ROLE OF SURGICAL INTERVENTION HEREDITARY SPHEROCYTOSIS WITH HAEMOLYTIC ANAEMIA, SPLENOMEGALY, JAUNDICE.

Abstract

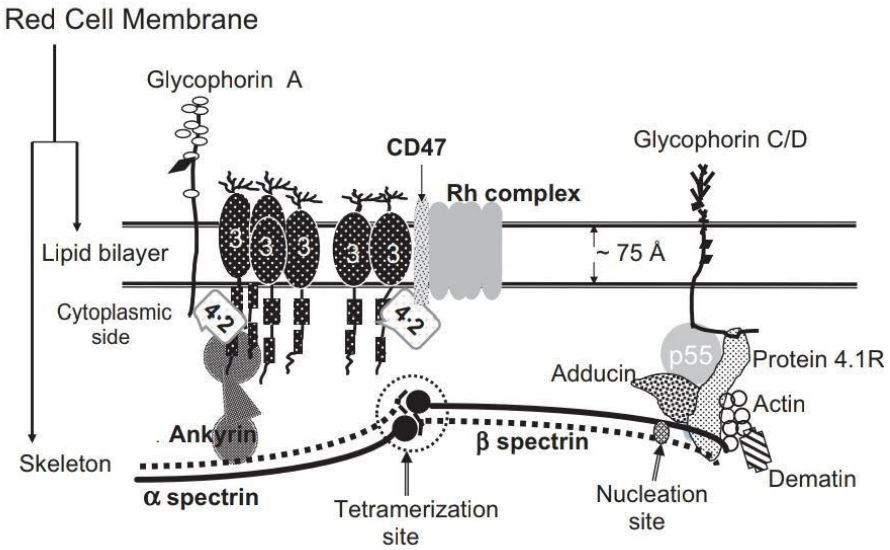
This case series includes surgical management in 3 cases of Hereditary spherocytosis presenting with haemolytic anaemia, splenomegaly and jaundice, admitted in Chettinad hospital and research institute during one year period from January 2019 to January 2020. Surgical intervention is indicated for selected patients with hereditary spherocytosis with haemolytic anaemia and jaundice to **abate the hemolytic process** after correction of anaemia with blood transfusion. Pigmented gallstones are seen in more than 50% cases for which incidence increases with severity of hemolysis and with age. Complications include aplastic anemia (most common after parvovirus B19 infection), haemolytic crisis during inter-current infection, megaloblastic crisis – during folic acid deficiency, cardiomyopathy, hematological malignancies.

Keywords: surgical management, hematological malignancies, hemolysis, spherocytosis

INTRODUCTION

Hereditary spherocytosis (HS) is a rare type of congenital haemolytic anaemia that occurs world wide. PREVALENCE : The estimated prevalence in Caucasian population ranges from 1:2000 to 1:5000 [1]HISTORY : Hereditary spherocytosis was described initially in 1871 [2]HS affects males and females equally. Age at diagnosis of HS is often between 3 – 7 years but can occur in infancy with severe disease or into adulthood with mild disease.INHERITANCE PATTERN : In 80% instances, the inheritance of HS is autosomal dominant and in others autosomal recessive. [3]

Fig 1: SCHEMATIC REPRESENTATION OF THE STRUCTURAL ORGANIZATION OF THE RED CELL CYTOSKELETON :



The molecular defect involves the genes encoding for spectrin, ankyrin, band 3, and protein 4.2 that link the bilayer of red cells to the membrane skeleton. Loss of this protein causes red blood cells to lack their characteristic biconcave shape.These proteins □□more susceptible to trapping and destruction by the spleen resulting in haemolysis, splenomegaly and cholelithiasis. The passage of these RBCs into sinusoids is difficult and gets phagocytosed resulting in extravascular haemolysis. Splenomegaly □□Mild to moderate; but the size

of spleen - not an indication for splenectomy. Direct / indirect bilirubin accumulates in gall bladder to form gallstones, altering liver function and causing hepatomegaly. Pigmented gallstones are seen in more than 50% cases for which incidence increases with severity of hemolysis and with age. Complications include aplastic anemia (most common after parvovirus B19 infection), haemolytic crisis during inter-current infection, megaloblastic crisis – folic acid deficiency, cardiomyopathy, hematological malignancies. during

