### Names for XG (ISBT 012) Blood Group Alleles

### Intro

General description: The XG blood group system consists of 2 antigens carried on 2 single-pass

glycoproteins, encoded by the genes XG and CD99, located in

pseudoautosomal region 1 (PAR1) of the sex chromosomes. The Xg glycoprotein is encoded by the X chromosome only, while the CD99

glycoprotein is encoded by both X and Y chromosomes. On Y, only exons 1-3 of XG exist. Both genes encode single pass glycoproteins of 180 and 185

amino acids, respectively.

Gene name: XG

Number of exons: 10

Initiation codon: Within exon 1
Stop codon: Within exon 10

Entrez Gene ID: 7499

LRG: LRG 805

LRG sequence: NG 011627.1 (genomic)

NM 175569.2 (transcript)

Reference allele: XG\*01 (shaded)

Reference allele  $\chi_g^a$ 

*XG\*01* encodes:

Antithetical antigens: n.a.

Additional information

Gene name: *CD99* 

Number of exons: 10

Initiation codon: Within exon 1
Stop codon: Within exon 10

Entrez Gene ID: 4267

LRG: LRG\_1023

LRG sequence: NG\_009174.1 (genomic)

NM\_002414.3 (transcript)

Reference allele: CD99\*01 (shaded)

Reference allele

*CD99\*01* encodes:

CD99

Antithetical antigens: n.a.

Additional information

| Phenotype       | Allele name | Nucleotide change                       | Exon<br>Intron | Predicted amino acid change | (Reference No.) PMID             | Accession number | rs number  |  |
|-----------------|-------------|---|----------------|-----------------------------|----------------------------------|------------------|------------|--|
| Xg <sup>a</sup> | XG*01       |   |                |                             |                                  |                  |            |  |
| Null phenotypes |             |   |                |                             |                                  |                  |            |  |
| Xg(a-)          |             | NC_000023.11:<br>g.2748343G>C           | 5' UTR         | p.0                         | PMID: 29748255<br>PMID: 30061310 | n.a.             | rs311103   |  |
| Xg(a-)          |             | NC_000023.11:<br>2,776,388_2,890783del  | i3             | p.0                         | PMID: 30938838                   | n.a.             | esv2662319 |  |
| Xg(a-)          |             | NC_000023.11:<br>2,776,662_2,891,056del | i3             | p.0                         |                                  | n.a.             | n.a.       |  |
| CD99            | CD99*01     |   |                |                             |                                  |                  |            |  |
| Null phenotypes |             |   |                |                             |                                  |                  |            |  |
| CD99-           |             | c.(100 + 1_101-1)_(361 +<br>1_362-1)del | 3–7            | p.0                         | (1), Abstract                    | n.a.             | n.a.       |  |
| CD99-           |             | c.(148 + 1_149-1)_(475 +<br>1_476-1)del | 4–8            | p.0                         | (1) Abstract                     | n.a.             | n.a.       |  |
| CD99-           |             | c.(67 + 1_68-1)_(475 +<br>1_476-1)del   | 2–8            | p.0                         | (1) Abstract                     | n.a.             | n.a.       |  |

#### References

- PMID 29748255 Möller M, Lee YQ, Vidovic K, Kjellström S, Björkman L, Storry JR, Olsson ML. Disruption of a GATA1-binding motif upstream of XG/PBDX abolishes Xga expression and resolves the Xg blood group system. Blood. 2018 Jul 19;132(3):334-338.
- PMID 30061310 Yeh CC, Chang CJ, Twu YC, Chu CC, Liu BS, Huang JT, Hung ST, Chan YS, Tsai YJ, Lin SW, Lin M, Yu LC. The molecular genetic background leading to the formation of the human erythroid-specific Xg<sup>a</sup>/CD99 blood groups. Blood Adv. 2018 Aug 14;2(15):1854-1864. doi: 10.1182/bloodadvances.2018018879. PMID: 30061310; PMCID: PMC6093725.
- PMID 30938838 Lee YQ, Storry JR, Karamatic Crew V, Halverson GR, Thornton N, Olsson ML. A large deletion spanning XG and GYG2 constitutes a genetic basis of the Xgnull phenotype, underlying anti-Xg<sup>a</sup> production. Transfusion. 2019 May;59(5):1843-1849.
- Abstract (1) Thornton NM, Karamatic Crew V, Muniz-Diaz E, Garcia- Arroba J, Nogues N, Lee E, Jones C, Schistal E, Jungbauer C, Allhoff W, Bullock T, Marais I, Daniels G. Four examples of anti-CD99 and discovery of the molecular bases of the rare CD99—phenotype. Vox Sang 2015;109(Suppl 1):50-1.
- PMID 36102098 Watanabe-Okochi N, Uchikawa M, Tsuneyama H, Ogasawara K, Shiraishi R, Masuno A, Onodera T, Tsuno NH, Muroi K. Genetic background of anti-Xga producers in Japanese blood donors. Vox Sang. 2022 Sep 14. doi: 10.1111/vox.13342. Online ahead of print.

# Track of changes

|     | Version            |                  | from<br>v3.0 30-OCT-2020 | to<br>v3.1 30-SEP-2022                   |
|-----|--------------------|------------------|--------------------------|--|
| 1 2 | Author<br>Reviewer | created reviewed | Jill Storry              | Jill Storry<br>C. Gassner                |
| 3   | Versioning         | LRG ID corrected |                          | LRG_802 corrected to LRG_805             |
| 4   | Allele Table       | corrected        |                          | XG*01N.02 nucleotide positions corrected |
| 5   | New alleles        | created          |                          | XG*01N.03                                |
| 6   | References         | renumbered       |                          | renumbered Abstract (4) to (1)           |
| 7   | References         | added            |                          | added PMID 36102098                      |
| 8   | End of changes     |                  | v3.0 30-OCT-2020         | v3.1 30-SEP-2022                         |

# Track of changes

|        | Version             |                     | from<br>v2.0 160630 | to<br>v3.0 30-OCT-2020   |
|--------|---------------------|---------------------|---------------------|--|
| 1<br>2 | Author<br>Reviewer  | created<br>reviewed | Geoff Daniels       | Jill Storry  |
| 3 4    | General<br>Intro    | Text changed        |                     | Changed and expanded: The XG blood group system consists of 2 antigens carried on 2 single-pass glycoproteins, encoded by the genes XG and CD99, located in pseudoautosomal region 1 (PAR1) of the sex chromosomes. The Xg glycoprotein is encoded by the X chromosome only, while the CD99 glycoprotein is encoded by both X and Y chromosomes. On Y, only exons 1-3 of XG exist. Both genes encode single pass glycoproteins of 180 and 185 amino acids, respectively. |
| 5      | Intro               | LRG ID line added   |                     | LRG_802  |
| 6      | Allele Table        |                     |                     | Inserted   |
| 7      | New alleles         |                     |                     | Inserted   |
| 8      | References          |                     |                     | References found for all alleles and collated  |
| 9      | End of changes v2.0 |                     | v2.0 160630         | v3.0 30-OCT-2020   |