musty odor associated with the disease. Another is phenylpyruvic acid, which is responsible for the term phenylketonuria.

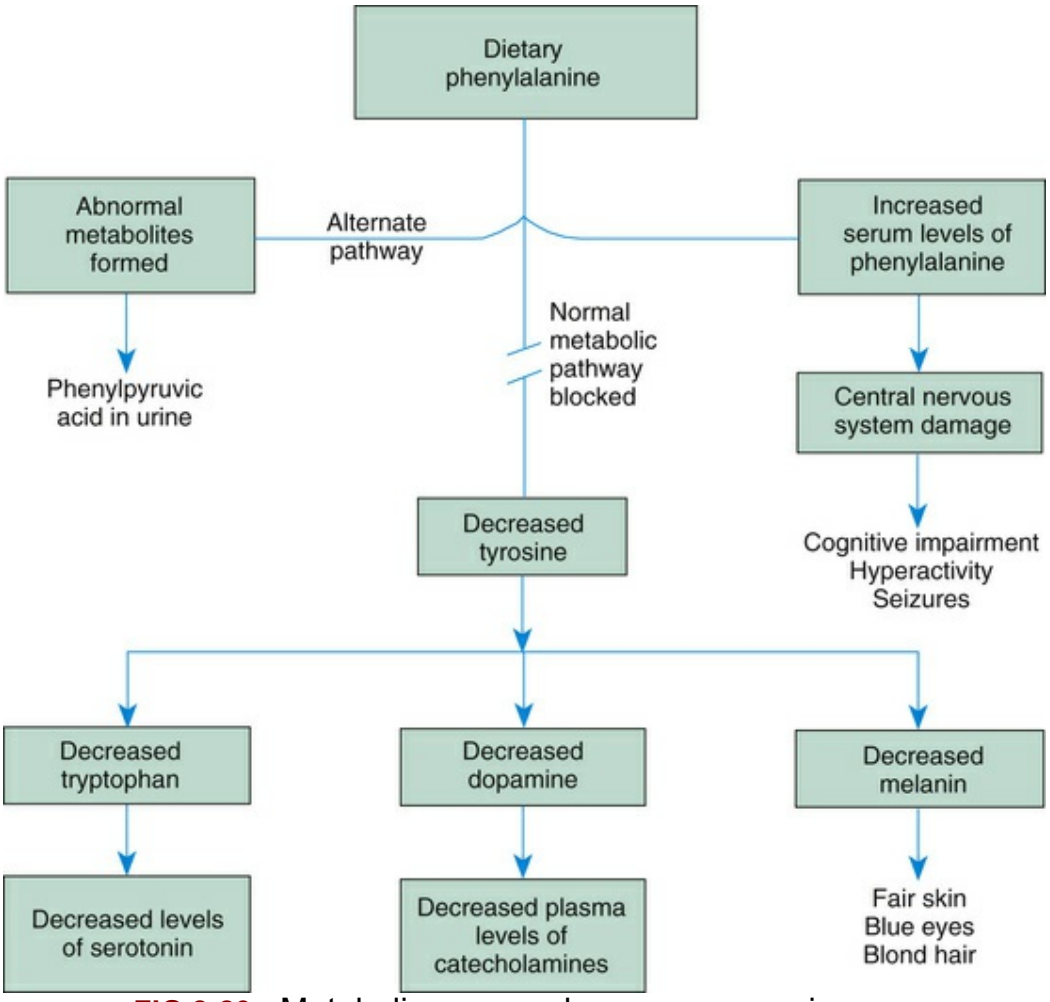

FIG 8-23 Metabolic error and consequences in phenylketonuria.

Tyrosine, the amino acid produced by the metabolism of phenylalanine, is absent in PKU. Tyrosine is needed to form the pigment melanin and the hormones epinephrine and T_4. Decreased melanin production results in similar phenotypes of most individuals with PKU, which is blond hair, blue eyes, and fair skin that is particularly susceptible to eczema and other dermatologic problems. Children with a genetically darker skin color may be red haired or brunette.

The prevalence of PKU varies widely in the United States because different states have different definition criteria for what constitutes