Id	Name/Reaction	Information	Kinetics
GLUT2	**Facilitated glucose transporter member 2** D-glucose (disse) [**glc_dis**] ↔ D-glucose (cytosol) [**glc**] D-galactose (disse) [**gal_dis**] ↔ D-galactose (cytosol) [**gal**] "Free galactose is transported into cells through a variety of transporters that are members of the glucose transporter (GLUT) family. Deficient transport of galactose into hepatocytes is evident in human patients with defective GLUT2 transporters (Fanconi-Bickel syndrome) resulting in galactose malabsorption/intolerance (Brown, 2000; Leslie, 2003)." "GLUT2 is not completely specific for glucose and is probably also involved in galactose transport (Brown, 2000)." "Data suggests that all transported sugars are likely to use the same outward-facing sugar-binding site on the transporter molecule. This is further indicated by the observation that each of the alternative substrates for each isoform is a competitive inhibitor of deGlc and that D-glucose is a competitive inhibitor of the transport of the alternative substrates. (Colville,	**Mechanism** TCDB:2.A.1.1 (glucose transporter subfamily) **Protein/Structure** UniProt:P11168 (GTR2_HUMAN) **Gene** SLC2A2, GLUT2 **Disease** MIM:227810 (Fanconi-Bickel syndrome; FBS)	Km(3-O-MG)=42.3±4.1mM (human liver) (Gould, et al., 1991; Walmsley, et al., 1998) Km(3-O-Methyl glc)=17.3 ± 4.3mM (rat liver) (Ciaraldi, et al., 1986) **Km(D-glc)=21.7 ± 1.8mM** (rat liver) (Ciaraldi, et al., 1986) **Km(D-glc)=66±14mM** (rat? hepatocytes) (Elliott and Craik, 1982) **Km(D-fruct) = 66mM** (Walmsley, et al., 1998) **Km(D-fruct)=212±32mM** (rat? hepatocytes) (Elliott and Craik, 1982) **Km(D-gal)=174±48mM** (rat? hepatocytes) (Elliott and Craik, 1982) **Km(D-gal) > 50mM** (GLUT2 enderocytes) (Walmsley, et al., 1998) **Km(D-gal) = 85.5 ± 10.7mM** (human, liver-type GLUT2) (Colville, et al., 1993) **Km(D-gal) = 92 ± 8.4mM** (human, liver-type GLUT2) (Arbuckle, et al., 1996) **accumulation rate** (human GLUT2)**:** v(deoxy-D-glc) = 4.33±0.15 pmol/min/oocyte v(gal) = 1.68±0.09 pmol/min/oocyte v(fru) = 0.78±0.09 pmol/min/oocyte **v(D-glc)=220±19**mmol/min/l of cell H2O (rat? hepatocytes) (Elliott and Craik, 1982) **v(D-gal)=288±48** mmol/min/l of cell H2O (rat? hepatocytes) (Elliott

	et al., 1993)"		and Craik, 1982) **v(D-fruct)=291±26** mmol/min/l of cell H2O (rat? hepatocytes) (Elliott and Craik, 1982) **Competitive inhibitor kinetics** (Colville, et al., 1993)
GALK	**Galactokinase** D-galactose [**gal**] + ATP [**atp**] ↔ D-galactose 1-phosphate [**gal1p**] + ADP [**adp**] EC:2.7.1.6 RHEA:13556 KEGG:R01092	**Protein** UniProt:P51570 (GALK1_HUMAN) homodimer P51570*2 **Gene** GALK, GALK1 **Disease** MIM:230200 (GALCT2 Galactosemia II)	SabioRK:P51570 **two-substrate ordered, ternary complex reaction** (Timson and Reece, 2003) **kcat(gal) = 8.7±5 1/s** (SABIORK:14785)(Timson and Reece, 2003) **km(atp) = 0.034±0.004mM** (SABIORK:14792)(Timson and Reece, 2003) **km(gal)=0.97±0.22 mM** (SABIORK:14785) (Timson and Reece, 2003) **km(gal) = 0.436mM** (SABIORK:45367), (Sangiuolo, et al., 2004) **Uncompetitive product inhibition** of GALK (adult rat liver) by gal1p with both 1mM and 5mM gal1p altering the Km for galactose from 0.150mM to 0.800mM. (Cuatrecasas and Segal, 1965) **ki(gal1p) = 5.3mM (5.0-5.7mM)** (Cuatrecasas and Segal, 1965) (1mM gal1p caused 15% inhibition, 5mM gal1p 50% inhibition) Galactokinase being rate limiting for galactose clearance (Schirmer, et al., 1986) -> [27,28, 50]
IMP	**Inositol monophosphatase** D-galactose 1-phosphate [**gal1p**] ↔ D-galactose [**gal**] + phosphate [**pi**]	**Protein** UniProt:P29218 (IMPA1_HUMAN) Homodimer P29218*2	**Competitive inhibition model:** **km(ino1p) = 0.320±0.050mM** (Slepak, et al., 2007) **km(gal1p) = 0.35mM** (similar kinetics gal1p to ino1p in vitro)

