A RARE **KEL**'02.17 | **KEL**'02N.06(**IVS3+1g>a**) COMPOUND HETEROZYGOUS INDIVIDUAL, PRONE TO ANTI-KEL11 IMMUNIZATION

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**Background**
The Kell blood group system includes some of the most immunogenic antigens among blood groups. Beside the well-known antigens Kell(*KEL*'01), Kpa(*KEL*'03), and Js(‘*KEL*’06), the antithetic antigens KEL11/17 further contribute to this list. However, KEL17 is considered as very rare, with an approximate frequency of one KEL*02.17 homozygote among 30,000 Europeans only. We recently observed an individual with a rare anti-K11 and describe here its unusual molecular mechanism of anti-K11 sensitization.

**Methode**
Standard serological methods for antigen- and antibody-detection and specification were used. **KEL** genotyping was performed using a commercially available test kit “**KELplus**” (Inno-Train, Germany) and in house KEL11/17 PCR-using Sequence Specific Priming technique (SSP) and **KEL** gene sequencing.

**Results**
By standard serological investigation, a 73 year old female was found positive for anti-KEL11 in her serum. Reasoned by the rarity of this observation, molecular confirmation was intended. A KEL11/17 PCR-SSP was performed, but resulted in an unexpected heterozygosity for KEL11/17. Further “**KELplus**” typing delivered KEL-1,2,3,4,6,7 (K, Kp, Js negative), and surprisingly KEL*02.06(**IVS3+1g>a**), for the investigated DNA. Finally, **KEL** gene sequencing of exon areas 3 and 8 confirmed the unusual **KEL** genotype of the patient:

Compound heterozygosity for an expressed **KEL**'02.17 and an unexpressed **KEL**'02N.06(**IVS3+1g>a**).

**Conclusion**
**KEL**’02.06(**IVS3+1g>a**) is the most frequent unexpressed **KEL** allele, encoding Kell<sub>a</sub> when present in homozygous, or compound heterozygous form. However, in inherited hemizygosity this Kell<sub>a</sub> allele will allow the second **KEL** allele to behave as seemingly homozygous, when expressed, as observed in our case. Such individuals might be expected at a frequency of 1 among 520,000 Europeans, only. Since this is the second observation of an anti-KEL11, beside another with true homozygosity for KEL*02.17, we assume elevated frequency of KEL17 in the Zurich area as compared to other European areas.

**References**