



Vac-Pac comfortably moulded to the body contour.

improvement in both general comfort and sleep. The sphere of application for Vac-Pac is probably far wider than just muscular dystrophy.

I would like to thank Mr A Glass for allowing me access to his patients and for helpful advice. The photograph is by the Department of Medical Illustration, North Manchester General Hospital. Vac-Pac is marketed by Howmedica (UK) Limited. Ambu Pumps are available from Oxylitre Limited of Manchester.

(Accepted 18 May 1977)

Booth Hall Hospital, Manchester

M A MORRIS, MB, FRCS, senior registrar in orthopaedics (present appointment: orthopaedic research fellow, Mayo Clinic, Rochester, Minnesota, USA)

Autoimmune haemolytic anaemia with anti-A autoantibody

In many cases of autoimmune haemolytic anaemia the target antigen corresponds to an antigenic entity defined by isoantibodies that have blood group specificity. Usually the Rh-complex is implicated,^{1,2} here we report a case in which the autoantibody showed specificity for A₁ and A₂ erythrocytes.

Case report

A 2-year-old girl was referred because of persistent listlessness after an upper respiratory tract infection. Examination showed pallor, jaundice, and hepatosplenomegaly. Results of investigations were: haemoglobin 4.1 g/dl; red cell count $1720 \times 10^9/l$ ($1\ 720\ 000/mm^3$); white cell count $5.2 \times 10^9/l$ (normal differential); platelet count $120 \times 10^9/l$; erythrocyte sedimentation rate 150 mm in first hour; reticulocytes 2.7%; 6 nucleated red cells/100 white cells; pronounced rouleaux formation; normoblastic hyperplasia in bone marrow; haemosiderinuria and methaemalbuminaemia detected; serum bilirubin concentration $58\ \mu mol/l$ ($3.4\ mg/100\ ml$); no evidence of intracorpular erythrocytic defects on screening; viral screen normal; IgG 96 (normal 43-142) U/ml, IgM 400 (47-220) U/ml, IgA 50 (18-115) U/ml, no M component.

Initial progress with steroid treatment was satisfactory, and a mild relapse after one month responded to a small increase in prednisolone.

On referral a direct antiglobulin test (DAGT) result was positive (titre 1/128) due to complement binding. Incubation produced lysis of the patient's and donor A₁ erythrocytes due to antibodies in the patient's serum. Tests with control serum gave negative results. The antibodies in the patient's serum showed specificity for A₁ and A₂ cells (see table). The patient was group A₁ and remained so after clinical remission. The specificity of the antibody for the A antigen was confirmed when absorption of the patient's serum with A₁ secretor saliva removed the autoanti-A antibodies and left the anti-B antibodies. Furthermore, the patient's erythrocyte eluate contained anti-A antibodies but no antibodies against B or O cells. Two factors suggested that the autoantibody was IgM—namely, incubation of the serum with dithiothreitol removed both the anti-B and autoanti-A antibodies, and Sephadex filtration showed maximal antibody activity in the 19S fraction.

The autoantibodies disappeared for a short time with treatment, but were present later when the DAGT result was positive. By then the autoantibodies had changed in reactivity, binding to group O and fetal cells in addition to group A erythrocytes. An erythrocyte eluate, however, showed activity against A₁ and A₂ cells but not against group O cells.

Comment

There has been one report³ of autoanti-A antibodies causing intravascular haemolysis, but their specificity has been questioned.⁴ The autoantibodies in our patient did not show any cross-reactivity with either group O or cord erythrocytes at the time of referral. The antiglobulin test result, though positive with specific anticomplement sera, was unexpectedly negative with anti-IgM sera. This may be explained either by a weak antiserum having been used or by the fact that IgM on the erythrocyte was masked by C3.

The antibody was IgM but was consistent with the type of blood group specificity shown, since naturally occurring anti-A and anti-B antibodies are of the IgM subclass. The lytic nature of the antibody shown in vitro also supports this conclusion and may explain the presence of the intravascular haemolysis, as evidenced by urinary haemosiderin and methaemalbuminaemia.

At follow-up the autoantibody that was initially specific for A₁ and A₂ appeared to be broadening in its reaction pattern. Whether this change was due to treatment or part of the natural evolution of the antibody type in this patient is unknown. Our findings, however, support those of the other reported case by showing that a major blood group antigen, group A, may be the target antigen in human autoimmune haemolytic anaemia.

We thank Miss E Manson for her technical expertise; Dr K James and Miss J Wallace for fractionating the serum; Dr J W Farquhar for allowing us to report this case; and Miss K Denholm for typing the manuscript.

¹ Hughes-Jones, H C, Polley, M J, and Telford, R, *Immunology*, 1964, **7**, 72.

² Vos, G H, Petz, C D, and Fudenberg, H H, *Journal of Immunology*, 1971, **106**, 1172.

³ Szymanski, I O, Roberts, P L, and Rosenfield, R E, *New England Journal of Medicine*, 1976, **294**, 995.

⁴ Stewart, K, *New England Journal of Medicine*, 1976, **295**, 281.

(Accepted 2 September 1977)

Department of Haematology, Royal Infirmary, Edinburgh EH3 9YW
A C PARKER, MRCP, MRCPATH, lecturer

Blood Transfusion Centre, Royal Infirmary, Edinburgh EH3 9YW
G WILLIS, AIMLS, technician
S J URBANIAK, BSC, MRCP, consultant

Royal Hospital for Sick Children, Edinburgh EH9 1LF
E M INNES, FRCP, consultant

Results of standard antibody screening of patient's serum and erythrocyte eluate on referral. Specificity for group A cells shown

	Presence in patient's serum of antibodies against:					Presence in erythrocyte eluate of antibodies against:				
	O erythrocytes (six separate donors)	Fetal cells	A ₁	A ₂	B	O erythrocytes (two separate donors)	Fetal cells	A ₁	A ₂	B
Saline (at 4°C)	-	-	+	+	+++	-	-	++	++	-
Saline (at room temperature)	-	-	±	±	+	-	-	++	++	-
Saline (at 37°C)	-	-	±	±	±	-	-	+	+	-
Enzyme (at room temperature)	-	-	-	-	±	-	-	++	++	-
Enzyme (at 37°C)	-	-	-	-	±	-	-	++	++	-
IAGT result	-	-	+	+	++	-	-	±	±	-

IAGT = Indirect antiglobulin test.