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Reply:

The letter from Dr. *Mandal* and his associates raises a number of important points which are worth emphasizing. My paper [1] had stated 'although splenectomy is indicated and is the definitive treatment where there is gross haemolysis in hereditary spherocytosis, its role in the mild form of the disease is not defined'. The presence of 'frequent attacks of jaundice' in the two patients of *Mandal* et al. would suggest that their patients had severe disease, and thus splenectomy was indicated. None of my patients had a history of jaundice.

It was fortunate that the patients of *Mandal* et al. had a good outcome after splenectomy. For even with splenectomy, problems may arise. Two cases of hereditary spherocytosis reported by *McElin* et al. [2] were splenectomised and had no operative complications, yet, one pregnancy ended in stillbirth and in the other the baby lived about 30 min. Although the unfortunate outcome in these 2 cases may have been unrelated to the splenectomy, it is important to remember that the outcome of pregnancy is influenced by a large number of factors. My first patient [1] (before she was investigated) had had two previous pregnancies which had ended in spontaneous abortion. She did not have a splenectomy, and had two subsequent pregnancies which ended in the birth of live, well children. Thus, hereditary spherocytosis and a bad obstetric history are not by themselves indicators for splenectomy. More important is the severity of the haemolysis - the principal benefit of splenectomy is on red cell survival, the benefits to the pregnancy are secondary.

My paper was concerned with patients with hereditary spherocytosis *presenting* in pregnancy. These are patients with mild disease and without a history of jaundice. In such patients, I have suggested a stepwise approach to their management, but I have assumed a high degree of medical and ante-natal care. In countries where these assumptions may be inappropriate, the letter of *Mandal* et al. raises the important point of symptomatic patients with hereditary spherocytosis who are not diagnosed until pregnancy. For these patients, I feel that it is most important that an attempt should be made to assses the severity of their disease prior to the pregnancy. If the evidence (family history, previous attacks, degree of haemolysis) suggests severe disease, splenectomy is indicated. If the patient has mild disease, the risks of splenectomy [3] have to be balanced against the potential benefits.

References

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