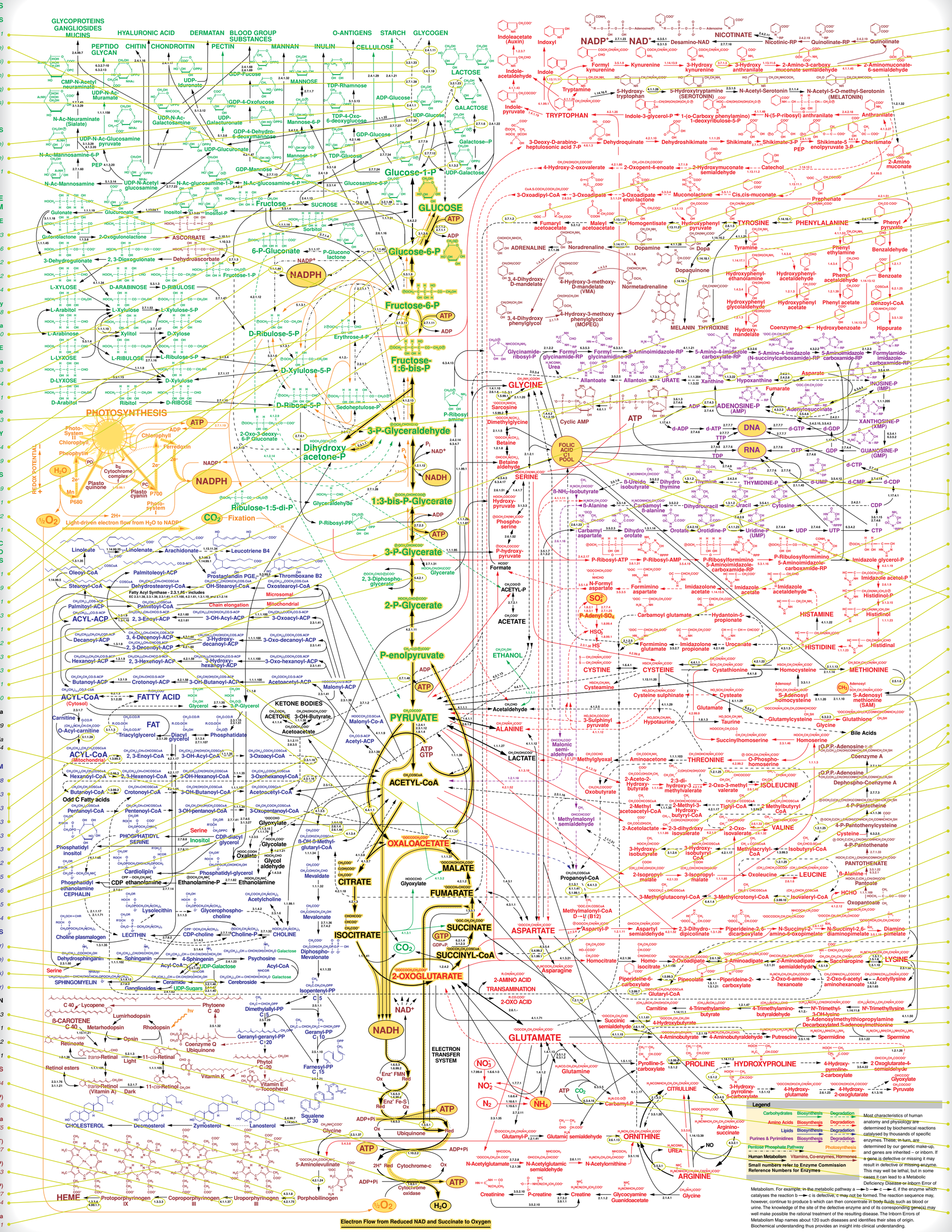
Inborn Errors of Metabolism



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OXIDOREDUCTASES

1.1.1.1 Alcohol dehydrogenase
1.1.1.8 Glycerol-3-phosphate dehydrogenase
1.1.1.9 D-Xylulose reductase
1.1.1.10 L-Xylulose reductase
1.1.1.14 L-Iditol dehydrogenase
1.1.1.15 D-Iditol dehydrogenase
1.1.1.19 Glucuronate reductase
1.1.1.21 Aldehyde reductase
1.1.1.22 UDPglucose dehydrogenase
1.1.1.23 Histidinol dehydrogenase
1.1.1.25 Shikimate dehydrogenase
1.1.1.27 Lactate dehydrogenase
1.1.1.28 D-Lactate dehydrogenase
1.1.1.29 Glycerate dehydrogenase
1.1.1.30 3-Hydroxybutyrate dehydrogenase
1.1.1.31 3-Hydroxyisobutyrate dehydrogenase
1.1.1.32 Mevaldate reductase
1.1.1.34 Hydroxymethylglutaryl-CoA reductase (NADPH)
1.1.1.35 3-Hydroxyacyl-CoA dehydrogenase
1.1.1.37 Malate dehydrogenase
1.1.1.38 Malate dehydrogenase (oxaloacetate-decarboxylating)
1.1.1.39 Malate dehydrogenase (decarboxylating)
1.1.1.41 Isocitrate dehydrogenase (NAD$^+$)
1.1.1.43 Phosphogluconate dehydrogenase
1.1.1.44 Phosphogluconate dehydrogenase (decarboxylating)
1.1.1.45 L-Gulonate dehydrogenase
1.1.1.49 Glucose-6-phosphate dehydrogenase
1.1.1.56 Ribitol dehydrogenase
1.1.1.59 3-Hydroxypropionate dehydrogenase
1.1.1.60 2-Hydroxy-3-oxopropionate reductase
1.1.1.61 4-Hydroxybutyrate dehydrogenase
1.1.1.79 Glyoxylate reductase (NADP$^+$)
1.1.1.81 Hydroxypyruvate reductase
1.1.1.82 Malate dehydrogenase (NADP$^+$)
1.1.1.85 3-Isopropylmalate dehydrogenase
1.1.1.86 Ketol-acid reductoisomerase
1.1.1.95 Phosphoglycerate dehydrogenase
1.1.1.100 3-Oxoacyl-[acyl-carrier-protein] reductase
1.1.1.102 3-Dehydrosphinganine reductase
1.1.1.105 Retinol dehydrogenase
1.1.1.155 Homoisocitrate dehydrogenase
1.1.1.157 3-Hydroxybutyryl-CoA dehydrogenase
1.1.1.158 UDP-N-acetylmuramate dehydrogenase
1.1.1.169 2-Dehydropantoate 2-reductase
1.1.1.204 Xanthine dehydrogenase
1.1.1.205 IMP dehydrogenase
1.1.3.8 L-Gulonolactone oxidase
1.1.3.15 (S)-2-Hydroxyacid oxidase
1.1.3.22 Xanthine oxidase
1.1.99.1 Choline dehydrogenase
1.1.99.5 Glycerol-3-phosphate dehydrogenase
1.2.1.3 Aldehyde dehydrogenase (NAD$^+$)
1.2.1.7 Benzaldehyde dehydrogenase (NADP$^+$)
1.2.1.8 Betaine-aldehyde dehydrogenase
1.2.1.11 Aspartate-semialdehyde dehydrogenase
1.2.1.12 Glyceraldehyde-3-phosphate dehydrogenase
1.2.1.13 Glyceraldehyde-3-phosphate dehydrogenase
1.2.1.16 Succinate-semialdehyde dehydrogenase (NAD(P)$^+$)
1.2.1.18 Malonate semialdehyde dehydrogenase (acetylating)
1.2.1.19 Aminobutyraldehyde dehydrogenase
1.2.1.21 Glycolaldehyde dehydrogenase
1.2.1.23 2-Oxoaldehyde dehydrogenase (NAD$^+$)
1.2.1.24 Succinate-semialdehyde dehydrogenase
1.2.1.25 2-Oxoisovalerate dehydrogenase (acylating)
1.2.1.26 2,5-Dioxovalerate dehydrogenase
1.2.1.27 Methylmalonate-semialdehyde dehydrogenase (acylating)
1.2.1.28 Benzaldehyde dehydrogenase (NAD$^+$)
1.2.1.31 L-Aminoadipate-semialdehyde dehydrogenase
