173. Rare Blood Type p and Pk in Japanese Families

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Miy. Family (p). The propositus of Miy. family was a 23-year-old male donor whose serum was found to contain an irregular hemolytic antibody in the screening tests for antibodies in donors, though he had never been transfused. His serum reacted well with all the cells tested other than two p cells at various temperatures. The serum agglutinin titer was 1:64 in saline, 1:128 with bromelin-treated cells and 1:64 in anti-globulin test. The serum when tested without inactivation gave a strong hemolytic reaction in saline at 37°C. These serological properties of the serum definitely suggested that the propositus' blood might be the extremely rare type p, the antibody in the serum being anti-Tja (anti-PP, P*). As might be expected, the serological examinations of the red cells confirmed the above presumption. As shown in Table I, the propositus' red cells were agglutinated with neither of two p sera (anti-Tja, Hata and Shimanouchi) nor one anti-Pk (anti-Tja, Hata, absorbed by P1 cells) nor two P* sera (anti-P, Kinoshita, and Mae., the propositus' serum of the P* family presented here) nor one anti-P1.

The pedigree studies (Figure 1) revealed that the propositus (II-3) had three sibs, two (II-2, II-4) of whom possessed p blood and the remaining one (II-5) was typed as normal P2. To our surprise, the father (I-1) of the propositus also possessed p blood.

Table I. Reactions of the propositus' red cells

| Anti-          | Red cells |       |
|---------------|-----------|-------|
|               | Miy.      | Mae.  |
| Tja (Hata)    | —         | +     |
| Tja (Shimanouchi) | —       | +     |
| Pk (anti-Tja, Hata, absorbed by P1 cells) | —     | +     |
| Tja, Hata, treated with hydatid cyst fluid | n.t.* | —     |
| P (Kinoshita) | —         | —     |
| P (Mae.)      | —         | n.t.* |
| P1            | —         | —     |

* not tested

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The deceased parents of the father (I-1) were first cousins. The parents of the propositus were not related, but both of them came from an island of the Inland Sea (Okayama Prefecture). All the p members of this family had anti-Tja of rather high titer in the serum.

The present p family seems unique in that the p blood looked as if it were transmitted as a dominant trait and that the p sister (II-2) of the propositus gave birth to a boy who unexpectedly escaped from hemolytic disease of the newborn, though her spouse (II-1) was typed as P1 and not related to her. Subsequently, another boy was born to her. The second son did not develop hemolytic disease either. The reason for the failure to develop hemolytic disease remains to be proved.

Mae. Family (P1). The propositus of the P1 family was a 22-year-old male donor. He was also found to have an irregular hemolytic antibody in his serum in the antibody detection test of donor's serum, though he had no history of transfusion.

His serum reacted well with all the cells tested other than one p and one P1 cells at various temperatures. The titer of the antibody was 1:32 independently of titrating methods. The red cells of the propositus reacted well with two p sera (anti-Tja) and one anti-Pk, but not with one Pk serum (anti-P), one anti-P1 and one p serum treated with hydatid cyst fluid (Table I). Thus, the propositus was typed as P1.

As shown in Figure 2, the propositus (II-2) was the only member with Pk blood. The parents (I-1, I-2) of the propositus, who
were first cousins belonged to normal $P_1$ and $P_2$ respectively. According to Race and Sanger,\(^1\) the antigen $P^*k$ is not produced by an allele at the $P$ locus, but by the hypothetical $Yy$ locus not closely linked to the $P$ locus, while the $p$ blood is assumed to occur in a homozygote for a rare allele $p$ at the $P$ locus. Thus, it appears that the propositus (II-2) is homozygous for an inactive allele $y$ and the parents (I-1, I-2) probably heterozygous. This $P^k$ family came from Hyogo Prefecture. (The name of this $P^k_2$ donor whose blood was re-examined at the WHO International Blood Group Reference Laboratory, London, was added to the International Panel of Donors of Rare Blood Types).

Blood groups data of the two family members were given in Table II.

