Introduction

Glycogen storage disease (glycogenosis) is one of several inherited inborn error of metabolism that result from enzymes defects that affect the processing of glycogen synthesis or breakdown within liver, muscle, and other cell types**.**

The overall GSD incidence is estimated 1 case per 20 000 - 43 000 live births **.**

There are over 12 types and they are classified based on the enzyme deficiency and the affected tissue.

Hypoglycemia and hepatomegaly is the primary manifestation of the hepatic glycogenoses, whereas weakness and muscle cramps are the predominant features of the muscle glycogenoses.

Hepatomegaly and mosaic pattern on liver biopsy are a feature of all hepatic GSDs. However, the distinction between these types is best done by enzyme studies of liver tissue obtained by wedge biopsy. However, the clinical, biochemical and histological features may suggest the type of GSDs.

Proper and early diagnosis of GSD is essential as dietary treatment with raw cornstarch and frequent high-starch feeds can prevent progression of this disease.

Growth retardation is a well-known problem for patients suffering from glycogen storage diseases (GSDs). However, the underlying mechanism, and therefore the management of growth impairment in these patients, remains controversial. Hyperlacticacidemia, recurrent hypoglycemia, growth hormone (GH) and/or insulin-like growth factor (IGF) deficiency, GH and/or IGF resistance, decreased insulin and increased cortisol secretions have all been suggested to explain growth retardation in GSDs.

This search aims at study endocrine and metabolic variables that affect the growth in patients with glycogen storage diseases receiving standard dietary therapy.

This is done by Observational study for twenty-five patients with GSDs, age range 1-12 years, are investigated on their usual dietary regimens. Data on height, weight, BMI and other laboratory investigations as CBC, liver enzyme, lipid profile and study of GH, IGF1 concentration profiles and are compared to twenty healthy subjects then the results statistically analyzed.