Cleveland Clinic

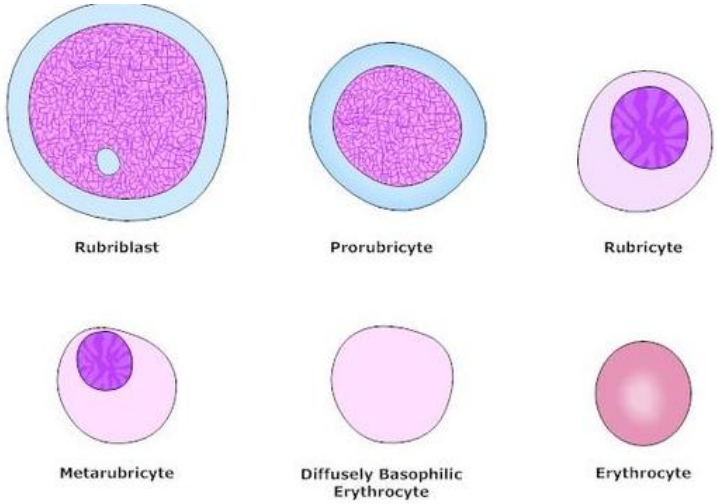
The Kell Blood Group System

Kell System

- 1946- Discovered in serum of Mrs. Keller
  - Found due to HDFN
  - First blood group discovered after introduction of AHG testing
- 32 antigens high and low prevalence
- Only found on RBCs
- Very immunogenic- 2nd after the RH system

