

Summary of pathways

- 1 Galactokinase (*mild galactosemia*)
- 2 Galactose-1-phosphate uridylyltransferase (*severe galactosemia*)
- 3 Hexokinase/glucokinase
- 4 Glucose-6-phosphatase (*von Gierke disease*)
- 5 Glucose-6-phosphate dehydrogenase
- 6 Transketolase
- 7 Phosphofructokinase-1
- 8 Fructose-1,6-bisphosphatase
- 9 Fructokinase (*essential fructosuria*)
- 10 Aldolase B (*fructose intolerance*)
- 11 Aldolase B (*liver*), A (*muscle*)
- 12 Triose phosphate isomerase
- 13 Pyruvate kinase
- 14 Pyruvate dehydrogenase
- 15 Pyruvate carboxylase
- 16 PEP carboxykinase
- 17 Citrate synthase
- 18 Isocitrate dehydrogenase
- 19 α -ketoglutarate dehydrogenase
- 20 Carbamoyl phosphate synthetase I
- 21 Ornithine transcarbamylase
- 22 Propionyl-CoA carboxylase
- 23 HMG-CoA reductase

- B Requires biotin cofactor
- T Requires thiamine cofactor (TPP)
- # Irreversible, important point of regulation

