

MAPLE SYRUP URINE DISEASE

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- the amino acids leucine, isoleucine and valine are collectively known as the branched-chain amino acids
- the disease name stems from the odor of “burnt maple syrup” caused by the accumulation of substances (ketoacids) that cannot be processed in this disease
- treatment: limiting the intake of the three amino acids by low protein diet and by consuming formulas containing amino acids but lacking the branched-chain amino acids