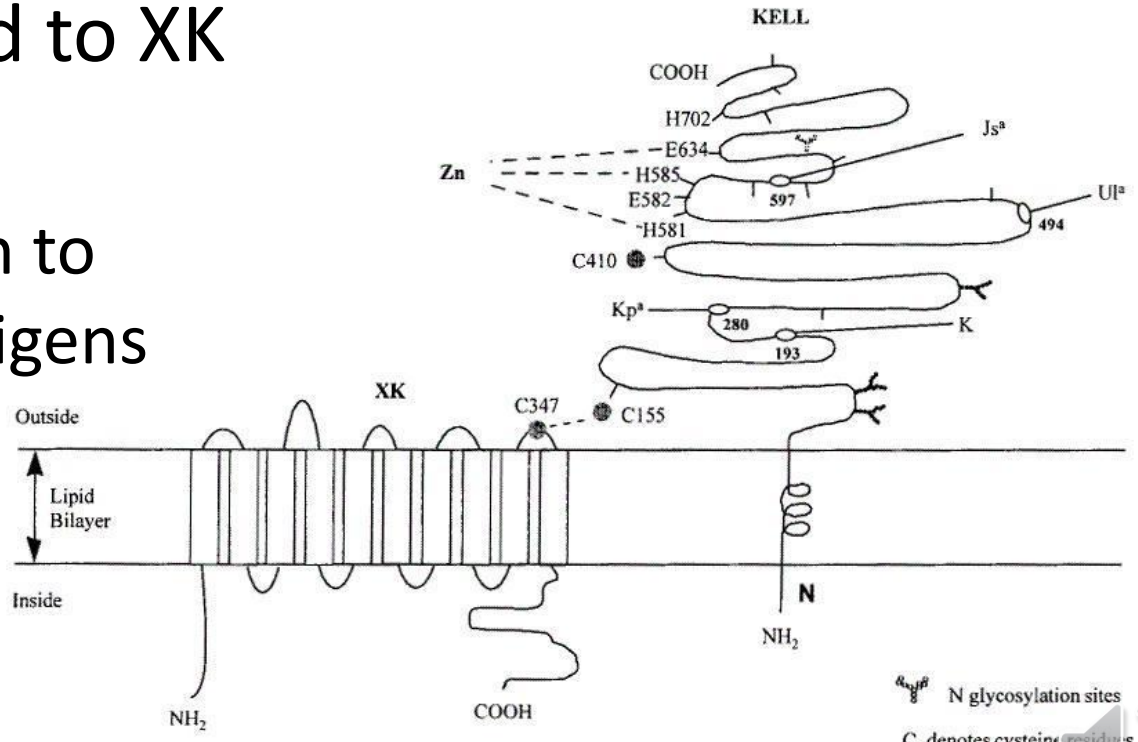
Fetal RBCs

- K antigen detected- 10 weeks
- k antigen detected- 7 weeks
- Extremely severe HDFN
  - Antigens very well expressed
  - Destroys erythroid precursor cells as well as mature cells
  - Titer of antibody can be low, but cause much destruction



Biochemistry

- Antigens on gylcoprotein spanning RBC once
- Covalently linked to XK protein
  - Need XK protein to express Kell antigens

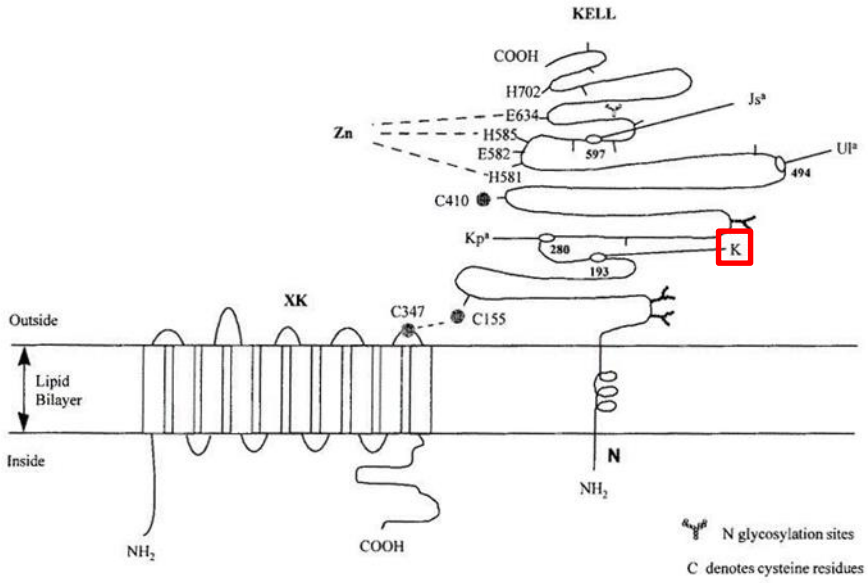


Genetics

- *KEL* gene- on chromosome 7
  - Single base mutation result in different antigens
- *XK* gene- on X chromosome
  - Codes for the Kx antigen
- Two genes independent

K and k Antigens

- Antithetical antigens
- 9% K positive
- >99% k positive
- High immunogenicity
  - If K-, 10% chance of making antibody if unit is K+



