

THIRD EDITION

THE
BLOOD GROUP
ANTIGEN

FactsBook

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Preface

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In compiling the entries for this text we were again impressed by the rapid pace with which new information became available. We have done our best to be consistent, accurate, and up-to-date (as of going to press). We encourage comments from readers on any errors, omissions, and improvements. Please email the authors:

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Abbreviations

AChE	Acetylcholinesterase
AET	2-aminoethylisothiuronium bromide
AIDS	Acquired immune deficiency syndrome
AIHA	Autoimmune hemolytic anemia
ART	ADP-ribosyltransferase
BCAM	Basal cell adhesion molecule
BGM	Blood group modified
bp	Base pair
CAT	Column agglutination technology
CCP	Complement control protein
CD	Cluster of differentiation
CDA	Congenital dyserythropoietic anemia
CDG	Congenital disorder of glycosylation
cDNA	complementary DNA
Cer	Ceramide
CHAD	Cold hemagglutinin disease
CHIP	Channel-forming integral protein
CHO	Carbohydrate moiety
CGD	Chronic granulomatous disease
COOH	Carboxyl terminus
CR1	Complement receptor 1
CSF	Cerebrospinal fluid
CTH	Ceramide trihexoside
DAF	Decay accelerating factor
DAT	Direct antiglobulin test
DNA	Deoxyribonucleic acid
DTT	Dithiothreitol
ep	epitope
ER-Golgi	Endoplasmic reticulum-Golgi apparatus
Fuc	L-fucose
Gal	D-galactose
GalNAc	<i>N</i> -acetyl-D-galactosamine
Glc	Glucose
GlcNAc	<i>N</i> -acetyl-D-glucosamine
GP	Glycophorin
GPI	Glycosylphosphatidylinositol
<i>GYP</i>	Glycophorin gene
HA	Hemolytic anemia
HDFN	Hemolytic disease of the fetus and newborn
HLA	Human leukocyte antigen
HEMPAS	Hereditary erythoblastic multinuclearity with positive acidified serum test
HUS	Hemolytic uremic syndrome
IAT	Indirect antiglobulin test

Ig	Immunoglobulin
ISBT	International Society of Blood Transfusion
ITP	Immune thrombocytopenia
kbp	Kilo base pair
LAD	Leukocyte adhesion deficiency
LISS	Low-ionic strength solution
MAb	Monoclonal antibody
MAIEA	Monoclonal antibody immobilization of erythrocyte antigens
2-ME	2-Mercaptoethanol
MDS	Myelodysplastic syndromes
M_r	Apparent relative molecular mass
NeuNAc	<i>N</i> -acetyl neuraminic acid
NH ₂	Amino terminus
NSAID	Nonsteroidal anti-inflammatory drug
NT/nt	Nucleotide
ORF	Open reading frame
PCH	Paroxysmal cold hemoglobinuria
PEG	Polyethylene glycol
PNH	Paroxysmal nocturnal hemoglobinuria
R	Remainder of carbohydrate chain
RBC	Red blood cell
RT	Room temperature
SCD	Sickle cell disease
SCR	Short consensus repeat
SDS-PAGE	Sodium dodecyl sulfate-polyacrylamide gel electrophoresis
Se/se	Secretor/non-secretor
SGP	Sialoglycoprotein
SLE	Systemic lupus erythematosus
SNP	Single nucleotide polymorphism
SSEA	Stage-specific embryonic antigen

Useful Websites

The easiest and quickest way is to access via a search engine.

dbRBC (for information about all blood group systems)

nybloodcenter.org (for information about *RHCE* alleles)

Rhesus Base (for information about *RHD* alleles)

www.genenames.org/ (for HUGO gene names)

www.isbt-web.org (for information about terminology and all blood group systems)

www.emina.med.uni-muenchen.de or <http://www.euro-hd.net/html/na/registry>

(for information about McLeod syndrome).

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Section I

The Introductory Chapters

Introduction

Aims of this FactsBook

The purpose of this FactsBook is to provide key information relating to the erythrocyte membrane components carrying blood group antigens, the genes encoding them, the molecular basis of the antigens and phenotypes, characteristics, and the clinical significance of blood group antibodies. Only key references are given to allow the interested reader to obtain more details. The book is designed to be a convenient, easy-to-use reference for those involved in the field of transfusion medicine, as well as medical technologists, students, physicians, and researchers interested in erythrocyte blood group antigens.

