### **­Supplementary Table 2** - Metabolites in hepatic galactose metabolism.

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| --- | --- | --- | --- |
| Id | Name (mass) Annotation | Initial Concentration | Concentrations, Comments |
| **glc** | **D-glucose**  C6H12O6  Charge: 0  (Mw 180.2)  CHEBI:4167  KEGG:C00031 | **5.5mM** ([König, et al., 2012](#_ENREF_7)) | [**glc**] = **5.5mM** ([König, et al., 2012](#_ENREF_7))  3-10mM (depending on physiological state) |
| **gal** | **D-galactose**  C6H12O6  Charge: 0  (Mw 180.2)  CHEBI:28061  KEGG:C0012c4 | **0.00012mM** (no galactose)  **0.00144mM** (GALT deficient)  **0.0013-0.0027mM** (GALE deficient) | **plasma of post-absorptive humans** (data considerable lower (3-18-fold) than conventional enzymatic assay) ([Schadewaldt, et al., 2000](#_ENREF_12))  **[gal] = 0.12±0.03µM** (n=16) healthy subjects  [**gal**] **= 1.44±0.54µM** (n=10) classical galactosemia (**GALT deficiency**)  **[gal] = 0.17±0.07µM (n=5)** obligate heterozygous parents of classical galactosemia  [gal] = 0.11±0.04µM (n=15) diabetic patients  **GALE deficient patients** (blood) ([Yamaguchi, et al., 1989](#_ENREF_17))  **[gal]=**24-29mg/L (**0.013-0.016mM**)  [**gal**]= 48mg/L (**0.027mM**)  **Neonatal control (blood):**  [gal]=13±6 mg/L (**0.0072±0.0033mM**) ([Yamaguchi, et al., 1989](#_ENREF_17))  **normal values**:  [gal]=**0.015±0.009mM** (range 0-0.044mM) ([Orfanos, et al., 1986](#_ENREF_10))  Cut-off values for newborn screening blood for galactosemias: “If gal > 60mg/L (0.033mM) or gal1P > 150mg/L (0.058mM).” ([Yamaguchi, et al., 1989](#_ENREF_17)) |
| **glc1p** | **D-glucose 1-phosphate**  C6H13O9P  Charge: 0  (Mw 260.1)  CHEBI:29042  KEGG:C00103 | **0.012mM** (no galactose)  **0.011mM** (1h galactose)  **0.012mM** (1h galactose, GALE inhibition) | [**glc1p**] = **0.012mM** ([König, et al., 2012](#_ENREF_7))  ([Keppler, et al., 1970](#_ENREF_6))  **[glc1p] =0.010 ±0.004µmol/gww** (~**0.011mM**)(starved + galactose 1h, rat, liver)  **[glc1p] =0.011 ±0.005µmol/gww** (**~0.012mM**) (ethanol, starved + galactose 1h, rat, liver)  ([Guynn, et al., 1974](#_ENREF_3))  **[glc1p] = 0.0075±0.0010 µmol/gWW** (**~0.0083mM**) (rat liver, starved)  **[glc1p] = 0.0115±0.008 µmol/gWW** (**~0.0127mM**) (rat liver, fed ad libitum)  **[glc1p] = 0.0132±0.0007 µmol/gWW** (**~0.0146mM**) (rat liver, meal fed)  **[glc6p]/[glc1p] ~10-12** |
| **glc6p** | **D-glucose 6-phosphate**  C6H11O9P  Charge: -2  (Mw 258.1)  CHEBI:58225  KEGG:C00668 | **0.12mM** (no galactose)  **0.29mM** (1h galactose)  **0.30mM** (1h galactose, GALE inhibition) | [**glc6p**] = **0.12mM** ([König, et al., 2012](#_ENREF_7))  ([Guynn, et al., 1974](#_ENREF_3))  **[glc6p] = 0.078±0.011 µmol/gWW** (**~0.086mM**) (rat liver, starved)  **[glc6p] = 0.147±0.012 µmol/gWW** (**~0.163mM**) (rat liver, fed ad libitum)  **[glc6p] = 0.157±0.007 µmol/gWW** (**~0.174mM**) (rat liver, meal fed)  **[glc6p]/[glc1p] ~10-12**  ([Keppler, et al., 1970](#_ENREF_6))  **[glc6p] =0.26 ±0.06µmol/gww** (**~0.29mM**) (starved + galactose 1h, rat, liver)  **[glc6p] =0.30 ±0.13µmol/gww** (**~0.33mM**) (ethanol, starved + galactose 1h, rat, liver)  **[glc6p]/[glc1p] =22.2 ±5.9** (starved + galactose 1h, rat, liver)  **[glc6p]/[glc1p] =22.8 ±5.9** (ethanol, starved + galactose 1h, rat, liver) |
