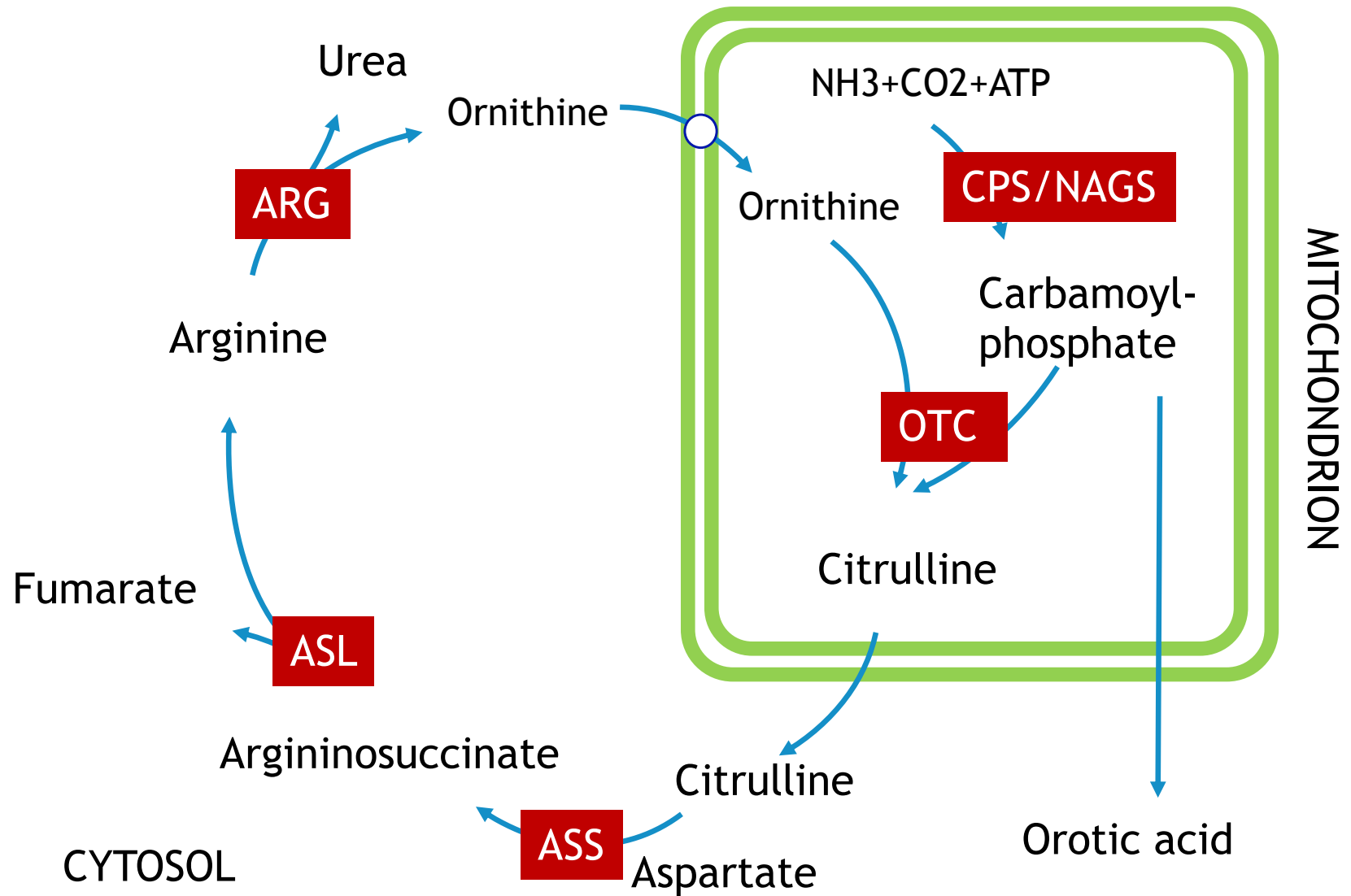


# Urea Cycle Disorders Pathway & Enzymes

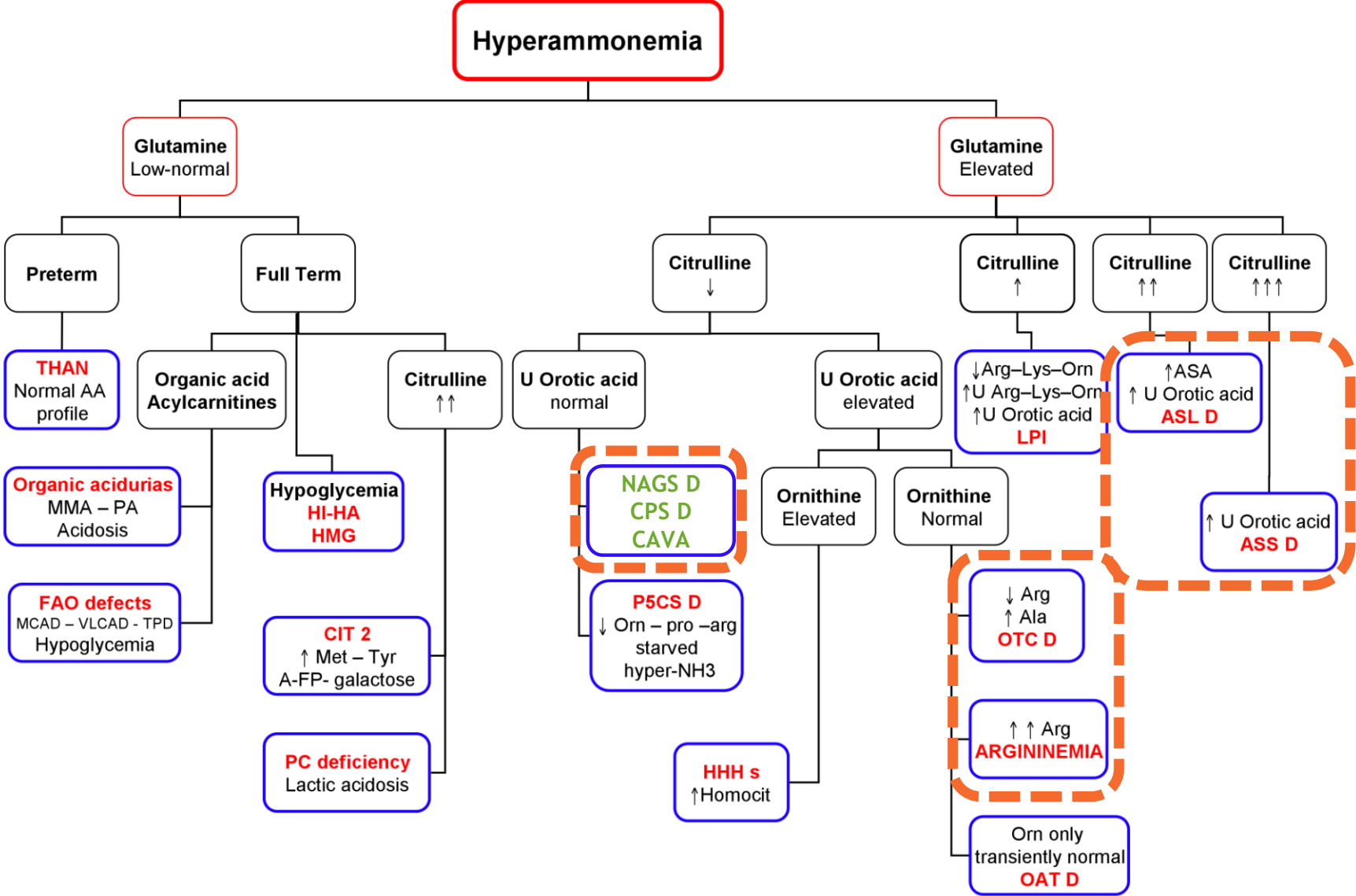


## Urea Cycle Disorders

### Summary of biochemical patterns

DEFECT	GLN	CIT	ARG-SUC ACID	ARG	OROTIC ACID
NAGS	High	Low	Low-normal	Low-normal	Low-normal
CPS 1	High	Low	Low-normal	Low-normal	Low-normal
OTC	High	Low	Low-normal	Low-normal	High
ARG-SUC SYNTH	High	Very high	Low-normal	Low-normal	High
ARG-SUC LYASE	High	High	High	Low-normal	High
ARGINASE	±High	Normal	Normal	High	±High

# Neonatal Hyperammonemia



 = Urea Cycle Disorders