Phenotype	Whites (%)	Blacks (%)
K-k+	91	98
K+k+	8.8	2
K+k-	0.2	<0.1

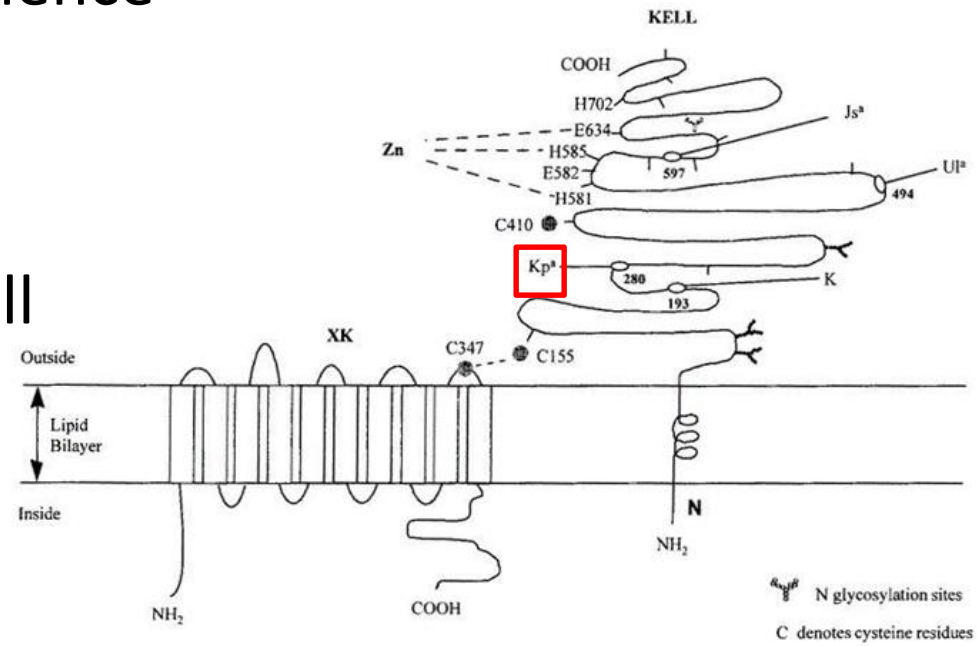
Anti-K

- Most common antibody outside ABO and Rh
- Severe HDFN and HTR
- From transfusion or pregnancy
- 20% bind complement

Enzymes	Resistant
IgM vs. IgG	IgG
Cold or Warm	37°C
Natural vs. Immune	Immune
HTR	Yes
HDN	Yes

Kp^a, Kp^b, and Kp^c Antigens

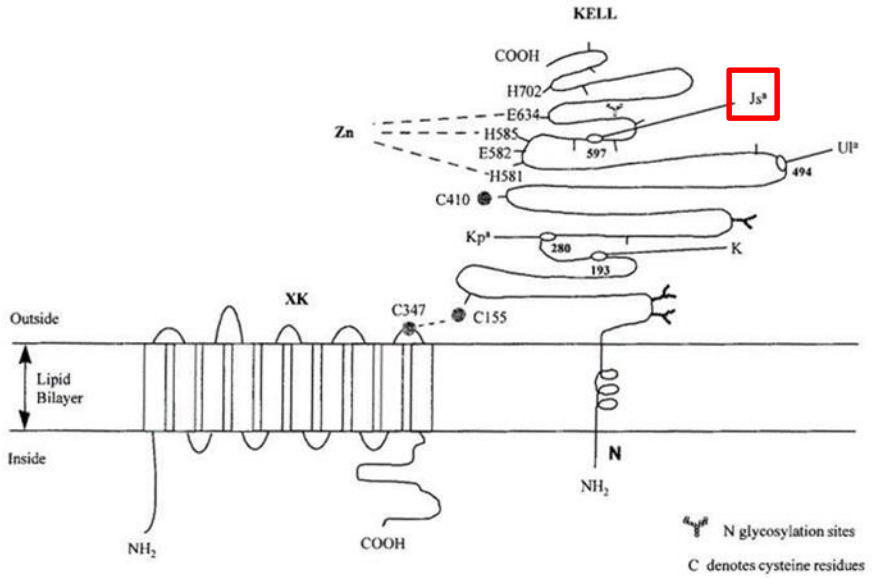
- Kp^a and Kp^c = low prevalence
  - Mutations of Kp^b
- Kp^b = high prevalence
- Kp^a associated with suppression of other Kell antigens
- Kp^c = extremely rare



Phenotype	Whites (%)	Blacks (%)
Kp(a+b-)	<0.1	0
Kp(a+b+)	2.3	Rare
Kp(a-b+)	97.7	100

