

found post mortem. The presence of a higher end-diastolic pressure in the left ventricle in our patient is very suggestive of myocardial restriction.⁴ Late onset pericardial constriction has been reported only in one case,⁵ and, like the case reported here, pericardiectomy led to improvement.

This case report shows that the absence of clinical findings within five years does not preclude the later development of significant radiation-induced heart disease.

We thank Dr W Whitaker for permission to report this case, and Mr M I Ionescu, who performed the pericardiectomy.

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Cardiac arrhythmias in thalassaemia major: evaluation of chelation treatment using ambulatory monitoring

Despite recent advances in treatment, thalassaemia major remains incurable. With repeated blood transfusion haemochromatosis develops, and death usually occurs from cardiac failure during the second decade.¹ Chelation treatment with desferrioxamine has been used to promote urinary excretion of iron. Hepatic iron accumulation is thereby reduced,² but its effectiveness in lowering cardiac iron deposition remains unproved. In this study ambulatory monitoring has provided data on the early development of cardiac arrhythmias in thalassaemia major, and their relation to chelation treatment.

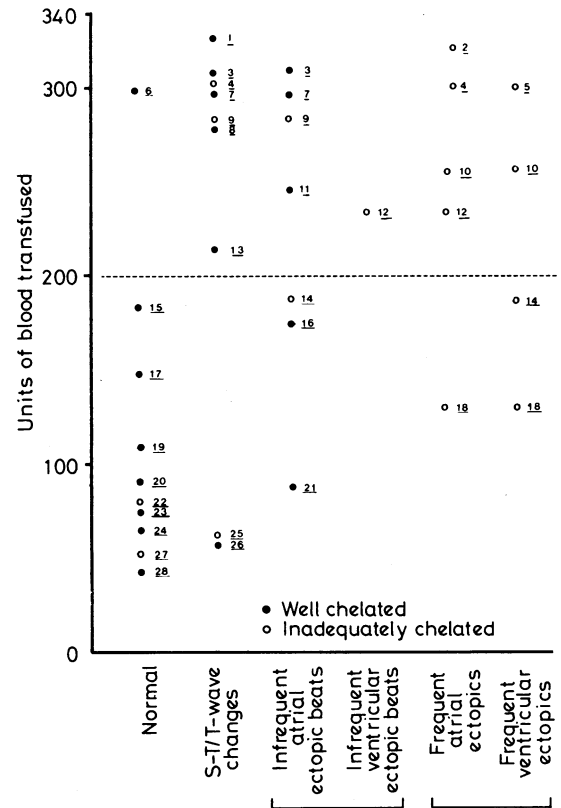
Patients, methods, and results

Twenty-eight patients with thalassaemia major, aged between 7 and 23 years, were treated on a high-transfusion regimen (total of 42-330 units). The lowest haemoglobin concentration at the time of recording was 10.4 g/dl. No patients showed signs or symptoms of cardiac failure, and only three had abnormalities on conventional electrocardiography. Seventeen patients were well chelated—that is, they had received desferrioxamine, 500 mg intramuscularly daily for at least two years. The remaining 11 patients received desferrioxamine very irregularly or not at all. Use of chelation treatment did not reflect the original clinical state of the patients.

The patients were studied by Holter 24-hour recordings, the Oxford Medilog recorder and replay system being used. Each patient was fully ambulant and asymptomatic during recording. Tapes were analysed with particular care to eliminate possible error induced by artefact.³

Four patterns were identified: (a) normal; (b) ST segment depression, sometimes associated with T-wave inversion, occurring independently of heart rate and exercise; (c) infrequent atrial or ventricular extrasystoles—that is, less than five in any one minute of record; and (d) frequent atrial or ventricular extrasystoles—that is, five or more in any one minute (including one example of episodic idioventricular rhythm). Eighteen patients had abnormal recordings, and eight of these showed two abnormalities (see figure).

Infrequent extrasystoles were noted in patients who had received as little as 90 units of blood. ST segment and T-wave changes were seen in two patients who had received 60-70 units and were also often noted in well-chelated patients with higher transfusion loads. Major arrhythmias (group (d)) occurred in seven of 11 inadequately chelated patients but in no well-chelated patients. This apparent benefit of chelation is particularly evident in the subgroup of 13 patients at greatest risk of developing cardiac iron toxicity—that is, those having received over 200 units of blood. These thirteen patients had similar clinical histories. The mean age of well-chelated



Results of ambulatory cardiac monitoring in 28 patients with thalassaemia major.

patients was 16.1 years (range 13-19) and of inadequately chelated patients 17.6 (range 15-23). The mean transfusion loads for the two groups were identical at 283 units (ranges 217-300 and 233-327). Four of six inadequately chelated patients showed major arrhythmias, while none were seen in seven well-chelated patients ($P < 0.001$).

Comment

Engle noted that cardiac arrhythmias in thalassaemia major were usually associated with terminal heart failure.⁴ Ambulatory monitoring provides a new means of investigating these patients, and it is apparent that arrhythmias occur more often and earlier than was recognised.

The correlation between abnormal electrocardiograms and cardiac iron deposition has been established by necropsy studies.⁵ In addition to atrial tachycardia, ST segment and T-wave changes were often observed, and these probably indicated early pathological change. With this technique, our results suggest that chelation treatment offers some protection against cardiac iron deposition. The investigation may find wider application in assessing children at risk at an early stage and in evaluating modified chelation regimens.

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