

Nucleotide sequence of the human α -galactosidase A gene

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Human α -galactosidase A (α -D-galactoside galactohydrolase; EC 3.2.1.22; α -Gal A) is a lysosomal hydrolase that cleaves terminal α -galactosyl residues from glycosphingolipids and glycopeptides and is the enzyme deficient in Fabry disease (1). The gene encoding this enzyme has been assigned to the region Xq21.33-Xq22 (1). Previously, we reported the sequence of the full-length cDNA, and the genomic organization and 5' flanking sequence of this 12 kb housekeeping gene (2,3). Here, we present the complete nucleotide sequence of the gene including 1179 bp of 5' flanking and 1169 bp of 3' flanking sequence. The 12,436 bp sequence derived from α -Gal A genomic clones (3) was determined on both strands in their entirety by the method of Sanger et al. (4). The seven exons are shown in bold and a CT-rich region in intron 1 is underlined.

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1  CCGTCTGTA  GGGGACAGA  GGTCTACTT  CATTACTGG  TCTCTGGGA  AGGCCATCAG  GACTGTGGC  TAAAGTGGG  ACCAGGACTC  TTTGTGAGTT
101  AAGAATTTGT  GTATTATAT  GTGTGTTAT  CACATTTTT  AAAAAACTGT  AACGCATCA  GGTGTAGCAG  TCGCTCCGG  GTGTGAATT  ATGTGTATT
201  TTAATATTTA  TACTATATT  TATTTTTCA  AATGTTGAA  ATGGAATAG  TAGATTGTTG  TTATCAGCAG  AAAAATAAAC  ATTATACAA  TACTCTATT
301  AGTAAAGTAA  TTTATGGGC  GCGTTTGCA  AGCAGCATT  TGCTTAGAT  TGACTCTACA  GATAAAATCT  ACTTGGGGCC  TCCCTTTACA  GACAATCAGG
401  CAGTGGAGAC  TGAATGCTC  ATAGATAGA  CACGACTCA  GACCACATT  TCTCAGTATT  ACTCACTTCA  ATCTAGAGCC  GTGGTTTTCA  GCGTCTTTTC
501  GCGTACAGGT  CACCTTAGT  GTCCCCGAG  ACCGGCCAG  ACAGACAGAT  ATACAAAAAC  ACATACACAG  TCATAGSGCT  CCACCAATTC  CCCACGACCC
601  GCAGCACAG  GCGCTTCCG  GCACTGAGT  GGGGGGAGG  AGGGAGAGAG  CCGCAGGGGG  GAGGGGAAAG  CAGAGAACGA  AAGAGGCGGA  GGGCCGCGGG
701  GAACCCCGCT  CTGGTCTCA  CTATACCAC  CCGCTGGTCC  CCAGTTCCCA  CCCACACACC  AACCTCTAAC  GATACCGGGT  AATTTTCCTC  CTCTCTCCCT