	EC:3.1.3.25	**Gene** IMPA1, IMPA	(Parthasarathy, et al., 1997) "Our kinetic analysis demonstrated that gal1p competitively inhibited human IMPase1 by increasing its Km for inositol-1p (ino1p) from 320±50μM to 980±70μM without changing the Vmax (Slepak, et al., 2007)."
GALT	**Galactose-1-phosphate uridyl transferase** UDP-D-glucose [**udpglc**] + D-galactose 1-phosphate [**gal1p**] ↔ D-glucose 1-phosphate [**glc1p**] + UDP-D-galactose [**udpgal**]. EC:2.7.7.12 RHEA:13992 KEGG:R00955 The catalytic mechanism of GALT is **ping-pong kinetics** with covalent intermediate UMP-enzyme (Facchiano and Marabotti, 2010).	**Protein** UniProt:P07902 (GALT_HUMAN) homodimer P07902*2 **Gene** GALT **Disease** MIM:230400 (GALCT Galactosemia)	**Mutation analysis** (Quimby, et al., 1996) **km(gal1p) = 0.57±0.14mM** (human, wildtype) (Quimby, et al., 1996) **km(udpglc) = 0.21±0.04mM** (human, wildtype) (Quimby, et al., 1996) **Mutation analysis** (Tang, et al., 2012) **km(gal1p) = 1.25±0.36mM** (human, wildtype) (Tang, et al., 2012) **km(udpglc) = 0.43±0.09mM** (human, wildtype) (Tang, et al., 2012) (?species, 4°C) (Geeganage and Frey, 1998) **km(udpglc) = 0.5±0.1mM** **v(glc1p) = 281± 18 1/s** **km(glc1p) = 0.37±0.18mM** **v(glc1p) = 226± 10 1/s** **km(gal1p) = 0.061±0.020mM** **v(glc1p) = 166± 13 1/s** **Potent linear competent inhibitors UTP and UDP of UDP-glucose** (Segal and Rogers, 1971)**:** **Ki(UTP) = 0.13mM** (rat, liver)

			Ki(UDP) = 0.35mM (rat, liver) Ki(UMP) = 2.3mM (rat, liver) Ki(UDP-glucuronic acid)=0.40mM (rat, liver)
GALE	**UDP-glucose 4-epimerase** UDP-D-glucose [**udpglc**] ↔ UDP-D-galactose [**udpgal**] EC:5.1.3.2 RHEA:22171 KEGG:R00291 Alternative activity with ! GlcNAc ? UDP-GalNAc <-> UDP-GlcNAc	**Protein** UniProt:Q14376 (GALE_HUMAN) homodimer Q14376*2 **Gene** GALE **Disease** MIM:230350	**Mutation analysis**(Timson, 2005) **km(udpgal)=0.069±0.012mM** (human, wildtype) (Timson, 2005) **kcat(udpgal) = 36±1.4 1/s** (human, wildtype) (Timson, 2005) **km(udpgal) = 0.15 ± 0.02mM** (human, wildtype) (Wohlers and Fridovich-Keil, 2000) km(udpgal, V94M) = 0.27 ± 0.01mM (human, V94M) (Wohlers and Fridovich-Keil, 2000) **km(udpgal)=0.140± 0.007mM** (human, wildtype) (SABIORK:19823) (Winans and Bertozzi, 2002) **km(udpgal)=0.120± 0.04mM** (human, wildtype) (SABIORK:46260) (Wasilenko, et al., 2005) **kcat= 33.8±11.2** (human, wildtype) (SABIORK:16222) (Thoden, et al., 2002) **km(udpgal) = 0.230±0.06mM** (human, wildtype) (SABIORK:46263) (Quimby, et al., 1997) “Ethanol treatment increases the NADH/NAD ratio in liver (Keppler, et al., 1970) -> [2-4] and by this inhibits the GALE ->[3,5]. Under these conditions oxidation and elimination ->[6-8] of galactose are impaired. Combined galactose+ethanol treatment results in accumulation of gal1p and udpgal in rat liver. The formation of high amounts of udpgal leads to a change in the distribution of liver uracil nucleotides. A marked decrease of udpglc,

