Cardiopulmonary bypass in hereditary spherocytosis: a case report

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A cardiopulmonary bypass procedure in a patient with hereditary spherocytosis is described. Details of *in vitro* tests carried out prior to surgery are given.

Hereditary spherocytosis is a genetically determined abnormality of red blood cells. It is likely that a defect of intracellular glycolysis results in the formation of spherical cells (spherocytes) which have a reduced life span and exhibit increased osmotic fragility in hypotonic saline solutions, increased susceptibility to mechanical trauma, and spontaneous lysis on incubation. Lysis is markedly reduced by the presence of added glucose (Dacie, 1963). Splenectomy usually results in clinical cure, although haematological signs of the disease may persist; for example, while osmotic fragility improves, it rarely returns to normal, and mechanical fragility of both fresh and incubated blood remains significantly abnormal.

The imposition of cardiopulmonary bypass on normal subjects is associated with some destruction of the formed elements of blood. Haemolysis occurs, and plasma haemoglobin levels rise at a rate which is seldom less than 0.25 mg per minute, consequent upon the trauma of bubble oxygenation, roller pumps and cardiotomy suction. Haemolysis of this degree is seldom important as the kidney is well able to tolerate plasma haemoglobin levels of up to 250 mg per 100 ml, and oxygen carrying capacity is not seriously deranged. However, the altered robustness of the red cells in a patient with hereditary spherocytosis may preclude an operation with cardiopulmonary bypass on the ground of excessive haemolysis. There are no available published reports of an operation with cardiopulmonary bypass in a patient with hereditary spherocytosis. We report a reconstructive operation on the mitral valve in such a patient, with particular reference to the conduct of cardiopulmonary bypass.

CASE REPORT

An African girl aged 16 years presented in early 1968 with haemolytic anaemia. On the basis of the micro-

scopic appearance of a blood smear, characteristic osmotic fragility and a negative response to the direct antiglobulin test (Coombs) a diagnosis of spherocytosis was made. Splenectomy was undertaken late in 1968 with relief of the clinical signs and symptoms.

The patient was re-admitted a year later with a history of increasing dyspnoea, in cardiac failure, and with the signs of mitral regurgitation. The form of management recommended was mitral valve repair. Haematological investigation at this time showed the haemoglobin to be 13 g%, PCV 36%, MCH 36%, WBC 10,000 per c.mm, and reticulocytes 0.5%. A blood smear showed evidence of haemolytic anaemia; serum bilirubin was 0.6%; osmotic fragility of the red blood cells in serial dilutions of saline showed fragility little different from that before splenectomy.

In view of the uncertain tolerance of the abnormal red cell corpuscles in hereditary spherocytosis to cardiopulmonary bypass, it was thought necessary to assess the response of a sample of the patient's blood to an experiment designed to simulate those aspects of cardiopulmonary bypass most damaging to blood. In the unit to which she was admitted bubble oxygenators of the disposable Travenol variety are used. The size of the bag selected allows an output of arterialized blood calculated on the basis of 2.5 litres per square metre of body surface area. Pumps are adjusted so that both rollers are just short of occlusive. The priming volume of Travenol bags varies with size, but, in general, when primed with a liquid other than blood, priming volume approximates calculated blood volume and, consequently, during bypass a 50% reduction in the haematocrit is expected.

In the experimental bypass a 'one-litre flow' Travenol bag primed with 450 ml of Ringer lactate and 30 mg of heparin was used. The circuit was similar to that which would be employed for the intended surgical operation. Priming fluid was drawn from the helix reservoir of the bag by the 'arterial' roller pump and circulated through a standard Sarnes heat exchange unit maintained at 37° C. The fluid was then pumped through a narrow metal cannula with an internal bore considered close to the size of the femoral arterial cannula which would be used for the ultimate operative procedure. The circuit was

TABLE

Red Cell Fragility		
% of Saline	% Haemolysis	
	Control	Test
0.9	0	0
0.75	0	0
0.65	0	0
0.6	0	4
0.55	0	25
0.5	1	75
0.45	7	87
0.4	20	92
0.35	43	95
0.3	70	97
0.2	100	100
0.1	100	100

completed by connection of the metal cannula to the venous inlet of the oxygenator column. Arterialization of the priming fluid was achieved by a 3 litres per minute flow of a gas mixture of 97% oxygenation and 3% carbon dioxide. A 450-ml aliquot of the patient's blood was freshly drawn into a heparinized container and added to the cardiopulmonary bypass circuit. Fluid in the circuit was thereby subjected to conditions which closely simulated those which would obtain, in terms of mechanical trauma, oxygenation, dilution, acid:base homeostasis and temperature, during the planned operation. Fluid from the pump was sampled every 5 minutes and investigations were made relevant to haemolytic anaemia, blood gas analysis, acid:base state, full blood count, free haemoglobin and electrolytes.

The results were compared with those obtained from a fresh specimen of the patient's blood, which acted as a control. Important destruction of blood did not occur during the study; in particular, the plasma haemoglobin level fell from 44 mg% to 31

mg% following addition of blood to the bypass machine, and remained at this level throughout the experiment. It was therefore considered safe to proceed with the planned procedure, during which a 6 litre flow Travenol bag primed with 2 litres of Ringer's lactate with 30 mg of heparin added, was used. The arterial pump was adjusted during bypass to deliver 3.6 litres per minute (i.e., at a rate of 2.5 litres per square metre of the patient's body surface area). During bypass the series of tests performed in the simulated bypass experiment were repeated, and again the abnormal red blood cells appeared to remain intact. Free plasma haemoglobin rose unimportantly from 24 mg% before bypass to 36 mg% after bypass, with return to previous levels 5 hours after bypass. At operation the cardiological findings were confirmed and competence of the valve was restored by plication of the mitral annulus. Convalescence was uneventful and the patient was discharged from hospital on the twenty-first post-operative day.

Samples of blood taken 12 weeks after operation showed the haemoglobin to be 12.5 g%, packed cell volume 39%, MCHC 32%, and plasma haemoglobin 34 mg%. The red cell fragility investigations undertaken at this time are appended in the Table.

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REFERENCE

Dacie, J. V. (1963). *The Haemolytic Anaemias*, 2nd ed. Churchill, London.