

Glycogen Storage Disease

These disorders principally affect the liver and have nine different types depending on the enzymes involved. As carbohydrate metabolism in liver is responsible for glucose homeostasis this group of disorders presents typically with hypoglycemia and hepatomegaly (not in all types). Treatment is designed mainly to maintain normoglycemia and is achieved by continuous nasogastric infusion of glucose or oral uncooked starch. Uncooked starch acts as a slow release form of glucose.¹² This is specially useful in Type I, III and IV but most demanding in Type I.

Acknowledgement

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Michael Brown



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