Haemoglobin levels and blood requirement in thalassaemia

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SUMMARY The relationship between blood requirement and the mean level of maintained haemoglobin was examined in 392 patients with homozygous β-thalassaemia. Pre- and post-transfusional haemoglobin levels and the amounts of blood transfused were measured during a 1-year period. No significant differences were noted in the blood requirements of patients (splenectomised or not) irrespective of the haemoglobin level. It may be supposed that if the mean haemoglobin level is high the haemopoietic activity is inhibited, and hence the bone marrow mass and total blood volume are reduced. High haemoglobin levels may thus be obtained with no increase in blood intake.

An understanding of the relationship between the amount of blood transfused and the mean haemoglobin (Hb) level maintained in patients with homozygous β-thalassaemia is important when assessing the risks and benefits of transfusion; it is important, too, in the clinical management of the patient. Data are scanty and conflicting. Modell reported a linear correlation between these two measurements, but Propper et al. found that the Hb level could be raised without increasing the amount of blood transfused, except during the initial few months. It was decided therefore to examine this question on a larger scale, using patients collected from three centres.

Material and methods

A total of 392 subjects aged between 2 and 23 years (166 from Turin, 158 from Ferrara, and 68 from Milan) were studied for 2 years. One hundred and eighty-four of them had been splenectomised. Any patient with hypersplenism or signs of isoinmunisation, or any patient who underwent splenectomy during the period of observation was excluded. Patients were transfused as outpatients every 25–30 days with one or two blood units. The amount that Hb levels increased by transfusion was 3–5–4 g/dl. The mean Hb level maintained was calculated as the average of the pre- and post-transfusional values during one year. The blood consumption was determined as the amount of blood (ml of concentrated red cells with haematocrit (75–85 %)) transfused during the same period, divided by the body weight in June.

The Hb levels were classified into groups so that the mean blood consumption could be statistically analysed by means of Student's t test; the paired-data t test was used to compare the requirement in each year of any patient in whom the Hb level increased.
Results

The Figure shows mean blood consumption plotted against Hb levels. It can be seen that consumption was significantly lower in the splenectomised patients (mean 154.4±9.8 compared with 182.2±10.6). No significant differences could be seen in the blood requirements of patients (splenectomised or not) in relation to maintained Hb levels. Of the 83 patients who had increases of more than 1 g/dl in the second year of follow-up only the 45 splenectomised subjects had a mean 12% decrease in blood requirement (t = 4.84; P < 0.001); the requirement of the non-splenectomised patients was unchanged (Table).

The blood requirement of our splenectomised patients with different Hb levels was compared with values observed by Modell in Britain after making adjustment for the differences in haematocrit of blood units infused in Italy and Britain. Blood consumption of patients with more than 10 g/dl is in the same range as that of Modell’s patients.

Discussion

Transfusion protocols that maintain a high mean Hb level give thalassaemic patients a life that is practically normal; there is also a decrease in skeletal deformity and less chance of damage caused by hypoxia.3

If there is a positive linear relationship between Hb level and blood requirement, as suggested by Modell,2 Hb levels can be raised only by increasing the amount of blood transfused, and hence by increasing the iron overload. Our results, like those of Propper et al.,2 show that all subjects maintained on between 9 and 14 g/dl had the same blood requirement. The reason for the differences between Modell’s data and our data could be that Modell’s correlation included patients kept at a lower Hb level (<9 g/dl) than ours. If this were to be the case, the maintained Hb level would be the result of endogenous erythropoiesis as well as of the transfusion. In this connection, it should be remembered that there are many β0 thalassaemics in Italy. In Britain, on the other hand, most patients are β+, and their erythropoiesis may be more efficient, owing to a less pronounced imbalance in Hb chain synthesis.

When the mean Hb level is raised to over 10 g/dl by periodic transfusions, endogenous production makes a non-significant contribution to its maintenance4-6 and there is a levelling off of the blood requirement. Moreover there was a significant reduction in blood requirement in splenectomised patients in whom mean Hb level was raised in 2 different years, pointing to more effective inhibition of haemopoietic overactivity, and hence a reduction in marrow mass and total blood volume.2-8 Our data suggest that the maintenance of high Hb levels in transfusion-dependent thalassaemic patients does not require a higher blood intake than that for standard Hb levels. Since it also reduces intestinal absorption of iron,7-8 supertransfusional protocols may prove to be the most beneficial.

We thank Dr B Modell for constructive criticism.

Table  Blood requirement (mean ± SD) of transfusion-dependent patients in whom an increase of more than 1 g/dl of the mean Hb level was achieved

<table>
<thead>
<tr>
<th>Patients</th>
<th>Blood requirement</th>
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<tr>
<td></td>
<td>First year</td>
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<tr>
<td>Splenectomised (n=45)</td>
<td>173.8 ± 43.0</td>
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<tr>
<td>Not splenectomised (n=38)</td>
<td>194.3 ± 97.1</td>
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References

Elusive blood clots and fluctuating ventricular dilatation after neonatal intraventricular haemorrhage

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SUMMARY Two cases are described of a previously unrecognised sequel of posthaemorrhagic ventricular dilatation. The first case documents freely mobile blood clots within the lateral ventricular system, the second variable asymmetry in the size of the dilated lateral ventricle. The unilateral ventricular dilatation depended on which side the infant was lying, the dependent ventricle being considerably larger than the upper one within 4 hours of head turning. Each of these conditions spontaneously resolved with no specific treatment.

Intracranial haemorrhage and consequent posthaemorrhagic dilatation are well-known entities in preterm babies and real-time ultrasonography has proved to be a safe and accurate method for diagnosing intracerebral haemorrhage and measuring ventricular size.

By using real-time ultrasonography routinely we have been able to follow the development of this condition and to note two unusual consequences of posthaemorrhagic dilatation.

Patients

Case 1. A boy was born at 30 weeks of gestation by spontaneous vaginal vertex delivery. Apgar score was 4 at one minute and 8 at ten minutes. Soon after birth he developed signs of respiratory distress and a chest x-ray film showed severe hyaline membrane disease. The baby was electively intubated at 4 hours of life for transporting to Hammersmith Hospital and then mechanically ventilated for 9 days. On arrival he was hypothermic (33°C) and acidicotic (pH 6.89). Two hours later pH was 7.27 and his condition appeared clinically to be improved. At age 48 hours his general condition deteriorated and he was noted to have grossly abnormal movements and thought clinically to have sustained intraventricular haemorrhage.

The occipitofrontal head circumference grew rapidly from the 10th day of life despite lumbar punctures and a ventriculocisternal shunt was inserted on the 40th day of life. When discharged at age 9 weeks, the infant had a pronounced head lag, abnormal eye movements, and a paralytic squint of the right eye. He was however responsive and both visually and auditorily quite alert.

Ultrasound scans showed bilateral intraventricular haemorrhage to have occurred at about age 72 hours with considerable intraparenchymal extension. Distended ventricles were first noted at 5 days of life and echogenic clots within the ventricles were clearly defined. By the 10th day of life, the posthaemorrhagic dilatation became very pronounced with considerable cerebral compression. Subsequent ultrasound examinations showed that when the infant was 30 days of age, the clots progressively became free within the ventricular system, and were from time to time either clearly visible or totally absent.

In order to determine the movements and the position of the clots in relation to the baby’s posture, we performed repeated scans, each time with the infant in a different position (Fig. 1).

When the infant was in the left lateral position, a clot was noted to be lying against the most lateral margin of the left lateral ventricle. The baby was then moved to lie supine, and the clots disappeared, although the most posterior part of the occipital poles could not be adequately visualised. When the
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