INTRODUCTION
Hereditary Spherocytosis (HS) is a rare inherited hemolytic anemia mainly affecting red blood cell (RBC) membrane which shows spherical shaped RBC on peripheral smear and cause cell lysis. It was first described in 1817. HS is inherited as autosomal dominant pattern in about 75% of cases whereas rest 25% of cases occur sporadically. HS is a product of heterogenous alteration in one of the six genes, most often in the ankyrin gene (ANK1) which encodes for ankyrin. Other commonly mutated genes are SPTB, SLC4A1, EPB42 and SPTA1 encoding for spectrin β-chain, anion exchanger 1 (band 3), protein 4.2 and the spectrin α-chain respectively. The characteristics of HS vary from asymptomatic to fulminant hemolytic anemia. The classical symptoms include anemia, jaundice, splenomegaly and elevated red blood cell osmotic fragility. HS is the most common under-recognized disorder which can present from infancy and throughout the life span.

CASE REPORT
A 22-year-old female visited a tertiary care hospital with the complaints of fever, yellow discoloration of eye and urine since 2 days. She was diagnosed with hereditary spherocytosis at the age of 12 with significant family history of HS. She has been taking blood transfusion since 10 years after visiting the hospital for regular checkup. She gets to know about her decreased hemoglobin level through routine checkup. On examination her pulse rate was 80 beats per minute, respiratory rate of 22 cycles per minute, blood pressure of 110/70 mm Hg and icterus positive. On day 1, her hemoglobin was 6.3 g/dL which falls under life-threatening anemia, total bilirubin was 7.9 g/dL, and direct bilirubin was 0.9 g/dL. She was given with blood transfusion for two days and hemoglobin level raised to 10 g/dL. This case presents life-threatening anemia and icterus associated with severe hereditary spherocytosis. The treatment in hereditary spherocytosis is quite challenging often because of its obscurity.

DISCUSSION
HS is most common inherited disease with deficiency in red cell membrane proteins like ankyrin and spectrin. Based on the severity of disease, HS is classified into mild, moderate and severe accounting for 10 to 20%, 60 to 75% and 5% of cases respectively. Mild HS is associated with non-anemic, reticulocytosis, splenomegaly or jaundice and undetectable until adolescence; moderate HS is associated with anemia, high reticulocyte count and elevated serum bilirubin and severe HS is associated with marked anemia, hemolysis, hyperbilirubinemia, splenomegaly, and regular blood transfusions. This case belongs to severe HS category due to the life-threatening anemia with total bilirubin levels of 7.9 mg/dL and 2 pint of blood transfusion.

ANK1 gene mutation were found in 45% of Asian patients according to the report by Yang et al., in 2018 which is the reason that HS is common in Asia than compared to other continents. Complications of HS are pigmented gallstones, anemia – hemolytic, megaloblastic and aplastic, decreased growth, gout, hematological malignancies, cardiomyopathy and skin disorders.

### Table 1: Treatment chart.

<table>
<thead>
<tr>
<th>Brand name</th>
<th>Generic name</th>
<th>Dose</th>
<th>Frequency</th>
<th>Day 1</th>
<th>Day 2</th>
<th>Day 3</th>
<th>Day 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inj. Xone</td>
<td>Ceftriaxone</td>
<td>1g</td>
<td>1-0-1</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
</tr>
<tr>
<td>Inj. Pan</td>
<td>Pantoprazole</td>
<td>40 mg</td>
<td>1-0-0</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
</tr>
<tr>
<td>Tab. Dolo</td>
<td>Paracetamol</td>
<td>625 mg</td>
<td>1-1-1</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
</tr>
<tr>
<td>Inj. Ondem</td>
<td>Ondansetron</td>
<td>4 mg</td>
<td>sos</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
</tr>
</tbody>
</table>

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According to the diagnosis and treatment guidelines, patient with positive family history of HS, typical clinical features like sphere shaped blood cells in peripheral smear can be diagnosed as HS. Other diagnostics include osmotic fragility test, acidified glycerol lysis test, cryohemolysis test, osmotic gradient ektacytometer and sodium dodecyl sulphate – polyacrylamide gel electrophoresis. Definitive treatment of HS includes splenectomy but is associated with increased risk of infection.

**CONCLUSION**

This is a case of 22 years old female suffering from severe HS. She had symptoms of severe HS that is life-threatening anemia, elevated levels of bilirubin, and icterus. Her management include 2 pint of blood transfusion with prophylactic usage of antibiotic and antiemetic. To reduce fever and prevent any gastric irritation she was prescribed with paracetamol and pantoprazole. Before transfusion she was given furosemide, pheniramine maleate and hydrocortisone to prevent transfusion related reactions. Splenectomy can be considered in this case to prevent further complications.

**CONFLICT OF INTEREST**

The author declares that there is no conflict of interest.

**REFERENCES**