801  CAACAGCGTA  TACGACGAG  TACGACGAG  ACCAGAATCA  CTTCTGCTCA  CSTAAGCGAG  TAATCAGCTG  AGCGCCTACG  TCATGTGAGA  TCTGGGTCA
901  GTGACCACT  CTCGCCATA  ACTCGGATC  ACTAATGCT  CCGCTCTCT  TCTGTATGG  AATTAGGGCC  GCTCAATCT  ARGAAAGGA  GAGGTGATT
1001  GGTAGCGGGA  ACCTCTTAG  TACTGATTA  TGGCTTACC  TCTGGGATA  ACCGTCCAG  TTCCGAGAGA  AACATATAG  TCATTTATA  ATAGCTCAT
1101  GGTGATTTGT  GCGCCCTCA  GTCTAATCT  ATAAGCCGAG  GTACCCCGGG  GAAATATAG  CTGCTCCGAG  ACTAGTACGA  TGCACTGTAG  GATCCAGTAA
1201  CTACACTGG  CTGCGCGCT  TGCGCTTCC  TCTCGGCC  TCCTTTCTG  GAACATCTCT  GGGGCTGAG  CATCTGCGA  TGATATGCA  AGAGCGCTA
1301  CTACGGGCT  CTGCGCTGG  GAGCGTCTA  TGTGCACTT  TGACTGCCG  GAGAGCCGAG  ATCTCCGAT  CAGGATACG  ATATGGGTA  CTCCCTCCC
1401  TTTGCTTTTC  CATGTGTTG  GGTGTGTTG  GGAAGTGA  GAGTCTCAAC  GGAACAGAT  GAGCCCGAG  GAGAGCTCCC  CCACCCGACT  CTCTGTCTC
1501  TTTTATTATC  CCAGCAACT  GTCCGAATC  AGGACTAGCC  CTAACCTTC  TCTGTGTAG  ATTCCTGGG  ATGGGGTCCC  GCGCCAGGCC  CCTGTTTTCT
1601  TTTCTCTCT  GCTCTCTCT  GCTCTCTCT  GCTCTCTCT  TCTCTCTCT  TCTCTCTCT  TCTCTCTCT  CCGTGGCGGG  TCTCTTTTT  TCACTGTCC
1701  TTTGACAGCA  GGGCCACCC  ATAGGCAGT  AGCCCTGCC  GGTTCATTC  AGACCCCTCT  TGTGAACCTG  TGCTTCTCT  TGCTGGGTT  TGCTGGTCA
1801  CTAACCGTAA  GAACATCTAG  GGTGGGTAG  AGGAATGGG  AACTAAGAT  CGTGGCATT  TTTCTCCTT  TGGGTCGTG  GATTTCTGG  CAGTATCTG
1901  AGGGAGTTAG  AGAGACATA  AGTTCGCTA  GATCTCTCC  ACCTGCGCCA  TAGCCCTGG  ATCAGGCTG  AAGGTTGACA  TGGAGAAC  TTATACATT
2001  ACACCTTTG  GTAGGGTTG  AGCTGGATT  AGATAGTAT  TGAACATAT  TGACCTCAC  AACTCTTAT  TGTAATTTG  GATTACACC  TTTTAATCT
2101  AGGGAGCTCA  CAATAAAAT  GTCAAAAATA  GTTCTTACT  CACACAGGTG  AGTTTTCAAG  GAGATAACCT  ATTTAAAGTA  CATAGACCA  GCGTGTACA
2201  TTTAAGCTGG  CTTACAGAG  AAATGTTCA  TGGGAAATG  AATGTAATC  TACAATATG  TGTAAATCT  TGATTTTTT  TGGAGAGAG  ATATTTACT
2301  TTTCTCAAT  TCTCAAGGG  CTTCTGAT  TAAAAGGT  TAGGAATCA  TGATAGATG  TGGTAAGG  AAGTAAGC  AGTACATTC  TGTTGCCAT
2401  AGTATTCTCT  ATGATATCT  TTTAGATA  AGTACAGAT  ATGTCAGAC  ATCCAGAG  AATGGCTT  TGTATGTT  ATGTAGACT  CTGTGACT