			utp, udp and ump is followed by an increase of the sum of uracil nucleotides. (Keppler, et al., 1970) "The GALE reaction is indicated as the rate-limiting step of galactose metabolism in rat liver by the ratio of galactose metabolites (Keppler, et al., 1970)". "The almost 4-fold increase of gal1p and updgal and the even stronger drop of the udpglc content in the ethanol treated liver avter a galactose load demonstrate the ethanol-induced inhibition of the GALE (Keppler, et al., 1970)". "Galactose provokes pronounced alterations of the uracil nucleotide contents in the liver, which are intensified by an inhibition of the GALE (Keppler, et al., 1970)"
UGP UGALP	**UDP-glucose pyrophosphorylase** D-glucose 1-phosphate [**glc1p**] + UTP [**utp**] ↔ UDP-glucose [**udglc**]+ diphosphate [**pp**] EC:2.7.7.9 RHEA:19892 KEGG:R00289 in both reaction directions, the enzyme displays simple Michaelis-Menten kinetics (Chang, et al., 1996) MgUTP is a product inhibitor that shows competitive inhibition with respect to UDP-Glc (Chang, et al., 1996) **UDP-galactose pyrophosphorylase**	**Protein** UniProt:Q16851 (UGPA_HUMAN) homooctamer Q16851*8 **Gene** UGP2, UGP1 "Previously we reported that stable transfection of human UDP-glucose pyrophosphorylase (hUGP2) rescued galactose-1-phosphate uridyltransferase (GALT)-deficient yeast from ``galactose toxicity." [Lai2002] "Previously, we showed that galactose-1-phosphate competitively	(human, liver, wildtype) (Chang, et al., 1996) **km(udpglc) = [0.031 - 0.051]mM** **km(pp) = [0.172 - 0.210] mM** **km(glc1p) = [0.172 - 0.174] mM** **km(utp) = [0.563 - 0.692] mM** **ki(utp) = 0.477± 41 mM** (competitive inhibition with respect to UDP-glc) **V_{fwd}/V_{rev} = 0.260** (human, liver, wildtype) (Duggleby, et al., 1996) **km(udpglc) = 0.049±0.004mM** **km(pp) = 0.166±0.013 mM** **km(glc1p) = 0.172±0.010 mM** **km(utp) = 0.563±0.115 mM** **ki(utp) = 0.643± 0.047 mM** (competitive inhibition with respect to UDP-glc)

	D-galactose-1-phosphate [**gal1p**] + UTP [**utp**] + ↔ UDP-D-galactose [**udpgal**] pyrophosphate [**pp**] EC:2.7.7.10 RHEA:14212 KEGG:R00502 Could circumvent GALT deficiency (Isselbacher) Appears to have very low activity, several magnitudes lower than GALDH [Segal1968] Activity locacted in glucose dependent enzyme. **1-2 % activity with gal1p (Km gal ~10-20mM)**	inhibited UDP-glucose pyrophosphorylase, leading to 66% reduction in UDP-glucose/galactose contents in GALT-deficient cells under galactose challenge [Slepak2007->Lai2002].” The formation of UDP-glucose is the major physiological function of UGP, however at slow rates, the enzyme also catalyzes the phosphorylation of UDP-galactose. (Knop and Hansen, 1970) “Under normal physiological conditions, this may not be significant, but, in a galactosemic patient, in which the normal galactose metabolism is impaired, the pyrophosphorylase may participate in an abnormal role.	**ki(udpglc) = 0.013± 4 mM** (competitive inhibition with respect to UTP?) (human, liver, wildtype) (Knop and Hansen, 1970) **keq**([udpglc][pp]/([UTP][glc1p])) = **0.15 – 0.16** **km(udpglc) = 50mM** **km(utp) = 48 mM** **km(glc1p) = 95±10 mM** **keq**([UTP][glc1p]/([udpglc][pp])) = 4.55±0.1 (Guynn, et al., 1974) (**0.22**) The saturating concentration for UDP-galactose is 10 times that of UDP-glucose: **km(udpgal)** = 10*km(udpglc) ~ **0.5mM** (human, liver, wildtype) (Knop and Hansen, 1970) **km(udpgal) = 0.420mM** (rabbit, liver, wildtype) (Turnquist, et al., 1974) udpgal was an adequate substrate at 10 times the concentration ofudpglc, showing 14.3% of udpglc (Calf) and 12.0% (Human). activity with udpgal 2-12% of udpglc (12% with 3mM udpgal) (human liver) (Turnquist, et al., 1974) The activity of UDPG:galactose-1-phosphate uridylyltransferase from rat liver under optimal conditions in vitro is less than 5% of the UDPG pyrophosphorylase activity (Keppler, et al., 1970)[Keppler1970 ->39,40]”
ALDR	**Aldose reductase (galactitol NAD 1-oxidoreductase)**	**Protein** UniProt:P15121 (ALDR_HUMAN)	SabioRK:P15121 **km(gal) = 40.0mM** (human brain) (SABIORK: 22893) (Wermuth, et

