All of the following statements regarding chronic autoimmune neutropenia of infancy are true, except:

1. Usually runs a benign course
2. Neutropenia is incidentally detected in most of the cases
3. Many children would have autoantibodies directed against surface antigens of neutrophils
4. Many patients require GCSF therapy

Correct answer: 4

Explanation:

Chronic autoimmune neutropenia of infancy and early childhood is a relatively common disorder and virtually always runs a benign course, despite very low absolute neutrophil counts (ANCs). It usually resolves spontaneously by age 3–5 years, with a mean duration of 17 months. In most cases, neutropenia is detected during the occurrence of an acute febrile illness. With follow-up, the neutropenia persists after resolution of the illness that led to testing. Systematic studies indicate that many, but not all, of these children have autoantibodies directed against surface antigens of neutrophils. From a clinical perspective the value of testing for autoantibodies in patients with moderate to severe neutropenia without evidence of recurrent fevers or infections is debatable. Testing is not widely available and, if done, it is best performed by a reference laboratory performing these assays frequently. Serial testing may give inconsistent results and patients with genetic as well as acquired neutropenia may have false positive test results.
In older children, chronic autoimmune neutropenia or multiple immune cytopenias should raise suspicion of a congenital immunological disorder such as autoimmune lymphoproliferative syndrome or common variable immunodeficiency. Screening for these disorders can be performed by measurement of circulating T-cell receptor alpha/beta positive, CD4/CD8 double-negative T cells, or serum immunoglobulins, respectively.

**Reference and suggested reading:**