2501  ATGTGTGCA  ATTTGCCA  TCAAGATT  TTTTATTG  GTATTTGAT  CTGATATA  AACTATGA  TGATCTCC  AAAATATG  GATAAGAG
2601  AGCAAAATG  AAATAAAT  TTTCCCCAC  CTTCCACCA  CCGAGAATA  ATCACTGTT  AATGTTAT  ATCACTCT  ACAATGTT  TCTATATA
2701  TGAACATCA  GATTTCTTA  TTTCAATT  TTTCAATA  AATGATCAT  GTTTATGTA  TGTTTGGTA  ATGGCAAGC  CTTGGCAC  AGTCTGGCT
2801  CAATCTCTG  CTAATGTTA  CTTACCTG  GAAATGGG  TAAATTAC  TTTTTTTTT  TTTAGACGG  TCTCGCTG  TCGCCAGCG  TGCCCGGTT
2901  TGGAGTAGC  TGGCAGACT  TGGCTTACT  GCAATCCGG  CTCTGGGT  CAGCCATT  TCTGTGCTA  GCTCCCGAG  TAGTGGGAC  TACAGCCGC
3001  TGCCACCAG  CTTGGCTTT  TTTTTTTTT  TAGTACAGC  GGGGTTTAC  CATGTGTACC  AGGTTGGT  CACTCTCTG  CTCTCTGAT  ACCCTGGAT
3101  TGCCGCCCT  CAGCTCCCA  AAGTGGCT  GTAGCCACC  GTTCCAGCC  TTACTTTTT  TTTTGAAGG  GGGTCTACT  CTGTACCA  GGTGTGAGT
3201  CAGTGGCGG  ACTCTGCTC  AGTCTGCT  CACCTCCCG  GGTTTAAGC  GTTCTCTGT  CGTAGTCT  GTGATCTCC  TGGCTAGCT  GGATTACAG  CACACACCA
3301  CGCCAGCTA  ATTTGTAT  TTTCACTA  CAGGGTTTC  ACCATGTGC  CCAAGCTGT  CTGCACTCT  TGGCTCAAG  TGATCTGCC  GCCTTGCCT
3401  CCGAGAGTC  TGGGATACA  GGTGTAGCC  ACCGCCCG  GCCTTTTT  TCTTTTTG  TCTATCAT  GTCATCTAG  CATTGCAAA  CAGTGTGCT
3501  TGGTTTTAG  ATTTATGTA  TCAAAACAT  CAGTTTTCC  TTTCTGATT  CTGACTTGG  GGTCACTG  AGAAGTCT  TTTCTACT  AAGATAATC
3601  AGTATAATG  TTTCTACT  TTTTITTT  GAAATTTAA  AATATTTAA  TCTTTAGTC  ATCTGAECT  GTTCTCTAT  CAGAATGCC  ACATTTAAT
3701  AATAATAAT  CCAATGAT  CAGATGGCT  GAAAGACT  TTTGCAACT  TGTTTAAT  CCAATTAAT  GTGATCTT  ATTCTAAT  TAATAGTTC
3801  ACACTAGCT  TTTTCTATT  TTTTCTTT  TTTTGGCTG  GAGTGTGCT  CTNTTGGCC  AGCTGGGAT  ACAATGTC  GATCTCGGT
3901  CACCCGAMC  TCGCCCTCC  AGTTCACG  AATCTCCCT  CCTCATCT  CCGGTAGCT  GGAATACAG  CCAATGTC  CTATTTGTA
4001  TTTTATAGT  AGATGGGTT  TCTCATGTT  GTTACAGCT  GTCTCAACT  CCGCCCTCA  GCTGATCT  CTGCTCGC  TCCCAAAAT  GCTGTATTA
4101  CAGCCCTGC  CCACAGCC  CAGCCTCAT  CTTTATGA  ATGATCAT  ATGTAATCT  TTAGGTGAC  TTTTGTAT  GTTGGCCA  GTTCTTAA
