

**STATEMENT OF THE  
IMMUNE DEFICIENCY FOUNDATION  
MEDICAL ADVISORY COMMITTEE**

**DEPARTMENT OF HEALTH AND HUMAN SERVICES  
ADVISORY COMMITTEE  
ON BLOOD SAFETY AND AVAILABILITY**

**NOVEMBER 24, 1998**

The primary immunodeficiency diseases are a group of nearly 80 different disorders that are intrinsic to the immune system and result in immunodeficiency. Most patients present clinically with an increased susceptibility to infection. These infections are marked by unusual severity, and are generally chronic or unremitting. A point especially relevant to today's discussion, since patients with primary immunodeficiency diseases may suffer severe complications from liver viral infections such as Hepatitis C.

The Immune Deficiency Foundation was founded in 1980 to further education and research into the primary immunodeficiency diseases and thereby improve clinical care and prognosis of these patients. The Foundation is comprised of over 20 chapters and represents nearly 50,000 US patients. The Foundation's Medical Advisory Committee is composed of 20 leading clinical immunologists who specialize in the care of patients with primary immunodeficiency diseases. Their function is to advise the Foundation on its many medical programs and develop position statements on issues related to the care and treatment of primary immunodeficient patients. It is on behalf of the IDF Medical Advisory Committee that I am making today's statement.

Of the nearly 50,000 US patients, who have primary immunodeficiency diseases, we have estimated from our survey data that some 20,000 to 30,000 currently receive IGIV antibody replacement therapy. Since the introduction of these products, these patients can look forward to a normal, or near normal life span.

However, adverse events associated with the administration of IGIV have occurred, and have forever changed or ended the lives of some patients. Most recently some of our patients have experienced an outbreak of Hepatitis C due to the use of IGIV. In July 1994, 112 cases of Hepatitis C were reported, but this was just the first wave. Because of the lack of surveillance within this community, we do not yet know how many people were ultimately infected. This has been a tragedy for the individual patients, but it is also unfortunate that we have not learned anything about the management and natural history of Hepatitis C in this vulnerable population. As recently as October 1998, the CDC's MMWR, Vol. 47/No. RR-19, neglected to mention IGIV recipients during the period from 1993 to 1994 as high risk individuals who should be screened for HCV, although the MMWR does mention the transmission via IGIV.

The establishment of a national registry of these cases would allow for comprehensive surveillance and thus we could learn about the natural history of Hepatitis C within the primary immunodeficiency diseases. Physicians are currently unable to counsel their patients concerning the best treatment, relate the disease severity to immune functions, give an estimate of the number of cases who have needed liver transplantation, or give any results about the outcome of this procedure and for which patient it proved the most useful. A national registry would be a most

valuable scientific resource for physicians who are still dealing with the aftermath of this outbreak, and for scientists who want to learn more about this common disease.

While it is presumed that transmission of Hepatitis C through IGIV is no longer a threat to our patient population due to additional viral screening and viral inactivation steps, we should not fail patients who may be unaware that they have contracted this disease or to improve the management of already diagnosed patients.

In summary the IDF Medical Advisory Committee makes the following recommendations:

1. The FDA, NIH, or CDC should establish a sufficient look-back and registry program to determine how many cases of Hepatitis C occurred in the United States, in recipients (both with and without primary immunodeficiency disease) of intravenous immunoglobulin during the relevant years.
2. A retrospective notification of primary immunodeficient patients and physicians should be conducted to alert these individuals to the possibility of a past Hepatitis C transmission.
3. Determine if the type of primary immunodeficiency which the patient has determines the clinical presentation, clinical course and eventual outcome of the Hepatitis C infection.
4. Determine what anti viral drugs have been efficacious, and to what degree they have been useful.
5. Determine the degree of liver disease that developed, if liver transplantation was done, and its outcome.

Thank you for the opportunity to comment.