

A Case of X-linked Agammaglobulinemia

by

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Case Presentation

Billy DeWitt was a normal, full-term baby at birth. Beginning at about 10 months of age, Billy suffered from a series of infectious processes such as sinusitis, otitis media, and pneumonia. All of these conditions were successfully treated with antibiotics, but within a few weeks of the resolution of one infection, another would occur.

Now at about four years of age, Billy is examined by a pediatrician who notes that Billy lacks palatine tonsils, although he does not have a history of tonsillectomy. Questioning of Billy's mother reveals that she had two male relatives who died in infancy from infectious disease. The physician orders laboratory tests that reveal that the quantity of immunoglobulin in Billy's serum is about one-fifth of normal, and that there is a marked deficiency in the number of circulating B-lymphocytes in Billy's blood. Tests to determine the functional state of Billy's T-lymphocytes are all normal.



Billy is diagnosed as having a genetic disorder called X-linked Agammaglobulinemia. He begins a course of monthly intravenous injections of gamma globulin which he will need to continue for the rest of his life. Billy should no longer suffer recurrent infections and should develop physically and mentally as a normal child. Aside from receiving monthly injections of gamma globulin, he should lead a normal life now that this course of treatment is in place.

Questions

1. What are the differences between nonspecific and specific (immunity) body defenses?
2. In what tissue do B- and T-lymphocytes originate and what are the two steps involved in lymphocyte "maturation"?
3. Describe the two "arms" of immunity.
4. Define the term *antigen* and state which class of organic molecules make the best antigens, and why.
5. What are the five classes of antibody?
6. What are the means by which antibody molecules exert a protective effect?
7. What are the basic differences between *active* and *passive* immunity?
8. Billy was free of infections for the first few months of life. Why?
9. Why did Billy lack tonsils?
10. Explain X-linked inheritance, and name other genetic diseases that are known to be X-linked?

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