HDN ABID CASE #3

1. What is the probable identity of this antibody? What is the relevance of the patient's ethnic origin?

   Probable anti-Jk3. Anti-Jk^3 reacts as an inseparable form of anti-Jk^a + anti-Jk^b. Jk^3 negative {Jk(a-,b-)} individuals are very rare. This patient presents the classic serologic findings of an antibody directed against a high frequency antigen, serum reactivity against all RBCs except the patient's own. The antigen phenotype then demonstrates that the patient lacks a high frequency antigen, in this case Jk^3.

   The Jk^3 negative phenotype is found most commonly in Polynesian populations (0.9% in one study), and its frequency is greater in other East Asian groups than in individuals of African or European descent. It is also noted that there are a number of Finns who are Jk^3 negative.

2. Is any further workup needed to prove it? What does the alloadsorption study demonstrate?

   In combination, the antigen phenotype and the adsorption study rule out other underlying allo-antibodies. That is, from the phenotype, the patient could make anti-K, -Fy^b, or –M. However, these should not have been adsorbed, since the absorbing cell was K negative and ficin treated.

   This workup does not prove the identity of the antibody; an antibody directed against another high frequency antigen would also be allo-adsorbed (as long as the antigen was not destroyed by ficin). To prove the anti-Jk^3 specificity, we would have to show that the antibody failed to react with 3 Jk^3 negative RBC samples. Nonetheless, given the patient's phenotype, this specificity is highly likely; we can certainly state this is the specificity with greater than 95% confidence, given the rarity of the Jk^3 negative state.

   One would also want to perform a titer. Although there are no standards against which such a titer can be compared, a very high titer, or a significant increase on serial sampling, would prompt measurement of middle cerebral artery blood flow velocity, or even amniocentesis or PUBS, all of which are more specific tests for the presence or absence of fetal hemolysis and anemia.

3. Does this antibody cause hemolytic transfusion reactions? *Yes*
   HDN? *Kidd antibodies cause relatively mild HDN in comparison to their severe consequences for incompatible transfusions.*

4. How might we find compatible blood for this patient?

   Test siblings and other blood relatives, or other individuals of Polynesian ethnic origin. Also, since the patient's risk for needing transfusion is in the future, she could donate autologous RBCs which could be frozen for future need. Similarly, if her infant IS affected by HDN, the frozen cells can be used for his or her transfusion.

5. What is the possible genetic basis of this patient's phenotype?

   The Kidd blood group system includes 3 antigens, Jk^a, Jk^b, and Jk^3. Family studies show that most Jk^3 negative phenotypes are inherited on a recessive basis, but there is also a dominant or "inhibitor" form in which Jk antigen expression is decreased. Individuals with inhibitor phenotypes do not form anti-Jk^3.

6. What is the biochemical nature of the antigen? Are there any health consequences of this phenotype?

   The Jk antigens are carried by the urea transport protein, which traverses the membrane 10 times. The Jk^a/Jk^b polymorphism is due to a single amino acid substitution. The Jk^3 negative phenotype has been shown to be due to multiple different deletions. Water reclamation by the kidneys is dependent on urea transport, and Jk^3 negative individuals cannot maximally concentrate their urine.