

Hyperammonemia

Glutamine
low-normal

Glutamine
elevated

Preterm

Full Term

Citrulline
↓

Citrulline
↑

Citrulline
↑↑

Citrulline
↑↑↑

THAN
normal AA
profile

Organic acid ↑↑
Acylcarnitines ↑

Citrulline
↑↑

U Orotic acid
normal

U Orotic acid
elevated

↓ Arg, Lys, Orn
↑ U Arg, Lys, Orn
↑ U Orotic acid
LPI

↑ ASA
↑ U Orotic acid
ASL D

↑ U Orotic acid
ASS D

Organic acidurias
MMA – PA
Acidosis

FAO defects
MCAD – VLCAD – TPD
Hypoglycemia

Hypoglycemia
HI-HA
HMG

CIT 2
↑ Met, Tyr
A-FP, galactose

PC deficiency
Lactic acidosis

NAGS D
CPS D

P5CS D
↓ Orn, Pro, Arg
starved
hyper-NH₃

HHH s
↑ Homocit

OAT D

Ornithine
elevated

Ornithine
normal

↓ Arg
↑ Ala
OTC D *

↑ ↑ Arg
ARG1 D

Orn can be
transiently normal
OAT D