1.2.1.32 Aminomuconate-semialdehyde dehydrogenase
1.2.1.36 Retinal dehydrogenase
1.2.1.39 Phenylacetaldehyde dehydrogenase
1.2.1.41 Glutamate-5-semialdehyde dehydrogenase
1.2.1.52 Oxoglutarate dehydrogenase
1.2.3.5 Glyoxylate oxidase
1.2.4.1 Pyruvate dehydrogenase (lipoamide)
1.2.4.2 Oxoglutarate dehydrogenase (lipoamide)
1.2.7.1 Pyruvate synthase
1.2.7.2 2-Oxobutyrate synthase
1.3.1.1 Dihydrouracil dehydrogenase (NAD$^+$)
1.3.1.2 Dihydropyrimidine dehydrogenase (NADP$^+$)
1.3.1.6 Fumarate reductase (NADH)
1.3.1.8 Acyl-CoA dehydrogenase (NADP$^+$)
1.3.1.9 Enoyl-[acyl-carrier-protein] reductase (NADH)
1.3.1.10 Enoyl-[acyl-carrier-protein] reductase (NADPH, B-specific)
1.3.1.13 Prephenate dehydrogenase (NADP$^+$)
1.3.1.14 Orotate reductase (NADH)
1.3.1.26 Dihydrodipicolinate reductase
1.3.1.35 Phosphatidylcholine desaturase
1.3.3.3 Coproporphyrinogen oxidase
1.3.3.4 Protoporphyrinogen oxidase
1.3.5.1 Succinate dehydrogenase (ubiquinone)
1.3.99.1 Succinate dehydrogenase
1.3.99.2 Butyryl-CoA dehydrogenase
1.3.99.3 Acyl-CoA dehydrogenase
1.3.99.7 Glutaryl-CoA dehydrogenase
1.3.99.10 Isovaleryl-CoA dehydrogenase
1.4.1.1 Alanine dehydrogenase
1.4.1.2 Glutamate dehydrogenase
1.4.1.7 Serine dehydrogenase
1.4.1.8 Valine dehydrogenase (NADP$^+$)
1.4.1.9 Leucine dehydrogenase
1.4.1.10 Glycine dehydrogenase
1.4.1.13 Glutamate synthase (NADPH)
1.4.1.14 Glutamate synthase (NADH)
1.4.1.19 Tryptophan dehydrogenase
1.4.3.1 D-Aspartate oxidase
1.4.3.2 L-Amino-acid oxidase
1.4.3.4 Amine oxidase (flavin-containing)
1.4.3.6 Amine oxidase (copper-containing)
1.4.3.8 Ethanolamine oxidase
1.4.4.2 Glycine dehydrogenase (decarboxylating)
1.5.1.1 Pyrroline-2-carboxylate reductase
1.5.1.2 Pyrroline-5-carboxylate reductase
1.5.1.3 Dihydrofolate reductase
1.5.1.5 Methylenetetrahydrofolate reductase
1.5.1.6 Formyltetrahydrofolate dehydrogenase
1.5.1.7 Saccharopine dehydrogenase (NAD$^+$), L-lysine-forming
1.5.1.8 Saccharopine dehydrogenase (NADP$^+$), L-lysine-forming
1.5.1.9 Saccharopine dehydrogenase (NAD$^+$), L-glutamate-forming
1.5.1.10 Saccharopine dehydrogenase (NADP, L-glutamate-forming*)
1.5.1.12 1-Pyrroline-5-carboxylate dehydrogenase
1.5.1.21 Δ^1-Piperideine-2-carboxylate reductase
1.5.3.1 Sarcosine oxidase
1.5.99.1 Sarcosine dehydrogenase
1.5.99.2 Dimethylglycine dehydrogenase
1.5.99.3 L-Pipecolate dehydrogenase
1.5.99.8 Proline dehydrogenase
1.6.4.1 Cystine reductase (NADH)
1.6.5.3 NADH dehydrogenase (ubiquinone)
1.6.6.1 Nitrate reductase (NADH)
1.6.6.2 Nitrate reductase [NAD(P)H]
1.6.6.3 Nitrate reductase (NADPH)
1.6.6.4 Nitrite reductase [NAD(P)H]
1.6.6.8 GMP reductase
1.7.3.3 Urate oxidase
1.7.7.1 Ferredoxin—nitrate reductase
1.7.99.4 Nitrate reductase
1.8.1.3 Hypotaurine dehydrogenase
1.8.1.4 Dihydrolipoamide dehydrogenase
1.8.2.1 Sulfite dehydrogenase
1.8.3.1 Sulfite oxidase
1.8.7.1 Sulfite reductase (ferredoxin)
1.8.99.1 Sulfite reductase
1.9.3.1 Cytochrome-c oxidase