| **gal1p** | **D-galactose 1-phosphate**  C6H11O9P  Charge: -2  (Mw 258.1)  CHEBI:58336  KEGG:C00446 | **0.001mM** (no galactose)  **0.20mM** (1h galactose)  **0.77mM** (1h galactose, GALE inhibition)  **1.2mM** (GALT deficient, glucose)  **5.2mM** (GALT deficient, galactose) | ([Lai, et al., 2003](#_ENREF_8)) (human cells)  [**gal1p**] **= ND** (not detectable) (Control glucose medium) [**gal1p**] **= 0.2±0.01mM** (Control galactose medium)  ([Keppler, et al., 1970](#_ENREF_6))  **[gal1p] =0.18 ±0.04µmol/gww** (**~0.2mM**)(starved + galactose 1h, rat, liver)  **[gal1p] =0.69 ±0.11µmol/gww** (**~0.77mM**) (ethanol, starved + galactose 1h, rat, liver)  ([Lai, et al., 2003](#_ENREF_8)) (human cells)  [**gal1p**] **= 1.2±0.4mM** (**GALT-deficient** glucose medium) [**gal1p**] **= 5.2±0.02mM** (**GALT-deficient** galactose medium)  **GALT deficiency** detected (blood)  [gal1p] > **3.0mM** (human cells) ([Diepenbrock, et al., 1992](#_ENREF_1))  **GALE deficient patients** (blood) ([Yamaguchi, et al., 1989](#_ENREF_17))  [gal1p]=330-360mg/L (**1.28-1.39mM**)  [gal1p]=474 mg/L (**1.84mM**) ([Yamaguchi, et al., 1989](#_ENREF_17))  **Neonatal control (blood):**  gal1P=15±11 mg/L (**0.058±0.042mM**) ([Yamaguchi, et al., 1989](#_ENREF_17))  **normal values:**  gal1P = **0.038±0.027 mM** (range 0-0.096µM) ([Orfanos, et al., 1986](#_ENREF_10))  Mean concentration of gal1p (blood) was **0.15mM** in cases below the cut-off of 0.74mM ([Diepenbrock, et al., 1992](#_ENREF_1)) |
| **udpglc** | **UDP-D-glucose**  (Mw 564.3)  [CHEBI:58885](http://www.ebi.ac.uk/chebi/searchId.do?chebiId=CHEBI:58885)  [KEGG:C00029](http://www.genome.jp/dbget-bin/www_bget?cpd:C00029) | **0.34mM** (no galactose)  **0.27mM** (1h galactose)  **0.17mM** (1h galactose, GALE inhibition) | [**udpglc**] = **0.38mM** ([König, et al., 2012](#_ENREF_7))  **[udpglc] = 0.32±0.05 µmol/gWW** (~**0.36mM**) (rat liver)([Keppler and Decker, 1969](#_ENREF_4))  **[udpglc] = 0.26±0.07 µmol/gWW** (**~0.29mM**) (rat liver)([Keppler, et al., 1969](#_ENREF_5))  ([Keppler, et al., 1970](#_ENREF_6))  **[udpglc] =0.32 ±0.04µmol/gww** (**~0.36mM**) (fed, rat, liver)  **[udpglc] =0.29 ±0.05µmol/gww** (**~0.32mM**) (starved, rat, liver)  **[udpglc] =0.24 ±0.09µmol/gww** (**~0.27mM**) (starved + galactose 1h, rat, liver)  **[udpglc] =0.15 ±0.03µmol/gww** (**~0.17mM**) (ethanol, starved + galactose 1h, rat, liver)  ([Guynn, et al., 1974](#_ENREF_3))  **[udpglc] = 0.342±0.024 µmol/gWW** (**~0.38mM**) (rat liver, starved)  **[udpglc] = 0.433±0.023 µmol/gWW** (**~0.48mM**) (rat liver, fed ad libitum)  **[udpglc] = 0.347±0.027 µmol/gWW** (**~0.39mM**) (rat liver, meal fed)  ([Lai, et al., 2003](#_ENREF_8)) (human cells, in µmol/100g(cell protein))  [udpglc] = 236**±**25 (Control glucose medium)  [udpglc] = 179**±**24 (**76% glucose**) (Control galactose medium)  ([Lai, et al., 2003](#_ENREF_8)) (human cells, in µmol/100g(cell protein))  [udpglc] = 157**±**10 (GALT-deficient glucose medium)  [udpglc] = 110**±**10 (**70% glucose**) (GALT-deficient galactose medium) |
