

The background of the slide features a large, circular seal of the Bangkok Metropolitan Administrative Medical College. The seal has a blue outer ring with white Thai text. The top arc reads 'สุขภาพ ชีวดี ปิยะ' (Health, Good Life, Piyas) and the bottom arc reads 'วิทยาลัยแพทยศาสตร์กรุงเทพมหานครและวชิรพยาบาล' (Bangkok Metropolitan Administrative Medical College). In the center of the seal is a golden emblem consisting of a stylized flame or flower-like shape above two green snakes coiled around a central pedestal.

# **Primary Immunodeficiency**

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TABLE 3

**Warning Signs of Primary Immunodeficiency Disorders****Medical history**

Eight or more ear infections in one year  
Two or more serious sinus infections in one year  
Two or more bouts of pneumonia in one year  
Two or more deep-seated infections, or infections in unusual areas  
Recurrent deep skin or organ abscesses  
Need for intravenous antibiotic therapy to clear infection  
Infections with unusual or opportunistic organisms  
Family history of primary immunodeficiency

**Physical signs**

Poor growth, failure to thrive  
Absent lymph nodes or tonsils  
Skin lesions: telangiectasias, petechiae, dermatomyositis, lupus-like rash  
Ataxia (with ataxia-telangiectasia)  
Oral thrush after one year of age  
Oral ulcers

*Adapted with permission from The 10 warning signs of primary immunodeficiency. The Jeffrey Model Foundation, Copyright 2003. Accessed October 6, 2003, at: [http://npi.jmfworld.org/patienttopatient/index.cfm?section=warning signs&CFID=4441749&CFTOKEN=89405863](http://npi.jmfworld.org/patienttopatient/index.cfm?section=warning%20signs&CFID=4441749&CFTOKEN=89405863), with additional information from references 6 and 16.*

TABLE 1

**Clinical Findings in the Major Subgroups of Primary Immunodeficiency Disorders**

Subgroup	Onset	Pattern of infection	Other features
Disorders of humoral immunity (B-cell differentiation and antibody production)	After 6 months of age; can present in adulthood	Encapsulated bacteria: <i>Haemophilus influenzae</i> , pneumococci, streptococci Fungi and parasites: <i>Giardia lamblia</i> , <i>Cryptosporidium</i> species Virus: enterovirus (especially with X-linked agammaglobulinemia)	Recurrent infections: sinus infections, otitis media, bronchiectasis Chronic gastrointestinal tract problems, including malabsorption Autoimmunity Postvaccination paralytic polio (with live oral poliovirus vaccine)
T-cell and mixed disorders (combined B-cell and T-cell defects)	Before 6 months of age	Various opportunistic infections: <i>Mycobacterium</i> species, cytomegalovirus, Epstein-Barr virus, varicella virus, enterovirus, <i>Candida</i> species, <i>Pneumocystis carinii</i> (pneumonia)	Failure to thrive Oral thrush Graft-versus-host disease from maternal lymphocytes Excess diarrhea Postvaccination disseminated bacille Calmette-Guerin infection or paralytic polio
Phagocytic disorders	Infancy or childhood	Bacteria: <i>Staphylococcus aureus</i> , <i>Pseudomonas</i> species, <i>Serratia</i> species, <i>Klebsiella</i> species Fungi and parasites: <i>Candida</i> species, <i>Nocardia</i> species, <i>Aspergillus</i> species	Unusually severe infections by common pathogens Granuloma formation, including granulomatous enteritis Poor wound healing Abscesses, skin infections Oral cavity infections Anorectal infections
Complement disorders	Any age	<i>Neisseria</i> infections, including meningococcal and gonococcal infections	Rheumatoid disorders Lupus-like syndrome Scleroderma

Information from references 4 through 6.

**TABLE I.** Infections in phagocyte immunodeficiencies

	Microorganisms	Localization
CGD	Common: <i>Staphylococcus aureus</i> ; <i>Serratia marcescens</i> ; <i>Burkholderia cepacia</i> ; <i>Nocardia</i> species; <i>Aspergillus</i> species Less common: <i>Chromobacterium violaceum</i> ; <i>Paecilomyces</i> species; <i>Exophiala</i> species; <i>Scedosporium</i> species	Lung, liver, lymph nodes, skin sepsis ( <i>Chromobacterium</i> species, <i>Burkholderia</i> species)
LADs	<i>Staphylococcus aureus</i> ; gram-negative rods (enteric)	Cutaneous, gastrointestinal tract, sepsis
IFN- $\gamma$ /IL-12 deficiencies	Common: <i>Mycobacteria</i> species (nontuberculous and TB complex); <i>Salmonella</i> species Less common: <i>Listeria monocytogenes</i> , viruses, <i>Histoplasma capsulatum</i>	Disseminated, bone (AD IFN- $\gamma$ R1), lymph nodes

# Evaluation of T-cell function

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- Quantitative evaluation
  - : T-cell subpopulation
- Qualitative evaluation
  - : Delayed-type hypersensitivity (DTH)  
skin testing