

DIAGNOSIS AND MANAGEMENT OF CHRONIC NEUTROPENIA DURING CHILDHOOD

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Chronic neutropenia is defined as an abnormally low absolute neutrophil count (ANC) lasting more than 6 months. Many attempts have been made to classify the different forms of chronic neutropenia during childhood, but unfortunately a comprehensive classification system is lacking. Pathophysiologically, neutropenia can occur as a result of decreased bone marrow production (e.g., Kostmann syndrome), failure to release mature neutrophils from the bone marrow (myelokathexis), increased margination of neutrophils, or increased destruction in the peripheral circulation (e.g., autoimmune neutropenia or hypersplenism) (Table 1).

The risk for infection is inversely proportional to the ANC. When the ANC falls below 1000 per mm³, stomatitis, gingivitis, and cellulitis dominate the clinical picture. More severe infections occur when the ANC is below 500 per mm³, with perirectal abscesses, pneumonia, and sepsis particularly common. *Staphylococcus aureus* is the most common isolated pathogen; however, in more severely affected patients, infections develop with gram-negative enteric bacilli, including *Klebsiella* species, *Escherichia coli*, and *Pseudomonas aeruginosa*. Uncommonly, fungal organisms also cause infection in patients with chronic neutropenia.

Considerable variability exists regarding susceptibility to infection for patients with similar circulating neutrophil counts. Monocytosis often occurs in patients with diverse types of chronic neutropenia, which

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