| **udpgal** | **UDP-D-galactose**  (Mw 564.3)  [CHEBI:66914](http://www.ebi.ac.uk/chebi/searchId.do?chebiId=CHEBI:66914)  [KEGG:C00052](http://www.genome.jp/dbget-bin/www_bget?cpd:C00052) | **0.11mM** (no galactose)  **0.36mM** (1h galactose)  **1.39mM** (1h galactose, GALE inhibition) | Both the levels and approximate ratio of 1:3 of udpgal and udpglc are very tightly controlled in normal human cells. ([Fridovich-Keil, 2006](#_ENREF_2); [Segal, 1995](#_ENREF_13)) (1:3 rule udpglc)  ([Keppler, et al., 1970](#_ENREF_6))  **[udpgal] =0.09 ±0.01µmol/gww** (**~0.10mM**) (fed, rat, liver)  **[udpgal] =0.09 ±0.01µmol/gww** (**~0.10mM**)(starved, rat, liver)  **[udpgal] =0.32 ±0.07µmol/gww** (**~0.36mM**) (starved + galactose 1h, rat, liver)  **[udpgal] =1.25 ±0.16µmol/gww** (**~1.39mM**) (ethanol, starved + galactose 1h, rat, liver)  ([Keppler, et al., 1970](#_ENREF_6))  **[udpgal]/[udpglc] =3.4 ±0.3 (**fed, rat, liver)  **[udpgal]/[udpglc] =3.3 ±0.3 (**starved, rat, liver)  **[udpgal]/[udpglc] =0.78 ±0.39 (**starved + galactose 1h, rat, liver)  **[udpgal]/[udpglc] =0.11 ±0.02 (**ethanol, starved + galactose 1h, rat, liver)  **[udpgal]/[gal1p] =1.94 ±0.35 (**starved + galactose 1h, rat, liver)  **[udpgal]/[gal1p] =1.85 ±0.27 (**ethanol, starved + galactose 1h, rat, liver)  ([Lai, et al., 2003](#_ENREF_8)) (human cells, in µmol/100g(cell protein))  [udpgal] = 82**±**10 (Control glucose medium)  [udpgal] = 46**±**4 (**56% glucose**) (Control galactose medium 24h)  ([Lai, et al., 2003](#_ENREF_8)) (human cells, in µmol/100g(cell protein))  [udpgal] = 25**±**5 (GALT-deficient glucose medium)  [udpgal] = 17**±**3 (**68% glucose**) (GALT-deficient galactose medium 24h) |
| **galtol** | **D-galactitol**  (Mw 182.2)  [CHEBI:16813](http://www.ebi.ac.uk/chebi/searchId.do?chebiId=CHEBI:16813)  [KEGG:C01697](http://www.genome.jp/dbget-bin/www_bget?cpd:C01697) | **0.001mM** (no galactose)  **~8mM** (GALT deficiency) | **[galtol]**=4.8-40µmol/g (**~5.3-44mM**) (occupational gray matter, human) [**galtol]**=17.6µmol/g (~)(basal ganglia, human) ([Wang, et al., 2001](#_ENREF_15))  **[galtol]=12.9µmol/g (~14.3mM)** ([Wang, et al., 2001](#_ENREF_15)) ([Wells, et al., 1965](#_ENREF_16))  **[galtol]=22.18µmol/g (~24.6mM)** ([Wang, et al., 2001](#_ENREF_15)) ([Quan-Ma, et al., 1966](#_ENREF_11))  Galactitol measured directly in **GALT-deficient mice** are lower (**2mM**) than levels detected by MRS in **human subjects** (**8mM**) ([Leslie, 2003](#_ENREF_9); [Wang, et al., 2001](#_ENREF_15)) |
