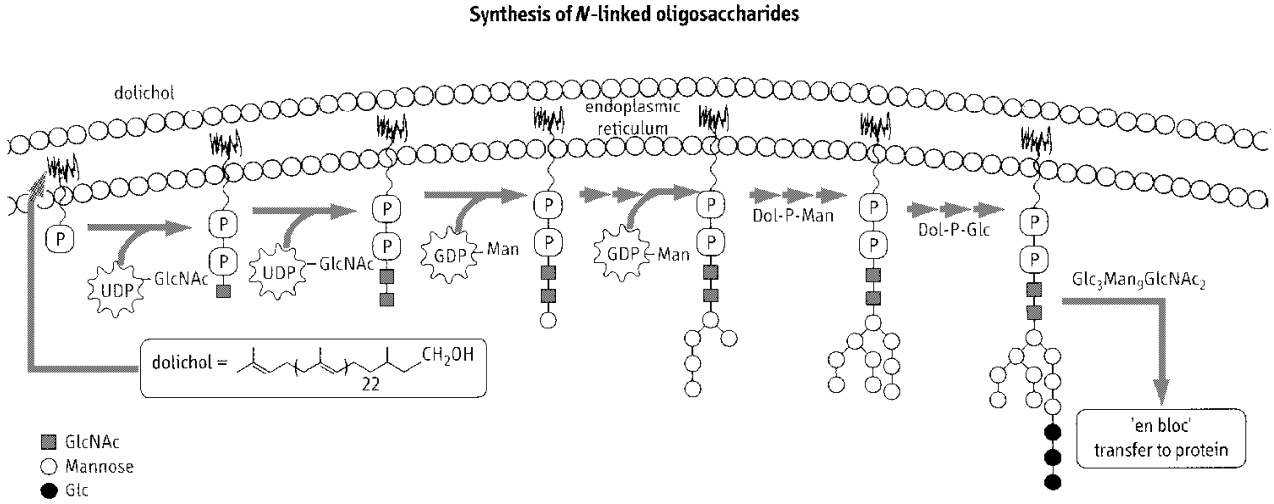
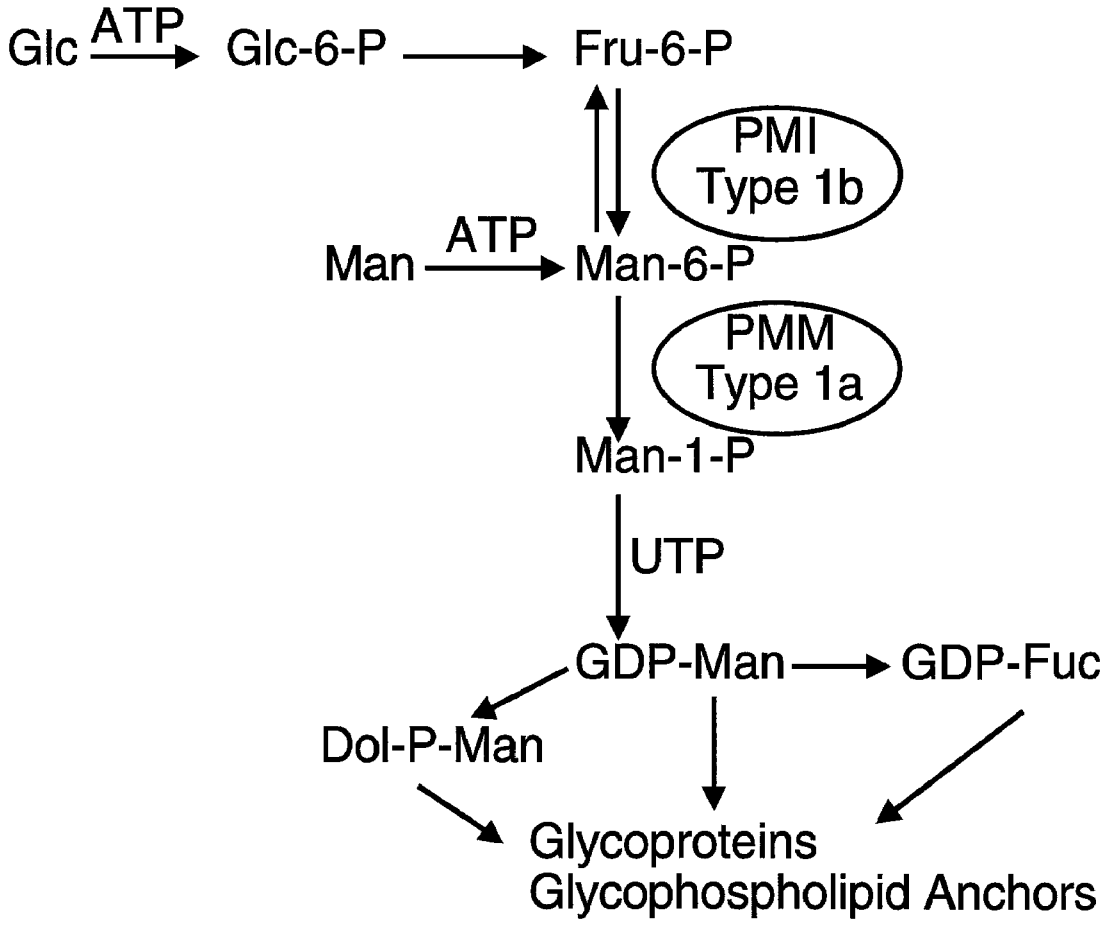