**Remarks.** In 1968 Race and Sanger\(^1\) noted that thirty-five $p$ and twelve $P^k$ families have been found. It is noteworthy that

| Table II. Blood groups of Miy. and Mae. families |
|------------------------------------------------|

| Miy. Family |  |
|-------------|-----------------------------------|
| I-1         | O NNss p CCDee Lu(a-b+) kk Le(a-b+) Fy(a+) Jk(a+) I |
| I-2         | A$_1$ NNss P$_1$ CCDee Lu(a-b+) kk Le(a-b-) Fy(a+) Jk(a-) I |
| II-1        | A$_1$ MNss P$_1$ CCDee Lu(a-b+) kk Le(a-b-) Fy(a+) Jk(a-) I |
| II-2        | A$_1$ NNss p CCDee Lu(a-b+) kk Le(a-b+) Fy(a+) Jk(a-) I |
| II-3*       | A$_1$ NNss p CCDee Lu(a-b+) kk Le(a-b+) Fy(a+) Jk(a-) I |
| II-4        | A$_1$ NNss p CCDee Lu(a-b+) kk Le(a-b+) Fy(a+) Jk(a+) I |
| II-5        | A$_1$ NNss P$_2$ CCDee Lu(a-b+) kk Le(a-b-) Fy(a+) Jk(a-) I |

| Mae. Family |  |
|-------------|-----------------------------------|
| I-1         | A$_1$ NNss P$_1$ CCDee . . kk Le(a-b-) Fy(a+) Jk(a-) . |
| I-2         | B MNss P$_2$ CCDee . . kk Le(a-b-) Fy(a+) Jk(a+) . |
| II-1        | A$_1$B MNss P$_2$ CCDee . . kk Le(a-b-) Fy(a+) Jk(a+) . |
| II-2*       | A$_1$B NNss P$_2^k$ CCDee Lu(a-b+) kk Le(a-b+) Fy(a+) Jk(a+) I |

* propositus
twelve of the 35 p families are Swedish, and nine of the 12 Pk families Finnish. In Japan, eight p\(^2\)\(^{1\text{--}8}\) and four Pk\(^9\)\(^{1\text{--}11}\) families had been reported including the present two families by the end of 1973. Recently, additional four p\(^{12\text{--}15}\) and one Pk\(^16\) examples followed the present two families. One of them was found to be related to our Miy. family. Earlier cases of p and Pk in Japan were detected mainly due to abortion and hemolytic disease of the newborn, while recent cases came to be brought to notice by ABO-grouping or antibody detecting tests. Four of the 12 p and four of the 5 Pk cases so far reported in Japan were found to occur in consanguineous families.

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References

1) Race, R. R., and Sanger, Ruth.: Blood Groups in Man 5th ed. (1968).
2) Iseki, S., Masaki, S., and Levine, P.: Nature, \(173\), 1193 (1954) cited by Race and Sanger in Blood Groups in Man 5th ed.
3) Yasuda, J., and Yokoyama, M.: Proc. 8th Congr. int. Soc. Blood Transf., Tokyo 1960, p. 203 (1962).
4) Hayashida, Y., Homma, T., Furukawa, K., and Watanabe, A.: Proc. 15th Congr. Japan Soc. Blood Transf., p. 18 (1967) and Furukawa, K.: J. Japan Soc. Blood Transf., \(14\), 158 (1967) (Abstract in Japanese).
5) Tomita, K., and Nakajima, H.: J. Japan Soc. Blood Transf., \(19\), 33 (1972) (in Japanese with English summary).
6) Furukawa, K., and Takizawa, H.: Jap. Jour. Human Genet., \(18\), 136 (1973) (Abstract in English).
7) Miwa, S., Matsumashi, T., Asai, I., and Seto, S.: Proc. 20th Congr. Japan Soc. Blood Transf., p. 28 (1972) (Abstract in Japanese).
8) Kawase, M., Hara, I., and Mochizuki, N.: Proc. 20th Congr. Japan Soc. Blood Transf., p. 28 (1972) (Abstract in Japanese).
9) Hayashida, Y.: J. Japan Soc. Blood Transf., \(15\), 181 (1968) (Abstract in Japanese).
10) Furukawa, K., and Takizawa, H.: Jap. Jour. Human Genet., \(18\), 136 (1973) (Abstract in English).
11) Yokota, T., Ohno, K., Sawai, H., Itoh, H., and Hamanaka, E.: Proc. 21st Congr. Japan Soc. Blood Transf., p. 2 (1973) (Abstract in Japanese).
12) Yokota, T., Ohno, K., Itoh, H., Hamanaka, E., Kosugi, T., and Nakajima, H.: J. Japan Blood Transf., \(20\), 31 (1974) (Abstract in Japanese).
13) Yamaguchi, H., Okubo, Y., and Tanaka, M.: unpublished data.
14) Honkawa, T.: Personal communication.
15) Sonoda, R.: Personal communication.
16) Furukawa, K., and Kishi, G.: Proc. 22nd Congr. Japan Soc. Blood Transf., p. 28 (1974) (Abstract in Japanese).