primary tumors, most often for AML, myelodysplastic syndromes, or breast cancer.

## CONGENITAL DISORDERS

Patients with some rare congenital chromosomal abnormalities have a higher risk of development of acute leukemia (e.g., Klinefelter's syndrome, Fanconi's anemia, Bloom's syndrome, ataxia-telangiectasia, and neurofibromatosis). Those with Down's syndrome have a twentyfold increased incidence of leukemia; ALL is increased in childhood and AML at an older age.

## INFECTIOUS AGENTS

No direct evidence implicates viruses as a major cause of human acute leukemia. However, viruses are involved in the pathogenesis of two lymphoid neoplasias. In the endemic African type of Burkitt's lymphoma, the Epstein-Barr virus, a DNA virus of the herpes family, has been implicated as a potential causative agent (see **Chap. 194**). Endemic infection with human T-cell leukemia virus I in Japan and the Caribbean has been shown to be an etiologic agent for rare cases of adult T-cell leukemia/lymphoma (see **Chap. 201**).

## DIAGNOSIS AND CLASSIFICATION

The diagnosis of acute leukemia is first made by examination of the peripheral blood and bone marrow. For further classification of the leukemic blast cells, cytochemical stains, immunologic markers, and cytogenetic and molecular analysis are required. The immunologic markers are still the major criteria to subdivide into B-cell lineage or T-cell lineage ALL leukemias.

## PERIPHERAL BLOOD

Peripheral blood counts and a differential count from a Wright-Giemsa–stained blood smear are essential at the time of presentation. The white blood cell (WBC) count in ~40% of ALL patients is reduced or normal **(Table 106-1)**. Only 16% of patients have a WBC above $100 \times 10^5$/L. It is noteworthy that in 8% of ALL patients, no circulating leukemic blast cells were observed. Thus, in