

**Leucine****Valine****Isoleucine****TRANSAMINATION***(Branched-chain amino acid aminotransferase)* $\alpha$ -Ketoiso-  
caproic acid $\alpha$ -Ketoiso-  
valeric acid $\alpha$ -Keto- $\beta$ -methyl-  
valeric acid**OXIDATIVE DECARBOXYLATION***(Branched-chain  $\alpha$ -keto acid dehydrogenase  
coenzymes: NAD<sup>+</sup>, CoA, TPP, lipoic acid, FAD)*

Oxidative decarboxylation of branched-chain amino acids is deficient in maple syrup urine disease.

Isovaleryl CoA

Isobutyryl CoA

 $\alpha$ -Methyl-  
butyryl CoA**FAD-linked DEHYDROGENATION** $\beta$ -Methyl-  
crotonyl CoA

Biotin

 $\beta$ -Methyl-  
glutaconyl CoA

HMG CoA

**ACETO-  
ACETATE**

+

**ACETYL CoA**

Propionyl CoA

Biotin

Methylmalonyl CoA

5'-Deoxyadenosyl-  
cobalamin  
(derivative of B<sub>12</sub>)**SUCCINYL CoA**