

## Metabolism sites

## Mitochondria

Fatty acid oxidation ( $\beta$ -oxidation), acetyl-CoA production, TCA cycle, oxidative phosphorylation, ketogenesis.

## Cytoplasm

Glycolysis, HMP shunt, and synthesis of cholesterol (SER), proteins (ribosomes, RER), fatty acids, and nucleotides.

## Both

Heme synthesis, urea cycle, gluconeogenesis. **Hugs take two** (both).

## 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 Phosphofruktokinase-1
- 8 Fructose-1,6-bisphosphatase 1
- 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  $\alpha$ -ketoglutarate dehydrogenase
- 20 Carbamoyl phosphate synthetase I
- 21 Ornithine transcarbamylase
- 22 Propionyl-CoA carboxylase
- 23 HMG-CoA reductase