1.10.2.1 L-Ascorbate—cytochrome-b_5 reductase
1.10.2.2 Ubiquinol—cytochrome-c reductase
1.10.3.3 L-Ascorbate oxidase
1.10.99.1 Plastoquinol—plastocyanin reductase
1.13.11.1 Catechol 1,2-dioxygenase
1.13.11.2 Catechol 2,3-dioxygenase
1.13.11.5 Homogentisate 1,2-dioxygenase
1.13.11.6 3-Hydroxyanthranilate 3,4-dioxygenase
1.13.11.11 Tryptophan 2,3-dioxygenase
1.13.11.20 Cysteine dioxygenase
1.13.11.21 ß-Carotene 15,15′-dioxygenase
1.13.11.27 4-Hydroxyphenylpyruvate dioxygenase
1.13.99.1 myo-Inositol oxygenase
1.13.99.2 Benzoate 1,2-dioxygenase
1.14.11.1 γ-Butyrobetaine dioxygenase
1.14.11.2 Procollagen-proline dioxygenase
1.14.11.8 Trimethyllysine dioxygenase
1.14.12.1 Anthranilate 1,2-dioxygenase (deaminating, decarboxylating)
1.14.12.10 Benzoate 1,2-dioxygenase
1.14.13.5 Imidazoleacetate 4-monooxygenase
1.14.13.9 Kynurenine 3-monooxygenase
1.14.13.12 Benzoate 4-monooxygenase
1.14.13.39 Nitric oxide synthase
1.14.16.1 Phenylalanine 4-monooxygenase
1.14.16.2 Tyrosine 3-monooxygenase
1.14.16.4 Tryptophan 5-monooxygenase
1.14.17.1 Dopamine ß-monooxygenase
1.14.18.1 Monophenol monooxygenase
1 14.99.1 Prostaglandin synthase
1 14.99.5 Stearoyl-CoA desaturase
1.14.99.7 Squalene monooxygenase
1.14.99.25 Linoleoyl-CoA desaturase
1.17.4.1 Ribonucleoside-diphosphate reductase
1.18.6.1 Nitrogenase

TRANSFERASES

2.1.1.1 Nicotinamide N-methyltransferase
2.1.1.2 Guanidinoacetate N-methyltransferase
2.1.1.3 Thetin—homocysteine S-methyltransferase
2.1.1.4 Acetylserotonin N-methyltransferase
2.1.1.5 Betaine—homocysteine S-methyltransferase
2.1.1.6 Catechol O-methyl transferase
2.1.1.10 Homocysteine S-methyltransferase
2.1.1.13 5-Methyltetrahydrofolate—homocysteine S-methyl transferase
2.1.1.14 5-Methyltetrahydropteroyltriglutamate — homocysteine S-methyltransferase
2.1.1.17 Phosphatidylethanolamine N-methyltransferase
2.1.1.20 Glycine N-methyltransferase
2.1.1.28 Phenylethanolamine N-methyltransferase
2.1.1.43 Histone-lysine N-methyltransferase
2.1.1.45 Thymidylate synthase
2.1.2.1 Glycine hydroxymethyltransferase
2.1.2.2 Phosphoribosylglycinamide formyltransferase
2.1.2.3 Phosphoribosylaminoimidazole carboxamide formyltransferase
2.1.2.5 Glutamate formiminotransferase
2.1.2.10 Aminomethyl transferase
2.1.3.1 Methylmalonyl-CoA carboxyltransferase
2.1.3.2 Aspartate carbamoyltransferase
2.1.3.3 Ornithine carbamoyltransferase
2.1.4.1 Glycine amidinotransferase
2.2.1.1 Transketolase
2.2.1.2 Transaldolase
2.3.1.1 Amino-acid N-acetyltransferase
2.3.1.4 Glucosamine-phosphate N-acetyltransferase
2.3.1.5 Arylamine N-acetyltransferase
2.3.1.6 Choline O-acetyltransferase
2.3.1.7 Carnitine O-acetyltransferase
2.3.1.8 Phosphate acetyltransferase
2.3.1.9 Acetyl-CoA C-acetyltransferase
2.3.1.12 Dihydrolipoamide S-acetyltransferase
2.3.1.15 Glycerol-3-phosphate O-acyltransferase
2.3.1.16 Acetyl-CoA C-acyltransferase
2.3.1.20 Diacylglycerol O-acyltransferase
