173. Rare Blood Type *p* and *P^k* in Japanese Families

By Hideo YAMAGUCHI,^{*)} Yasuto OKUBO,^{**)} Masayoshi TANAKA,^{**)}
Wataru MURAKAMI,^{***)} and Tadashi HONKAWA^{***)}

(Comm. by Ren KIMURA, M. J. A., Nov. 12, 1974)

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-Tj^a (anti-PP₁P^k). 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-Tj^a, Hata and Shimanouchi) nor one anti-P^k(anti-Tj^a, Hata, absorbed by P₁ cells) nor two P^k sera (anti-P, Kinoshita, and Mae., the propositus' serum of the P^k 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.
Tj ^a (Hata)	—	+
Tj ^a (Shimanouchi)	—	+
P ^k (anti-Tj ^a , Hata, absorbed by P ₁ cells)	—	+
Tj ^a , Hata, treated with hydatid cyst fluid	n.t.*	—
P(Kinoshita)	—	—
P(Mae.)	—	n.t.*
P ₁	—	—

* not tested

^{*)} Osaka Red Cross Hospital and Osaka Red Cross Blood Center, Osaka.

^{**)} Osaka Red Cross Blood Center, Osaka, Japan.

^{***)} Hyogo Red Cross Blood Center, Kobe, Japan.

Fig. 1

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 P_1 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 (P^k). The propositus of the P^k 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 P^k 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-Tj^a) and one anti-P^k, but not with one P^k serum (anti-P), one anti-P₁ and one p serum treated with hydatid cyst fluid (Table I). Thus, the propositus was typed as P₂^k.

As shown in Figure 2, the propositus (II-2) was the only member with P^k blood. The parents (I-1, I-2) of the propositus, who

Fig. 2

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_2^k 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 Sanger¹⁾ 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 ₂	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 P^k families Finnish. In Japan, eight p²⁾⁻⁸⁾ and four P^{k 9)-11)} families had been reported including the present two families by the end of 1973. Recently, additional four p¹²⁾⁻¹⁵⁾ and one P^{k 16)} examples followed the present two families. One of them was found to be related to our Miy. family. Earlier cases of p and P^k 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 P^k cases so far reported in Japan were found to occur in consanguineous families.

Acknowledgement. We wish to offer our thanks to Prof. R. Kimura for his revision of this manuscript and to Prof. T. Kikuchi for his constant encouragement. The p and P^k blood samples used here which were obtained from the Japanese who had been registered as the WHO International Panel of Donors of Rare Blood Types were kindly supplied from Prof. S. Miwa and Mr. T. Yokota.

A report of this investigation was read at the 18th Annual Meeting of the Japan Society of Human Genetics (Tokushima, 1973).

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., Matsushashi, 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).