INTRODUCTION

Glycogen storage diseases (GSDs) are inherited metabolic disorders of glycogen metabolism. In postprandial period, blood glucose level increases and endogenous glucose production is suppressed. Glycogen is primarily stored in liver and muscle. Hypoglycemia is the primary manifestation of the hepatic glycogenoses, whereas weakness and muscle cramps are the predominant features of the muscle glycogenoses. Different hormones, including insulin, glucagon, cortisol and others regulate the relationship or glycolysis, gluconeogenesis and glycogen synthesis ***(Roach, 2002).***

Although it is hard to estimate accurately, based on several studies, the overall GSD incidence is approximately 1 case per 20000-43000 live births and the most common one is type IX ***(Chen et al., 2001).***

Disorders or glycogen degradation may affect primarily the liver, the muscle, or both. Episodes or hypoglycemia and enlargement of the liver are the main findings in all hepatic GSD, but apart from this the heterogeneity or their presentation is considerable. There are over 12 types and they are classified based on the enzyme deficiency and the affected tissue. Eighty percent of hepatic GSD is formed by types I, III, and IX ***(Saltik et al. ,2000).***

Type Ia involves the liver, kidney and intestine (and Ib also

leukocytes), and the clinical manifestations are hepatomegaly, failure to thrive, hypoglycemia, hyperlactatemia, hyperuricemia and hyperlipidemia. Type IIIa involves both the liver and muscle, and IIIb solely the liver. The liver symptoms generally improve with age. Type IV usually presents in the first year of life, with hepatomegaly and growth retardation. The disease in general progresses to cirrhosis. Type VI and IX are a heterogeneous group of diseases caused by a deficiency of the liver phosphorylase and phosphorylase kinase system. There is no hyperuricemia or hyperlactatemia. Type XI is characterized by hepatic glycogenesis and renal Fanconi syndrome. Type II is a prototype of inborn lysosomal storage diseases and involves many organs but primarily the muscle. Types V and VII involve only the muscle ***(Ozen, 2007).***

Insulin stimulates the liver to store glucose in the form of glycogen. A large fraction or glucose absorbed from the small intestine is immediately taken up by hepatocytes, which convert it into the storage polymer glycogen. The net effect is clear: when the supply of glucose is abundant, insulin "tells" the liver to bank as much of it as possible for use later. A well-known effect of insulin is to decrease the concentration of glucose in blood. In the absence of insulin, glycogen synthesis in the liver ceases and enzymes responsible for breakdown~ of glycogen become active. Glycogen breakdown is stimulated not only by the absence of insulin but by the presence of glucagon, which is secreted when blood glucose levels fall below the normal range ***(Mundy et al., 2003).***

Patients with GSDIb have an increased prevalence of thyroid autoimmunity and hypothyroidism, although patients with GSDIa have little evidence or thyroid abnormalities. Concomitant damage at the level or the hypothalamus or pituitary gland might be hypothesized on the basis or the slightly elevated thyrotropin levels, even in patients with overt hypothyroidism ***(Melis et al.,2007).***

AIM OF WORK

The aim of the present study is to estimate the thyroid hormones and insulin levels in previously diagnosed patients with glycogen storage disease and evaluation of their growth.