2.3.1.21 Carnitine O-palmitoyltransferase
2.3.1.23 Lysolecithin acyltransferase
2.3.1.24 Sphingosine N-acyltransferase
2.3.1.30 Serine O-acetyltransferase
2.3.1.32 Lysine N-acetyltransferase
2.3.1.35 Glutamate N-acetyltransferase
2.3.1.37 5-Aminolevulinate synthase
2.3.1.38 [Acyl-carrier-protein] S-acetyltransferase
2.3.1.39 [Acyl-carrier-protein] S-malonytransferase
2.3.1.41 3-Oxoacyl-[acyl-carrier-protein] synthase
2.3.1.46 Homoserine O-succinyltransferase
2.3.1.50 Serine C-palmitoyltransferase
2.3.1.51 1-Acylglycerol-3-phosphate O-acyltransferase
2.3.1.76 Retinol O-fatty-acyltransferase
2.4.1.1 Phosphorylase
2.4.1.4 Amylosucrase
2.4.1.9 Inulosucrase
2 4.1.11 Glycogen (starch) synthase
2.4.1.13 Sucrose synthase
2.4.1.17 Glucuronosyltransferase
2.4.1.21 Starch synthase
2.4.1.22 Lactose synthase
2.4.1.23 Sphingosine ß-galactosyltransferase
2.4.1.29 Cellulose synthase (GDP-forming)
2.4.1.32 Glucomannan 4-ß-mannosyltransferase
2.4.1.41 Polypeptide N-acetylgalactosaminyltransferase
2.4.1.62 Ganglioside galactosyltransferase
2.4.1.68 Glycoprotein 6-α-L-fucosyltransferase
2.4.1.69 Galactoside 2-α-L-fucosyltransferase
2.4.1.101 α-1,3-Mannosyl-glycoprotein ß-1,2-N-acetylglucosaminyltransferase
2.4.2.1 Purine-nucleoside phosphorylase
2.4.2.2 Pyrimidine-nucleoside phosphorylase
2.4.2.4 Thymidine phosphorylase
2.4.2.8 Hypoxanthine phosphoribosyltransferase
2.4.2.9 Uracil phosphoribosyltransferase
2.4.2.10 Orotate phosphoribosyltransferase
2.4.2.11 Nicotinate phosphoribosyltransferase
2.4.2.14 Amidophosphoribosyltransferase
2.4.2.15 Guanosine phosphorylase
2.4.2.17 ATP phosphoribosyltransferase
2.4.2.18 Anthranilate phosphoribosyltransferase
2.4.2.19 Nicotinate-nucleotide pyrophosphorylase (carboxylating)
2.4.99.7 Sialyltransferase
2.4.99.1-11 Sialyl transferases
2.5.1.1 Dimethylallyltransferase
2.5.1.6 Methionine adenosyltransferase
2.5.1.10 Geranyl*trans*transferase
2.5.1.16 Aminopropyltransferase
2.5.1.19 3-Phosphoshikimate 1-carboxyvinyl-transferase
2.5.1.21 Farnesyltransferase
2.5.1.22 Spermine synthase
2.5.1.29 Farnesyl*trans*transferase
2.5.1.32 Geranylgeranyl-diphosphate geranylgeranyl transferase
2.6.1.1 Aspartate transaminase
2.6.1.2 Alanine transaminase
2.6.1.4 Glycine transaminase
2.6.1.5 Tyrosine transaminase
2.6.1.6 Leucine transaminase
2.6.1.9 Histidinol-phosphate transaminase
2.6.1.11 Acetylornithine transaminase
2.6.1.13 Ornithine—oxo-acid transaminase
2.6.1.16 Glutamine—fructose-6-phosphate transaminase
2.6.1.17 Succinyldiaminopimelate transaminase
2.6.1.18 ß-Alanine—pyruvate transaminase
2.6.1.19 4-Aminobutyrate transaminase
2.6.1.22 L-3-Aminoisobutyrate transaminase
2.6.1.23 4-Hydroxyglutamate transaminase
2.6.1.27 Tryptophan transaminase
2.6.1.32 Valine—3-methyl-2-oxovalerate transaminase
2.6.1.36 L-Lysine 6-transaminase
2.6.1.39 2-Aminoadipate transaminase
2.6.1.42 Branched-chain-amino-acid transaminase
2.6.1.51 Serine—pyruvate transaminase
