

## **Step 2: Formation of Citrulline**

- ⦿ **The second reaction is also mitochondrial.**
- ⦿ **Citrulline is synthesized from carbamoyl phosphate & ornithine by ornithine transcarbamoylase.**
- ⦿ **Ornithine is regenerated & used in urea cycle.**

- ⦿ **Ornithine & citrulline are basic amino acids.**  
(Never found in protein structure **due to lack of codons**).
- ⦿ **Citrulline is transported to cytosol by a transporter system.**
- ⦿ **Citrulline is neither present in tissue proteins nor in blood; but it is present in milk.**

### **Step 3: Formation of Arginosuccinate**

- ⦿ **Citrulline condenses with aspartate to form arginosuccinate by the enzyme Arginosuccinate synthetase.**
- ⦿ **Second amino group of urea is incorporated.**
- ⦿ **It requires ATP, it is cleaved to AMP & P<sub>Pi</sub>**
- ⦿ **2 High energy bonds are required.**
- ⦿ **Immediately broken down to inorganic phosphate (P<sub>i</sub>).**

## **Step:4 Formation of Arginine or cleavage of Arginosuccinate**

- ⦿ **The enzyme Argininosuccinase or argininosuccinate lyase cleaves arginosuccinate to arginine & fumarate (an intermediate in TCA cycle)**
- ⦿ **Fumarate provides connecting link with TCA cycle or gluconeogenesis.**

- ⊙ **The fumarate is converted to oxaloacetate via fumarase & MDH & transaminated to aspartate.**
- ⊙ **Aspartate is regenerated in this reaction.**



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