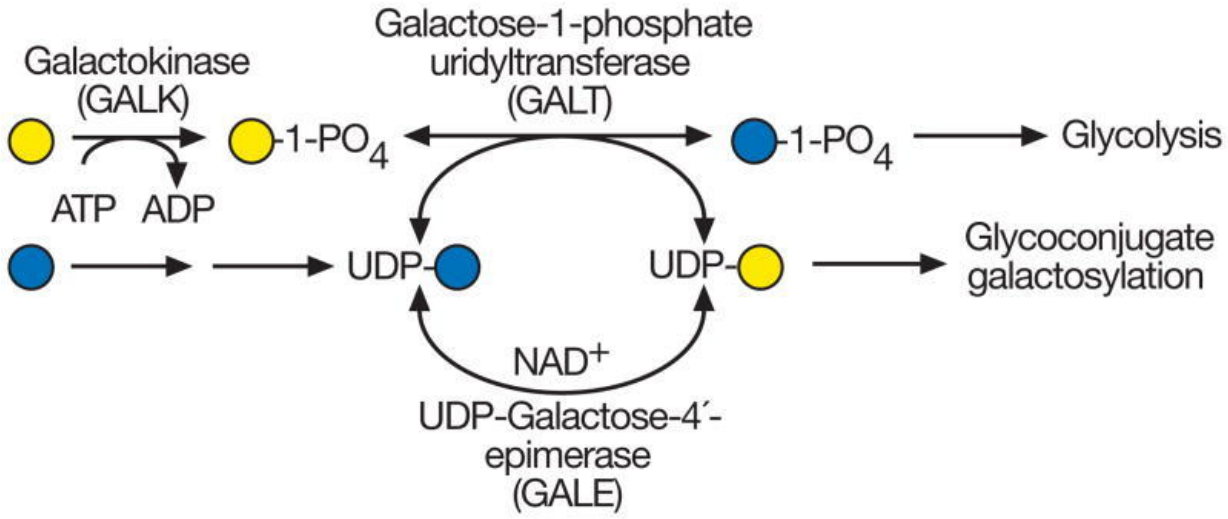
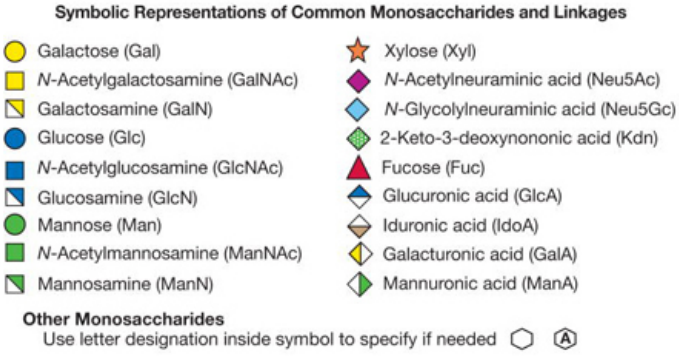


FIGURE 42.3

UDP-galactose synthesis and galactosemia. The most common form of galactosemia is due to a deficiency of galactose-1-phosphate uridyltransferase (GALT). This enzyme normally utilizes galactose-1-phosphate derived from dietary galactose. In the absence of GALT, galactose-1-phosphate accumulates, along with excessive galactose and its oxidative and reductive products galactitol and galactonate (not shown). UDP-galactose synthesis may also be impaired in the absence of GALT, but not completely because UDP-galactose-4′-epimerase (GALE) can form UDP-galactose from UDP-glucose and can supply the donor to galactosyltransferases required for normal glycoconjugate biosynthesis.

Symbol Key:



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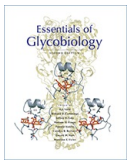

Essentials of Glycobiology. 2nd edition.
Varki A, Cummings RD, Esko JD, et al., editors.
Cold Spring Harbor (NY): Cold Spring Harbor Laboratory Press; 2009.

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