4201  AGCCCTTCT  GGAAGTGG  GAGTGGCTA  CAGCTTAT  CCGACATT  TGGGAGTCT  AGGCAAGT  ATCACTGAG  CCGAGGAT  CAGACTAGC
4301  CTAGCCAAA  TGCAAAACC  TCTCTACT  AAGATACAA  AATATAGCC  GATGCGATG  CACATGCTT  TATCTCAG  TACTGGGAG  GCTAGGTAG
4401  AAGAATGCT  TGAACCGGG  AGGCAAGT  TCGACTGAC  AAGATGGCC  CACTGCATC  CAGCCTGCT  CAGCAGGGA  GACTCTCAT  CAATAAAAA
4501  AAAAAAATA  AAGATAAAA  GGAACTATA  GTACTTGG  GCTTTGTA  AGGATTTGT  AANATACAA  GATTTAGGA  GGAAATTA  CTATTTTTA
4601  ATATTGAGTA  TGCTATCCA  AGAGCAAA  AATATTCT  CATTATCA  AATCAATT  GAGCATATA  TTTTAACT  ATGGCCCTG  CAGCTATCT
4701  AAAATTAICT  CTAGCATAT  TAGTGTGTC  ATTTGTTCT  GTGAATGGG  TCTTTTCT  CAATAGGAT  GATTTGAT  ACTTGTCT  TAGTAAACT
4801  TGTAGTATT  TGACTTACT  GACTGTCT  TTAGATCTA  ATACTCTTT  CAATTTACT  ATATATTT  CATTTACT  GATTTCTA  TTTTGGGG  TTTTATGGC
4901  GGGAAATTA  ACCGGATAG  AGAGACAAA  GAAATCTGG  AAAAAAAT  CATTTTACT  TACATTGCT  CATTTACTA  CCACACTAT  ACTGGGTTG
5001  AAAAAATGT  GAAATCCAA  GGTGCTAAT  AAATGGGAG  TACTTAAGT  TTCAATTA  ATATTGAT  GATTTAGGA  ATTTCTTT  ACTGGGTTG
5101  CTTCTCAGG  AAGATGGGA  GCTCACGTC  TCGAAGGCT  GAGAGAGG  AGCTGAGG  TACTGATCA  TACTGATCA  ATTTCTTT  CCGGAGGCT
5201  ATCTCAGG  CAGCTCTG  TCAAGACT  TGTTAAGA  CAGCCCTAT  GGGCCGGCT  GGTGGCTCA  GCTGTAICT  CAACACTT  GGAGCCGAG
5301  GAGCTAGTA  AGCACTCT  TCAAGACT  GTTTAAGA  CAGCCCTAT  GGGCCGGCT  GGTGGCTCA  GCTGTAICT  CAACACTT  GGAGCCGAG
5401  CGGGTGGTA  CACTGAGT  CAAAGTCT  AGGCAACT  GGCACAGT  GCGCCAGCT  AACCTTATA  AAGTACAA  AATTAGTC  TGAGTGTG  GGCATGGTG
5501  TGAACAGCT  TAACCCAGC  TACTTGGAG  GCTGAGCCG  GAGAATGCT  TGAACCCAG  AGGTGAGT  TCACTGAGC  TGAGACTAG  CCACTGCAT
5601  CTAGCCTGG  CAACAAAAG  GCGTCCAT  CTTCAAAAA  AAAACAAAG  AGCCCTCAT  GAGCCTCAT  CAGCATAG  AATAGATA  GCTGCTGTT
5701  ATCTGAACAT  TGAATGTAA  GCTTATCAG  GTGGACTTT  CATTCTACA  CGAGACAAT  TTTTTTTT  TTTTTTTT  AGATGGAGT  TCATCTCTC
5801  TCCAGCGAG  GAGGCGAGT  GGCTCAGCT  GCTCAAAAA  AAAACAAAG  AGCCCTCAT  GAGCCTCAT  CAGCATAG  AATAGATA  GCTGCTGTT
