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## ISBT Working Party on Terminology for Red Cell Surface Antigens

### S o Paulo Report

The responsibility of the ISBT Working Party on Terminology for Red Cell Surface Antigens is to maintain and monitor a machine- and eye-readable terminology in keeping with the genetic basis of blood groups. The Working Party published a report in *Vox Sanguinis* in 1990 outlining the recommended terminology and providing other relevant information [1]. A supplement to this report was published in 1991 following the Los Angeles meeting of the Working Party [2]. On 11 October, 1992, the Working Party met in S o Paulo, Brazil, during the 22nd ISBT Congress. In the light of new findings, a number of additions or changes to the 1990 classifications were made and are reported here. Tables 1-5 include most modifications agreed since 1990, so that the present report can be used in conjunction with the 1990 report alone. It is envisaged that a full revision of the ISBT terminology will be published following the Amsterdam meeting in 1994.

#### Existing Blood Group Systems (table 1)

**002, The MNS System.** Five new low incidence antigens have been added since the 1991 report [2]. MNS33 (TSEN) and MNS34 (MINY) represent abnormal amino acid sequences on glycophorin (GP) hybrid molecules, GP(A-B), resulting from a GPA<sup>58</sup> to GPB<sup>27</sup> junction; MNS33 is expressed only when the GPB portion of the molecule expresses S [3, 4]. MNS35 (MUT) represents an antigen

**Table 1.** Additions to existing blood group systems since 1990 [1]

|          |          |                     |                     |
|----------|----------|---------------------|---------------------|
| 002 MNS  | 004 RH   | 005 LU              | 014 DO              |
| 032 DANE | 049 STEM | 018 Au <sup>a</sup> | 003 Gy <sup>a</sup> |
| 033 TSEN |          | 019 Au <sup>b</sup> | 004 Hy              |
| 034 MINY |          | 020 Lu20            | 005 Jo <sup>a</sup> |
| 035 MUT  |          |                     |                     |
| 036 SAT  |          |                     |                     |
| 037 ERIK |          |                     |                     |

defined by antibodies which generally behave as inseparable anti-MNS10 and anti-MNS19 (anti-Mur/Hut), although there are some exceptional phenotypes [5]. MNS36 (SAT) is associated with a unique GP(A-B) hybrid in one family and an abnormal M antigen in another [6]. MNS37 (ERIK) is located on a GPA molecule with a Gly<sup>59</sup>→Arg substitution; exon skipping within the abnormal *GYP A* gene also results in a shortened GPA molecule expressing MNS15 (St<sup>a</sup>) [7, 8].

**004, The Rh System.** RH49 (STEM) is a low incidence antigen which may be associated with the absence of RH19 (hr<sup>S</sup>) or RH31 (hr<sup>B</sup>). The assignment to Rh is supported by family evidence ( $\hat{z}$  5.21 at  $\hat{\theta}$  = 0) [9].

**005, The Lutheran System.** A high incidence antigen absent from LU:-1,-2 cells and located on the Lutheran glycoproteins is numbered LU20 [10].

**Table 2.** New blood group systems since 1990 [1]

| Name    | Symbol | No. | No. within system |                 |                  |                 |                 |                 |                 |                  |                  |     |  |
|---------|--------|-----|-------------------|-----------------|------------------|-----------------|-----------------|-----------------|-----------------|------------------|------------------|-----|--|
|         |        |     | 001               | 002             | 003              | 004             | 005             | 006             | 007             | 008              | 009              | 010 |  |
| Gerbich | GE     | 020 | ...               | Ge2             | Ge3              | Ge4             | Wb              | Ls <sup>a</sup> | An <sup>a</sup> | Dh <sup>a</sup>  |                  |     |  |
| Cromer  | CROMER | 021 | Cr <sup>a</sup>   | Tc <sup>a</sup> | Tc <sup>b</sup>  | Tc <sup>c</sup> | Dr <sup>a</sup> | Es <sup>a</sup> | IFC             | WES <sup>a</sup> | WES <sup>b</sup> | UMC |  |
| Knops   | KN     | 022 | Kn <sup>a</sup>   | Kn <sup>b</sup> | McC <sup>a</sup> | Sl <sup>a</sup> | Yk <sup>a</sup> |                 |                 |                  |                  |     |  |

**Table 3.** Chromosomal assignments of blood group system genes since 1990 [1]

| Locus               | Chromosome and region |
|---------------------|-----------------------|
| <i>CROMER (DAF)</i> | 1q32                  |
| <i>KN (CRI)</i>     | 1q32                  |
| <i>GE (GYPC)</i>    | 2q14→q21              |
| <i>CO</i>           | 7p                    |
| <i>KEL</i>          | 7q33                  |
| <i>YT (ACHE)</i>    | 7q22                  |

014, *The Dombrock System*. Serological and biochemical evidence [11] has led to the antigens of collection 206 (Gregory) and the high incidence antigen 901004 (Jo<sup>a</sup>) being elevated to the Dombrock system. 206001 (Gy<sup>a</sup>) has become DO3; 206002 (Hy) is DO4; 901004 (Jo<sup>a</sup>) is DO5. Red cells lacking the DO3 antigen represent a null phenotype and are DO:-1,-2,-3,-4,-5. DO:-4 cells are DO:-1,2,3 with weak DO2 and DO3 antigens. DO1, DO3, DO4, and DO5 have all been shown to be on the same membrane glycoprotein. DO2 awaits appropriate testing [11, 12].

