TABLE 102-1 CONDITIONS CAUSING IMMUNE DEFICIENCY

NEUTROPHILS AND PHAGOCYTES

Neutropenia
- Congenital syndromes
- Drug-related neutropenia (e.g., chemotherapy, antimicrobial agents, antipsychotics, anticonvulsants)
- Autoimmune neutropenia
- Cyclic neutropenia
- Myelodysplastic syndrome
- Fanconi's anemia
- Aplastic anemia
- Myeloproliferative disorders (e.g., acute myeloid leukemia)

Neutrophil dysfunction
- Chédiak-Higashi syndrome
- Hyperimmunoglobulin E syndrome (Job's syndrome)
- Chronic granulomatous disease
- Leukocyte adhesion deficiency
- Immunosuppressive medications (e.g., mycophenolate, azathioprine)
- Warts, hypogammaglobulinemia, infections, and myelokathexis (WHIM)
- Viral infections (e.g., human immunodeficiency virus, human herpesvirus 6)

CELL-MEDIATED IMMUNITY

Immunosuppressive agents for transplantation
- Cyclosporine, tacrolimus, sirolimus
- Daclizumab, basiliximab
- Mycophenolate, azathioprine
- Antilymphocyte therapies (e.g., Thymoglobulin, alemtuzumab)

Corticosteroids
Cytotoxic drugs (e.g., cyclophosphamide)
Fludarabine
Anti–tumor necrosis factor-α agents (e.g., adalimumab, etanercept, infliximab, certolizumab, golimumab)
Graft-versus-host disease (GVHD)
DiGeorge syndrome (i.e., thymic hypoplasia)
Severe combined immunodeficiency (SCID)
Ataxia-telangiectasia
Wiskott-Aldrich syndrome
End-stage renal disease
Malnutrition
Human immunodeficiency virus (HIV) infection
T-cell lymphoma
Idiopathic $CD4^+$ lymphopenia

HUMORAL IMMUNITY

Common variable immune deficiency (CVID)
Splenectomy, splenic aplasia (e.g., sickle cell disease)
Nephrotic syndrome
Protein-losing enteropathy
Multiple myeloma
B-cell lymphoma
Chronic lymphocytic leukemia
Waldenstrom's macroglobulinemia
Severe combined immune deficiency
Ataxia-telangiectasia
Wiskott-Aldrich syndrome
Hyperimmunoglobulin M syndrome
Selective IgA deficiency
X-linked agammaglobulinemia
Immunosuppressive therapies (e.g., cyclophosphamide, azathioprine, mycophenolate)
Medications (e.g., rituximab, azathioprine, sulfasalazine, gold, cyclosporine, carbamazepine, valproic acid, phenytoin, alemtuzumab, chloroquine)
Hypogammaglobulinemia complicating solid organ and hematopoietic stem cell transplantation

COMPLEMENT DEFICIENCY

C2 deficiency
Mannose-binding lectin deficiency
C3 deficiency
Factor H deficiency
Factor I deficiency
Terminal pathway (C5-C9) deficiency

TABLE 102-2 ORGANISMS ASSOCIATED WITH IMMUNE DYSFUNCTION

NEUTROPHILS AND PHAGOCYTES

Staphylococcus auerus
Pseudomonas aeruginosa
Enterobacteriaceae
Streptococcus mitis, viridans streptococci
Aspergillus species
Candida species
Mucorales order fungi (cause mucormycosis)
Fusarium species
Herpes simplex virus (HSV)

CELL-MEDIATED IMMUNITY

Herpesviuruses (HSV, varicella-zoster virus, Epstein-Barr virus, human herpesviruses 6 and 8)
JC virus
BK virus (especially in kidney transplants)
Human papilloma virus (HPV)
Respiratory viruses (e.g., influenza, metapneumovirus, parainfluenza, respiratory syncytial virus)
Listeria monocytogenes
Nocardia species
Salmonella species
Mycobacterium species (*M. avium complex* in human immunodeficiency virus infection)
Cryptococcus neoformans
Aspergillus species
Candida species
Pneumocystis jirovecii
Strongyloides stercoralis
Cryptosporidium species
Toxoplasma gondii
Leishmania species

HUMORAL IMMUNITY

Mycoplasma species
Streptococcus pneumoniae
Haemophilus influenzae
Campylobacter jejuni
Ureaplasma urealyticum
Chlamydia pneumoniae
Salmonella species
Giardia lamblia
Echovirus
Varicella-zoster virus

COMPLEMENT DEFICIENCY

Recurrent sinopulmonary infections
Streptococcus pneumoniae
Haemophilus influenzae
Neisseria gonorrhoeae
Neisseria meningitidis

order fungi (which cause mucormycosis) and other molds, may be inhaled, causing sinopulmonary infections in neutropenic hosts. Infection may be complicated by dissemination to sites such as the skin and brain.

Bacterial infection with organisms such as *Nocardia* and *Legionella* and parasitic or protozoal infections such as *Strongyloides stercoralis* and *Babesia* are common in patients with cell-mediated immune defects.

Humoral Immunity

Humoral immunity is critical to control infection by encapsulated bacteria such as *Neisseria meningitidis*, *Streptococcus pneumoniae*, and *Haemophilus influenzae*. Patients with hypogammaglobulinemia, protein-losing conditions such as enteropathy or nephrotic syndrome, splenectomy, or chronic lymphocytic leukemia have significant defects in humoral immunity, predisposing them to infection with these organisms (see Table 102-1). Transplant recipients on immunosuppressive therapy for many years may develop hypogammaglobulinemia, predisposing them to similar infections. The use of agents such as rituximab, a monoclonal antibody against CD20, in malignancy and transplantation may result in significant B-cell defects and infection with encapsulated bacteria.

Complement Deficiency

Patients deficient in complement factors have a higher risk of autoimmune disease and can develop recurrent infections. Sinopulmonary infections, particularly from *S. pneumoniae* and *H. influenzae*, are common. Patients with deficiencies of terminal