Js^a and Js^b Antigens

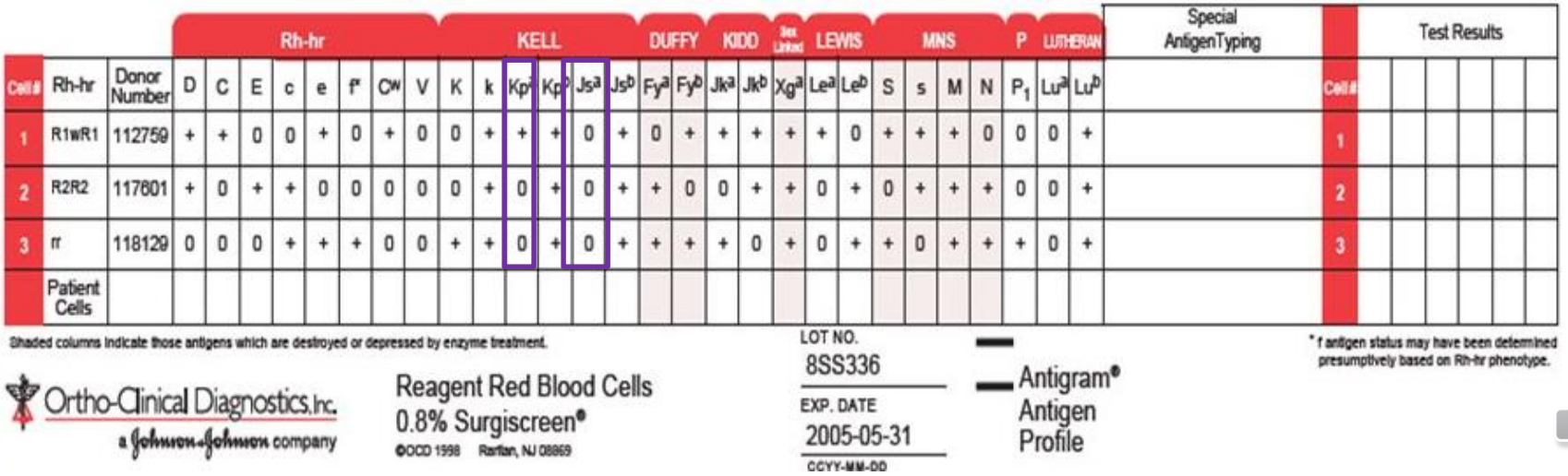
- Js^a – found in 20% of blacks, but <0.1% of whites
- Js^b – high prevalence



Phenotype	Whites (%)	Blacks (%)
Js(a+b-)	0	1
Js(a+b+)	Rare	19
Js(a-b+)	100	80

Anti-Kp^a, Js^a

- Low prevalence antigens
- Rare- few people exposed to antigen
- Rarely detected on screen/panel
- Detected through crossmatch/HDFN
- Same characteristics/significance as K



			Rh-hr									KELL				DUFFY		KIDD		Sex Linked	LEWIS		MNS				P	LUTHERAN		Special Antigen Typing		Test Results						
Cell #	Rh-hr	Donor Number	D	C	E	c	e	f*	Cw	V	K	k	Kp^a	Kp^b	Js^a	Js^b	Fy^a	Fy^b	Jk^a	Jk^b	Xg^a	Le^a	Le^b	S	s	M	N	P$_1$	Lu^a	Lu^b		Cell #						
1	R1wR1	112759	+	+	0	0	+	0	+	0	0	+	+	+	0	+	0	+	+	+	+	+	0	+	+	+	0	0	0	+		1						
2	R2R2	117601	+	0	+	+	0	0	0	0	0	+	0	+	0	+	+	0	0	+	+	0	+	0	+	+	+	0	0	+		2						
3	rr	118129	0	0	0	+	+	+	0	0	+	+	0	+	0	+	+	+	+	0	+	0	+	+	0	+	+	+	0	+		3						
	Patient Cells																																				

Shaded columns indicate those antigens which are destroyed or depressed by enzyme treatment.