This FactsBook contains information about the blood group antigens that have been numbered by the International Society of Blood Transfusion Working Party on Red Cell Immunogenetics and Blood Group Terminology¹⁻⁴. The blood group systems and the antigens within each system are listed by their traditional name, and are arranged in the same order as described by the ISBT Working Party for Red Cell Immunogenetics and Blood Group Terminology. See Table 1.1 for an overview of the blood group systems. Those antigens not in a blood group system are accommodated in Collections (200 series), in the 700 Series of Low-Incidence Antigens or in the 901 Series of High-Incidence Antigens (for latest ISBT terminology, see www.isbt-web.org).

Selection of entries

Blood group antigens are surface markers on the outside of the red blood cell (RBC) membrane. They are proteins and carbohydrates attached to lipid or protein. A model for the types of membrane components carrying blood group antigens is shown in Figure 1.1. A blood group antigen is defined serologically by antibodies made by a human, and in order to be assigned a number by the ISBT Working Party the antigen must be shown to be inherited. Historically, antigens associated with forms of polyagglutination have not been numbered by the ISBT; however, in Section III we have included a table summarizing the characteristics of T, Tn, Tk, and Cad.

TABLE 1.1 Blood group systems with gene name and chromosome location

System name	ISBT symbol ^a	ISBT number	Number of antigens	Gene names	Chromosome location	CD number
ABO	ABO	001	4	ABO	9q34.2	
MNS	MNS	002	46	GYPB, GYPB, GYPE	4q31.21	CD235
P1PK	P1PK	003	3	A4GALT	22q13.2	CD77 (P ^k)
Rh	RH	004	52	RHD, RHCE	1p36.11	CD240
Lutheran	LU	005	20	LU, BCAM	19q13.32	CD239
Kell	KEL	006	34	KEL	7q34	CD238
Lewis	LE	007	6	FUT3	19p13.3	
Duffy	FY	008	5	FY, DARC	1q23.2	CD234
Kidd	JK	009	3	JK, SIC14A1	18q12.3	
Diego	DI	010	22	DI, SIC4A1	17q21.31	CD233
Yt	YT	011	2	YT, ACHE	7q22.1	
Xg	XG	012	2	XG	Xp22.33	CD99
Scianna	SC	013	7	SC, ERMAP	1p34.2	
Dombrock	DO	014	8	DO, ART4	12p12.3	CD297
Colton	CO	015	4	CO, AQP1	7p14.3	
Landsteiner-Wiener	LW	016	3	LW, ICAM4	19p13.2	CD242
Chido/Rodgers	CHI/RG	017	9	CHI/RG, C4A, C4B	6p21.32	
H	H	018	1	FUT1	19q13.33	CD173

System name	ISBT symbol [^]	ISBT number	Number of antigens	Gene names	Chromosome location	CD number
Kx	XK	019	1	XK	Xp21.1	
Gerbich	GE	020	11	GE, GYP	2q14.3	CD236
Cromer	CROM	021	17	CROM, CD55	1q32.2	CD55
Knops	KN	022	9	KN, CR1	1q32.2	CD35
Indian	IN	023	4	IN, CD44	11p13	CD44
Ok	OK	024	3	OK, BSG	19p13.3	CD147
Raph	RAPH	025	1	RAPH, CD151	11p15.5	CD151
John Milton Hagen	JMH	026	6	JMH, SEMA7A	15q24.1	CD108
I	I	027	1	GCNT2	6p24.2	
Globoside	GLOB	028	1	B3GALNT1	3q26.1	
Gill	GIL	029	1	GIL, AQP3	9p13.3	
Rh-associated glycoprotein	RHAG	030	4	RHAG	6p12.3	CD241
Forssman	FORS	031	1	GBGTT1	9q34.2	
JR	JR	032	1	JR, ABCG2	4q22.1	CDw338
LAN	LAN	033	1	LAN, ABCB6	2q36	