2.6.1.52 Phosphoserine transaminase
2.6.1.65 N^6-Acetyl-ß-lysine transaminase
2.6.1.66 Valine—pyruvate transaminase
2.7.1.1 Hexokinase
2.7.1.2 Glucokinase
2.7.1.3 Ketohexokinase
2.7.1.4 Fructokinase
2.7.1.6 Galactokinase
2.7.1.7 Mannokinase
2.7.1.11 6-Phosphofructokinase
2.7.1.15 Ribokinase
2.7.1.16 Ribulokinase
2.7.1.17 Xylulokinase
2.7.1.19 Phosphoribulokinase
2.7.1.23 NAD$^+$ kinase
2.7.1.24 Dephospho-CoA kinase
2.7.1.25 Adenylylsulfate kinase
2.7.1.28 Triokinase
2.7.1.30 Glycerol kinase
2.7.1.31 Glycerate kinase
2.7.1.32 Choline kinase
2.7.1.33 Pantothenate kinase
2.7.1.34 Pantetheine kinase
2.7.1.36 Mevalonate kinase
2.7.1.39 Homoserine kinase
2.7.1.40 Pyruvate kinase
2.7 1.47 D-Ribulokinase
2.7.1.53 L-Xylulokinase
2.7.1.60 N-Acylmannosamine kinase
2.7.1.64 myo-Inositol 1-kinase
2.7.1.71 Shikimate kinase
2.7.1.80 Pyrophosphate—serine phosphotransferase
2.7.1.107 Diacylglycerol kinase
2.7.2.1 Acetate kinase
2.7.2.3 Phosphoglycerate kinase
2.7.2.4 Aspartate kinase
2.7.2.6 Formate kinase
2.7.2.11 Glutamate 5-kinase
2.7.3.2 Creatine kinase
2.7.4.2 Phosphomevalonate kinase
2.7.4.3 Adenylate kinase
2.7.4.4 Nucleoside-phosphate kinase
2.7.4.6 Nucleoside-diphosphate kinase
2.7.4.9 dTMP kinase
2.7.4.14 Cytidylate kinase
2.7.6.1 Ribose-phosphate pyrophosphokinase
2.7.7.3 Pantetheine-phosphate adenylyltransferase
2.7.7.4 Sulfate adenylyltransferase
2.7.7.6 RNA nucleotidyltransferase (DNA-directed)
2.7.7.7 DNA nucleotidyltransferase (DNA-directed)
2.7.7.9 UTP—glucose-1-phosphate uridylyltransferase
2.7.7.10 UTP—hexose-1-phosphate uridylyltransferase
2.7.7.12 UDP glucose—hexose-1-phosphate uridylyltransferase
2.7.7.13 Mannose-1-phosphate guanylyltransferase
2.7.7.14 Ethanolamine-phosphate cytidylyltransferase
2.7.7.15 Choline-phosphate cytidylyltransferase
2.7.7.18 Nicotinate-mononucleotide adenylyltransferase
2.7.7.22 Mannose-1-phosphate guanylyltransferase (GDP)
2.7.7.23 UDP-N-acetylglucosamine pyrophosphorylase
2.7.7.24 Glucose-1-phosphate thymidylyltransferase
2.7.7.27 Glucose-1-phosphate adenylyltransferase
2.7.7.34 Glucose-1-phosphate guanylyltransferase
2.7.7.41 Phosphatidate cytidylyltransferase
2.7.7.43 N-Acylneuraminate cytidylyltransferase
2.7.8.1 Ethanolamine phosphotransferase
2.7.8.2 Diacylglycerol cholinephosphotransferase
2.7.8.3 Ceramide cholinephosphotransferase
2.7.8.5 CDPdiacylglycerol—glycerol-3-phosphate 3-phosphatidyltransferase
2.7.8.8 CDPdiacylglycerol—serine O-phosphatidyltransferase
2.7.8.11 CDPdiacylglycerol—inositol 3-phosphatidyltransferase
2.7.8.13 Phospho-N-acetylmuramoyl-pentapeptide-transferase
2.8.3.1 Propionate CoA-transferase
2.8.3.5 3-Oxoacid CoA-transferase
2.8.3.6 3-Oxoadipate CoA-transferase

HYDROLASES

3.1.1.3 Triacylglycerol lipase
3.1.1.4 Phospholipase A_2
3.1.1.5 Lysophospholipase
3.1.1.7 Acetylcholinesterase
3.1.1.17 Gluconolactonase
3.1.1.21 Retinyl-palmitate esterase
3.1 1.24 3-Oxoadipate enol-lactonase