	D-galactose [**gal**] + NADPH [**nadph**] + H ↔ galactitol [**galtol**] + NADP [**nadp**] EC:1.1.1.21 RHEA:12792 -> RHEA:37967 KEGG:R01095 Aldolase reductase is specific for NADPH as cofactor (NADH ~10% of NADPH-dependent activity) (Wermuth and von Wartburg, 1982). "Aldolase reductase catalyzes the conversion of aldoses and a number of other aldehydes to the corresponding alcohol metabolites. It is one of several cytosolic, monomeric, NADPH-dependent aldehyde and ketone reductases of wide substrate specificity (Wermuth, et al., 1982)".	monomer P15121*1 **Gene** AKR1B1, ALDR1 Tissue distribution: Aldolase reductase activity has been demonstrated in brain, kidney, placenta, testis, lens, lung, heart, and pancreas, but appears to be absent from the liver. The enzyme is localized in the cytoplasm of the cell. [Wermuth1982].	al., 1982) **kcat(gal) = 0.40 1/s** (human brain) (SABIORK: 22893) (Wermuth, et al., 1982) **km(gal) = 110.0mM** (human brain) (SABIORK:15695) (Wermuth and von Wartburg, 1982)
PGM1	**Phosphoglucomutase-1** D-glucose 1-phosphate [**glc1p**] ↔ D-glucose 6-phosphate [**glc6p**] EC:5.4.2.2 KEGG:R00959 RHEA:23539	**Multiple isoforms (PGM1, PGM2)** **Protein** UniProt:P36871 (PGM1_HUMAN) monomer P36871*1 main isoform for glc1p ↔ glc6p reaction **Gene** PGM1 **Disease** Glycogen storage disease 14 (GSD14) [MIM:612934]	The equilibrium lies strongly toward G6P (Keq) and the reaction proceeds through a **ping-pong mechanism** involving aspartyl-phosphoenzyme and glucose-1,6-bisphosphate intermediates. [glc6p]/[glc1p] ~10-12 (Guynn, et al., 1974) DeltaG =-7.1 kJ/mol [Koenig2012] **km(glc1p) = 0.049mM** (human, RBC) (Quick, et al., 1974) **km(glc1p) = 0.045mM** (rat, heart) (Kashiwaya, et al., 1994) **km(glc6p) = 0.67mM** (rat, heart) (Kashiwaya, et al., 1994)