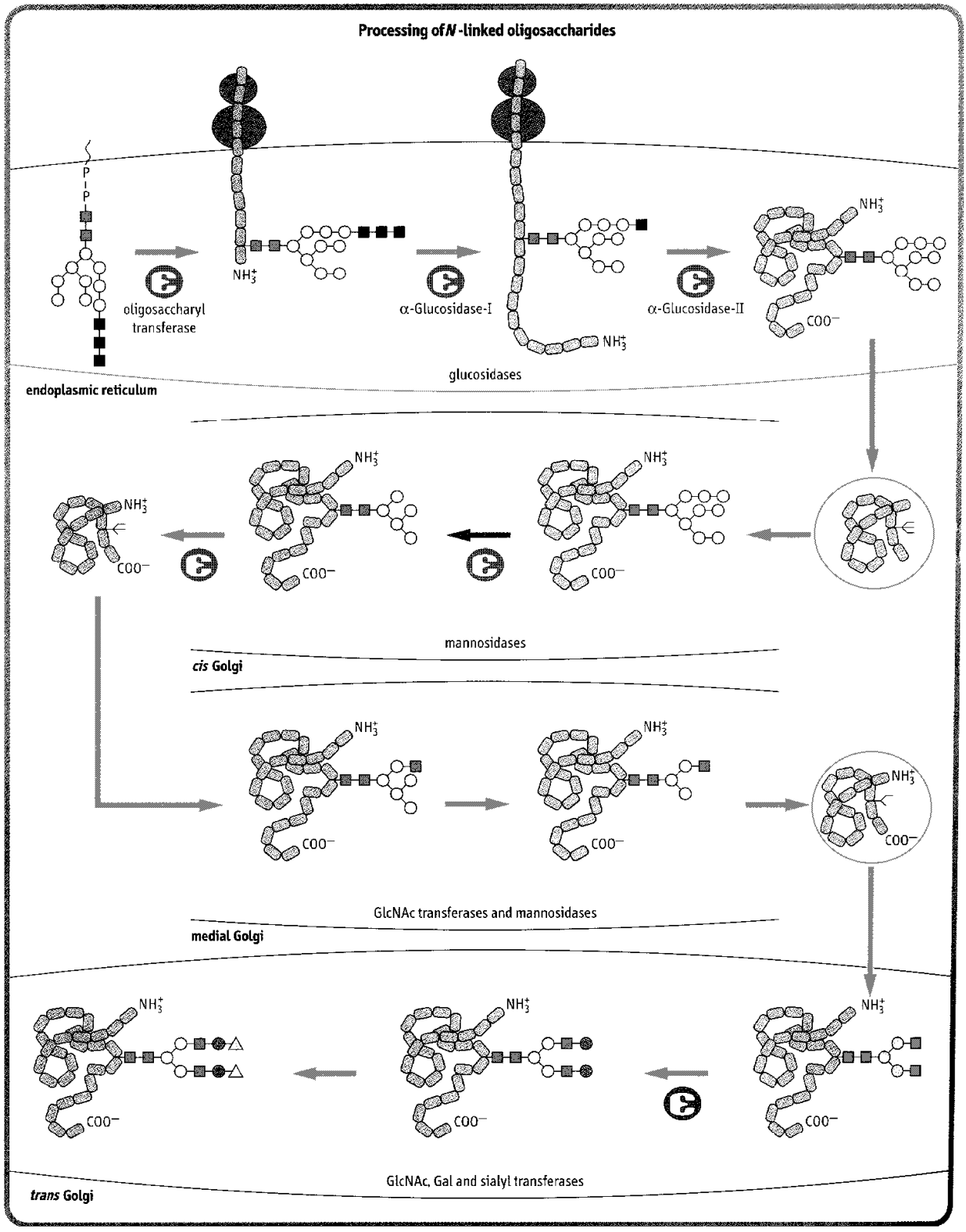
CARBOHYDRATE DEFICIENT GLYCOPROTEIN SYNDROME AND FUNCTION OF GLYCOPROTEINS