* f antigen status may have been determined presumptively based on Rh-hr phenotype.

Ortho-Clinical Diagnostics, Inc. a Johnson & Johnson company

Reagent Red Blood Cells
0.8% Surgiscreen®
©OCD 1998 Raritan, NJ 08869

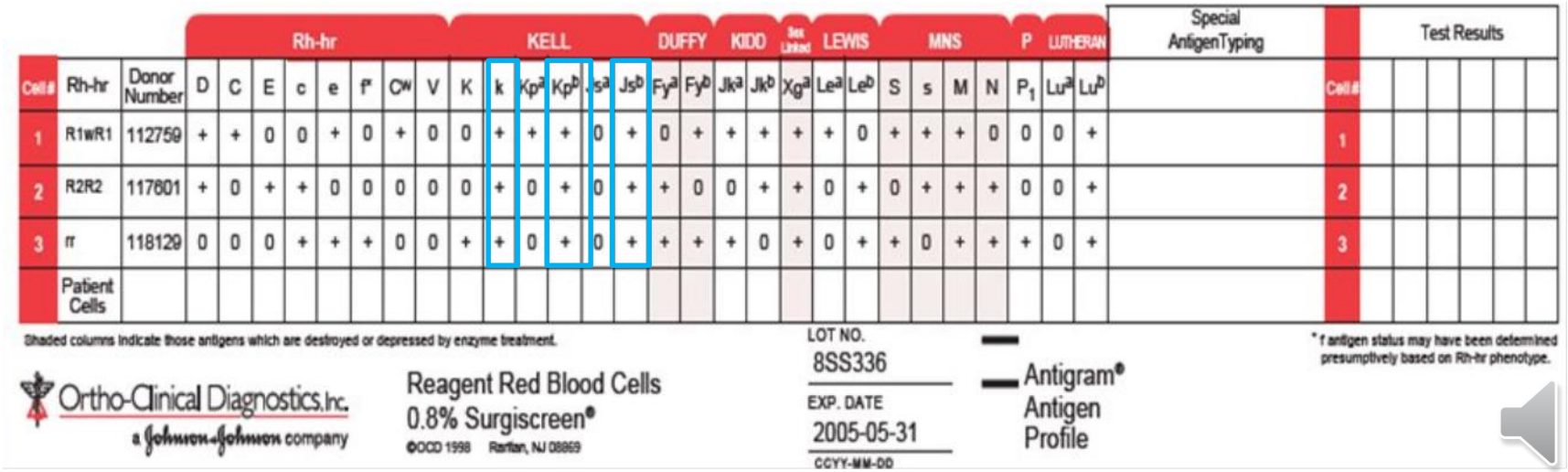
LOT NO. 8SS336

EXP. DATE 2005-05-31
CCYY-MM-DD

Antigram® Antigen Profile

Anti- k, Kp^b, Js^b

- High prevalence antigens
- Rare- so few people lack them
- Easy to detect- all screen cells positive
  - Hard to rule-out other antibodies
  - DTT can destroy Kell, leaving other antibodies
- Difficult to find compatible units
  - Autologous units
  - Rare donor units