3.1.1.28 Acylcarnitine hydrolase
3.1.1.31 6-Phosphogluconolactonase
3.1.1.32 Phospholipase A_1
3.1.2.1 Acetyl-CoA hydrolase
3.1.2.3 Succinyl-CoA hydrolase
3.1.2.4 3-Hydroxyisobutyryl-CoA hydrolase
3.1.2.11 Acetoacetyl-CoA hydrolase
3.1.3.2 Acid phosphatase
3.1.3.3 Phosphoserine phosphatase
3.1.3.4 Phosphatidate phosphatase
3.1.3.5 5′-Nucleotidase
3.1.3.9 Glucose-6-phosphatase
3.1.3.11 Fructose-bisphosphatase
3.1.3.15 Histidinol-phosphatase
3.1.3.25 myo-Inositol-1(or 4)-monophosphatase
3.1.3.27 Phosphatidylglycerophosphatase
3.1.3.29 N-Acylneuraminate-9-phosphatase
3.1.3.31 Nucleotidase
3.1.4.2 Glycerophosphocholine phosphodiesterase
3.1.4.3 Phospholipase C
3.1.4.4 Phospholipase D
3.1.4.10 1-Phosphatidylinositol phosphodiesterase
3.1.4.12 Sphingomyelin phosphodiesterase
3.2.1.3 Glucan 1,4-α-glucosidase
3.2.1.21 ß-Glucosidase
3.2.1.23 ß-Galactosidase
3.2.1.26 ß-Fructofuranosidase
3.2.1.45 Glucosylceramidase
3.2.1.46 Galactosylceramidase
3.2.1.48 Sucrose α-glucosidase
3.3.1.1 Adenosylhomocysteinase
3.5.1.1 Asparaginase
3.5.1.2 Glutaminase
3.5.1.6 ß-Ureidopropionase
3.5.1.7 Ureidosuccinase
3.5.1.8 Formylaspartate deformylase
3.5.1.9 Aryl-formamidase
3.5.1.16 Acetylornithine deacetylase
3.5.1.18 Succinyl-diaminopimelate desuccinylase
3.5.1.19 Nicotinamidase
3.5.1.20 Citrullinase
3.5.1.22 Pantothenase
3.5.1.23 Ceramidase
3.5.1.32 Hippurate hydrolase
3.5.2.2 Dihydropyrimidinase
3.5.2.3 Dihydroorotase
3.5.2.5 Allantoinase
3.5.2.7 Imidazolonepropionase
3.5.2.10 Creatininase
3.5.3.1 Arginase
3.5.3.4 Allantoicase
3.5.3.5 Formiminoaspartate deiminase
3.5.3.6 Arginine deiminase
3.5.4.1 Cytosine deaminase
3.5.4.3 Guanine deaminase
3.5.4.6 AMP deaminase
3.5.4.9 Methenyltetrahydrofolate cyclohydrolase
3.5.4.10 IMP cyclohydrolase
3.5.4.12 dCMP deaminase
3.5.4.19 Phosphoribosyl-AMP cyclohydrolase
3.5.4.22 1-Pyrroline-4-hydroxy-2-carboxylate deaminase
3.6.1.3 Adenosine-triphosphatase
3.6.1.15 Nucleoside-triphosphatase
3.6.1.31 Phosphoribosyl-ATP pyrophosphatase
3.7.1.2 Fumarylacetoacetase
3.7.1.3 Kynureninase
3.7.1.9 2-Hydroxymuconate-semialdehyde hydrolase
3.9.1.1 Phosphoamidase

LYASES

4.1.1.1 Pyruvate decarboxylase
4.1.1.3 Oxaloacetate decarboxylase
4.1.1.4 Acetoacetate decarboxylase
4.1.1.9 Malonyl-CoA decarboxylase
4.1.1.11 Aspartate 1-decarboxylase
4.1.1.12 Aspartate 4-decarboxylase
4.1.1.15 Glutamate decarboxylase
4.1.1.17 Ornithine decarboxylase
4.1.1.20 Diaminopimelate decarboxylase
4 1.1.21 Phosphoribosylaminoimidazole carboxylase
4.1.1.22 Histidine decarboxylase
4.1.1.23 Orotidine-5′-phosphate decarboxylase
4.1.1.25 Tyrosine decarboxylase
4.1.1.28 Aromatic-L-amino-acid decarboxylase
4.1.1.29 Sulfoalanine decarboxylase
4.1.1.31 Phospho*enol*pyruvate carboxylase
4.1.1.32 Phospho*enol*pyruvate carboxykinase (GTP)
4.1.1.33 Diphosphomevalonate decarboxylase
4.1.1.34 Dehydro-L-gulonate decarboxylase
4.1.1.36 Phosphopantothenoylcysteine decarboxylase
4.1.1.37 Uroporphyrinogen decarboxylase
