Change of blood group from $A_2$ to $A_x$ in a child with congenital abnormalities

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SYNOPSIS  A blood group change from $A_2$ to $A_x$ is described in a boy who had Fallot’s tetralogy, oesophageal atresia, and a tracheo-oesophageal fistula. The change of blood group created a transfusion problem. The unusual serology is discussed.

$A_x$ ($A_4$, $A_0$, etc.) is a rare variant (Race and Sanger, 1968) of the human blood group antigen A. $A_x$ red cells are not agglutinated, or sometimes weakly agglutinated, by group B sera (anti-A) but are strongly agglutinated by group 0 sera (anti-A+B). It is important to use many anti-B and anti-A+B sera to distinguish between $A_2$ and $A_x$ because some anti-B sera react fairly well with $A_x$ cells. Group $A_x$ secretors secrete H but usually no A-substance or, in rare instances, faint traces of it. The serum of $A_x$ people is said usually to contain anti-A\textsubscript{1}.

In most families $A_x$ is controlled by an $A_x$ gene. In others there is evidence of the effect of modifying genes or, more likely, of allelic enhancement (C. Salmon, personal communication). Two families have been reported (van Loghem and van der Hart, 1954; Beckers \textit{et al}, 1955) in which both parents were group 0 and the child $A_x$. In another family an $A_2B$ man transmitted $A_x$ to his daughter (Cahan \textit{et al}, 1957).

We here report a blood transfusion problem which arose in an unusual $A_x$ patient.

Case Report

SURGICAL

P.W. (male, born 27 June 1971) was first admitted to hospital on 27 June 1971. In was his mother’s second pregnancy (first child normal), and the child was delivered at 37 weeks’ gestation. His mother had had hydramnios during pregnancy. A radiograph of the child’s chest at this time showed a Fallot-type heart and, in addition to oesophageal atresia, there was a tracheo-oesophageal fistula. The tracheo-oesophageal fistula was disconnected from the trachea (K.D.R.) on 27 June 1971, and it was decided to carry out a delayed reconstruction of the oesophagus.

The child was again admitted in March 1972 for reconstruction of the oesophagus, in November 1972 for cardiac assessment, and again in October 1973 for corrective surgery for the cardiac lesion. This was carried out on 16 October using cardiopulmonary bypass and a diluted blood prime of the heart/lung machine. The boy’s progress after operation was uneventful and he was discharged home on 25 October 1973, at which time he had full activity and no signs of cardiac failure.

SEROLOGY

P.W.’s mother had been investigated antenatally and was known to be a typical $A_x$; a subsequent family study showed that she had inherited this character from her father (figure). At birth P.W. was a typical $A_2$, his red cells were agglutinated by several examples of both anti-A and anti-A+B sera; they reacted very weakly with the \textit{Dolichos biflorus} lectin. We were not disturbed by this observation because, as stated above, the inheritance of $A_x$ is not always straightforward. Before cardiac surgery, however, P.W., who previously grouped as $A_2$, was found to be a typical $A_x$; his red cells were negative with all of 12 anti-A sera, strongly positive with 12 anti-A+B, and negative with \textit{Dolichos biflorus}. Since antigens of the ABO system are usually weaker at birth than later in life, the change from $A_2$ to $A_x$ is remarkable. Furthermore, his serum was found to react with $A_1$ cells to a titre of 64 and $A_2$ cells to a titre of 16.

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Absorption tests showed that both anti-A and anti-A\sub{1} were present.

**Transfusion Problem**

This case was complicated enough from the purely surgical point of view; the unusual serological features presented an additional problem. Cardiac surgery requires a relatively large amount of blood. Because of strong anti-A antibodies in the patient's plasma, group A blood could not be used. Group 0 whole blood was also contraindicated because group 0 plasma strongly agglutinates group A\sub{x} cells. The heart/lung machine was therefore primed with thrice-washed group 0 cells resuspended in sterile pyrogen-free Hartmann's solution. There were no complications.

**Discussion**

The apparent change of P.W.'s blood group from A\sub{2} to A\sub{x} suggests the possibility that, as he grew older, a conformational change took place on the red cell surface, so that his red cells were no longer agglutinated by anti-A\sub{2} sera. This idea is not without precedent; a conformational change in the erythrocyte surface is believed to explain the change from i to I, which normally occurs at the age of about 18 months. An alternative possibility is somatic mutation with either loss or modification of the blood group gene.

Spivey and Widmann (1974) have reported a similar change, A\sub{2}B to A\sub{x}B, in a 50-year-old woman after a successful operation for carcinoma of the cervix, and refer to two other examples, both associated with malignant disease. There was, however, no evidence of malignancy in our patient. In Spivey and Widmann's patient, a genetic A\sub{2}B changed to A\sub{x}B; our patient, however, appears to have reverted from A\sub{2} to his true genetic group A\sub{x}.

Spivey and Widmann (1974) suggest that the change from A\sub{2}B to A\sub{x}B can be explained by postulating systemic loss of N-acetylgalactosaminyl transferase (in their case A\sub{2}-gene specific transferase). We intend to do transferase studies on our patient and his family when he is older.

Although it is widely held that the antibody usually present in A\sub{x} plasma is anti-A\sub{1}, inspection of the protocols of tests on our A\sub{x} blood donors and on another A\sub{x} person (Dunsford, 1955) show that the antibody is often not only anti-A\sub{1}, that is, an antibody specific for A\sub{1} cells, but also anti-A, an antibody which acts more strongly on A\sub{1} than on A\sub{2} cells. This is also the experience of H. G. Koster (personal communication). The patient of Spivey and Widmann (1974) also had both anti-A and anti-A\sub{1} in her serum. The anti-A antibody in P.W.'s plasma was much stronger than any we have previously seen in an A\sub{x} person.

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**References**


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