The Carbohydrate Deficient Gycoprotein Syndrome (CDGS) are caused by the impairment in the biosyntehsis of Asn-linked sugar chains.



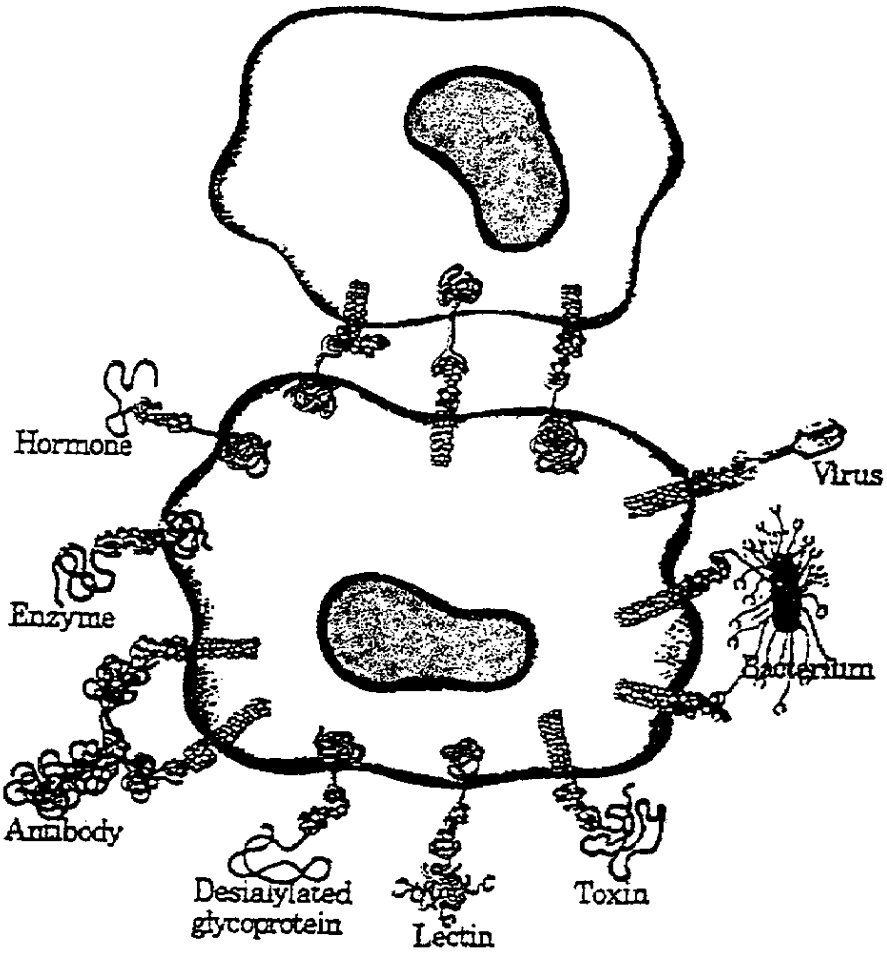
CDGS and Biosynthesis of GDP-Man




Processing of N-linked oligosaccharides
P
P
oligosaccharyl transferase
α-Glucosidase-I
α-Glucosidase-II
glucosidases
endoplasmic reticulum
mannosidases
cis Golgi
GlcNAc transferases and mannosidases
medial Golgi
GlcNAc, Gal and sialyl transferases
trans Golgi

