### **Supplementary *Table 1*** - Reactions and transporters in human galactose metabolism and kinetic parameters.

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| Id | 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]  Mechanism  TCDB:2.A.1.1 (glucose transporter subfamily)  Protein/Structure  [UniProt:P11168](http://www.uniprot.org/uniprot/P11168) (GTR2\_HUMAN)  Gene  SLC2A2, GLUT2  Disease  [MIM:227810](http://omim.org/entry/227810) (Fanconi-Bickel syndrome; FBS)  Galactose and glucose transported via GLUT2 (competitive inhibition kinetics) ([Brown, 2000](#_ENREF_4); [Colville, et al., 1993](#_ENREF_7))  Deficient transport of galactose into hepatocytes in human patients with defective GLUT2 transporters (Fanconi-Bickel syndrome) resulting in galactose malabsorption/intolerance ([Brown, 2000](#_ENREF_4); [Leslie, 2003](#_ENREF_27)). | **km(D-glc)=21.7 ± 1.8mM** (rat liver) ([Ciaraldi, et al., 1986](#_ENREF_6))  **km(D-glc)=66±14mM** (rat hepatocytes) ([Elliott and Craik, 1982](#_ENREF_12))  **km(D-glc)=17mM** (perfused rat liver, cited) ([Elliott and Craik, 1982](#_ENREF_12))  **km(D-glc)=30mM** (rat hepatocytes, cited) ([Elliott and Craik, 1982](#_ENREF_12))  km(3-O-MG)=42.3±4.1mM (human liver) ([Gould, et al., 1991](#_ENREF_17); [Walmsley, et al., 1998](#_ENREF_43))  km(3-O-Methyl glc)=17.3 **±** 4.3mM (rat liver) ([Ciaraldi, et al., 1986](#_ENREF_6))  **Vmax(D-glc)=220±19**mmol/min/l of cell H2O (rat hepatocytes) ([Elliott and Craik, 1982](#_ENREF_12))  **Vmax(D-glc)=345**mmol/min/l of cell H2O (perfused rat liver, cited) ([Elliott and Craik, 1982](#_ENREF_12))  **Vmax(D-glc)=70**mmol/min/l of cell H2O (rat hepatocytes, cited) ([Elliott and Craik, 1982](#_ENREF_12))  **km(D-gal)=174±48mM** (rat hepatocytes) ([Elliott and Craik, 1982](#_ENREF_12))  **km(D-gal)=100mM** (rat hepatocytes, cited) ([Elliott and Craik, 1982](#_ENREF_12))  **km(D-gal)>50mM** (GLUT2 enderocytes) ([Walmsley, et al., 1998](#_ENREF_43))  **km(D-gal)=85.5 ± 10.7mM** (human, liver-type GLUT2) ([Colville, et al., 1993](#_ENREF_7))  **km(D-gal)=92 ± 8.4mM** (human, liver-type GLUT2) ([Arbuckle, et al., 1996](#_ENREF_2))  **km(D-gal)~27.7mM** (dog liver, multiple indicator dilution curves ([Goresky, et al., 1973](#_ENREF_16))  **Vmax (D-gal)=288±48** mmol/min/l of cell H2O (rat hepatocytes) ([Elliott and Craik, 1982](#_ENREF_12))  **Vmax (D-gal)=160**mmol/min/l of cell H2O (rat hepatocytes, cited) ([Elliott and Craik, 1982](#_ENREF_12))  Km(D-fru)=66mM ([Walmsley, et al., 1998](#_ENREF_43))  Km(D-fru)=67mM (perfused rat liver, cited) ([Elliott and Craik, 1982](#_ENREF_12))  Km(D-fru)=>100mM (rat hepatocytes, cited) ([Elliott and Craik, 1982](#_ENREF_12))  v(D-fru)=291±26 mmol/min/l of cell H2O (rat hepatocytes) ([Elliott and Craik, 1982](#_ENREF_12))  Vmax (D-fru)=50mmol/min/l of cell H2O (perfused rat liver, cited) ([Elliott and Craik, 1982](#_ENREF_12))  Vmax (D-fru)=>160mmol/min/l of cell H2O (rat hepatocytes, cited) ([Elliott and Craik, 1982](#_ENREF_12))  **Accumulation rate** (human GLUT2)  v(deoxy-D-glc) = 4.33**±**0.15 pmol/min/oocyte  v(D-gal) = 1.68**±**0.09 pmol/min/oocyte  v(D-fru) = 0.78**±**0.09 pmol/min/oocyte |