5901  ACAGGCCACC  GCCACATGC  CAAATTAAT  TTTTATATT  TAGTAGAGA  GGGGTTTCA  CCAATTTAG  CAAAGTGT  TGATCTCT  GACCTGTGA
6001  TCCGCCCCC  AAGTGTGTT  GATTACAGC  ATGAGCCACC  GCGCCTAGC  TCAAAATGTT  TTGTAATAG  TCTGTAGCC  CATCTGGAG
    
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Nucleic Acids Research

6101	TTCTCTTTTT	GCTAAAACCA	CTGAAGTCTC	TAGGAGAAA	AAGGAAGTGG	GTTCTTGACA	TATGTGTGCA	TGTATTCCCA	TATAACCTTT	AGGAAGCTAT
6201	TGCAATGGTA	CTATAAAGTA	GAATTTTAGA	AGATAGAAGG	AAAATATTTT	GGAGATCATT	GAAGCAAAAT	GGAGTCCAAC	AGGTATTTAA	GATGATGAAG
6301	ACAGATTTTT	TTTTTTAGC	GAGTCTCGCT	CTGTGCCCA	GCGTGGAGTG	CAGTGGCACA	ATTCAGACCT	ACTGCCAACC	TCCACTCTTT	GGGTCAAGT
6401	GATTCCTAGC	CTCCAGCTCG	CCAAAGTAGCT	GGGACTACAG	GGCCACACCA	CCACGCCCGG	CTAATTTTTG	TATTTTTAGT	AGAGCAACGG	TTTCACCATA
6501	TTGCCACCGG	TGGTCTGCAA	CTCTGACCT	TGTAATCCG	CCACCTTGGC	CTCCCAAAGT	GCTGGGATAT	CAGGCAATCC	CCACCCACGG	CGGCCGATGA
6601	AGACAGATTT	TATTCAGTAC	TACCACAGTA	GAGGAAAGAG	CCAAGTTCAA	TTCCAAATAC	AACAAGAGCA	GGTGAGATTT	TATAGCCAA	GAGCAGATTT
6701	AGGGGGTCAG	TGGATGGAA	ATTTAAGAA	ACATCAAGGG	TAGGGAGCTT	CTTGCTAAG	CTTCATGTAC	TAAACAAGA	AGGGTGGGG	ATGAGGGAAA
6801	TTGATCGAGT	ATCAATGGT	GCAGTATTA	CTTAGCAGGA	TTCTTGCTAA	GAGGTCTTGC	TAGGACAGAC	ATAGGAAGCC	AAAGTGGAG	TCTAGTTCGA
6901	AAGAAGGCTC	ATCAGAGAT	CTTAAGTAA	GTAAGAGTCT	GAAAGAGTCT	TGTCAGGTA	GAATATATC	TTCCTCAAA	AGGTATTTAA	CAGATTCGAA
7001	TCAGAGAGAT	TAGCATGGCT	GCTAGCTTTT	TCCTCAGTTC	TGGGCTATAG	CTCACATGCC	TAGTTGTAA	TAGCTCAGCA	GACTCGGGG	ATTTATCTTT
7101	TGTCCTTCGA	CAAACTCAT	TGATGATTTT	TGGGGCTTGG	TGGGGAAAG	CCCCCAATAC	CTGTGTAGT	ACCTTTGTCT	CTTCCCGAC	CTCGGATGG
7201	TTCTCTCTTT	CTGTCTACTC	AGGATCTGTC	TTCTACAAAT	GTGACTCTTT	CTCTCCCTCT	CTTTTCAAGT	CTACAGCAAA	CTCTTCAAG	TAGGATTTA
7301	TGCGAGTCTT	GGAAAPAAA	CTCGCGCAGG	CTTCCCTGGG	AGTTTTGGAT	ACTAAGACAT	TGATGCCCA	ACCTTTGCTG	ACTCGGGAT	AGATTCGTA
7401	AAATATTTG	GTGTTACTG	TGACAGTCT	GAAATTTGG	CAGATGCTAA	TGTTTTATTC	CAGAGATTTA	GCCACAAGG	AAAGACTTT	GAGGCCATGG
7501	TAGCTGAGCC	AAAGAACCNA	TCCTCAGAAT	TTAAATATCC	CTGTCCAGAT	ACTGGAAATA	ATTATTTCTC	ATGTGCCAGA	GCTCCCATCT	CTTCTCTTTC
