Hereditary Spherocytosis

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

HS is an autosomal dominant, or recessive autosomal inherited blood disease (Huq, Pieterno, Rahman & Alam, 2010). HS happens in 1 in 2,000 of the Caucasian population in or from Northern Europe (MedlinePlus, 2013).

The proteins involved in HS are ankyrin, band 3, and protein 4.2. Hyperbilirubinemia is a common symptom in neonates with HS. According to Chistensen and Henry (2010) hyperbilirubinemia is sometimes missed as a diagnosis for HS (Christensen & Henry, 2010).

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

HS symptoms:
- Jaundice
- In older children: varies from fatigue, palor of the skin, enlarged spleen or weakness
- In some cases: gallstones, (Huq, Pieterno, Rahman & Alam, 2010)
- Aplastic crisis due to Parvovirus B19 infection (Bhate, Gowler, Dias, 2012).

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