			Rh-hr								KELL					DUFFY		KIDD		Xg	LEWIS		MNS				P	LUTHERAN		Special Antigen Typing		Test Results						
Cell #	Rh-hr	Donor Number	D	C	E	c	e	f	C^w	V	K	k	Kp^a	Kp^b	Js^a	Js^b	Fy^a	Fy^b	Jk^a	Jk^b	Xg^a	Le^a	Le^b	S	s	M	N	P_1	Lu^a	Lu^b		Cell #						
1	R1wR1	112759	+	+	0	0	+	0	+	0	0	+	+	+	0	+	0	+	+	+	+	+	0	+	+	+	0	0	0	+		1						
2	R2R2	117601	+	0	+	+	0	0	0	0	0	+	0	+	0	+	+	0	0	+	+	0	+	0	+	+	+	0	0	+		2						
3	rr	118129	0	0	0	+	+	+	0	0	+	+	0	+	0	+	+	+	+	0	+	0	+	0	+	+	0	+	+	0	+		3						
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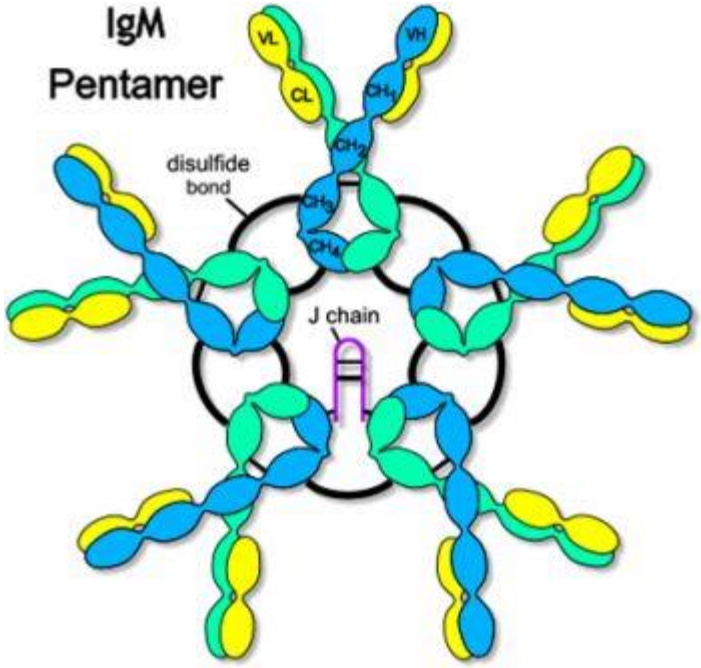
Antigram® Antigen Profile

DTT (Dithiothreitol)

- Distinguish between IgM and IgG antibodies
- IgM is removed leaving only IgG
- DTT dissociates pentameric form of IgM
  - Cleaves covalent bond between subunits and J chain
- Also breaks down antigens of Kell system