7601	AGTTCTACCA	TTAATTAATT	AAATCCATGG	ATACCTAAC	ATAGCTAATA	TTGTGCACAT	ATAATTCAG	AGGGCTCAA	GAGTTAATTA	GAATTAATTA
7701	GTAATTTGTA	CTCTCTTATA	CATCATATTG	GGGAGTCCAG	GTGTCATCT	GGTCACAGAG	AAAAGAGACT	CTTCACTTCT	GCCTTCTCTC	AAATATACAA
7801	CCATCTCTGC	ACTACTTTCT	CCAGAACAAT	CCAGAGTCT	GGGAGGTACT	CTACACAAAT	TAAACAGAG	CGAATGCCT	GTCCCTGAT	CTAGTTTAAA
7901	CATAGACTTT	CCAGGTAGCC	TCTTCTTAAA	ATATACAGCC	CCAGCTGGCC	ATGATGGCTC	ATGGCTGTA	TCCTAGCACT	TGGGAGGCT	GAGGCCGGGT
8001	GATAGACTTA	GCTCAGGAGT	TGGAGACCAC	CCCTGCCAAC	ATGGTGAJAA	CCCATCTCA	GAAAAATATC	AAAATTTAG	TGACTTTGT	GGCACATGCC
8101	TGTAATCCCA	GCTACTTGG	AACTGTAGAC	AGAAGAGTCA	CTTGAACCTG	GGAAACAGAG	TGTCGATG	GCCAGATGT	CACCATGCA	CTCCACCCTG
8201	GATGACAGAC	TGAACCCCT	CTCAAAGAT	TAAAATAAAA	TAAAATAAAA	TAACTATATA	TATAGCCCCA	GCTGGAAAT	CAITTTCTTC	CTTTATTTTA
8301	CCCAATGTTT	TCTCATACAG	GTTPAABCA	CAATGCTTGG	GCCTCGAATA	GGACTGCGAG	AAGCAATTTG	TACTCCCTGG	AGTGGCCCT	TTATTCGCG
8401	CCCTCTTCGA	AGGTGAGATA	GGAGGCCAG	AACTCAATAG	AACTGTACTG	ATAGATAGAA	CTTGAACAA	AAGGAACCA	AGCTTAACT	CAATAGAGCA
8501	CCCTCTCTTT	CTGCTCTCTT	CTTCCCTTTT	ATTTCCACT	TCATGTCTG	TCTATCAACA	GTCCCTCCAC	CAGTATCTCT	AAAATATCTC	CTGATCAGC
8601	CCACTTCTAC	TACATGACC	CTGGCTTTC	AAGTACTACT	CGGCTCTCAA	CCGACTGCT	CCGACTGCT	ACTCTCTCT	CTTCACTCT	GTCTCAACCC
8701	CCATCTATT	TCCAGCAGC	ACTAGACTG	TCAATATAA	ATGTAAATAT	CAGTTTTTTT	TTTTAAGAAA	AAAACCTTGA	GACTTAAACG	AGTTATAAAA
8801	AAATATAAAT	TCACTACAG	TCTCCGTTT	AAAACCTTAA	ACTCGCTTCC	AAATGCACT	GGAAATGAAC	CAAACTGCAG	TGATCCAGC	CTTCCGCTGC
8901	TCCCAAAAGT	CAAGGGCTG	ATGGCTCTTT	CCCTGGCTAC	ACTGGTTTTT	TTTCTGCTCC	TCAACACTGC	AAGCTATATG	CTGCCCCAGG	GGCTTTACAC
9001	TGCTTTTTTT	TCTGCCAGA	ACAGTCTTC	CCCAAGATTT	TTTTAAGGGC	CGGGCTCTCT	AACTATGAG	CGGACAGCA	AAGCCGACAT	ATCCAGACAG
9101	TTCTCTCTTA	ACTACTTTAA	AATGGCCCTC	TGTCATTTA	TTCTTCTATCA	CATTAACCTG	TTTTAATTTT	TTCTAGAGC	TCCACATAT	TTGGAAGTAT
9201	TTGTTGACTT	TTTACCAAT	CTCCCACTA	GAGTGTAAAT	TTCATGAGGG	CAGGACCTT	GTTCGATCTT	GACTGTATCT	CTCCGATATG	CTTAAAGTGT
