



















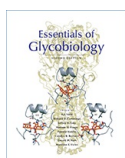


FIGURE 42.3

UDP-galactose synthesis and galactosemia. The most common form of galactosemia is due to a deficiency of galactose-1-phosphate uridylyltransferase (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.

Symbolic Representations of Common Monosaccharides and Linkages			
	Galactose (Gal)		Xylose (Xyl)
	N-Acetylgalactosamine (GalNAc)		N-Acetylneuraminic acid (Neu5Ac)
	Galactosamine (GalN)		N-Glycolylneuraminic acid (Neu5Gc)
	Glucose (Glc)		2-Keto-3-deoxynononic acid (Kdn)
	N-Acetylglucosamine (GlcNAc)		Fucose (Fuc)
	Glucosamine (GlcN)		Glucuronic acid (GlcA)
	Mannose (Man)		Iduronic acid (IdoA)
	N-Acetylmannosamine (ManNAc)		Galacturonic acid (GalA)
	Mannosamine (ManN)		Mannuronic acid (ManA)
Other Monosaccharides			
Use letter designation inside symbol to specify if needed  			

Symbol Key:Use letter designation inside symbol to specify if needed  
[Download Teaching Slide](#) (PPT, 299K)

 From: [Chapter 42, Genetic Disorders of Glycosylation](#)


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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