

**ACUTE ILLNESS PROTOCOL**  
**ORGANIC ACIDEMIAS**  
**MAPLE SYRUP URINE DISEASE (MSUD)**

Maple syrup urine disease (MSUD) is an organic acid disorder\* but is often classified as an amino acid disorder because of increased levels of branched chain amino acids as well as the organic acids. The central emergency features of MSUD are profound metabolic ketoacidosis and brain edema with lethargy progressing to coma.

*\*Organic acids are distinguished from amino acids in that they do not contain nitrogen.*