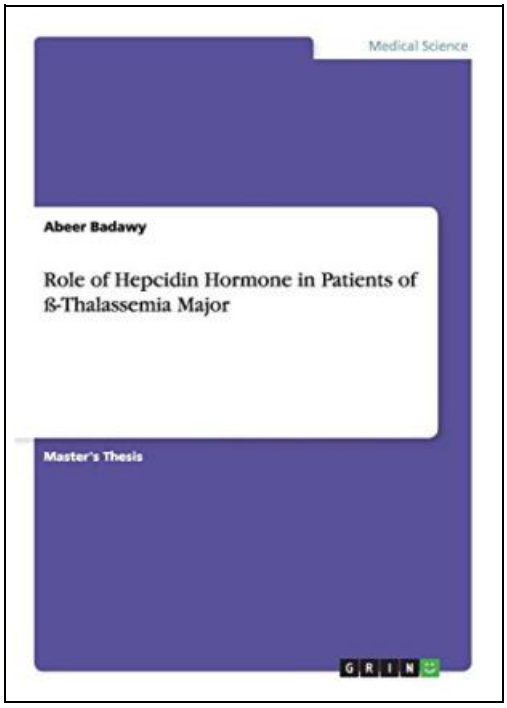
Role of Hepcidin Hormone in Patients of ß-Thalassemia Major



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ROLE OF HEPCIDIN HORMONE IN PATIENTS OF &SZLIG;-THALASSEMIA MAJOR

GRIN Verlag Gmbh Mrz 2015, 2015. Taschenbuch. Condition: Neu. Neuware - Master's Thesis from the year 2010 in the subject Medicine - Pharmacology, , language: English, abstract: The thalassemias are a heterogeneous group of genetic disorders of haemoglobin synthesis, occurring more frequently in the Mediterranean region, the Indian subcontinent, Southeast Asia, and West Africa .The thalassemias are divided according to their severity into major which is severe and transfusion dependent, intermediate and minor forms of illness. The -thalassemias are the most important types of thalassemia because they are so common and usually produce severe anemia in their homozygous and compound heterozygous states (Hillman et al., 2005). In -thalassemia major, the neonate is well at birth but develops severe anemia, bone abnormalities, failure to thrive, and life-threatening complications. In many cases, the first signs are pallor, yellow skin and scleras in infants ages 3 to 6 months. Later clinical features, in addition to severe anemia, include splenomegaly or hepatomegaly, with abdominal enlargement, frequent infections, bleeding tendencies (especially toward epistaxis), and anorexia (Fucharoen et al., 2000). Transfusional iron overload is the most important complication of -thalassemia and is a major focus of management, which can be prevented by adequate iron chelation. Extensive iron deposits are associated with cardiac hypertrophy and dilatation, degeneration of myocardial fibers (Aessopos et al., 1995; Du et al., 1997). Hepcidin is a 25-amino-acid iron peptide hormone. Initially identified in human plasma and urine as an anti-microbial molecule. Hepcidin is the key regulator of systemic iron homeostasis and a pathogenic factor in anemia of inflammation and hereditary hemochromatosis. Hepcidin inhibits iron influx into plasma from duodenal enterocytes that absorb dietary iron, from macrophages that recycle iron from senescent erythrocytes and from hepatocytes that store iron (Park et al., 2001). Iron-Loading anemias are characterized by ineffective erythropoiesis and increased intestinal...

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