Hereditary Spherocytosis in Pregnancy: Prognostic Factors, Maternal and Fetal Outcomes

Z Arain[1] **R Abdul Kadir**[1&2]

1 Department of Obstetrics and Gynaecology, Royal Free London NHS Foundation Trust, London,UK
2 Katharine Dormandy Haemophilia and Thrombosis Centre Royal Free London NHS Trust, Royal Free London NHS Foundation Trust, London,UK

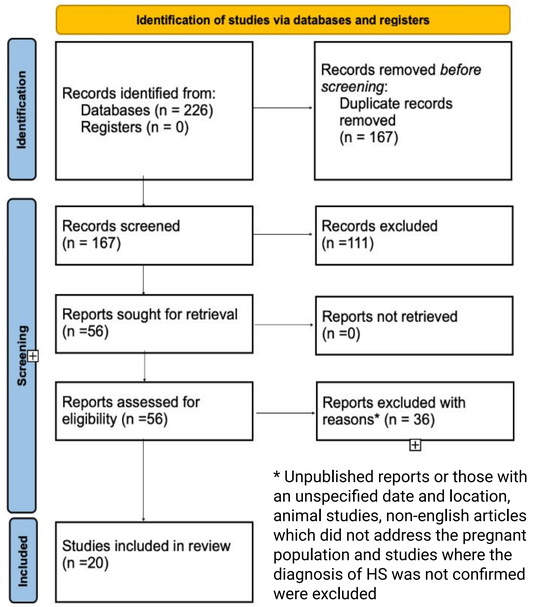
Royal Free London NHS Foundation Trust

INTRODUCTION

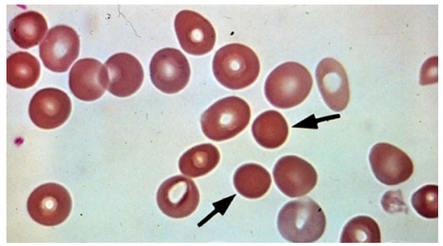
There is limited data on the course of pregnancy and maternal and fetal outcomes in women with hereditary spherocytosis.
We aimed to:
1) Evaluate maternal and fetal outcomes in women with Hereditary Spherocytosis
2) Establish the prognostic significance of disease severity and pre-pregnancy splenectomy

METHODS

Cochrane, PubMed, Ovid Medline and Embase were searched for literature on Pregnant women with Hereditary Spherocytosis from database inception until January 16th 2022 using a combination of search terms. Included studies were scored using National Institutes of Health Quality Assessment Tools to assess the quality of methodology



* Unpublished reports or those with an unspecified date and location, animal studies, non-english articles which did not address the pregnant population and studies where the diagnosis of HS was not confirmed were excluded



Blood film from a patient with hereditary spherocytosis showing evidence of spherocytes

Source: Hereditary Spherocytosis. Shafqat Shah, Roger Vega. Pediatrics in Review May 2004, 25 (5) 168-172; DOI: 10.1542/pir.25-5-168

RESULTS

Included Studies: 20 Observational studies (13 case reports, 5 cases series, 1 retrospective cohort study and 1 retrospective cross-sectional study).

National Institutes of Health Quality Assessment Tools scoring: 18 studies scored fair, 2 studies scored poor

Clinical characteristics

Maternal outcomes were recorded for 104 pregnancies in 53 women

HS was diagnosed in 24 women (45%) prior to pregnancy and 29 women (55%) during pregnancy

14 women (26%) had undergone splenectomy prior to pregnancy following HS diagnosis

Baseline disease severity (as per Eber classification) could be identified in 28 women (53%) as mild (n =16), moderate (n=9) and severe (n=3)

Data recording for ethnicity, BMI and smoking status was poor.

Included women were aged 18-34 with a mean age of 25

Table: MATERNAL OUTCOMES

	number of pregnancies where outcome observed (n=101)	Cases stratified by maternal baseline disease severity			
		mild	moderate	severe	unknown
Haemolytic crisis	24 (24%)	9	4	1	10
Blood transfusion	20 (20%)	8	3	1	8
Sepsis	7 (7%)	1	2	0	4

5% of women underwent splenectomy antenatally or in the immediate postpartum period.
Data was retrieved for 101 pregnancies 53 women

Outcomes in splenectomised women
14 women were splenectomised prior to pregnancy
1 woman experienced sepsis in 2 pregnancies. She had a past history of aplastic crisis prior to pregnancy. 2 women experienced significant anaemia.

FETAL OUTCOMES

3 infants required NICU admission and phototherapy, all were confirmed to have hereditary spherocytosis. 9 cases of preterm delivery and 2 cases of Small for Gestational Age were reported. There were no reported cases of infants requiring blood transfusion.

CONCLUSION

Women with hereditary spherocytosis are at risk of a number of adverse pregnancy outcomes irrespective of baseline disease severity. Splenectomy prior to pregnancy may be protective.