Thirteen Children from Twelve Pregnancies in Sick-cell Thalassaemia*

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CASE REPORT

A Fante housewife aged 40 years, known to have sickle-cell thalassaemia and to have a husband with haemoglobin SC disease, presented in May 1968 with amenorrhoea of three to four months' duration. The details of 11 previous pregnancies and the haematological and haemoglobin electrophoretic findings in her family are fully reported elsewhere (Konotey-Ahulu and Ringelhann, 1969).

Examination showed her to be well nourished but clinically anaemic. There was pronounced frontal bossing of her skull and a large diffuse non-toxic goitre was noted. The pulse and blood pressure were normal, but a precordial systolic bruit was present. On palpation of the abdomen the liver and spleen were not felt; there was enlargement of the uterus compatible with a pregnancy of 14 to 16 weeks. The haemoglobin level was 9.2 g./100 ml., the reticulocyte count 6%, and the sickling test positive. The electrophoretic pattern was that of sickle-cell thalassaemia.

The patient was followed up fortnightly until November, when she was admitted to hospital with a suspected loss of liquor, but this was not confirmed. She had been well throughout the pregnancy except for occasional "rheumatic" pains. Labour began spontaneously on 25 November and she was delivered of a healthy 8-lb. (3,620-g.) male infant. The placenta (see Fig.) was described

The placenta, showing pale areas of infarction.

as "unhealthy-looking," with ragged incomplete membranes and areas of infarction. There was excessive postpartum blood loss, for which she received three units of blood.

At the age of 4 months the baby was found to have sickle-cell thalassaemia. Contraceptive advice was earlier refused by the mother; later she agreed to use an intrauterine device.

COMMENT

Although sickle-cell anaemia and its variants do not always present a great obstetric problem as in this patient, there are

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risks to both mother and child. Curzen (1964) described a case of "leather pouch" uterus due to extensive myometrial necrosis in a second pregnancy associated with sickle-cell anaemia. Konotey-Ahulu and Korsah (1964) observed what they termed "intermittent claudication of the pregnant uterus"—that is, lower abdominal cramps on walking which disappeared on stopping—in an individual with sickle-cell trait who had a poor obstetric history. Woodruff (1961) believes that pregnancy is a hazard even in the sickle-cell trait. We believe that the patient whose case is reported here might have died outside hospital because blood would not have been readily available after delivery of her *thirteenth* child.

More and more haemoglobinopathy patients are passing through pregnancy (see Table); we therefore stress the need for contraceptive measures for them, not only because of the hazards of pregnancy (Bannerman and White, 1957; Smith and Conley, 1957; Hook and Cooper, 1958; Smith and Krevans, 1959) but also to prevent dissemination of the genes.

Number of Patients with Various Sickle-cell Genotypes Followed up Through Pregnancy at Korle Bu Teaching Hospital (1964-8)

Year	Hb SS	Hb SC	Hb S-Thal.	Hb SFhighgene	Hb S Korle Bu	Total
1964	1	8	—	—	—	9
1965	1	11	—	—	1*	13
1966	2	6	—	—	—	8
1967	2	7	1	1	—	11
1968	3	16	2	1	1	23
Total	9	48	3	2	2	64

* In retrospect.

This is the first recorded case of 12 pregnancies with 13 children (9 alive) in sickle-cell thalassaemia. The other related facts of a husband with haemoglobin SC disease and hence the completely haemoglobinopathic offspring also constitute a record.

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