

# Mitral valve replacement using cold cardioplegia in a patient with sickle cell trait

I HUDSON, I A DAVIDSON, AND C G A MCGREGOR

From the Department of Cardiac Surgery, Royal Infirmary, Edinburgh

## Case reports

A 13-year-old negro girl from Sierra Leone was referred for mitral valve replacement. Her weight was 40 kg and she had a history of progressive exertional dyspnoea and paroxysmal nocturnal dyspnoea. Medication consisted of digoxin 0.25 mg/day, frusemide 40 mg/day, and potassium supplements.

At cardiac catheterisation the mean simultaneous mitral valve gradient was 17 mmHg with a cardiac index of 2.8 l/m<sup>2</sup>/min, and left ventriculography showed good left ventricular function with gross reflux into an enlarged left atrium. The haemoglobin was 10.8 g/dl.

The presence of sickling had been noted on a blood film. The SICKLEDEX test was positive and electrophoretic examination of the haemoglobin revealed one band in the A and one band in the S position. The haemoglobin F fraction was 1.3%. Quantitative estimation of haemoglobin S was not available.

Mitral valve replacement was performed on 13 September 1978. Before the institution of cardiopulmonary bypass 1400 ml of fresh whole blood was exchange-transfused with the addition of 50 ml 8.4% sodium bicarbonate solution. Bypass flow rate was 2.4 l/m<sup>2</sup>/min using a disposable Harvey oxygenator. The initial haematocrit on bypass was 19%. The patient was not systemically cooled but the temperature was allowed to fall to 33°C. The aorta was cross-clamped and 500 ml of cold (4°C) solution of dextrose 5% in 0.9% saline containing 10 mmol potassium chloride was infused into the aortic root. Topical hypothermia was achieved by pouring cold (4°C) normal saline over the heart and by continuous pericardial irrigation.

There was no clot in the left atrium. The grossly abnormal mitral valve was excised, and replaced by a 31 mm Carpentier-Edwards porcine prosthesis. The patient was rewarmed and the heart beat in sinus rhythm after one direct current shock (10 joules). The heart maintained a good output without inotropic support after the discontinuation of cardiopulmonary bypass. The total bypass time was 65 minutes and the aortic cross-clamp time 43 minutes.

Creatine kinase MB isoenzyme, total creatine kinase, urea, stable lactate dehydrogenase, aspartate transaminase, alanine aminotransferase, and plasma

Table Results of plasma haemoglobin and CKMB estimations

Time	Plasma Hb g/l	CK-MB μl	% CKMB of total CK μl
Preoperative	0.002	< 10	
Prebypass	0.117	7	7
End of bypass	0.048	16	9
3 hours	0.18	24	5
6 hours	0.08	20	4
9 hours	0.1	15	3
12 hours	0.08	12	1
24 hours	0.076	17	1

haemoglobin were measured before operation and at 3, 6, 9, 12, and 24 hours after cardiopulmonary bypass. Electrocardiograms were obtained before operation and daily afterwards for one week. The table shows the results of the plasma Hb and CKMB estimations.

Recovery was uneventful and the patient was discharged on the thirteenth day after operation. There were no electrocardiographic changes indicative of operative myocardial injury.

## Discussion

In 1967 Leachman *et al*<sup>1</sup> reported a case of systemic sickle cell thrombi in the lungs, kidneys, heart and brain in a patient with sickle cell trait complicating aortic valve replacement and resulting in the patient's death. Since then, several authors have reported successful cardiopulmonary bypass in patients with sickle cell disease.<sup>2-4</sup> These writers have stressed the need for close monitoring of pO<sub>2</sub>, pH, and temperature to avoid hypoxia, acidosis, and hypothermia, factors known to precipitate the "vicious circle" of vaso-occlusion and irreversible capillary obstruction resulting in tissue injury.

The use of tissue valves has been advocated to avoid the increased blood trauma caused by mechanical prostheses but Craenen inserted a Starr-Edwards mitral prosthesis in a child with sickle cell anaemia and noted no increased haemolysis. Wenham *et al*<sup>5</sup> showed no significant haemolysis in a patient with sickle cell trait after aortic valve replacement with a Björk-Shiley prosthesis. Szentpetery felt on a theoretical basis that topical hypothermia was contraindicated in patients with sickle cell disease and that local sickling and myocardial injury might result.<sup>4</sup>

Address for reprint requests: Mr CGA McGregor, Department of Clinical Surgery, Royal Infirmary, Edinburgh EH3 9YW.

In the present case there is no evidence either clinically, electrocardiographically, or enzymatically that significant myocardial injury occurred. Levels of CKMB of  $<25 \mu\text{l}$  found in our patient indicate that no significant myocardial injury had occurred. The non-specific enzymes were likewise not significantly increased. Plasma free haemoglobin levels were no higher than would be expected during the course of normal cardiopulmonary bypass.

We therefore conclude that potassium-induced cardioplegia and topical hypothermia in conjunction with pre-bypass exchange transfusion and haemodilution can be safely used during cardiopulmonary bypass in patients with the sickle cell trait.

#### Post scriptum

A second patient with sickle cell trait has undergone valve replacement in this hospital using the techniques of myocardial protection described in this paper. There was no evidence of any myocardial injury using enzyme and isoenzyme estimation and the patient had a negative postoperative technetium pyrophosphate scan. There were likewise no changes

*I Hudson, I A Davidson, and C G A McGregor*

indicative of myocardial injury in the electrocardiogram over the operative period.

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#### References

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