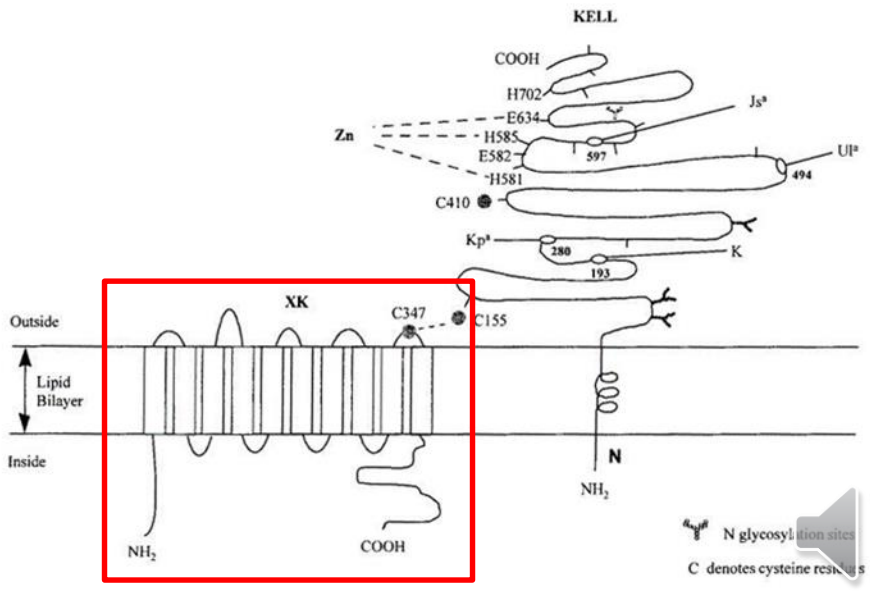


Autoantibodies

- Most against unidentified high prevalence Kell antigens

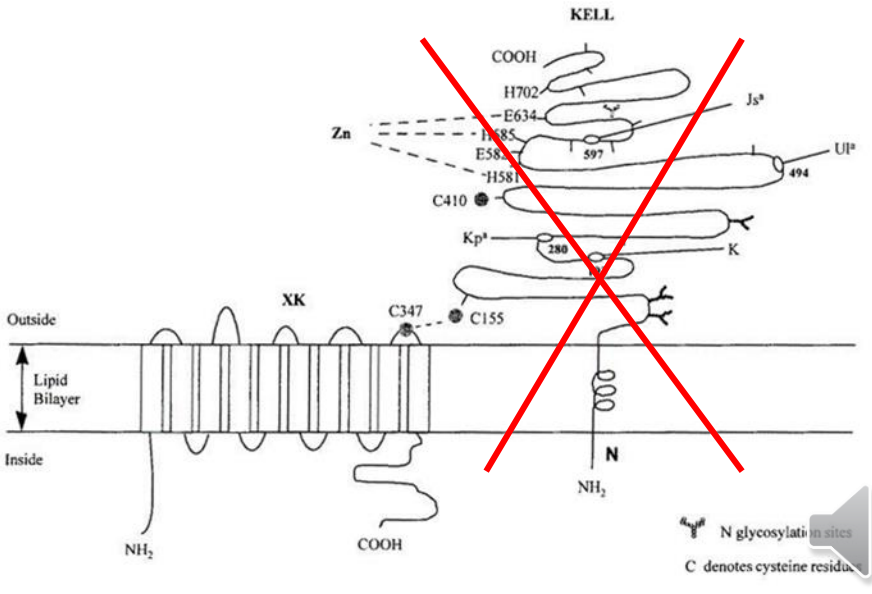
Kx Antigen

- On all RBCs except those with McLeod phenotype
- Separate protein encoded by *XK* gene
- Separate blood group system: XK System
  - Only antigen in system



K_0 Phenotype

- K_0 – silent Kell allele
  - Null phenotype when K_0K_0
- Lack expression of Kell antigens
- Normal RBC survival
- Rare 1:25,000