9301	AAATAGTTAT	TTATGGTAT	AAATCCCTAT	ATTCCTCAT	TATCTTGCA	AAATAGTCTT	TTTTCTCAAC	ATCTTAAAC	TGATATCCCA	CTCCGCTATC
9401	TAGCAAAAT	TTTTTTGCA	CAGAGCTCA	CTGTCAACCA	GGCTAGAGTG	CAGTGGCCGC	ATCTGGCTC	ACTGCAACT	CCGCCTCCG	GGTTTAAAGG
9501	TTCTCTTGC	CTAGCCCTG	CTAGCTCTG	GATTAAGGC	GTGGCTACC	ACATTTGGCT	TTTTTTTTGA	TTTTTATGA	AGATGGTTTC	ACCATGTTGG
9601	CCAGCCCTGT	CTGAACTCT	TGACTCTAGA	TGATCCACT	CCCTCGCCCT	CCCAAGAGTG	TGGGATTTA	GGTTATGAGC	ACCGTGGCCA	CCCTCTACAA
9701	ACTTTTTTAT	CCATTAAGCA	ACTATATGCT	GGATTTAAG	TTTTCTTAA	ACTTGAATGA	CTTGAACAA	TTTTTCGAG	TTTTTACCT	ATTATAAGG
9801	TTTTATGACT	TTTTAGGAG	TTTTAGGAT	TTTTAGGAT	TTTTAGGAT	TTTTAGGAT	TTTTAGGAT	TTTTAGGAT	TTTTAGGAT	TTTTAGGAT
10001	TTAGCAGCT	CTTAGGAGA	CTTCACTCT	GTAACCTCA	CAGAAGGCTA	CAGCTGCCTC	CTTAAAGTGT	TTTTCACTCT	ACAAGATGCT	TAGTCAAGG
10101	TAAACAGAA	ACTCATATCT	CTTTTACAG	CCCAATATA	CAGAARCCG	ACAGTACTCG	ATACCACTCG	GAAATTTGAC	TGACATTTAT	GATTCCTGGA
10201	AACTATAAAA	GACTACTCTG	CTGGACACT	CTTTAABCA	GGAGAGAAAT	GGTGTATGTT	GGTGTATGTT	GGTGTATGTT	GGTGTATGTT	GGTGTATGTT
10301	TTAGGCCCTC	CTTGTTCAG	ACCTCGGGT	AGGCTGTFTT	CCATTTTTGA	CATTCAGGTT	AAATACAGTT	AAAGTTCCTG	GAGGAGGCT	TTATGTGAGA
10401	GACTATTAG	CAGGATGTG	TGAAAGGCT	TTTCTCATG	TGGTCTCAT	AGGTAACTTT	AAAGATGTTT	CTCTCTCTCT	TTTTGAATTT	ATTTCATTT
10501	TTTTCTCAGT	TAGTATGTC	GAACTTTGGC	CTCAGCTGGA	ATCCAGAGT	AACTCAGAG	GGCTCTGGG	CTATCAGGG	TGCTCTTTA	TTGATCTCA
10601	AAAGCTCCG	ACAGATCAG	AAAGCTCCG	AAAGCTCCG	AAAGCTCCG	AAAGCTCCG	AAAGCTCCG	AAAGCTCCG	AAAGCTCCG	AAAGCTCCG
10701	TAGACAGTA	AATAAGAGTA	TATATTTTAA	GATGGCTTTA	TATACCAAT	ACCAACTTGT	TCTTGGGCT	AAATCTTTT	TTTTCCCTGT	CTCTTGATGT
10801	TACTATCAGT	AATAAGAGT	CTTGCAGAA	ACATTACTT	ATTTCCAAA	TAATGTCTACA	GGATCTTTT	TAATTTCTT	ACAAGTGGCT	GATGTTCTGT
10901	ACATTAAGAA	TGAATGCCA	ACTAACAGGG	CCACTATCA	CTAGTTGCTA	AGCAACCA	CTTCTTGGT	TTTTAGGGA	GAAACTTTG	AAAGTGGGA
11001	ACGAGCTCTC	TCGAGCTCTC	TCGAGCTCTC	TCGAGCTCTC	TCGAGCTCTC	TCGAGCTCTC	TCGAGCTCTC	TCGAGCTCTC	TCGAGCTCTC	TCGAGCTCTC
11101	GGAGTGGCT	GTAACTCTG	CTCTCTCTC	CTCTCTCTC	CTCTCTCTC	CTCTCTCTC	CTCTCTCTC	CTCTCTCTC	CTCTCTCTC	CTCTCTCTC
