Hereditary Spherocytosis

Dolores Loeser
Otterbein University, dolores.loeser@otterbein.edu

Follow this and additional works at: https://digitalcommons.otterbein.edu/stu_msn

Part of the Hemic and Lymphatic Diseases Commons, Medical Pathology Commons, and the Nursing Commons

Recommended Citation

This Project is brought to you for free and open access by the Student Research & Creative Work at Digital Commons @ Otterbein. It has been accepted for inclusion in Nursing Student Class Projects (Formerly MSN) by an authorized administrator of Digital Commons @ Otterbein. For more information, please contact digitalcommons07@otterbein.edu.
**Hereditary Spherocytosis**

**Dolores P Loeser, RN**
Department of Pediatric Surgery, Nationwide Children’s Hospital.

Otterbein University
Westerville, Ohio

Columbus, Ohio

---

**Definition**

Hereditary Spherocytosis (HS) is a hemolytic anemia where red blood cell membranes are spherical unlike common red blood cells, which are flat and round. In individuals with HS, the spleen does not recognize these amorphous cells and destroys—rather filters—they, making the individual anemic (MedlinePlus, 2013).

HS is an autosomal dominant, or recessive autosomal inherited blood disease (Hsiao, Sthya, Nathens, de Mestral, Hill, & Langer, 2013). HS happens in 1 in 2,000 of the Caucasian population or in from Northern Europe (MedlinePlus, 2013).

The proteins involved in HS are ankyrin-A (Christensen & Henry, 2010), spectrin (Pietroni, 2013), and acidic glycerol-phosphatidylcholine (AGLC) (Wormann, 2009). Hereditary Spherocytosis (HS) is a common symptoms in neonates with HS. According to Clotistensen and Henry (2010) hyperbilirubinemia is sometimes miss as a diagnose for HS (Christensen & Henry 2010).

**Diagnosis**

- **Peripheral blood smear**
- **Family history**
- **Hemolytic fragility test**
- **Acidified glycerol lysis time test (AGLT)**
- **Cytoskeleton test**
- **Eosin-5'-maleimide (EMA)-binding test** (King & Zanella, 2013).

**Pathophysiology**

Cells in HS have distorted Na+/K+ fluctuations. The membrane proteins in the red blood cells are changed. The normal intracellular K+ and Na+ are reversed in plasma. Spectrin, a structural protein of the membrane skeleton is reduced, which leads to increased loss of the outside area of the cell and makes it spherical instead of a biconcave disc. The spleen does not recognize these new transformed cells and destroys them (King & Zanella, 2013).

**Symptoms**

- Jaundice
- Normal

**Additional Sources**