| **atp** | **ATP**  (Mw 503.2)  [CHEBI:30616](http://www.ebi.ac.uk/chebi/searchId.do?chebiId=CHEBI:30616)  [KEGG:C00002](http://www.genome.jp/dbget-bin/www_bget?cpd:C00002) | **2.7mM** (no galactose)  **2.9mM** (1h galactose)  **2.9mM** (1h galactose, GALE inhibition) | [**atp**] = **2.8mM** ([König, et al., 2012](#_ENREF_7))  ([Guynn, et al., 1974](#_ENREF_3))  **[atp] = 2.49±0.12 µmol/gWW** (**~2.77mM**) (rat liver, starved)  **[atp] = 2.56±0.09 µmol/gWW** (**~2.84mM**) (rat liver, fed ad libitum)  **[atp] = 2.32±0.07 µmol/gWW** (**~2.58mM**) (rat liver, meal fed)  **[atp] = 2.42±0.50 µmol/gWW** (**~2.69mM**) (rat liver) ([Keppler, et al., 1969](#_ENREF_5))  ([Keppler, et al., 1970](#_ENREF_6))  **[atp] =2.60 ±0.16µmol/gww** (**~2.89mM**) (starved + galactose 1h, rat, liver)  **[atp] =2.81 ±0.15µmol/gww** (**~3.12mM**) (ethanol, starved + galactose 1h, rat, liver)  **[atp]/[adp] =3.14 ±0.52** (starved + galactose 1h, rat, liver)  **[atp]/[adp] =3.10 ±0.53** (ethanol, starved + galactose 1h, rat, liver) |
| **adp** | **ADP**  (Mw 424.2)  [CHEBI:456216](http://www.ebi.ac.uk/chebi/searchId.do?chebiId=CHEBI:456216)  [KEGG:C00008](http://www.genome.jp/dbget-bin/www_bget?cpd:C00008) | **1.2mM** (no galactose)  **1.0mM** (1h galactose)  **1.0mM** (1h galactose, GALE inhibition) | [**adp**] = **0.8mM** ([König, et al., 2012](#_ENREF_7))  ([Guynn, et al., 1974](#_ENREF_3))  **[adp] = 1.38±0.08µmol/gWW** (**~1.53mM**) (rat liver, starved)  **[adp] = 1.06±0.03µmol/gWW** (**~1.18mM**) (rat liver, fed ad libitum)  **[adp] = 1.24±0.04µmol/gWW** (**~1.38mM**) (rat liver, meal fed)  **[adp] = 1.08±0.12 µmol/gWW** (**~1.20mM**) (rat liver) ([Keppler, et al., 1969](#_ENREF_5))  ([Keppler, et al., 1970](#_ENREF_6))  **[adp] =0.88 ±0.17µmol/gww** (**~0.98mM**) (starved + galactose 1h, rat, liver)  **[adp] =0.97 ±0.19µmol/gww** (**~1.08mM**) (ethanol, starved + galactose 1h, rat, liver) |
| **utp** | **UTP**  (Mw 480.1)  [CHEBI:46398](http://www.ebi.ac.uk/chebi/searchId.do?chebiId=CHEBI:46398)  [KEGG:C00075](http://www.genome.jp/dbget-bin/www_bget?cpd:C00075) | **0.27mM** (no galactose) | [**utp**] = **0.27mM** ([König, et al., 2012](#_ENREF_7))  ([Guynn, et al., 1974](#_ENREF_3))  **[utp] = 0.362±0.014 µmol/gWW** (**~0.40mM**) (rat liver, starved)  **[utp] = 0.494±0.038 µmol/gWW** (**~0.55mM**) (rat liver, fed ad libitum)  **[utp] = 0.443±0.039 µmol/gWW** (**~0.49mM**) (rat liver, meal fed) |
| **udp** | **UDP**  (Mw 401.1)  [CHEBI:58223](http://www.ebi.ac.uk/chebi/searchId.do?chebiId=CHEBI:58223)  [KEGG:C00015](http://www.genome.jp/dbget-bin/www_bget?cpd:C00015) | **0.09mM** (no galactose) | [**udp**] = **0.09mM** ([König, et al., 2012](#_ENREF_7))  **[utp+udp] = 0.35±0.07 µmol/gWW** (**~0.39mM**) (rat liver) ([Keppler, et al., 1969](#_ENREF_5))  **[utp+udp] = 0.35±0.05 µmol/gWW** (**~0.39mM**) (rat liver)([Keppler and Decker, 1969](#_ENREF_4))  ([Keppler, et al., 1970](#_ENREF_6))  **[utp+udp] =0.34 ±0.05µmol/gww** (**~0.38mM**) (fed, rat, liver)  **[utp+udp] =0.23 ±0.05µmol/gww** (**~0.26mM**) (starved, rat, liver)  **[utp+udp] =0.15 ±0.03µmol/gww** (**~0.17mM**) (starved + galactose 1h, rat, liver)  **[utp+udp] =0.11 ±0.02µmol/gww** (**~0.39mM**) (ethanol, starved + galactose 1h, rat, liver)  Marked decrease in [utp+udp] under galactose challenge. |