4.1.1.38 Phospho*enol*pyruvate carboxykinase (pyrophosphate)
4.1.1.39 Ribulose-bisphosphate carboxylase
4.1.1.41 Methylmalonyl-CoA decarboxylase
4.1.1.43 Phenylpyruvate decarboxylase
4.1.1.45 Aminocarboxymuconate-semialdehyde decarboxylase
4.1.1.47 Tartronate-semialdehyde synthase
4.1.1.48 Indole-3-glycerol-phosphate synthase
4.1.1.49 Phosphoenolpyruvate carboxykinase (ATP)
4.1.1.50 Adenosylmethionine decarboxylase
4.1.1.53 Phenylalanine decarboxylase
4.1.1.65 Phosphatidylserine decarboxylase
4.1.1.71 2-Oxoglutarate decarboxylase
4.1.2.5 Threonine aldolase
4.1.2.9 Phosphoketolase
4.1.2.12 Ketopantoaldolase
4.1.2.13 Fructose-bisphosphate aldolase
4.1.2.14 2-Dehydro-3-deoxyphosphogluconate aldolase
4.1.2.15 2-Dehydro-3-deoxyphosphoheptonate aldolase
4.1.3.1 Isocitrate lyase
4.1.3.2 Malate synthase
4.1.3.4 Hydroxymethylglutaryl-CoA lyase
4.1.3.5 Hydroxymethylglutaryl-CoA synthase
4.1.3.7 Citrate (*si*)-synthase
4.1.3.8 ATP citrate (*pro-S*)-lyase
4.1.3.16 4-Hydroxy-2-oxoglutarate aldolase
4.1.3.18 Acetolactate synthase
4.1.3.20 N-Acylneuraminate-9-phosphate synthase
4.1.3.21 Homocitrate synthase
4.1.3.22 Citramalate lyase
4.1.3.27 Anthranilate synthase
4.1.99.1 Tryptophanase
4.2.1.2 Fumarate hydratase
4.2.1.3 Aconitate hydratase
4.2.1.4 Citrate dehydratase
4.2.1.9 Dihydroxy-acid dehydratase
4.2.1.10 3-Dehydroxyquinate dehydratase
4.2.1.11 Phosphopyruvate hydratase (enolase)
4.2.1.12 Phosphogluconate dehydratase
4.2.1.13 L-Serine dehydratase
4.2.1.16 Threonine dehydratase
4.2.1.17 Enoyl-CoA hydratase
4.2.1.18 Methylglutaconyl-CoA hydratase
4.2.1.19 Imidazoleglycerol-phosphate dehydratase
4.2.1.20 Tryptophan synthase
4.2.1.22 Cystathionine ß-synthase
4.2.1.24 Porphobilinogen synthase
4.2.1.33 3-Isopropylmalate dehydratase
4.2.1.34 (S)-2-Methylmalate hydratase
4.2.1.36 Homoaconitate hydratase
4.2 1.46 dTDPglucose 4,6-dehydratase
4.2.1.47 GDPmannose 4,6-dehydratase
4.2.1.49 Urocanate hydratase
4.2.1.51 Prephenate dehydratase
4.2.1.52 Dihydrodipicolinate synthase
4.2.1.55 3-Hydroxybutyryl-CoA dehydratase
4.2.1.58 Crotonoyl-[acyl-carrier-protein] hydratase
4.2.1.59 3-Hydroxyoctanoyl-[acyl-carrier protein] dehydratase
4.2.1.60 3-Hydroxydecanoyl-[acyl-carrier protein] dehydratase
4.2.1.61 3-Hydroxypalmitoyl-[acyl-carrier protein] dehydratase
4.2.1.75 Uroporphyrinogen-III synthase
4.2.1.80 2-Oxopent-4-enoate hydratase
4.2.99.2 Threonine synthase
4.2.99.8 Cysteine synthase
4.2.99.9 O-Succinylhomoserine (thiol)-lyase
4.3.1.1 Aspartate ammonia-lyase
4.3.1.2 Methylaspartate ammonia-lyase
4.3.1.3 Histidine ammonia-lyase
4.3.1.4 Formiminotetrahydrofolate cyclodeaminase
4.3.1.8 Hydroxymethylbilane synthase
4.3.2.1 Argininosuccinate lyase
4.3.2.2 Adenylosuccinate lyase
4.4.1.1 Cystathionine γ-lyase
4.4.1.8 Cystathionine ß-lyase
4.6.1.1 Adenylate cyclase
4.6.1.3 3-Dehydroquinate synthase
4.6.1.4 Chorismate synthase
4.99.1.1 Ferrochelatase

ISOMERASES

