Correction of Fallot's Tetralogy in a Patient Suffering from Hereditary Spherocytosis

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SUMMARY

The use of cardiopulmonary bypass in a patient with Fallot's tetralogy and hereditary spherocytosis is described. A method of evaluating the fragility of the red cells is given.

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Hereditary spherocytosis is a genetically determined abnormality of the red blood cells. A defect in intracellular glycolysis results in a spherical cell which has a shorter life span than normal and exhibits increased osmotic fragility in hypotonic saline solutions. Lysis is reduced in glucose solutions.[1] Normal formed elements in blood are prone to destruction during cardiopulmonary bypass, due to trauma, cardiotomy suction, bubble oxygenation and roller pumping.

The problem of the imposition of cardiopulmonary bypass on patients suffering from excessive fragility of their red cells gives rise to concern for the safe conduct of such a bypass.[2] Haemolysis normally occurs at a rate which is seldom less than 0,25 mg of free haemoglobin per minute, but renal function is usually able to tolerate plasma haemoglobin levels of up to 250 mg/100 ml. The oxygen-carrying capacity is not seriously deranged.

A case report is presented on the conduct of cardiopulmonary bypass in a patient suffering from hereditary spherocytosis.

CASE REPORT

An Indian boy, aged 5 years, presented with Fallot's tetralogy and a typical history of tiring on exertion and squatting. He was cyanosed. The clinical diagnosis was confirmed by cardiac catheterisation. This showed marked infundibular narrowing of the right ventricle, with a gradient of 67 mmHg; a patent foramen ovale; a right-to-left shunt of 56% with a satisfactory pulmonary artery. The pulmonary vascular resistance to the systemic vascular resistance ratio was calculated at 11%. The child's father and 4-year-old sibling were known cases of hereditary spherocytosis. The diagnosis of hereditary spherocytosis was established by the microscopic appearance of the blood smear, characteristic osmotic fragility and a negative response to direct antiglobulin tests. Splenectomy had not been undertaken in this child. The serum bilirubin was elevated at 3,8% and the reticulocyte count ranged at about 3%.

In view of possibly excess fragility of the blood, a 'mock' bypass was set up, using an infants' Temptrol oxygenator, primed with 200 ml of Ringer's lactate and heparinised with 1 500 units of heparin; 125 ml of the patient's blood was extracted into a donor bag containing 1 ml heparin and the mixture was circulated through a circuit similar to that to be used for the surgical operation. The priming fluid was drawn from the helix reservoir of the bag by the 'arterial' roller pump, and pumped via a 6-mm diameter tubing back into the 'venous' side of the oxygenator. Arterialisation of the priming fluid was achieved by 3-litres flow of 97% oxygen in 3% carbon dioxide mixture. Samples were taken every five minutes, and haemoglobin, packed cell volume, platelets and electrolytes, sodium, potassium and chloride were estimated. The osmotic fragility was estimated at the end of the procedure. Cardiotomy was simulated by increasing the pump speed in combination with a low fluid level, allowing an air/blood mixture to be sucked through the pump during the last 30 minutes of the 'mock' procedure.

The results (Table I) show that there was no important blood destruction and that the plasma haemoglobin rose from 6 to 64 mg/100 ml. Osmotic fragility was not altered at the conclusion of the procedure.

It was decided to proceed with the planned operation. A 3-litre Temptrol oxygenator was primed with 1 litre Ringer's lactate and 500 ml donor blood. The arterial pump was adjusted to deliver 2,5 litres/m²/min of body surface area—in this case 1,8 litres/min. The series of tests performed in the simulated bypass were repeated and the abnormal red cells appeared to remain intact. Free plasma haemoglobin rose from 12 before to 45 mg/100 ml after bypass, the level at 6 hours after bypass being 18 mg/100 ml.

The operation confirmed the cardiological findings, with the exception of the patent foramen ovale, which was not found. The ventricular septal defect was closed by routine patch closure and the infundibulum of the right ventricle opened by infundibulectomy. The postoperative course of the patient was uneventful and he was discharged from hospital on the 20th postoperative day.

After 6 months a sample of blood showed the haemoglobin to be 10,9 g/100 ml with a reticulocyte count of 3%, an excessive fragility of the red cells still being present.

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TABLE I. RESULTS (MCHC IN ALL SPECIMENS ± 40%)

Specimen No.	Time on bypass (in min)	Hb (mg/100 ml)	PCV (%)	Platelets	Na (mEq/L)	K (mEq/L)	Cl (mEq/L)	Plasma Hb (mg/100 ml)
1	Prime	4,4	10	42 000	137	4,5	111	6
2	5	3,9	10	—	—	—	—	7
3	10	3,7	10	—	—	—	—	8
4	15	3,7	10	—	131	4,7	116	11
5	30	3,5	10	23 000	—	—	—	14
6	45	3,7	10	—	131	4,85	118	24
7	60	3,7	10	25 000	129	4,85	118	34
8	75	4,1	10	24 000	—	—	—	47
9	90	3,7	10	26 000	135	5,1	118	64

CONCLUSION

Cardiopulmonary bypass appeared to be well tolerated in this patient with hereditary spherocytosis. The performance of a 'mock' bypass with an aliquot of the patient's blood, gave a good indication of damage to the red cells.

REFERENCES

1. Dacie, J. V. (1963): *The Haemolytic Anaemias*. 2nd ed. London: J. & A. Churchill.
2. Moyes, D. G., Rogers, M. A. and Coleman, A. J. (1971): Thorax, **26**, 131.