### New Blood Group System (table 2)

022, *The Knops System*. The following antigens of collection 205 (Cost) are located on the C3b/C4b receptor, complement receptor 1 (CR1, CD35): 205004 (Kn<sup>a</sup>); 205005 (Kn<sup>b</sup>); 205006 (McC<sup>a</sup>); 205007 (Sl<sup>a</sup>); and 205003 (Yk<sup>a</sup>) [13, 14]. The *CRI* locus belongs to the regulators of complement activation (RCA) gene cluster on chromosome 1q32, which also includes the *DAF* (Cromer system) locus. Consequently, like *DAF*, *CRI* is distinct from genes controlling systems 001 to 021 [2]. The Knops system is shown in table 2. The antigens 205001 (Cs<sup>a</sup>) and 205002 (Cs<sup>b</sup>) do not appear to be on CR1 and remain in the Cost collection (205) [14, 15].

### Chromosome Location of Blood Group System Genes (table 3)

Close linkage of *YT* to two DNA polymorphisms has confirmed the assignment of *YT* to chromosome 7q [16]. Furthermore, Yt<sup>a</sup> and Yt<sup>b</sup> are carried on the acetylcholinesterase protein [17] and *YT* polymorphism is determined by the amino acid occupying position 322 [18]. The recent assignment of the acetylcholinesterase locus to 7q22 thus provides *YT* regional localization [19]. The *KEL* locus was assigned to 7q32–q36 through close linkage with the prolactin-inducible protein locus (*PIP*) [20] and confirmed by linkage with three DNA markers tightly linked to the cystic fibrosis locus [21]. In situ hybridization studies have refined the *KEL* location to 7q33 [22]. Location of system 22 (Knops) antigens on CR1 assigns the locus governing them to 1q32 [13, 14]. Association of DI1 with a band-3 variant suggests that the Diego system (010) may be controlled by the *EPB3* locus on chromosome 17q [23].

### Blood Group Collections (table 4)

Collection 205 (Cost) now consists of 205001 (Cs<sup>a</sup>) and 205002 (Cs<sup>b</sup>) only; 205003 to 205007 comprise the new system 22 (Knops). Collection 206 (Gregory) is obsolete as both antigens previously within that collection are now part of system 14 (Dombrock).

### The 700 Series (table 5)

There have been no changes in the 700 series since the 1991 report.

### The 901 Series (table 5)

901004 (Jo<sup>a</sup>) has become 014005 and the number 901004 is obsolete.

**Table 4.** Changes to blood group collections since 1990 [1]

| Collection |          | Specificity |                  | Comments             |
|------------|----------|-------------|------------------|----------------------|
| No.        | name     | No.         | symbol           |                      |
| 201        | Gerlich  |             |                  | Obsolete, now 020    |
| 202        | Cromer   |             |                  | Obsolete, now 021    |
| 204        | Auberger |             |                  | Obsolete             |
|            |          | 204001      | Au <sup>a</sup>  | Now 005018           |
|            |          | 204002      | Au <sup>b</sup>  | Now 005019           |
| 205        | Cost     | 205003      | Yk <sup>a</sup>  | Obsolete, now 022005 |
|            |          | 205004      | Kn <sup>a</sup>  | Obsolete, now 022001 |
|            |          | 205005      | Kn <sup>b</sup>  | Obsolete, now 022002 |
|            |          | 205006      | McC <sup>a</sup> | Obsolete, now 022003 |
|            |          | 205007      | SI <sup>a</sup>  | Obsolete, now 022004 |
| 206        | Gregory  |             |                  | Obsolete             |
|            |          | 206001      | Gy <sup>a</sup>  | Now 014003           |
|            |          | 206002      | Hy               | Now 014004           |
| 210        |          | 210001      | Le <sup>c</sup>  |                      |
|            |          | 210002      | Le <sup>d</sup>  |                      |
| 211        | Wright   | 211001      | Wr <sup>a</sup>  |                      |
|            |          | 211002      | Wr <sup>b</sup>  |                      |

**Table 5.** Changes to 700 and 901 series since 1990 [1]

| No.    | Name      | Symbol          | Comments             |
|--------|-----------|-----------------|----------------------|
| 700001 | Wright    | Wr <sup>a</sup> | Obsolete, now 211001 |
| 700012 | Griffiths | Gf              | Obsolete             |
| 700020 | Ahonen    | An <sup>a</sup> | Obsolete, now 020007 |
| 700031 | Duch      | Dh <sup>a</sup> | Obsolete, now 020008 |
| 700032 | POLLIO    | POLL            | Obsolete             |
| 700048 | FPTT      | FPTT            |                      |
| 700049 | HJK       | HJK             |                      |
| 700050 | HOFM      | HOFM            |                      |
| 700051 | ELO       | ELO             |                      |
| 901004 | Joseph    | Jo <sup>a</sup> | Obsolete, now 014005 |
| 901010 | Fritz     | Wr <sup>b</sup> | Obsolete, now 211002 |

## Applications for ISBT Numbers

The 1990 report [1] should be consulted for the criteria and procedures required for acquisition of ISBT numbers. The necessary forms will be found in appendices 2, 3, and 4 [1]. MNS32 to MNS37 and 700048 to 700051 must be added to appendix 2 in which 700.12 and 32 should be deleted. LU20 must be added to appendix 3. Other changes in numerical designations resulting from this report and the 1991 report [2] should be made, or revised application forms should be requested from the appropriate Working Party member:

Prof. Dr. W. Dahr for an MNS number

Dr. P. D. Issitt for an Rh number

Dr. J. Jørgensen for a number in other systems

Dr. D. J. Anstee for a number in collections

Dr. A. Lubenko for a 700 number

Dr. G. L. Daniels for a 901 number

For addresses and Fax numbers see appendix 5 [1]. The following address and Fax numbers have been changed:

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