

**SUBCOMMITTEE ON LABOR, HEALTH AND HUMAN  
SERVICES, EDUCATION AND RELATED AGENCIES**

**TESTIMONY PRESENTED BY**

**JOANNE BAKKEN PEASE  
IMMUNE DEFICIENCY FOUNDATION**

**JANUARY 28, 1998  
2:00 P.M.**

I thank you for the opportunity to speak here today on behalf of the Immune Deficiency Foundation. My three sons and my nephew have primary immunodeficiency diseases which means that they were born with incomplete immune systems. My name is Joanne Bakken Pease and I am a volunteer with the Washington State Chapter of the Immune Deficiency Foundation.

As I tell my story today, I am mindful of the significant work being done by the Immune Deficiency Foundation on behalf of the approximately 40,000 individuals affected by these genetic disorders of the immune system. The IDF is developing a national registry of US patients suffering from the most common of these disorders, funded by a grant from NIAID. The registry is providing an important resource to physicians and investigators by giving them access to a more complete clinical understanding of these diseases. The clinical information contained in these registries will help determine the frequency of complications, long term prognosis and possibly open the gateway for gene therapy. As a mother of three affected children, I am proud of the work of the Foundation, and in sharing my story I hope to express to you the need for this work to continue.

In November of 1985 my eldest son Curtis received his vaccine for measles mumps and rubella. Curtis got the mumps from the vaccination in February 1986, and the Doctor told me - *these things happen*. After three years of constant colds and pneumonia - Curtis was diagnosed with X-linked Agammaglobulinemia. I was thankful when I learned there was treatment available for him. However, the treatment consisted of a very painful injection administered intramuscularly every ten days. This treatment, a pooled plasma derivative, replaced portions of his incomplete immune system. However, the pain involved caused this therapy to be a source of terror in our household, requiring four adults for administration.

When my second son Jeff was ten months old we learned he had contracted polio from the oral vaccine, signaling the presence of the same immunodeficiency. He was left with a withered right leg and a terribly deformed foot. Now both my beautiful boys received these painful shots. Three years later they both started getting their medicine intravenously. Intravenous immune globulin was less painful and more effective, it reduced our infusions to once a month.

Jeff has had six very painful orthopedic surgeries including, tendon transfers and releases, hip reconstruction, leg rotations and ankle repositioning. He is now ready for his seventh and eighth operations which will be complicated leg lengthening.

My nephew Joshua was born in 1988 and my third son Mitchell in 1990, both were diagnosed with XLA at birth and began their treatments.

We have learned to cope with the care necessary to keep our children alive. However, issues related to the safety of the blood supply make me feel that my children are so vulnerable. Between the four boys they receive 70 infusions per year.

My children's lives are not what I envisioned for them. The pain that I feel for the loss of my dream will never go away. We need to continue the focus on medical research so perhaps future generations will have the option of gene therapy and not have to suffer years of chronic care.

For fiscal year 1999, the Immune Deficiency Foundation recommends the National Institute of Allergy and Infectious Diseases receive a 15% increase over last year. This percentage translates into \$202.7 million over FY 98 and would bring NIAID's total appropriation to \$1.55 billion. In addition, Mr. Chairman, I would like to make clear that although IDF strongly supports the concept of doubling NIH's overall budget in the next five years, we do not believe that these increases should come at the expense of other important public health service programs.

As you regard your decision to increase funding please remember my boys and the approximately 40,000 primary immunodeficient patients affected by your vote.

Thank you.