28 Er Antigens

28.1 Introduction

Er^a and Er^b, the products of alleles, are high and low frequency antigens, respectively; Er3 is defined by an antibody produced by an Er(a–b–) individual. These three antigens constitute collection 208 of the ISBT terminology, the Er collection: Er^a is ER1 (208001); Er^b is ER2 (208002); and Er3 is ER3 (208003).

28.2 Er antigens

28.2.1 Er^a and Er^b (ER1 and ER2): inheritance and frequencies

Families of two of the original three Er(a–) propositi described by Daniels *et al.* [1] in 1982 showed Er(a–) to be an inherited character. In 1988, Hamilton *et al.* [2] described an antibody to a low frequency antigen, named Er^b, that reacted with five of six Er(a–) red cell samples. In one family the presence of Er(a–) in two generations resulted from an Er(a–b+) × Er(a+b+) mating [2]. Er^a and Er^b, therefore, appear to be inherited regularly as co-dominant alleles. Family studies have shown that Er is not part of the ABO, MNS, P1PK, Duffy, Kidd, or Dombrock systems [1–3].

Er(a–) phenotype has only been found in people of European origin ([1,4–6] and several other unpublished examples), including a Mexican family [7], although an abnormal Er(a–) phenotype was identified in a Japanese family [3] (see below). No Er(a–) individual was found in tests on red cells from 63 762 mostly white [1,4,8] and 13 521 Japanese [3] blood donors.

Four of 605 random white donors were Er(b+) and the frequency of the Er^b allele is calculated as 0.0033 [2]. If the existence of a third allele is disregarded, the Er^a allele has a frequency of 0.9967 and Er(a–) would only be expected in about 1 in 100 000 white people.

Red cells of a Japanese woman and two of her siblings were negative with five anti-Er^a (including the original), but reacted with three others [3]. Positive and negative results were confirmed by adsorption techniques. The serum of the propositus, who had been transfused twice and pregnant three times, contained an antibody that resembled anti-Er^a: it reacted with all cells except Er(a–b+) cells and those of the propositus and two of her siblings.

28.2.2 Er3

Er(a–b–) phenotype has been identified in two unrelated individuals and an Er(a+b–) daughter had an Er(a–b+) mother, suggesting the presence of a third allele [2,9]. One of the Er(a–b–) individuals, a Caucasian man with consanguineous parents, had an antibody, named anti-Er3, that reacted with all red cells tested, including Er(a–) cells [9]. Anti-Er3 reacted with the red cells of the other Er(a–b–) person, suggesting that the man with anti-Er3 may have an Er-null phenotype, whereas the other Er(a–b–) individual could have another active allele that produces neither Er^a nor Er^b.

28.2.3 Antigen characteristics

Er^a is fully expressed on cord cells and is not sensitive to the treatment of red cells with proteases (trypsin, chymotrypsin, papain, ficin, pronase), sialidase, or the

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disulphide bond reducing agent AET. Incubation of red cells in low pH EDTA/glycine buffers, often used in antibody-elution tests, resulted in loss of Er^a and Er3 [9,10]. There was total loss of Er^a at pH 2.0, partial loss at pH 2.5, and no apparent loss at pH 3.0 [10].

Er^b is resistant to treatment of red cells with ficin, papain, or DTT [2].

28.3 Antibodies

All the recorded producers of anti-Er^a have been transfused and/or pregnant [1,3–7,11]. The antibodies are IgG and do not fix complement [1,3,4,7]. In two patients with anti-Er^a, Er(a+) red cells gave a positive DAT after transfusion, but there were no signs of haemolysis [1,4]. The patient with anti-Er3 showed signs suggesting mild haemolysis following transfusion of one unit of incompatible red cells [9]. Monocyte phagocytosis assays and *in vivo* red cell survival studies provided additional evidence that Er^a antibodies are not clinically significant [1,4,7], but that the anti-Er3 was potentially significant [9]. Red cells of three babies born to women with anti-Er^a gave positive DATs, but none had HDFN [1,6,11].

The producers of the only two known anti-Er^b had been pregnant, but not transfused; both had Er(a+b+) husbands [2,12]. Er(b+) red cells from babies of both of the women with anti-Er^b gave strongly positive DATs, but there were no other indications of HDFN.

References

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