

## Introduction to Epstein-Barr Virus and Lymphoproliferative Diseases in Immunodeficient Individuals

David T. Purtilo and George Klein

Department of Pathology and Laboratory Medicine, University of Nebraska Medical Center, Omaha, Nebraska 68105 and Department of Tumor Biology, Karolinska Institute, 104 01 Stockholm 60, Sweden

EBV<sup>1</sup> is an ubiquitous B-cell-tropic virus responsible for infectious mononucleosis (5) and is one possible etiological agent of Burkitt lymphoma and nasopharyngeal carcinoma (6, 7). When the virus infects a young child, silent seroconversion is the rule. In contrast, primary infection of young adults results in infectious mononucleosis in about one-half of the cases (6). The observations of fatal infectious mononucleosis, agammaglobulinemia, and aplastic anemia associated with infectious mononucleosis and malignant B-cell lymphoma in 6 maternally related males in the Duncan kindred (11) and other similar families (1-4, 8, 9) had suggested to Purtilo (10) that immunodeficient responses to EBV were responsible for a variety of diseases in males with the X-linked lymphoproliferative syndrome. Primary infection by EBV normally provokes potent immune responses which efficiently cope with the B-cell proliferation and provide lifelong latency (6). Persons with inherited or acquired immune defects, depending on the type and degree of the deficiency, are more or less vulnerable to life-threatening B-cell proliferation.

The articles published in this symposium focus on the possible roles of EBV and immunodeficiency in lymphoproliferative disorders. Some of the major objectives of the reports are to: describe various diseases possibly associated with EBV in immunodeficient patients; demonstrate inherited or acquired cellular and humoral immune defects to EBV-specific antigens;

and document the presence of viral genome in lymphoid tissues from the immunodeficient patients. The authors have sought to develop criteria for establishing involvement or noninvolvement of EBV in the lymphoproliferative diseases in immunodeficient patients.

### References

1. Banihashemi, A., Nasr, K., Hedayateh, H., and Mortazavee, H. Familial lymphoma including a report of familial primary upper small intestine lymphoma. *Blut*, 26: 363-368, 1973.
2. Bar, R. S., DeLor, C. J., Clausen, K. P., Hurtubise, P., Henle, W., and Hewetson, J. F. Fatal infectious mononucleosis in a family. *N. Engl. J. Med.*, 290: 363-367, 1974.
3. Falletta, J. M., Fernbach, D. J., Singer, D. B., South, M. A., Landing, B. H., Heath, C. W., Shore, N. A., and Barrett, F. F. X-linked recessive reticulo-endotheliosis with hyperglobulinemia in relatives. *J. Pediatr.*, 83: 549-555, 1973.
4. Hambleton, G., and Cottom, D. G. Familial lymphoma. *Proc. R. Soc. Med.*, 62: 1095, 1969.
5. Henle, G., Henle, W., and Diehl, V. Relation of Burkitt's tumor-associated herpes-type virus to infectious mononucleosis. *Proc. Natl. Acad. Sci. U. S. A.*, 59: 94-101, 1968.
6. Henle, W., Henle, G., and Lennette, E. L. The Epstein-Barr virus. *Sci. Am.*, 241: 48-59, 1979.
7. Klein, G. The Epstein-Barr virus and neoplasms. *N. Engl. J. Med.*, 293: 1353-1354, 1975.
8. Maurer, H. S., Gotoff, S. P., Allen, L., and Bolan, S. P. Malignant lymphoma of the small intestine in multiple family members: association with an immunologic deficiency. *Cancer (Phila.)*, 37: 2224-2231, 1976.
9. Pevzner, S., and Leef, F. Childhood abdominal lymphoma in two brothers. *Isr. J. Med. Sci.*, 9: 914-917, 1973.
10. Purtilo, D. T. Pathogenesis and phenotypes of an X-linked lymphoproliferative syndrome. *Lancet*, 2: 882-885, 1976.
11. Purtilo, D. T., Cassel, C., Yang, J. P. S., Stephenson, S. R., Harper, R., Landing, G. H., and Vawter, G. F. X-linked recessive progressive combined variable immunodeficiency (Duncan's disease). *Lancet*, 1: 935-941, 1975.

<sup>1</sup> The abbreviation used is: EBV, Epstein-Barr virus.  
Received July 8, 1981; accepted July 22, 1981.

# Cancer Research

The Journal of Cancer Research (1916–1930) | The American Journal of Cancer (1931–1940)

**AACR** American Association  
for Cancer Research

## Introduction to Epstein-Barr Virus and Lymphoproliferative Diseases in Immunodeficient Individuals

David T. Purtilo and George Klein

*Cancer Res* 1981;41:4209.

**Updated version** Access the most recent version of this article at:  
[http://cancerres.aacrjournals.org/content/41/11\\_Part\\_1/4209.citation](http://cancerres.aacrjournals.org/content/41/11_Part_1/4209.citation)

**E-mail alerts** [Sign up to receive free email-alerts](#) related to this article or journal.

**Reprints and Subscriptions** To order reprints of this article or to subscribe to the journal, contact the AACR Publications Department at [pubs@aacr.org](mailto:pubs@aacr.org).

**Permissions** To request permission to re-use all or part of this article, use this link [http://cancerres.aacrjournals.org/content/41/11\\_Part\\_1/4209.citation](http://cancerres.aacrjournals.org/content/41/11_Part_1/4209.citation). Click on "Request Permissions" which will take you to the Copyright Clearance Center's (CCC) Rightslink site.