GSK** **GHF** **ESG** **SKV** **SVT** **SGK** **GPT**  
951 **QSGA** **EGSG** **SGPK** **VKGP** **GAPE** **IT** **DGE** **ESS** **STST** **GDK** **SGG** **KPAD** **KSD** **NKN**  
1001 **VPKT** **GDKN** **PDIT** **TDGED** **STSET** **SGGE** **QGP** **KGK** **SKG** **QPP** **GDK** **GSE** **VKK** **P** **T**  
1051 **EVD** **GPGN** **LSGT** **KGK** **SNV** **PLK** **PTDL** **PEEG** **SGIL** **TSS** **GGK** **NST** **FEH** **G** **T** **K** **L** **E**

1101 RLPPKTEDKSSETPQLGLEISAGKKPEPEDGTSKEVGLLEILWESTTPGST  
1151 TLDSDSVGLLEISGSDLTKATKKPHVEIEGSGTGDEEITATTRDVSKSTKK  
1201 PRVEVDGGDNGETSGVDGKPTTPAPTSSSAESSTSRIPPTTSEASPEGGS  
1251 GEAGVPESPDGSGESSTSAPDGVSPPTSSATAPEVPTTSASSTPDAVEESG  
1301 IPSTSKPTAEPLETTAPSTEVTSPGSGTEESTLPPTEGSGESTTSSAPT  
1351 VEPATVLPQNRNEKPEPTKDTFALPTTTTGAPQANDSSVENTKCTSSDE  
1401 CGLDALCERRTGVCRCPEPGFEGAPPKKS CVDVD ECATGDHNC HESARCQN  
1451 YVGGYACFCPTGFRKADDGSCQDIDEC TEHNSTCCGANAKCVNKP GTYSC  
1501 ECENGLFGDGYQCVPTTKKPCDSTQSSKSHCSESNMSC EVDTV DGSVECK  
1551 ECMGGYKKS GKVCEDINECVAEKAPCSLNANCVNMNGTFSCSCKQGYRGD  
1601 GFMCTDINECDERHPCHPHAECTNLEGSFKCECHSGFEGDGIKKCTNPLE  
1651 RSCEDVEKF CGRVDHVSCLSVRIYNGSLSSVCECEPGFRFEKESNSCVDI  
1701 DECEESRNNCDPASAVCVNTEGSYRCECAEGYEGEGGVCTDIDECDRGMA  
1751 GCDSMAMCINRMGSCGCKCMAGYTG DGATCIKIEEEP KSDKTA CTDEWSR  
1801 LCELEKKQCTVDEEEVPOCGACLPGHHPINGTCSLQISGLCAQKND CNK  
1851 HAECDIDIHPDSHFCSCP DGFIGDGMICDDVDECNNAGMCD DENTKCENTI  
1901 GSFNCVCLEGFKKVDEKCVVDEKKQPNREKIEIDEENSSSSNSGQEKPTT  
1951 KGDTNSTTPSTSSLASVKSTS APEERTTVSTTVSTTTPTEETTSES LIL  
2001 TAAPSKPTESTTESSEAPTPAKTSETKSLTTPQPPPTTTTAKPATTSG  
2051 KRGPPIQPPAEMFTTPAPPPPSNGGYGEETNQEEEQVTSTTTTEAPS LC  
2101 STVTC HSLATCEQSTGVCICRDGFIGDGT TACS KKSTAD C I S L P S L C A D K  
2151 AKCDNSTRSCECDAGYIGDGYVCS PHPQDCVLRDNLCSPEAVCQNRRCQC  
2201 LPGFTGDGVKCVSIHERASNC SQCDANAHC VGGTTCKCNPGYFGNGLCCV  
2251 PDPLDCVHFTGICHPNAV CNSERQCQCSSGFSGNGVSCFPQKSCRTDKS

2301 **VCAKNAICLPTGSCICR**HGFKGDPFYK**CS**SSLVSKEPANQQDLSDV**SCVT**  
 2351 **PCDSSTQLCISGECIC**KSGFRRNSTLSG**SETCAD**IDE**CAEKSHKCDRVAT**  
 2401 **CRNTFGSHVCTCPDGHVGDGITCV**PHVNQGKLSVY**CEADGMTLVLGNETA**  
 2451 **DFEGKIFVKGQAENPYCSKSFSSLLNSRKPYPVFKVVFQHC**DVQLLDN**HTM**  
 2501 **ASTVVVQKHAMFLTNKADSYDLRCQYPIGSRAVESHVNVSE**LATTSTL**TE**  
 2551 **KNSTLAPICRLSVSNDQHSSISSAMVGD**TLKLAL**EVTPSE**NGILPR**NCF**  
 2601 **AINIESGERYTTLTDDM**GCAID**ESLFPQWSAATSSKVQAVFRTFKWP**DSS**M**  
 2651 **IRFQCDCNPCVGCSTPTC**ISS**ARFR**HQTTSPVVNDEIRQ**EMVLM**SGVE  
 2701 SLAVSSIINVHDSHDMNTSDDDDVTSS**HVDATSKESK**SICV**KMAPLLIAL**  
 2751 **GSFAVCSAILLYLCS**KKPKTIDLESELRF

**Ca-binding EGF-like**

**EGF-like**

**RGD-integrin binding**

**Zona pellucida**

**Furin cleavage**

**Transmembrane**

## Signal Peptide

### FBN-1 Isoform A

Region(a.a.)	Predicted Domain		
1-27	SP	1609-1646	Ca-binding EGF-like
94-130	Ca-binding EGF-like	1652-1698	Ca-binding EGF-like
136-171	Ca-binding EGF-like	1702-1740	Ca-binding EGF-like
175-219	Ca-binding EGF-like	1741-1781	Ca-binding EGF-like
223-262	Ca-binding EGF-like	1796-1834	Ca-binding EGF-like
265-304	Ca-binding EGF-like	1843-1878	Ca-binding EGF-like
308-343	EGF-like	2098-2133	Ca-binding EGF-like
344-381	Ca-binding EGF-like	2139-2174	Ca-binding EGF-like
387-418	Ca-binding EGF-like	2179-2212	EGF-like
410-412	RGD	2220-2250	EGF-like
421-457	Ca-binding EGF-like	2255-2290	EGF-like
517-559	Ca-binding EGF-like	2294-2329	EGF-like
1394-1430	Ca-binding EGF-like	2347-2383	Ca-binding EGF-like
1433-1472	Ca-binding EGF-like	2387-2424	Ca-binding EGF-like
1476-1514	Ca-binding EGF-like	2436-2672	Zona pellucida domain
1530-1564	Ca-binding EGF-like	2674-2677	Furin cleavage site
1568-1607	Ca-binding EGF-like	2743-2765	Transmembrane domain
1598-1600	RGD		

**Figure 4–figure supplement 2.** Annotated peptide sequence for FBN-1 (a isoform). Locations of domains and genetic mutations are indicated.