[^]For up to date HUGO gene names see <http://www.genenames.org/>

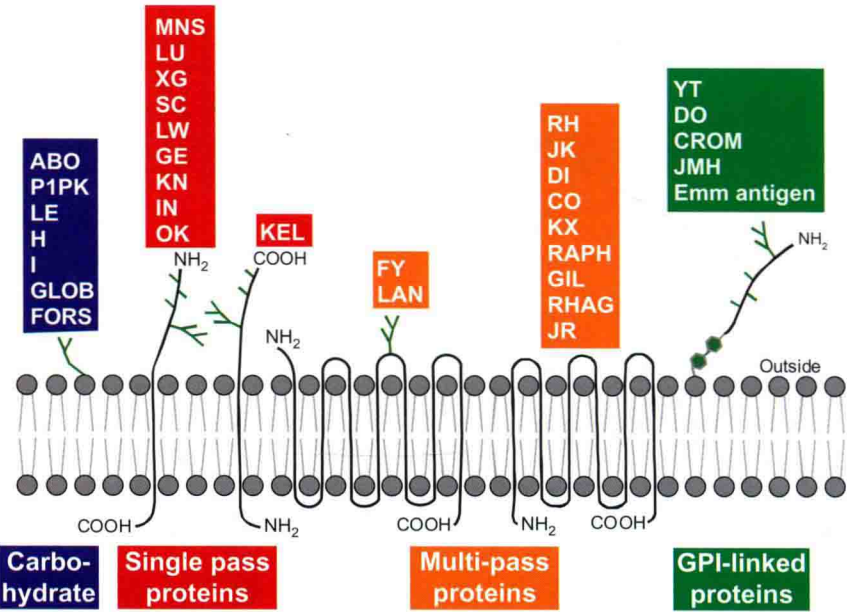


FIGURE 1.1 Model of RBC membrane components that carry blood group antigens. Not shown is the Ch/Rg blood group system. Ch/Rg antigens are carried on C4d, which is adsorbed from plasma onto RBC membrane components.

Terminology

The nomenclature used for erythrocyte blood group antigens is inconsistent. While several antigens were named after the proband whose RBCs carried the antigen or who made the first known antibody, others were assigned an alphabetical or a numerical notation. Even within the same blood group system, antigens have been named using different schemes, and this has resulted in a cumbersome terminology for describing phenotypes. The ISBT Working Party established a system of upper case letters and numbers to represent blood group systems and blood group antigens in a format that will allow both infinite expansion and computer-based storage. These symbols and numbers are designed for use in computer databases (no lower case letters) and are short (for column headings). A comprehensive review of terminology and its recommended usage can be found in Garratty, et al.⁵

Throughout this book, the systems and antigens are named by the traditional name, but we also give the ISBT symbol, the ISBT number, and obsolete names that have been used in the literature. We have included a brief description of how the blood group systems and antigens were named.

The following are examples of how to write antigens, antibodies, phenotypes, and genotypes.

List of antigens:	M, N, P1, K, Kp ^b , K11, Fy ^a , Fy ^b , Lu3
List of antibodies:	Anti-M, anti-K, anti-Fy ^a , anti-Jk3 or Anti-M, -K, -Fy ^a , -Jk3 or antibodies directed against M, K, Fy ^a , and Jk3 antigens
Phenotype:	D+C-E-c+e+; M+N-S-s+; Vw+; K+k-K11-; Fy(a+b+); Jk(a+b-) or RH:1,-2,-3,4,5; MNS:1,-2,-3,4,9; KEL:1,-2,-11; FY:1,2; JK:1,-2

	Traditional	ISBT
Antigen	Fy ^a	FY1, 008001 or 8.1
Phenotype	Fy(a+b-)	FY:1,-2
Gene	Fy ^a	FY*01 or FY*A
	FY	FY*N, FY*01N or FY*02N
Genotype	Fy ^a Fy ^a	FY*01/FY*01 or FY*A/FY*A
	Fy ^a Fy	FY*01/FY*N or FY*A/FY*N

In addition to the ISBT terminology for antigens and traditional allele names, the ISBT Working Party recently agreed on a proposed terminology for blood group alleles⁴. Since the introduction of a new naming system for thousands of alleles is a complicated and cumbersome procedure, it is anticipated that certain changes to the new terminology are unavoidable. Despite this, we have used the currently agreed nomenclature to familiarize the reader with the new allele names. However, we encourage the use of the official and constantly updated lists of allele names found at www.isbt-web.org. These allele names are restricted to those with a phenotypic effect and intended for use in Transfusion Medicine. Their use does not require sequencing of the entire allele. The rules for naming alleles and for obtaining a number for a new allele may be found at www.isbt-web.org.

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