Normal	6-8	
Normal (<3%)	>10	

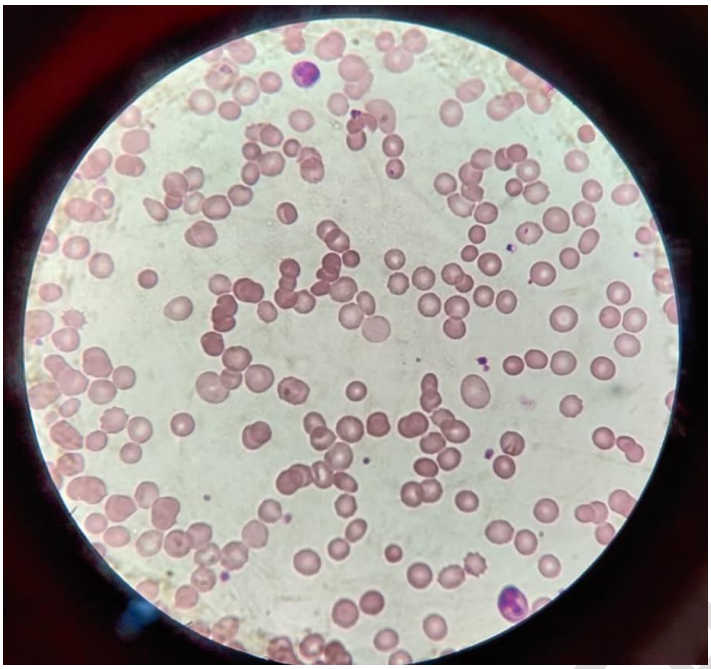
MATERIALS AND METHODS - This case series includes surgical management in 3 cases of Hereditary spherocytosis presenting with haemolytic anaemia, splenomegaly and jaundice, admitted in Chettinad hospital and research institute during one year period from January 2019 to January 2020, treated by Elective splenectomy with concomitant cholecystectomy in two patients (56-year male with obstructive jaundice post-ERCP status, and 30-year old female with cholelithiasis) and Only splenectomy in one patient (10 year-old boy with no features of gall stone disease). All three patients had Grade III splenomegaly (Hackett's grading score) and diagnosed with hereditary spherocytosis according to hematological criteria.

Fig 2: CLASSIFICATION OF HEREDITARY SPHEROCYTOSIS AND INDICATIONS FOR SPLENECTOMY :	<17	>51	
	100	40-60	
	Not required	Necessary – delay until 6 years if possible	

DIAGNOSIS : Laboratory diagnosis mainly involves peripheral blood smear – abnormally small and lack central pallor, Howell-Jolly bodies maybe seen, MCHC & reticulocyte count.

The presence of spherocytes is confirmed by an Osmotic Fragility Test (Sensitivity - 48 to 95%) ; Acidified Glycerol Lysis Test (Sensitivity – 99%) and a Negative Coombs test.

USG Abdomen to quantify size of spleen, to rule out cholelithiasis.



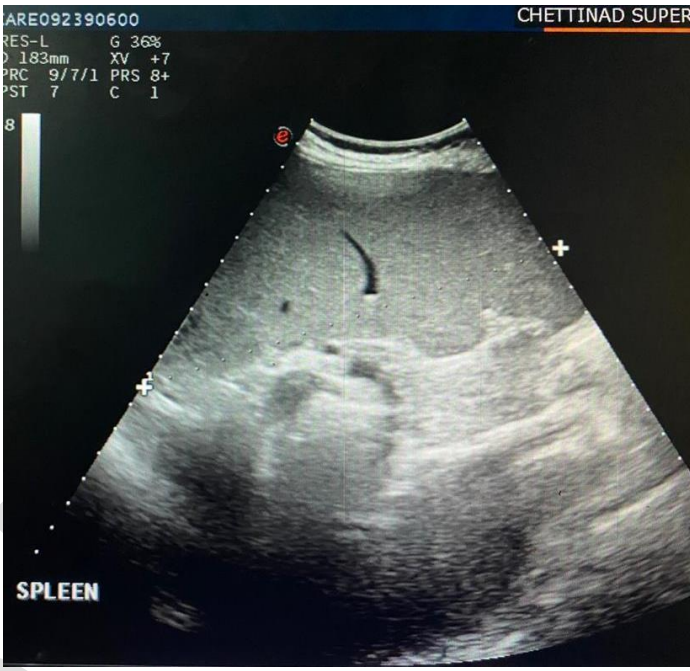
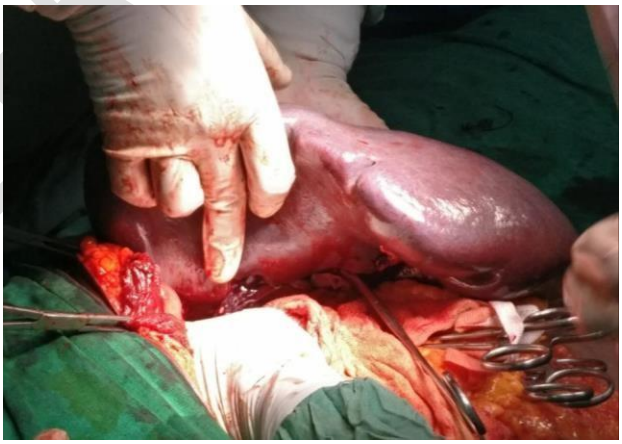


