

ACUTE ILLNESS PROTOCOL ORGANIC ACID DISORDERS

ISOVALERIC ACIDEMIA

INTRODUCTION

The organic acidemias comprise a group of metabolic disorders in which the defect produces an accumulation of organic acids*. The central emergency features of the organic acid disorders are profound metabolic **ketoacidosis** and **hypoglycemia**. The inheritance of Isovaleric acidemia is autosomal recessive. It is often referred to as the “sweaty foot syndrome” due to the characteristic odor body and body fluids odor produced by it.

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