Anaesthesia, 1984, Volume 39, pages 1121–1123

CASE REPORT

Hypothermic cardiopulmonary bypass in a patient with sickle-cell trait

M. A. FOX AND T. R. ABBOTT

Summary

A male patient, aged 7.5 years, with known sickle cell trait, presented for correction of Fallot's tetralogy. The successful management under hypothermic cardiopulmonary bypass is described and the behaviour of sickle haemoglobin under hypothermic conditions is discussed.

Key words

Surgery, cardiac; cardiopulmonary bypass, hypothermia.
Complications; sickle cell trait.

Hypothermia is frequently mentioned as one of the factors to be avoided when anaesthetising patients suffering from sickle-cell disease or sickle trait.[1–4]

During the surgical correction of cardiac defects, it is desirable to employ hypothermia for both myocardial and cerebral protection. There are, in the literature, examples both of the uneventful use of this technique,[5–6] and of the use of hypothermia in patients with sickle-cell trait causing profound sickling.[7]

Case history

The patient was a Nigerian male, resident in this country for the preceding year, having spent the rest of his life in Nigeria. At the age of 6 months he presented with failure to thrive and recurrent chest infections. A systolic murmur was noted and a clinical diagnosis of a ventricular septal defect (VSD) was made and treatment started with digoxin, which was continued for the first 5 years of life. In Nigeria the patient received antimalarial prophylaxis.

On admission, aged 7 years 6 months and weighing 23 kg, the patient was not in heart failure and was not receiving any medication. Angiography was compatible with Fallot's tetralogy and haematological investigation revealed a haemoglobin concentration 11.4 g/dl with an electrophoresis showing HbS 40% and HbA 60%.

Surgical correction of Fallot's tetralogy was performed. After premedication with trimeprazine 30 mg, morphine 6 mg and atropine 0.6 mg, induction of anaesthesia was achieved with intravenous thiopentone (100 mg) and tracheal intubation facilitated with tubocurarine 23 mg. Anaesthesia was maintained with morphine 6 mg, lorazepam 0.6 mg, 50% oxygen and 50% nitrous oxide mixture, and lung ventilation was controlled. An intravenous route was established and monitoring lines including central venous and arterial pressures, and oesophageal and naso-

M.A. Fox, MB, BS, FFARCS, Registrar, T.R. Abbott, MB, ChB, FFARCS, Consultant Anaesthetist, Royal Liverpool Children's Hospital, Myrtle Street, Liverpool L7 7DG.

0003–2409/84/111121 + 03 $03.00/0

Table 1. Results of blood gas analysis

Time (mins)	pH	$Paco_2$ (kPa)	Hco_2	Base excess	Pao_2 (kPa)	T(C)	HbS (%)	PCV
Prebypass	7.48	3.35	23.2	−1.7	38.5	37	40	34
On bypass +10	7.36	5.1	22.8	−2.8	41.2	35	17	33
+30	7.32	5.4	21.9	−4.0	51.9	26	17	33
+50	7.33	5.1	21.6	−4.4	67.2	26	17	33
+80	7.38	4.8	23.2	−2.3	60.8	30	17	33
Post-bypass	7.33	6.0	23.9	−1.4	38.6	37	10	40
First postoperative day	7.35	4.8	21.	−5.0	10.4	37	10	40

pharyngeal temperatures were inserted.

Once surgery had commenced and cardiovascular stability had been established the patient was vasodilated with phenoxybenzamine 20 mg, the circulatory blood volume was expanded by infusing 400 ml crystalloid and 100 ml blood.

Cardiopulmonary bypass was established using a roller pump and a bubble oxygenator which was primed with 4 units of heparinised CPD blood, 400 ml 4% dextrose 0.18% saline and 37 ml 8.4% sodium bicarbonate.

Cardiopulmonary bypass time was 1 hour 34 minutes during which the patient was cooled to 26°C. The aorta was cross clamped for a period of 49 minutes, the heart having previously been perfused with asanguinous cold cardioplegia (30 ml/kg).

The stenotic muscle bands were excised and the VSD repaired with a Dacron patch. Weaning from cardiopulmonary bypass was achieved without difficulty, the residual blood in the oxygenator was discarded and subsequent transfusion was with CPD blood. The patient required 36 hours of postoperative ventilation and then went on to make an uneventful recovery.

Blood was withdrawn at regular intervals for estimation of haemoglobin, presence of any sickle cells and blood gases. The results are summarised in Table 1. At no time was any sickling detected. The fall in the percentage of HbS may be accounted for by dilution with blood from the pump prime and subsequent transfusion.

Discussion

Formation of sickle haemoglobin can only occur in the deoxygenated form and therefore adequate oxygenation is of paramount importance. In spite of adequate arterial oxygenation, there will inevitably be some deoxygenation occurring in the capillary beds.