11201	AGCCGAGGG	CTCTCTCTG	CTCTCTCTG	CTCTCTCTG	CTCTCTCTG	CTCTCTCTG	CTCTCTCTG	CTCTCTCTG	CTCTCTCTG	CTCTCTCTG
11301	CTCCACTTT	TTCTCTCTT	TTCTCTCTT	TTCTCTCTT	TTCTCTCTT	TTCTCTCTT	TTCTCTCTT	TTCTCTCTT	TTCTCTCTT	TTCTCTCTT
11401	TGCGGACTTG	GAGACCCGG	TGCAACACT	GGTGAACCC	CATTTCTAAT	AAAAATATA	AAATAGCCA	GGTGTGGTG	CGCAGCTGTG	GTCCACGTA
11501	CTCTGGGGG	TAGGCGATGA	TAGGCGATGA	TAGGCGATGA	TAGGCGATGA	TAGGCGATGA	TAGGCGATGA	TAGGCGATGA	TAGGCGATGA	TAGGCGATGA
11601	ATCTCAAAA	AAAAAAAACA	GGCAGGACA	GTGGCTCAT	CCCTGAACTC	CAGCCTCTT	GGAGCTGAG	CGAGCCAGT	ACTCTTAGT	TAGGATTTCA
11701	AGACCAGGCT	GGCTAAATA	GGCAGGACT	GTCTCTACTA	AAAAACAAA	AATTAGCCAG	TTAGGTGGG	GAGCTCTCTG	ACCCCTTAGT	ACTCAGGAGA
11801	CTGAGGCGG	AGAATCACT	GAACCCGGGA	AGTGGGGGG	TGCGACTGAC	CAAGTACAG	CCACTGACTT	CCAGCTGGG	CAACAGACA	AGACTCATC
11901	TCAAAAATAA	AAAGTTCAAT	TCCTTGAATA	AAATTTTCCG	AAATTTTAA	TTTAGGAATA	AAACTATTA	ACCCTGATTT	ACTACTCAG	ATACCACCC
12001	CCCTGTGTA	GATCTCTCT	CAATTAACAA	AAATGTGAG	ATATTTAAT	ACCAGAGCT	AAACATCAT	AAAGACTGAA	TGATATAAG	AGGATGATA
12101	GGCAGGAC	GGTGTCTAC	GGCTGTAA	CCAACACTT	GGGAGGCCAA	GTGGGGCCAA	GTGGGGCCAA	GTGGGGCCAA	GAGCATCTG	GGCAACATGG
12201	TGAAGACCC	TCTCTACTA	AAATACAAA	ATTAGCCAG	CAGTGGGAC	GCACCTGTAA	TCCCAGCTAC	TCCCAGCTAC	GAGGCAGAC	AAATCCTTGA
12301	ACCTGGGAG	CAGAGGCTG	AAATGACTG	GTTTGTACA	ATGTGACTC	AGCCTAGTA	ACAGGACAA	CTCCAGCTCT	AAAAAGAAA	AAAAAAGAA
12401	TGTAATAATT	GGAACTGTA	AGAGGCAATT	TAAAGA						

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References

- Desnick, R.J. and Bishop, D.F. in Scriver, C.R., Beaudet, A.L., Sly, W.S. and Valle, D. (eds.) *The Metabolic Basis of Inherited Disease*, 6th edn., McGraw Hill, New York, in press.
- Bishop, D.F., Calhoun, D.H., Bernstein, H.S., Hantzopoulos, P., Quinn, M. and Desnick, R.J. (1986) *Proc. Natl. Acad. Sci. USA*, **83**, 4859-4863.
- Bishop, D.F., Kornreich, R. and Desnick, R.J. (1988) *Proc. Natl. Acad. Sci. USA*, **85**, 3903-3907.
- Sanger, F., Nicklen, S. and Coulson, A.R. (1977) *Proc. Natl. Acad. Sci. USA*, **74**, 5463-5467.