		Congenital disorder of glycosylation 1T (CDG1T) [MIM:614921]: A multisystem disorder caused by a defect in glycoprotein biosynthesis and characterized by under-glycosylated serum glycoproteins. **Protein** UniProt:Q96G03 (PGM2_HUMAN) **Gene** PGM2	**km(glc1p) = 0.083mM** (human, RBC, PGM1) (Accorsi, et al., 1989) ki(fru16bp) = 0.092mM (human, RBC, PGM1) (Accorsi, et al., 1989) The kinetic properties of PGM1 and PGM2 are essentially the same. PGM1 is specific for mutation of glucose, whereas PGM2 also has phosphoribomutase activities. (human, RBC) (Accorsi, et al., 1989)
PPASE	**Pyrophosphatase** Pyrophosphate [**pp**] + H2O → 2 phosphate [**pi**] EC:3.6.1.1 RHEA:24579 KEGG:R00004	**Protein** UniProt:Q15181 (IPYR_HUMAN) homodimer Q15181*2 **Gene** PPA1, IOPPP, PP	**km(pp) = 0.005mM** (rat liver) (Yoshida, et al., 1982) **km(pp) = 0.14mM** (human erythrocyte) (Thuillier, 1978) **km(pp) = 0.07mM** (rat liver) (Irie, et al., 1970) **Delta G0 = -23.56 kJ/mol** (Thuillier, 1978) **Delta G0 = -19.2 kJ/mol** (Guynn, et al., 1974)
NDKU	**Nucleoside diphosphokinase (ATP:UDP phosphotransferase)** ATP [**atp**] + UDP [**udp**] ↔ ADP [**adp**] + UTP [**udp**] EC: 2.7.4.6 RHEA:25101 KEGG:R00156	**Multiple isoforms**	Compulsory-order substituted-enzyme (**Ping Pong Bi Bi**) mechanism (Lam and Packham, 1986) **km(atp) = 0.38mM** (human, platelets) (Lam and Packham, 1986) **km(adp) = 0.024mM** (human, platelets) (Lam and Packham, 1986) **km(gtp) = 0.12mM** (human, platelets) (Lam and Packham, 1986) **km(atp) = 1.33mM** (rat, liver) (Kimura and Shimada, 1988) **km(adp) = 0.042mM** (rat, liver) (Kimura and Shimada, 1988) **km(udp) = 0.19mM**(rat, liver) (Kimura and Shimada, 1988)

			km(atp) = 1.80 mM (rat, liver) (Fukuchi, et al., 1994) **km(adp) = 0.066 mM** (rat, liver) (Fukuchi, et al., 1994) **km(utp) = 27.00mM** (rat, liver) (Fukuchi, et al., 1994) km(gtp) = 0.15mM (rat, liver) (Fukuchi, et al., 1994) km(gdp) = 0.049mM (rat, liver) (Fukuchi, et al., 1994)
NADPR	**NADP reductase** NADP [**nadp**] + H2→NADPH [**nadph**] Modeled via pentose phosphate pathway glucose-6-phosphate dehydrogenase D-glucose 6-phosphate [**glc6p**] + NADP [**nadp**] → 6-phospho-D-glucono-1,5-lactone + NADPH [**nadph**] + H EC: 1.1.1.49 RHEA:15844 KEGG:R00835	**Protein** UniProt:P11413 (G6PD_HUMAN) homotetramer (dimer of dimer) P11413*4 **Gene** G6PD	Delta G0 = -19.6 kJ/mol [Schuster1995] **km(glc6p) = 0.040±0.008 mM** (human, placenta) (Ozer, et al., 2001) **km(nadp) = 0.020±0.010 mM** (human, placenta) (Ozer, et al., 2001) **ki(nadph) = 0.0171±0.0032 mM** (human, placenta) (Ozer, et al., 2001) **km(glc6p) = 0.072 mM** (human, RBC) (Bautista, et al., 1992) **km(glc6p) = 0.069±0.003 mM** (human, recombinant) (Bautista, et al., 1992) **km(nadp) = 0.013 mM** (human, RBC) (Bautista, et al., 1992) **km(nadp) = 0.012±0.002 mM** (human, recombinant) (Bautista, et al., 1992) **km(nadph) = 0.015±0.002 mM** (human, RBC) (Bautista, et al., 1992) **km(nadph) = 0.014±0.003 mM** (human, recombinant) (Bautista, et al., 1992) **km(glc6p) = 0.326mM** (rat, liver) **km(glc6p) = 0.157mM** (rat, liver) (Corpas, et al., 1995; Corpas, et al., 1995) **km(nadp) = 0.108 mM** (rat, liver)

			km(nadp) = 0.258 mM (rat, liver) (Corpas, et al., 1995; Corpas, et al., 1995) **ki(nadhp) = 0.010 mM**(rat, liver) **ki(nadhp) = 0.021 mM**(rat, liver) (Corpas, et al., 1995; Corpas, et al., 1995)

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