141. A Rare Phenotype Blood Jr(a−) Occurring in Two Successive Generations of a Japanese Family

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The propositus was a 24-year-old female who had an induced labor owing to the intrauterine death of her first fetus which occurred at the 26th week of pregnancy. The death of the fetus was thought to be probably due to the severe omphalocele which was noticed at the delivery. Since the autopsy was not done, there remained another possible cause of death, hemolytic disease in utero. Thus, the serum of the proposita was examined for irregular antibodies.

The serum agglutinated all the group O cell samples tested (30 samples selected at random and all the “panel cells” Ortho), but did not react with one Jr(a−) sample (provided from Yamaguchi Red Cross Blood Center). The antibody was only active by the anti-globulin test with the titer of 1:4.

As was expected, the red cells of the proposita gave negative reactions against 4 samples of anti-Jra. The direct antiglobulin test of the proposita’s cells was negative. These serological findings strongly suggested that the proposita belonged to the Jr(a−) phenotype with anti-Jra in her serum.

As she had never received blood transfusion before, her anti-Jra was supposed to be formed by pregnancy, but the titer of the antibody was so low that it seemed unreasonable to regard this antibody as a cause of the fetus' death.

The blood groups of the proposita and her pedigree members were shown in Table I.

Pedigree studies indicated that the proposita (II-3, Fig. 1) and two sibs, one sister (II-2) and one brother (II-4), both of whom were also Jr(a−). Unexpectedly, the proposita’s mother (I-2) had the Jr(a−) blood. The parents of the proposita were not related, but the natives of the same district (Okayama Prefecture). Two (II-2 and II-3) of the four Jr(a−) members of this pedigree had anti-Jra in

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their sera, whereas the remaining two had no such antibody. The cross-matching tests carried out between the blood samples of these two carriers of anti-Jra gave negative results, as was expected. As the Jr (a-) has been considered to be inherited as a recessive condition, the 4 members of the Jr (a-) (I-2, II-2-4) are assumed to be homozygous for a rare silent allele provisionally called Jr, while the father (I-1) and the 3 children (III-1-3) of the proposita’s sister (II-2) should be heterozygous for, let us say, Jra a common allele responsible for the antigen Jra. Furthermore, the proposita’s father (I-1) who happens to be doubly heterozygous for both Diego and Jr loci (DiaDi, JrJr) as shown in Table I and Figure 1, has one Di (a+) Jr (a-) and two Di (a-) Jr (a-) children. Thus, this family clearly demonstrates that the two systems are inherited independently.

At all events, the present pedigree seems quite unique in that the Jr (a-) condition is transmitted as if it were a dominant trait.
and that there is evidence for separate segregation of the genes responsible for the antigen Jr\textsuperscript{a} and for the Diego system.

**Remarks.** The antigen Jr\textsuperscript{a} was first recognized by Stroup and MacIlroy (1) in 1970 as one of the very frequent antigens. Since then, according to Race and Sanger (2), the following points have been made clear:—1) 7 of the 18 Jr(a−) propositi (who lack the antigen Jr\textsuperscript{a}) tested by the workers of the Ortho Laboratory are Japanese. No random Jr(a−) person has yet been found. 2) Jr(a−) appears to be a straightforward recessive condition. 3) Jr\textsuperscript{a} is shown to be independent of almost all the established blood group loci (no information about Diego). 4) Anti-Jr\textsuperscript{a} is not regularly occurring in Jr(a−) people: it can be stimulated by pregnancy but has not so far been the cause of hemolytic disease of the newborn.

In 1971 the authors (3) tested 464 Osaka donors with the anti-Jr\textsuperscript{a} delivered from the Ortho Laboratory and detected only one negative sample (0.2%). This year the authors (4) found one example in testing 530 random donors (0.19%) with another anti-Jr\textsuperscript{a} sample, whereas Yamada et al. (5) reported from Niigata that 8 of the 460 samples (1.74%) tested were compatible in cross-matching tests carried out with an anti-Jr\textsuperscript{a} from a Jr(a−) patient. The Jr(a−) condition seems less rare in some limited districts of Japan, but its conclusive incidence among Japanese population remains to be proved.

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