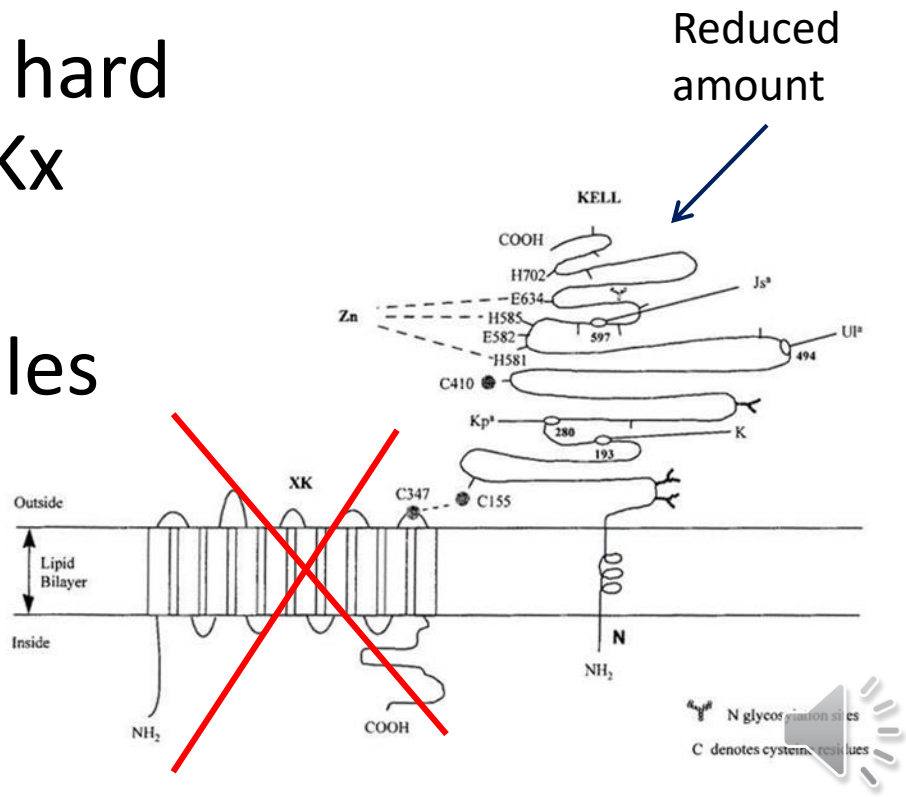


K_0 Phenotype

- Can make antibody anti-Ku (K5) – universal Kell antigen
  - Can not be separated into different antibodies
- Can make all other Kell antibodies
- Cause HDN and HTR

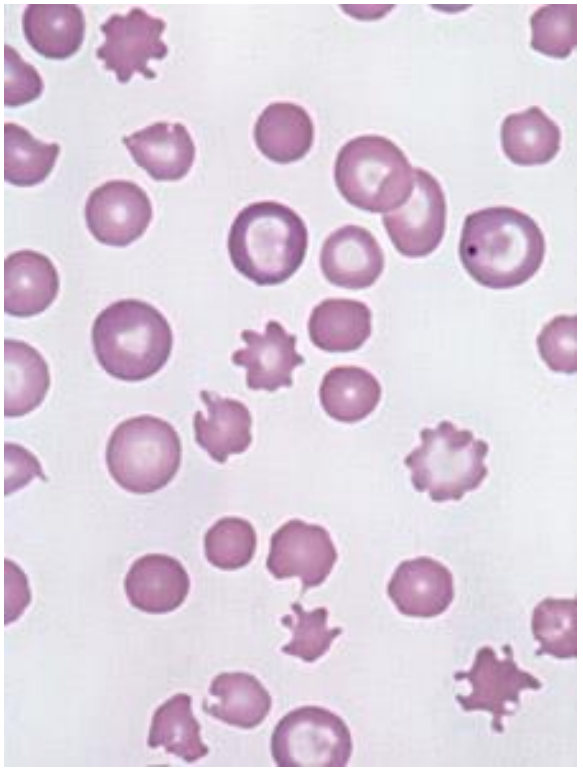
McLeod Phenotype

- RBCs lack Kx and Km antigens
  - Mutations and deletions at *XK* locus
- Very few Kell antigens- hard time bonding without Kx
- Very rare- X-linked inheritance, mostly males
- Named after student discovered in



McLeod Syndrome

- RBCs acanthocytic (irregular shape)
- Reduced in vivo RBC survival
- Many have chronic hemolytic anemia
- Muscle and nerve disorders
  - Muscular dystrophy at 40-50 yrs.
  - Cardiomegaly



McLeod and CGD

- Chronic Granulomatous Disease (CGD)
  - Phagoctyes can't make NADH oxidase- need to make H_2O_2 –kill bacteria
  - Die early from untreated infections
- Near Kx on X chromosome
- May be related, but you don't need one to have the other

McLeod and CGD

- McLeod and CGD = make anti-Kx and Km
- McLeod only = Anti-Km
- Female Carriers- one X chromosome shuts down- only 1 active in every cell
  - Exhibit 2 cell populations- 1 normal, 1 McLeod
  - Vary from 5-85% McLeod populations

Altered Kell Expression

- Weaker expression:
  - McLeod Phenotype
  - Kp^a gene expression
  - Gerbich-negative phenotype
  - AIHA- directed against Kell
- Acquired K:
  - *Streptococcus faecium* can convert K- to K+ cells
  - Return to normal after infection

Cleveland Clinic

Every life deserves world class care.