The MNSs and Kk Blood Group Systems in the Disease of Mljet (mal de Meleda)

T.		Salamon ^a
R.		Hadžiselimović ^b
G.		Hajdić ^c

^aDepartment of Stomatopathology, Faculty of Stomatology, and ^bFaculty of Natural Sciences and Mathematics, University of Sarajevo, Bosnia; ^cDepartment of Transfusion, Regional Medical Center, Dubrovnik, Croatia

In a previous paper we reported on the ABO and Rhf:Rh and Rh:CDE/cde systems in 9 patients affected by the disease of Mljet, in 20 of their family members, as well as in 50 persons born in Mljet, but not related. The small number of persons affected did not allow to draw any firm conclusion from the results [1]. We now report on the MNSs and Kk (Kell-Cellano) blood group systems in 9 patients affected by the disease of Mljet, as well as in 63 controls from the island, not related to the patients. Six patients were female, 3 male. Seven were from the island of Mljet, 2 from the island of Lopud (Lafodia) near Mljet. In all, the blood group systems MNSs and Kk (Kell-Cellano) were examined.

There were 8 patients showing the MN allele; 1 was NN. Seven patients had the ss allele, 1 the SS and another the Ss allele (table 1). All patients had the kk allele; K was absent. There were 63 controls (44 male, 19 female). The MNSs and Kk systems in unaffected autochthonous inhabitants of Mljet not related to the patients were 27 showing MN, 20 MM and 16 NN; 15 showing SS, 27 ss and 2 Ss; 55 showing kk; 8 Kk (87.30 vs. 12.70%).

These results indicate a possible association of the disease of Mljet with the MN system, since the difference was statistically significant (χ test $p < 0.05$; table 2) between affected and controls. The locus of the MN system resides on the long arm of chromosome 4 in the region 40q28-q32 [2].

It is interesting that all affected persons had the kk allele, while K was absent. In various populations, K has a low frequency: there is no population in which this allele has a frequency over 13%. In the autochthonous population of Australia, in Eskimos and Mongols, the antigen K is absent. Its frequency in Negroes is essentially lower than in Whites. In European populations, the frequency is from 5 to 10% [3]. In Croatia, K is present in about 11% [4], Mljet being in that country.

According to Zelinski et al. [5], the Kell blood group locus (Kell) is tightly linked to the locus of prolactine-inducible protein. This has been established through family studies. 'However, any assignment is considered to be provisional until the data have been duplicated by another group of investigators or until linkage to other loci in the region has been established' [5].

The frequency of K in the unaffected autochthonous population of Mljet is 12.70%, which is very near to the possible maximum of K, while no patient with the disease of Mljet had it.

However, it must be emphasized: (a) that the number of persons affected by the disease of Mljet was small, and (b) that the difference between the number of affected with the homozygous pair kk and that of controls was not statistically significant ($p > 0.05$; Fisher's exact test; table 2).

Anyway we think that the problem requires further investigations. In this context, it is noteworthy that in 4 cases of the Richner-Hanhart syndrome, published by Salamon et al. [6], MN was present in 3 cases, NN in 1 case, whereas K was absent in all cases.

We found no statistically significant difference in the frequencies of Ss between the group with the disease of Mljet and the controls ($p > 0.05$; χ^2 test; table 2).

We think that the problem of linkage between the MNSs and Kk blood group systems and the disease of Mljet (and other palmo-plantar keratoses) deserves further investigations.

Acknowledgment

We are deeply indebted to Prof. Dr. Davor Ivankovic from the Institute of Statistics, Epidemiology and Informatics of the Medical Faculty of the University Zagreb for the statistical data processing.

Dr. Tibor Salamon Bukovacka ccsta 55 41000 Zagreb (Croatia)

© 1994 S. KargerAG, Basel

1018-8665/94/1881-0078 \$ 5.00/0

Table 1. MNSs and Kk (Kell-Cellano) blood group systems in patients affected by the disease of Mljet (mal de Meleda)

Table 2. Observed frequencies

References

1 Hajdic G, Hadziselimovic R, Cezarovic B, Prokop O, Göhler W: Die menschlichen Blut-Salamon T: Autochthonous mal de Meleda and the ABO and Rh blood group system. *Acta Dermatol Croatica*, in press. McKusick VA: Mendelian inheritance of man, ed 10. London, Johns Hopkins, 1992, pp 155–156.

gruppen. Jena, Fischer, 1986. Polak A: Krv; in *Medicinska Enciklopedija*. Zagreb. 1969. vol 4. Zelinski T, Coghlan G

Myal Y

Shin RPC. et al: Genetic linkage between the Kell blood group system and prolactin-inducible protein loci: Provisional assignment of Kell to chromosome 7. *Ann Hum Genet* 1991;55:137–140.