| **GALK** | **Galactokinase**  D-galactose [gal] + ATP [atp] ↔ D-galactose 1-phosphate [gal1p] + ADP [adp]  **Reaction**  EC:2.7.1.6  [RHEA:13556](http://www.ebi.ac.uk/rhea/reaction.xhtml?id=13556)  [KEGG:R01092](http://www.genome.jp/dbget-bin/www_bget?rn:R01092)  Protein  [UniProt:P51570](http://www.uniprot.org/uniprot/P51570)(GALK1\_HUMAN)  homodimer P51570\*2  **Gene**  GALK, GALK1  Disease  [MIM:230200](http://www.omim.org/entry/230200) (GALCT2 Galactosemia II)  Galactokinase being rate limiting for galactose clearance ([Schirmer, et al., 1986](#_ENREF_34)) | **Two-substrate ordered, ternary complex reaction** ([Timson and Reece, 2003](#_ENREF_41))  **kcat(gal) = 8.7±5 1/s** (SABIORK:14785)([Timson and Reece, 2003](#_ENREF_41))  **km(atp) = 0.034±0.004mM** (SABIORK:14792)([Timson and Reece, 2003](#_ENREF_41))  **km(atp) = 0.12mM** (adult, rat liver){Cuatrecasas1965}  **km(gal)=0.97±0.22mM** (SABIORK:14785) ([Timson and Reece, 2003](#_ENREF_41))  **km(gal) = 0.436mM** (SABIORK:45367), ([Sangiuolo, et al., 2004](#_ENREF_33))  **km(gal) = 0.15mM** (adult, rat liver){Cuatrecasas1965}  km(gal) = 0.65mM (newborn, rat liver){Cuatrecasas1965}  km(gal) = 0.91mM (18 day fetal, rat liver){Cuatrecasas1965}  **km(gal) = 0.14±0.01mM** (SEM, N=6, adult rat liver) {Walker1968}  km(gal) = 0.15±0.01mM (SEM, N=4, neonatal rat liver) {Walker1968}  km(gal) = 0.14±0.01mM (SEM, N=4, foetal rat liver) {Walker1968}  **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 (1mM gal1p caused 15% inhibition, 5mM gal1p 50% inhibition)  **ki(gal1p) = 5.3mM (5.0-5.7mM)** (adult rat liver) ([Cuatrecasas and Segal, 1965](#_ENREF_10))  **km(gal)<0.83mM** (dog liver, multiple indicator dilution curves) ([Goresky, et al., 1973](#_ENREF_16)) |
| **IMP** | **Inositol monophosphatase**  D-galactose 1-phosphate [gal1p] ↔ D-galactose [gal] + phosphate [pi]  **Reaction**  EC:3.1.3.25  Protein  [UniProt:P29218](http://www.uniprot.org/uniprot/P29218) (IMPA1\_HUMAN)  homodimer P29218\*2  Gene  IMPA1, IMPA  Normal substrate inositol-1p (ino1p) | Competitive inhibition model  Kinetic analysis demonstrated that gal1p competitively inhibited human IMP1 by increasing Km for inositol-1p (ino1p) from 320±50µM to 980±70µM without changing the Vmax ([Slepak, et al., 2007](#_ENREF_36))  km(ino1p) = 0.320±0.050mM ([Slepak, et al., 2007](#_ENREF_36))  km(gal1p) = 0.35mM (similar kinetics gal1p to ino1p in vitro) ([Parthasarathy, et al., 1997](#_ENREF_29)) |
| **GALT** | **Galactose-1-phosphate uridyl transferase**  UDP-D-glucose [**udpglc**] + D-galactose 1-phosphate [**gal1p**] ↔ D-glucose 1-phosphate [**glc1p**] + UDP-D-galactose [**udpgal**].  **Reaction**  EC:2.7.7.12  [RHEA:13992](http://www.ebi.ac.uk/rhea/reaction.xhtml?id=13992)  [KEGG:R00955](http://www.genome.jp/dbget-bin/www_bget?rn:R00955)  Protein  [UniProt:P07902](http://www.uniprot.org/uniprot/P07902) (GALT\_HUMAN)  homodimer P07902\*2  Gene  GALT  Disease  [MIM:230400](http://www.omim.org/entry/230400) (GALCT Galactosemia) | The catalytic mechanism of GALT is **ping-pong kinetics** with covalent intermediate UMP-enzyme ([Facchiano and Marabotti, 2010](#_ENREF_13)).  **Mutation analysis** ([Quimby, et al., 1996](#_ENREF_32))  **km(gal1p) = 0.57±0.14mM** (human, wildtype) ([Quimby, et al., 1996](#_ENREF_32))  **km(udpglc) = 0.21±0.04mM** (human, wildtype) ([Quimby, et al., 1996](#_ENREF_32))  **Mutation analysis**  ([Tang, et al., 2012](#_ENREF_37))  **km(gal1p) = 1.25±0.36mM** (human, wildtype) ([Tang, et al., 2012](#_ENREF_37))  **km(udpglc) = 0.43±0.09mM** (human, wildtype) ([Tang, et al., 2012](#_ENREF_37))  (?species, 4°C) ([Geeganage and Frey, 1998](#_ENREF_15))  **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](#_ENREF_35))**:**  **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**]  **Reaction**  EC:5.1.3.2  [RHEA:22171](http://www.ebi.ac.uk/rhea/reaction.xhtml?id=22171)  [KEGG:R00291](http://www.genome.jp/dbget-bin/www_bget?rn:R00291)  **Protein**  [UniProt:Q14376](http://www.uniprot.org/uniprot/Q14376) (GALE\_HUMAN)  homodimer Q14376\*2  **Gene**  GALE  Disease  [MIM:230350](http://www.omim.org/entry/230350) (GALE deficiency)  Alternative activity with GlcNAc:  UDP-GalNAc ↔ UDP-GlcNAc  “Ethanol treatment increases the NADH/NAD ratio in liver ([Keppler, et al., 1970](#_ENREF_21)) [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](#_ENREF_21))  “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](#_ENREF_21))“.  “The almost 4-fold increase of gal1p and updgal and the even stronger drop of the udpglc content in the ethanol treated liver after a galactose load demonstrates the ethanol-induced inhibition of the GALE” ([Keppler, et al., 1970](#_ENREF_21)).  “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](#_ENREF_21)) | **Mutation analysis**([Timson, 2005](#_ENREF_40))  **km(udpgal)=0.069±0.012mM** (human, wildtype) ([Timson, 2005](#_ENREF_40))  **kcat(udpgal) = 36±1.4 1/s** (human, wildtype) ([Timson, 2005](#_ENREF_40))  **km(udpgal) = 0.15 ± 0.02mM** (human, wildtype) ([Wohlers and Fridovich-Keil, 2000](#_ENREF_48))  km(udpgal, V94M) = 0.27 ± 0.01mM (human, V94M) ([Wohlers and Fridovich-Keil, 2000](#_ENREF_48))  **km(udpgal)=0.140± 0.007mM** (human, wildtype) (SABIORK:19823) ([Winans and Bertozzi, 2002](#_ENREF_47))  **km(udpgal)=0.120± 0.04mM** (human, wildtype) (SABIORK:46260) ([Wasilenko, et al., 2005](#_ENREF_44))  **kcat= 33.8±11.2** (human, wildtype) (SABIORK:16222) ([Thoden, et al., 2002](#_ENREF_38))  **km(udpgal) = 0.230±0.06mM** (human, wildtype) (SABIORK:46263) ([Quimby, et al., 1997](#_ENREF_31)) |
| **UGP**  **UGALP** | **UDP-glucose pyrophosphorylase**  D-glucose 1-phosphate [**glc1p**] + UTP [**utp**] ↔ UDP-glucose [**udglc**]+ diphosphate [**pp**]  **Reaction**  EC:2.7.7.9  [RHEA:19892](http://www.ebi.ac.uk/rhea/reaction.xhtml?id=19892)  [KEGG:R00289](http://www.genome.jp/dbget-bin/www_bget?rn:R00289)  **Protein**  [UniProt:Q16851](http://www.uniprot.org/uniprot/Q16851) (UGPA\_HUMAN)  homooctamer Q16851\*8  **Gene**  UGP2, UGP1  **UDP-galactose pyrophosphorylase**  D-galactose-1-phosphate [gal1p] + UTP [utp] + ↔ UDP-D-galactose [udpgal] pyrophosphate [pp]  **Reaction**  EC:2.7.7.10  [RHEA:14212](http://www.ebi.ac.uk/rhea/reaction.xhtml?id=14212)  [KEGG:R00502](http://www.genome.jp/dbget-bin/www_bget?rn:R00502)  **Protein**  [UniProt:Q16851](http://www.uniprot.org/uniprot/Q16851) (UGPA\_HUMAN)  homooctamer Q16851\*8  **Gene**  UGP2, UGP1  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](#_ENREF_24)) [Segal1968].  