| **phos** | **Phosphate**  (Mw 96.0)  [CHEBI:43474](http://www.ebi.ac.uk/chebi/searchId.do?chebiId=CHEBI:43474)  [KEGG:C00009](http://www.genome.jp/dbget-bin/www_bget?cpd:C00009) | **5.0mM (**[**König, et al., 2012**](#_ENREF_7)**)** | [**pi**] = **5.0mM** ([König, et al., 2012](#_ENREF_7))  ([Guynn, et al., 1974](#_ENREF_3))  **[pi] = 4.37±0.16 µmol/gWW** (**~4.86mM**) (rat liver, starved)  **[pi] = 3.64±0.32 µmol/gWW** (**~4.04mM**) (rat liver, fed ad libitum)  **[pi] = 4.41±0.10 µmol/gWW** (**~4.90mM**) (rat liver, meal fed)  **[pi] = 3.18±0.56 µmol/gWW** (**~3.53mM**) (rat liver)([Keppler and Decker, 1969](#_ENREF_4)) |
| **ppi** | **Pyrophosphate**  (Mw 175.0)  [CHEBI:33019](http://www.ebi.ac.uk/chebi/searchId.do?chebiId=CHEBI:33019)  [KEGG:C00013](http://www.genome.jp/dbget-bin/www_bget?cpd:C00013) | **0.008mM(**[**König, et al., 2012**](#_ENREF_7)**)** | [**pp**] = **0.008mM** ([König, et al., 2012](#_ENREF_7))  ([Guynn, et al., 1974](#_ENREF_3))  **[pp] = 0.0023±0.0003 µmol/gWW** (**~0.0026mM**) (rat liver, starved)  **[pp] = 0.0038±0.0004 µmol/gWW** (**~0.0042mM**) (rat liver, fed ad libitum)  **[pp] = 0.0049±0.0006 µmol/gWW** (**~0.0054mM**) (rat liver, meal fed)  **[pp] = 0.0065±0.00086 µmol/gWW** (**~0.0072mM**) (rat total liver) |
| **nadp** | **NADP**  (Mw 740.4)  [CHEBI:58349](http://www.ebi.ac.uk/chebi/searchId.do?chebiId=CHEBI:58349)  [KEGG:C00006](http://www.genome.jp/dbget-bin/www_bget?cpd:C00006) | **0.1mM** |  |
| **nadph** | **NADPH**  (Mw 741.4)  [CHEBI:57783](http://www.ebi.ac.uk/chebi/searchId.do?chebiId=CHEBI:57783)  [KEGG:C00005](http://www.genome.jp/dbget-bin/www_bget?cpd:C00005) | **0.1mM** |  |
| **hydron** | **H+**  CHEBI:15378  KEGG:C00080 |  |  |
| **suc** | **Sucrose**  (Mw 342.3)  [CHEBI:17992](http://www.ebi.ac.uk/chebi/searchId.do;B9D50010EF7CAD66B236DA3B6207693C?chebiId=CHEBI:17992)  [KEGG:C00089](http://www.genome.jp/dbget-bin/www_bget?cpd:C00089) |  |  |
| **h2oM** | **H2O M**  [CHEBI:15377](http://www.ebi.ac.uk/chebi/searchId.do;B9D50010EF7CAD66B236DA3B6207693C?chebiId=CHEBI:15377)  [KEGG:C00001](http://www.genome.jp/dbget-bin/www_bget?cpd:C00001) |  |  |
| **alb** | **albumin**  PR:000003918 |  |  |
| **rbc** | **red blood cell**  BTO:0000424 |  |  |
| **galnat** | **D-galactonate**  (Mw 195.1)  [CHEBI:12931](http://www.ebi.ac.uk/chebi/searchId.do?chebiId=CHEBI:12931)  [KEGG:C00880](http://www.genome.jp/dbget-bin/www_bget?cpd:C00880) |  |  |
| **galn** | **galactosamine** |  | Uptake of galactosamine by rat liver is a~0.4µmol/g(liver)/min as measured by the disappearance of galactosamine from the medium ([Keppler, et al., 1969](#_ENREF_5))  Time-dependent decrease in uridine nucleotides in isolated perfused rat livers after galactosamine addition. ([Keppler, et al., 1969](#_ENREF_5)) |
| **amp** | **AMP** |  | **[amp] = 0.28±0.06 µmol/gWW** (**~0.31mM**) (rat liver) ([Keppler, et al., 1969](#_ENREF_5))  ([Keppler, et al., 1970](#_ENREF_6))  **[amp] =0.15 ±0.09µmol/gww** (**~0.167mM**) (starved + galactose 1h, rat, liver)  **[amp] =0.19 ±0.07µmol/gww** (**~0.21mM**) (ethanol, starved + galactose 1h, rat, liver) |
| **ump** | **UMP** |  | [**ump**] = **0.04 µmol/gWW** (**~0.044mM**) (rat liver) ([Segal and Rogers, 1971](#_ENREF_14)) |

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