Amino acid	Primary metabolic disorder	Secondarily increased (decreased) in:
Taurine	Not described	Catabolic state; high protein intake; acute liver failure; sulphite-oxidase deficiency
Phosphoethanolamine	Hypophosphatasia (bone alkaline-phosphatase deficiency)	Some endocrine disorders; hypertension; bone diseases
Aspartylglycosamine	Aspartylglucosaminuria (lysosomal aspartylglycosylaminase deficiency)	Not described
Aspartic acid	Dicarboxylic aminoaciduria (transport disorder in combination with glutamic acid and proline)	Decomposition of asparagine
4-Hydroxyproline	Hydroxyprolinaemia (hydroxyproline oxidase deficiency)	Newborn infants; chronic uraemia; bone disease
Threonine	Not described	Vitamin B6 deficiency; liver cirrhosis
Serine	Not described	Vitamin B6 deficiency; (Decreased in folic acid deficiency, and bacterial decomposition of the urine)
Asparagine	Not described	Not described
Glutamic acid	1. Dicarboxylic aminoaciduria (transport disorder together with aspartic acid and proline)	Decomposition of glutamine
	2. Glutamic acidaemia	
Glutamine	Not described	All conditions with hyperammonaemia together with alanine; acute sick newborn infants
Sarcosine	Hypersarcosinaemia (sarcosine dehydrogenase deficiency)	Glutaric aciduria Type 2; folic acid deficiency
2-Aminoadipic acid	2-Aminoadipic aciduria (no enzyme defect detected)	2-Ketoadipic aciduria; severe convulsive conditions
Proline	1. Hyperprolinaemia Type 1 (proline oxidase deficiency)	Iminodipeptiduria (prolidase deficiency); newborn infants
	2. Hyperprolinaemia Type 2 (A'-pyrroline-5-carboxylic acid dehydrogenase deficiency)	
	3. Iminoglycinuria (transport defect for proline, OH-proline and glycine)	
Glycine	Nonketotic hyperglycinaemia (deficiency of the cleavage enzyme)	1. Ketotic hyperglycinaemia due to (a) propionic acidaemia,
		(b) methylmalonic acidaemia, (c) isovaleric acidaemia, or
		(d) 2-methylacetoacetic aciduria
		2. D-Glyceric aciduria
		3. Hyperprolinaemia Type 1, or iminoglycinuria
		4. Hypersarcosinaemia

Amino acid	Primary metabolic disorder	Secondarily increased (decreased) in:
		5. Infants up to 6 months of age
		6. Starvation
		7. Bacterial decomposition of hippuric acid
		8. Valproic acid therapy
		(Decreased in folic acid deficiency)
Alanine	Not described	All conditions with lactic acidaemia; together with glutamine in hyperammonaemia; bacterial contamination of the urine; (Decreased in ketotic hypoglycaemia)
Citrulline	Citrullinaemia (argininosuccinate synthetase deficiency)	Argininosuccinic aciduria; saccharopinuria; renal insufficiency; hyperargininaemia
2-Aminobutyric acid	Not described	Hyperaminoaciduria
Valine	1. Hypervalinaemia (transaminase defect)	Not described
	2. Maple syrup urine disease	
Cystine	1. Isolated cystinuria (transport defect)	Conditions of homocystinuria due to a remethylation defect; infants during the first months of life; renal defects; (Decreased in homocystinuria and cystathioninuria)
	2. Dibasic aminoaciduria together with ornithine, lysine and arginine (transport defect)	
	3. (Increased in leukocytes and cultured fibroblasts in cystinosis)	
Homocitrulline	Not described	Young infants; various hyperammonaemia syndromes
Cystathionine	Cystathioninuria (P-cystathioninase deficiency)	Vitamin B_6 deficiency; neuroblastoma and hepatoblastoma; liver cirrhosis
Methionine	1. Hypermethioninaemia (methionine-adenylotransferase deficiency)	Homocystinuria; tyrosinosis
	2. Methionine malabsorption syndrome	(Decreased in homocystinuria due to remethylation defects)
Alloisoleucine	Not described	Maple syrup urine disease
Isoleucine	Maple syrup urine disease	Not described
Leucinc	Maple syrup urine disease	Not described
Argininosuccinic acid	Argininosuccinic aciduria (argininosuccinate lyase deficiency)	Detectable in hyperornithinaemia
Tyrosine	1. Tyrosinosis Type 1 (fumarylacetoacetase deficiency)	Transient tyrosinosis in newborns; high protein intake; galactosaemia; fructose intolerance; liver disease (hepatitis, Wilsons disease)

Amino acid	Primary metabolic disorder	Secondarily increased (decreased) in:
	2. Tyrosinosis Type 2 (cytosolic tyrosine-aminotransferase defiency	
β-Alanine	Hyper-P-alaninaemia (β-alanine-2-oxo-glutarate-amino-transferase deficiency	After kidney transplantation, when the kidney is rejected, carnosinuria
Phenylalanine	1. Phenylketonuria (phenylalanine hydroxylase deficiency)	Newborns (especially in maternal PKU); high protein intake; tyrosinosis
	2. Hyperphenylalaninaemia (dihydropteridine reductase deficiency)	
	3. Hyperphenylalaninaemia (various disorders of dihydro- pteridine biosynthesis	
β-Aminoisobutyric acid	β-Aminoisobutyric aciduria - occurs in 6-10 % of the normal population in N.W. Europe (not considered as a disease)	Various types of neoplastic disease; excessive tissue break-down
Homocystine	1. Homocystinuria (cystathionine-β-synthase deficiency)	Vitamin B_{12} malabsorption or deficiency together with methylmalonic acid; vitamin B_6 deficiency
	2.Homocystinuria (N^5,N^{10}-methylene-THF-reductase defiency)	
	3. Homocystinuria (N^5-methyl-THF-methyl-transferase defiency	
y-Aminobutyric acid	Not described with an expression in urine	β-Alaninaemia; intestinal dysbacteriosis
Tryptophan	Tryptophan pyrrolase deficiency (?)	Hartnup disease; hypoalbuminaemia: (Decreased in intestinal tryptophan malabsorption; chronic diarrhoea or constipation)
Ethanolamine	Ethanolaminosis (hepatic ethanolamine kinase deficiency)	Liver cirrhosis; hyperlysinaemia; hypersarcosinaemia; new-borns
Ornithine	1. Hyperornithinaemia (ornithine-2-oxo-acid-amino-transferase deficiency)	Not described
	2. Dibasic aminoaciduria (transport defect)	
Lysine	1. Hyperlysinaemia (lysine-2-oxoglutarate reductase deficiency	Newborns together with cystine; hyperammonaemia (sometimes together with homocitrulline and arginine); saccharopinuria
	2. Dibasic aminoaciduria (transport defect)	
	3. Lysinuric protein intolerance (transport defect)	
1-Methylhistidine	Not described	Renal insufficiency; by alimentary cause (chicken)
Histidine	Histidinaemia	Newborns; folic acid deficiency
3-Methylhistidine	Not described	Malnutrition, starvation

Amino acid	**Primary metabolic disorder**	**Secondarily increased (decreased) in:**
Carnosine	Carnosinaemia (carnosinase deficiency)	High meat diet (especially chicken, turkey, etc.); infants up to 2 years of age
Anserine	Carnosinaemia (carnosinase deficiency)	High intake of meat, especially poultry
Arginine	1. Hyperargininaemia (arginase deficiency)	Lysinaemia; ornithinaemia
	2. Dibasic aminoaciduria (transport defect)	
	3. Lysinuric protein intolerance (transport defect)	