Not significant in normal physiological conditions, but in galactosemic patients could circumvent GALT deficiency (Isselbacher ?).  Stable transfection of human UGP (hUGP2) rescued galactose GALT deficient yeast from “galactose toxicity [Lai2002]. | Enzyme displays simple Michaelis-Menten kinetics in both directions ([Chang, et al., 1996](#_ENREF_5))  MgUTP is a product inhibitor that shows competitive inhibition with respect to UDP-Glc ([Chang, et al., 1996](#_ENREF_5))  (human, liver, wildtype) ([Chang, et al., 1996](#_ENREF_5))  **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)  **Vfwd/Vrev = 0.260**  (human, liver, wildtype) ([Duggleby, et al., 1996](#_ENREF_11))  **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)  **ki(udpglc) = 0.013± 4 mM** (competitive inhibition with respect to UTP?)  (human, liver, wildtype) ([Knop and Hansen, 1970](#_ENREF_24))  **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](#_ENREF_18)) (**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](#_ENREF_24))  **km(udpgal) = 0.420mM** (rabbit, liver, wildtype) ([Turnquist, et al., 1974](#_ENREF_42))  udpgal was an adequate substrate at 10 times the concentration of udpglc, 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](#_ENREF_42))  “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](#_ENREF_22))[Keppler1970 ->39,40]  **gal1p as competitive inhibitor of glc1p**  “Previously, we showed that galactose-1-phosphate competitively inhibited UDP-glucose pyrophosphorylase, leading to 66% reduction in UDP-glucose/galactose contents in GALT-deficient cells under galactose challenge” [Slepak2007->Lai2002]. |
| **ALDR** | **Aldose reductase (galactitol NAD 1-oxidoreductase)**  D-galactose [**gal**] + NADPH [**nadph**] + H ↔ galactitol [**galtol**] + NADP [**nadp**]  **Reaction**  EC:1.1.1.21  [RHEA:12792](http://www.ebi.ac.uk/rhea/reaction.xhtml?id=12792) -> RHEA:37967  [KEGG:R01095](http://www.genome.jp/dbget-bin/www_bget?rn:R01095)  **Protein**  [UniProt:P15121](http://www.uniprot.org/uniprot/P15121)(ALDR\_HUMAN)  monomer P15121\*1  **Gene**  AKR1B1, ALDR1  Aldolase reductase is specific for NADPH as cofactor (NADH ~10% of NADPH-dependent activity) ([Wermuth and von Wartburg, 1982](#_ENREF_46)).  “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](#_ENREF_45))”. | km(gal) = 40.0mM (human brain) (SABIORK:22893) ([Wermuth, et al., 1982](#_ENREF_45))  kcat(gal) = 0.40 1/s (human brain) (SABIORK:22893) ([Wermuth, et al., 1982](#_ENREF_45))  km(gal) = 110.0mM (human brain) (SABIORK:15695) ([Wermuth and von Wartburg, 1982](#_ENREF_46)) |
