

CONQUERING INHERITED IMMUNE DEFICIENCY— CHILDREN WHO MADE MEDICAL HISTORY

The importance of T cells in establishing and maintaining immunity is obvious in AIDS, which is acquired, and in severe combined immune deficiency (SCID), which is inherited. The 20 types of SCID disrupt receptors on T cells or hamper cytokine production. SCID is called “combined” because both T and B cells are affected. Medical technology has evolved to treat SCID. Here is a look at some young pioneers.

David

David Vetter was born in Texas in 1971 without a thymus. He therefore could not make mature T cells or activate B cells. He spent his short life in a vinyl bubble that protected him from infection, awaiting a treatment that never came (fig. 16B). Before AIDS, living without immunity was very unusual.

As David reached adolescence, he desperately wanted to leave the bubble. After receiving a bone marrow transplant, he did so. But the transplant hadn't worked, and within days, David began vomiting and developed diarrhea, both signs of infection. He soon died.

Laura

For her first few years, Laura Cay Boren didn't know what it felt like to be well (fig. 16C). Ever since her birth in July 1982, she fought infection after infection. Colds landed her in the



FIGURE 16B

David Vetter was born without a thymus. Because his T cells could not mature, he was virtually defenseless against infection.

hospital with pneumonia, and routine vaccines caused severe skin abscesses. Laura had inherited a form of SCID in which the body lacks an enzyme, adenosine deaminase (ADA). Lack of ADA blocks a biochemical pathway that breaks down a metabolic toxin, which instead builds up and destroys T cells. The T cells in turn can no longer activate B cells. Immunity fails.

Laura underwent two bone marrow transplants, which temporarily restored her immune defenses, and blood transfusions helped. But by the end of 1985, Laura was near death. Then she was chosen to receive experimental injections of ADA altered to



FIGURE 16C

Laura Cay Boren spent much of her life in hospitals until she received the enzyme her body lacks, adenosine deaminase (ADA). Here, she pretends to inject her doll as her mother looks on.

remain in the bloodstream long enough to help T cells survive. It worked! Within hours of the first treatment, Laura's ADA level increased twenty-fold. After three months, her immune function neared normal, and stayed that way, with weekly ADA shots. By the following year, Laura began school. She is healthy today.

Ashi

In the late 1980s, the DeSilvas did not think that their little girl, Ashanthi (“Ashi”), would survive. She suffered near-continual coughs