Fig 3: PREOPERATIVE WORKUP :



Preoperative preparation of the patients included

Correction of anaemia with blood transfusion,

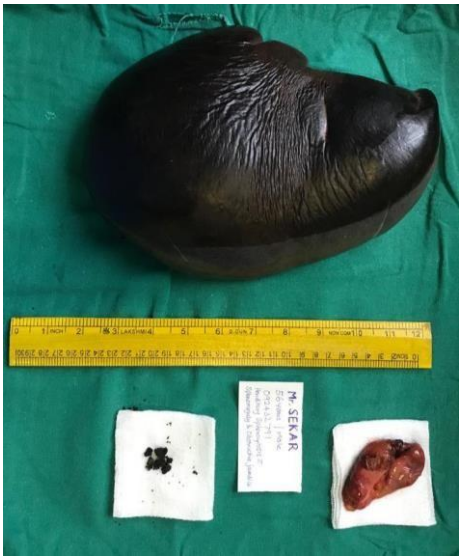
Cardiac status evaluation, and

Prophylactic administration of vaccines to prevent OPSI – Polyvalent Pneumococcal vaccine (PPV23), H. influenzae type b conjugate, and meningococcal [4] polysaccharide vaccine.

ELECTIVE SPLENECTOMY WITH CHOLECYSTECTOMY :

Left subcostal(1)/ upper midline incision(2).

Entry into abdominal cavity □□blunt and sharp dissection along convex surface of organ.



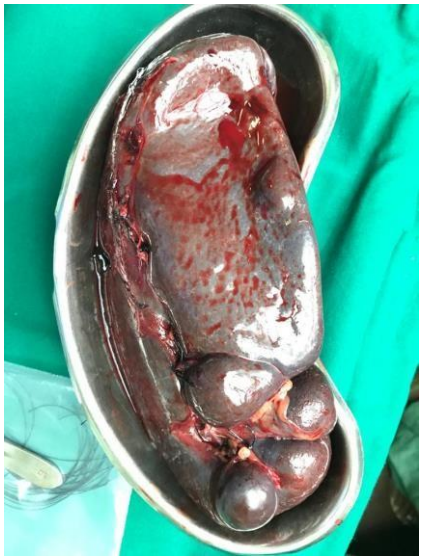


Table 57-1 Centers for Disease Control and Prevention Vaccine Recommendations for Asplenic Patients

VACCINE	RECOMMENDATION
Tetanus (Td/Tdap)	One dose every 10 years
Human papilloma virus (HPV)	Three doses for women through age 26 (0, 2, 6 mo)
Measles, mumps, rubella (MMR)	One or two doses
Varicella	Two doses (0, 4-8 wk)
Zoster	One dose
Influenza	One dose annually
Pneumococcal polysaccharide	One or two doses
Hepatitis A	Two doses (0, 6-12 mo or 0, 6-18 mo)
Hepatitis B	Three doses (0, 1-2 mo, 4-6 mo)
Meningococcal	One dose

Avascular peritoneal attachments and ligaments incised (Splenophrenic, splenocolic and splenorenal).

Identify hilum □□Splenic Artery and Vein double ligated and transfixed close to spleen.

Short gastric vessels ligated and divided.

Blunt dissection of posterior attachments.

Hemostasis checked : Left subphrenic area, Greater curvature of stomach, Splenic hilum, Tail of pancreas.

Look for accessory spleen : Hilum, Gastrocolic, gastrosplenic ligament, Greater omentum, Mesenteric region, presacral space.

Drain may/maynot be kept.

+/- Cholecystectomy.

Intraoperative periods were uneventful.

Postoperative febrile episodes required escalation of antibiotics.

Normal hematocrit values were achieved 4 weeks postoperatively in all patients.

There was no evidence of postoperative abdominal or wound infections, and no post-splenectomy sepsis.

DISCUSSION- Surgical intervention is indicated for selected patients with hereditary spherocytosis with haemolytic anaemia and jaundice to **abate the hemolytic process** after correction of anaemia with blood transfusion. Cholecystectomy is indicated in patients with pigment gallstones. Splenectomy is very effective in reducing hemolysis, leading to a significant prolongation of the red cell lifespan and should be performed in patients with severe HS, considered in those who have moderate disease, and should probably not be performed in those with mild disease. If splenectomy is done in a patient with symptomatic gallstones, cholecystectomy should be done concomitantly. [5]

CONCLUSION- To conclude, in patients with Hereditary Spherocytosis (ModerateSevere) presenting with splenomegaly and jaundice, splenectomy with or without cholecystectomy can be performed with minimal morbidity, to relieve symptoms due to hemolysis and to prevent further episodes of jaundice.

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