| **PGM1** | **Phosphoglucomutase-1**  D-glucose 1-phosphate [**glc1p**] ↔ D-glucose 6-phosphate [**glc6p**]  **Reaction**  EC:5.4.2.2  [KEGG:R00959](http://www.genome.jp/dbget-bin/www_bget?rn:R00959)  [RHEA:23539](http://www.ebi.ac.uk/rhea/reaction.xhtml?id=23539)  **Protein (multiple isoforms PGM1, PGM2)**  [UniProt:P36871](http://www.uniprot.org/uniprot/P36871) (PGM1\_HUMAN)  monomer P36871\*1  main isoform for glc1p ↔ glc6p reaction  **Gene**  PGM1  **Disease**  [MIM:612934](http://www.omim.org/entry/612934) (Glycogen storage disease 14)  [MIM:614921](http://www.omim.org/entry/614921) (Congenital disorder of glycosylation 1T CDG1T)  **Protein**  [UniProt:Q96G03](http://www.uniprot.org/uniprot/Q96G03) (PGM2\_HUMAN)  **Gene**  PGM2  CDG1T - A multisystem disorder caused by a defect in glycoprotein biosynthesis and characterized by under-glycosylated serum glycoproteins. | The equilibrium lies strongly toward glc6p and reaction proceeds through **ping-pong mechanism** ([Guynn, et al., 1974](#_ENREF_18))  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](#_ENREF_1))  **[glc6p]/[glc1p] ~10-12** ([Guynn, et al., 1974](#_ENREF_18))  DeltaG =-7.1 kJ/mol ([König, et al., 2012](#_ENREF_25))  **km(glc1p) = 0.049mM** (human, RBC) ([Quick, et al., 1974](#_ENREF_30))  **km(glc1p) = 0.045mM** (rat, heart) ([Kashiwaya, et al., 1994](#_ENREF_20))  **km(glc6p) = 0.67mM** (rat, heart) ([Kashiwaya, et al., 1994](#_ENREF_20))  **km(glc1p) = 0.083mM** (human, RBC, PGM1) ([Accorsi, et al., 1989](#_ENREF_1))  ki(fru16bp) = 0.092mM (human, RBC, PGM1) ([Accorsi, et al., 1989](#_ENREF_1)) |
| **PPASE** | **Pyrophosphatase**  Pyrophosphate [**pp**] + H2O → 2 phosphate [**pi**]  **Reaction**  EC:3.6.1.1  [RHEA:24579](http://www.ebi.ac.uk/rhea/reaction.xhtml?id=24579)  [KEGG:R00004](http://www.genome.jp/dbget-bin/www_bget?rn:R00004)  **Protein**  [UniProt:Q15181](http://www.uniprot.org/uniprot/Q15181) (IPYR\_HUMAN)  homodimer Q15181\*2  **Gene**  PPA1, IOPPP, PP | **km(pp) = 0.005mM** (rat liver) ([Yoshida, et al., 1982](#_ENREF_49))  **km(pp) = 0.14mM** (human erythrocyte) ([Thuillier, 1978](#_ENREF_39))  **km(pp) = 0.07mM** (rat liver) ([Irie, et al., 1970](#_ENREF_19))  **Delta G0 = -23.56 kJ/mol** ([Thuillier, 1978](#_ENREF_39))  **Delta G0 = -19.2 kJ/mol** ([Guynn, et al., 1974](#_ENREF_18)) |
| **NDKU** | **Nucleoside diphosphokinase (ATP:UDP phosphotransferase)**  ATP [**atp**] + UDP [**udp**] ↔ ADP [**adp**] + UTP [**udp**]  **Reaction**  EC: 2.7.4.6  [RHEA:25101](http://www.ebi.ac.uk/rhea/reaction.xhtml?id=25101)  [KEGG:R00156](http://www.genome.jp/dbget-bin/www_bget?rn:R00156)  **Multitude of isoforms** | Compulsory-order substituted-enzyme (**Ping Pong Bi Bi**) mechanism ([Lam and Packham, 1986](#_ENREF_26))  **km(atp) = 0.38mM** (human, platelets) ([Lam and Packham, 1986](#_ENREF_26))  **km(adp) = 0.024mM** (human, platelets) ([Lam and Packham, 1986](#_ENREF_26))  **km(gtp) = 0.12mM** (human, platelets) ([Lam and Packham, 1986](#_ENREF_26))  **km(atp) = 1.33mM** (rat, liver) ([Kimura and Shimada, 1988](#_ENREF_23))  **km(adp) = 0.042mM** (rat, liver) ([Kimura and Shimada, 1988](#_ENREF_23))  **km(udp) = 0.19mM**(rat, liver) ([Kimura and Shimada, 1988](#_ENREF_23))  **km(atp) = 1.80 mM** (rat, liver) ([Fukuchi, et al., 1994](#_ENREF_14))  **km(adp) = 0.066 mM** (rat, liver) ([Fukuchi, et al., 1994](#_ENREF_14))  **km(utp) = 27.00mM** (rat, liver) ([Fukuchi, et al., 1994](#_ENREF_14))  km(gtp) = 0.15mM (rat, liver) ([Fukuchi, et al., 1994](#_ENREF_14))  km(gdp) = 0.049mM (rat, liver) ([Fukuchi, et al., 1994](#_ENREF_14)) |