Function

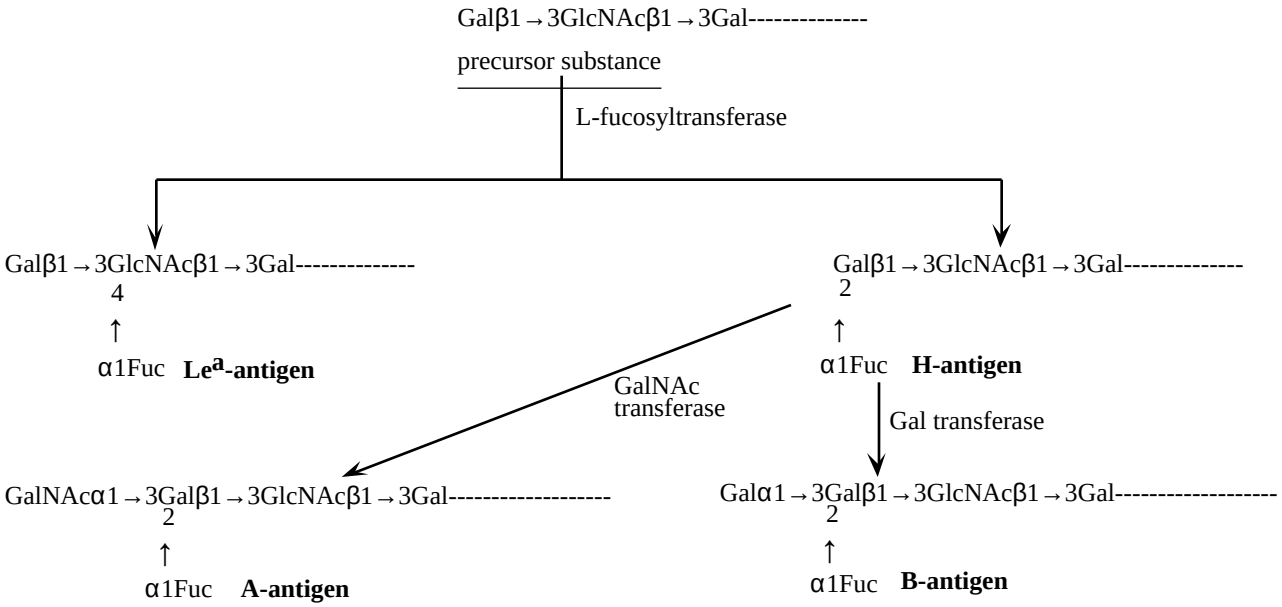
In contrast to nucleotides and amino acids, which can interconnect in only one way, monosaccharides can be joined to one another through multiple linkages. For example, two Glc residues can form 11 different disaccharides whereas two alanines can make only one dipeptide. This potential structural diversity makes sugar polymers effective carriers of information. It has been shown that glycoconjugates are involved in biological recognition. For carbohydrate groups to serve as recognition signals, there must be proteins that bind to them specifically. In addition to immunoglobulins, another major class of saccharide binding proteins is the **lectins**. The lectins were first recognized in plant tissues (such as concanavalin A and wheat germ agglutinin), but we now know *they are also widely distributed in animal tissues* and have many functions. The figure below illustrates how glycoconjugates (proteoglycans, glycosphingolipids, and glycoproteins) on the cell surface can interact with microorganisms, adjacent cells, toxins, enzymes, and lectins etc.



Complex carbohydrate chains in glycoconjugates have been found to play many important and intriguing biological functions. The following are some medically relevant examples:

(1) **They are involved in host -pathogen interaction.** Pathogen must attach to host tissues in order to colonize and establish infection. Complex carbohydrates on cell surfaces can serve as specific sites for bacterial colonization. For example, urinary tract infection can be caused by the specific attachment of p-fimbriated *E. coli* to the disaccharide, Galα1→4Gal, in $GbOse_3Cer$ and $GbOse_4Cer$ (p-blood substances) found in uroepithelial cells.

(2) They determine the blood group activities.



(3) They regulate the catabolism of circulating glycoproteins by the liver.