In vitro, the solubility of deoxygenated sickle haemoglobin increases as the temperature decreases, making the formation of a gel and subsequent sickling less likely as blood is cooled.[8] From the formation of deoxygenated haemoglobin to the onset of aggregation of the deoxygenated haemoglobin into a gel there is a delay time, which is inversely proportional to temperature.[9] The presence of significant amounts of polymerised sickle haemoglobin does not invariably produce a sickle cell; as the cell returns to a less hypoxic environment there is reversal of the polymerisation.[10]

It has been postulated by Eaton *et al.*[11] that the probability of sickling inside capillaries is determined by the delay time of gelation and that sickling occurs when the delay times are shortened enough to be less than the capillary transit times or conversely, capillary transit time is lengthened to exceed delay time.

This would suggest that hypothermia should to some extent have a protective action in preventing sickling as the red blood cells pass through an hypoxic environment provided care is taken to keep the capillary transit time as short as possible.

Consequently, hypothermia is not harmful in itself to patients with sickle-cell disease, but it is the subsequent vascular stasis associated with peripheral vasoconstriction which precipitates sickling.

It would seem that hypothermic techniques can reasonably be used in patients with sickle cell trait provided care is taken to ensure adequate peripheral perfusion. This would involve the administration of a vasodilating agent, phenoxybenzamine in our patient's case, and the control of the packed cell volume to reduce blood viscosity and therefore optimise oxygen delivery. These considerations need to be continued into the postoperative period until cardiovascular and

temperature homeostasis has been restored.

Many centres use a blood prime for paediatric cardiac surgery. In the case of a patient with sickle cell trait this has the advantage of diluting those erythrocytes at risk of sickling under conditions of low oxygen tension. In adult cardiopulmonary bypass it is usual to allow some degree of haemodilution to occur and although this decreases oxygen carrying capacity, it also decreases viscosity which improves tissue perfusion.[12] From the limited number of case reports in the literature adults with sickle cell trait do not seem to tolerate this decrease in oxygen carrying capacity which accompanies haemodilution.[7,13] When this sudden haemodilution is avoided by using a blood containing prime the cardiopulmonary bypass does not seem to be associated with an episode of sickling.[14]

Whether the patient has pre-operative exchange transfusion or the concentration of sickle haemoglobin is reduced by dilution whilst going onto cardiopulmonary bypass, there will still be a small but nevertheless significant mass of sickle haemoglobin which may sickle in an hypoxic environment. From our experience of this case and a review of the literature it would appear that this can be prevented by maintaining arterial oxygen tensions and adequate capillary perfusion with short capillary transit times during the per operative and postoperative periods.

References

1. Fullerton MW, Philippart AI, Sarnaik S, Lusher JM. Preoperative exchange transfusion in sickle cell anemia *Journal of Pediatric Surgery* 1981; **16:** 297–300.
2. Janik J. Seeler RA. Peri-operative management of children with sickle haemoglobinopathy. *Journal of Pediatric Surgery* 1980; **15:** 117–20.
3. Hudson I, Davidson IA, McGregor CGA. Mitral valve replacement using cold cardioplegia in a patient with sickle cell trait. *Thorax* 1981; **36:** 151–2.
4. Szentpetery S, Roberston L, Lower RR. Complete repair of tetralogy associated with sickle cell anemia and G-6-PD deficiency. *Journal of Thoracic and Cardiovascular Surgery* 1976; **72:** 276–9.
5. Somanathan S. Anaesthesia and hypothermia in sickle cell disease. *Anaesthesia* 1976; **31:** 113.
6. Ingram CT, Floyd JB, Santora AH. Aortic and mitral valve replacement after sickle cell crisis. *Anesthesia and Analgesia* 1982; **61:** 802.
7. Heiner M, Teasdale SJ, David T, Scott AA, Glynn MFX. Aorto-coronary bypass in a patient with sickle cell trait. *Canadian Anaesthetists' Society Journal* 1979; **26:** 428–34.
8. Noguchi CT, Schechter AW. The intracellular polymerization of sickle hemoglobin and its relevance to sickle cell disease. *Blood* 1981; **58:** 1057–68.
9. Malfa R, Steinhardt J. A temperature-dependent latent-period in the aggregation of sickle-cell deoxyhaemoglobin. *Biochemical and Biophysical Research Communications* 1974; **59:** 887–93.
10. Serjeant GR, Serjeant BE, Milner PF. The irreversibly sickled cell; determinant of haemolysis in sickle cell anaemia. *British Journal of Haematology* 1969; **17:** 527–33.
11. Eaton WA, Hofrichter J, Ross PD. Delay time of gelation: a possible determinant of clinical severity in sickle cell disease. *Blood* 1978; **47:** 621–7.
12. Branthwaite MA. *Anaesthesia for cardiac surgery and allied procedures* 2nd ed. Oxford: Blackwell Scientific Publications, 1980.
13. Leachman RD, Miller WT, Atias IM. Sickle cell trait complicated by sickle cell thrombi after open-heart surgery. *American Heart Journal* 1967; **74:** 268–70.
14. Yacoub MH, Baron J, Et-Etr A, Kittle CF. Aortic homograft replacement of the mitral valve in sickle cell trait. *Journal of Thoracic and Cardiovascular Surgery* 1970; **59:** 568–73.