| **NADPR** | **NADP reductase**  NADP [**nadp**] + H2→NADPH [**nadph**]  Modeled via glucose-6-phosphate dehydrogenase in pentose phosphate pathway  D-glucose 6-phosphate [**glc6p**] + NADP [**nadp**] → 6-phospho-D-glucono-1,5-lactone + NADPH [**nadph**] + H  **Reaction**  EC: 1.1.1.49  [RHEA:15844](http://www.ebi.ac.uk/rhea/reaction.xhtml?id=15844)  [KEGG:R00835](http://www.genome.jp/dbget-bin/www_bget?rn:R00835)  **Protein**  [UniProt:P11413](http://www.uniprot.org/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](#_ENREF_28))  km(nadp) = 0.020±0.010 mM (human, placenta) ([Ozer, et al., 2001](#_ENREF_28))  ki(nadph) = 0.0171±0.0032 mM (human, placenta) ([Ozer, et al., 2001](#_ENREF_28))  km(glc6p) = 0.072 mM (human, RBC) ([Bautista, et al., 1992](#_ENREF_3))  km(glc6p) = 0.069±0.003 mM (human, recombinant) ([Bautista, et al., 1992](#_ENREF_3))  km(nadp) = 0.013 mM (human, RBC) ([Bautista, et al., 1992](#_ENREF_3))  km(nadp) = 0.012±0.002 mM (human, recombinant) ([Bautista, et al., 1992](#_ENREF_3))  km(nadph) = 0.015±0.002 mM (human, RBC) ([Bautista, et al., 1992](#_ENREF_3))  km(nadph) = 0.014±0.003 mM (human, recombinant) ([Bautista, et al., 1992](#_ENREF_3))  km(glc6p) = 0.326mM (rat, liver)  km(glc6p) = 0.157mM (rat, liver)  ([Corpas, et al., 1995](#_ENREF_8); [Corpas, et al., 1995](#_ENREF_9))  km(nadp) = 0.108 mM (rat, liver)  km(nadp) = 0.258 mM (rat, liver) ([Corpas, et al., 1995](#_ENREF_8); [Corpas, et al., 1995](#_ENREF_9))  ki(nadhp) = 0.010 mM(rat, liver)  ki(nadhp) = 0.021 mM(rat, liver) ([Corpas, et al., 1995](#_ENREF_8); [Corpas, et al., 1995](#_ENREF_9)) |
| **ATPS** | **ATP synthesis**  ADP [**adp**] + phosphate [**pi**] → ATP [**atp**]  Modeled via general ATP producing reaction representative for ATP production via glycolysis and oxidative phosphorylation |  |
| **GTF**  **GTFGAL**  **GTFGLC** | **Glycosyltransferase**  Aceptor [**gac**] + UDP-glucose [**udpglc**] -> Acceptor-glucose[**gacglc**] + UDP [**udp**]  Aceptor [**gac**] + UDP-glucose [**udpgal**] -> Acceptor-glucose[**gacgal**] + UDP [**udp**]  Enzymes that transfer mono- or oligosaccharides from donor molecules to growing oligosaccharide chains or proteins are called glycosyltransferases (Gtfs) |  |
| **GALDH** | **Galactose 1-dehydrogenase**  D-galactose + NAD+ ↔D-galactono-1,4-lactone + NADH + H+  EC.1.1.1.48 (Brenda only bacteria)  D-galactose -> galactonate  (first enzyme in oxidative pathway)  [Segal1968 -> Cuatrecasas1966,15]  Alternative pathway to xylulose.  D-Galactose + Oxygen + H2O <=> D-Galactonate + Hydrogen peroxide  EC:1.1.3.9  [KEGG:R01098](http://www.genome.jp/dbget-bin/www_bget?rn:R01098)  (only bacteria) |  |

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