5.1.3.1 Ribulose-phosphate 3-epimerase
5.1.3.2 UDPglucose 4-epimerase
5.1.3.4 L-Ribulose-phosphate 4-epimerase
5.1.3.6 UDPglucuronate 4-epimerase
5.1.3.7 UDP-N-acetylglucosamine 4-epimerase
5.1.3.12 UDPglucuronate 5′-epimerase
5 1.3.13 dTDP-4-Dehydrorhamnose 3,5-epimerase
5.1.3.14 UDP-N-acetylglucosamine 2-epimerase
5.1.99.1 Methylmalonyl-CoA epimerase
5.2.1.2 Maleylacetoacetate isomerase
5.2.1.3 Retinal isomerase
5.2.1.7 Retinol isomerase
5.3.1.1 Triose-phosphate isomerase
5.3.1.3 Arabinose isomerase
5.3.1.4 L-Arabinose isomerase
5.3.1.5 Xylose isomerase
5.3.1.6 Ribose-5-phosphate isomerase
5.3.1.8 Mannose-6-phosphate isomerase
5.3.1.9 Glucose-6-phosphate isomerase
5.3.1.15 D-Lyxose ketol-isomerase
5.3.1.16 N-(5′-Phospho-D-ribosylformimino)-5-amino-1-(5′′-phosphoribosyl)-4-imidazolecarboxamide isomerase
5.3.3.2 Isopentenyl-diphosphate Δ-isomerase
5.3.3.4 Muconolactone Δ-isomerase
5.3.99.3 Prostaglandin-E synthase
5.3.99.5 Thromboxane-A synthase
5.4.2.1 Phosphoglycerate mutase
5.4.2.2 Phosphoglucomutase
5.4.2.3 Phosphoacetylglucosamine mutase
5.4.2.8 Phosphomannomutase
5.4.3.8 Glutamate-1-semialdehyde 2,1-aminomutase
5.4.99.1 Methylaspartate mutase
5.4.99.2 Methylmalonyl-CoA mutase
5.4.99.5 Chorismate mutase
5.4 99.7 Lanosterol synthase
5.5.1.1 Muconate cycloisomerase
5.5.1.4 myo-Inositol-1-phosphate synthase

LIGASES

6.2.1.4 Succinate-CoA ligase (GDP-forming)
6.2.1.25 Benzoate—CoA ligase
6.3.1.1 Aspartate—ammonia ligase
6.3.1.2 Glutamate—ammonia ligase
6.3.1.4 Aspartate—ammonia ligase (ADP-forming)
6.3.1.5 NAD$^+$ synthetase
6.3.2.1 Pantoate—ß-alanine ligase
6.3.2.2 Glutamate—cysteine ligase
6.3.2.3 Glutathione synthase
6.3.2.5 Phosphopantothenate—cysteine ligase
6.3.2.6 Phosphoribosylaminoimidazole-succinocarboxamide synthase
6.3.2.7 UDP-N-Acetylmuramoyl-L-alanyl-D-glutamate—lysine ligase
6.3.2.8 UDP-N-acetylmuramate—alanine ligase
6.3.2.9 UDP-N-acetylmuramoylalanine—D-glutamate ligase
6.3.2.10 UDP-N-acetylmuramoylalanyl-D-glutamyl-lysine—D-alanyl-D-alanine ligase
6.3.2.13 UDP-N-Acetylmuramoylalanyl-D-glutamate—2,6-diaminopimelate ligase
6.3.3.1 Phosphoribosylglycinamidine cyclo-ligase
6.3.4.1 GMP synthase
6.3.4.2 CTP synthase
6.3.4.3 Formate—tetrahydrofolate ligase
6.3.4.4 Adenylosuccinate synthase
6.3.4.5 Argininosuccinate synthase
6.3.4.7 Ribose-5-phosphate—ammonia ligase
6.3.4.13 Phosphoribosylamine—glycine ligase
6.3.4.16 Carbamoyl-phosphate synthase (ammonia)
6.3.4.17 Formate—dihydrofolate ligase
6.3.5.1 NAD$^+$ synthetase (glutamine-hydrolysing)
6.3.5.2 GMP synthetase (glutamine-hydrolysing)
6.3.5.3 Phosphoribosylformylglycinamidine synthetase
6.3.5.4 Asparagine synthase (glutamine-hydrolysing)
6.3.5.5 Carbamoyl-phosphate synthase (glutamine-hydrolysing)
6.4.1.1 Pyruvate carboxylase
6.4.1.2 Acetyl-CoA carboxylase
6.4.1.3 Propionyl-CoA carboxylase
6.4.1.4 Methylcrotonoyl-CoA carboxylase

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