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Pictorial Review

Respiratory disease in common variable immunodeficiency and other primary immunodeficiency disorders

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Respiratory disease is a significant cause of morbidity and mortality amongst patients with primary immunodeficiency disorders. Computed tomography (CT) plays an important role in the multidisciplinary approach to these conditions, in detecting, characterizing, and quantifying the extent of lung damage and in directing treatment. The aim of this review is to classify the primary immunodeficiency disorders and describe the thoracic complications and the associated CT findings whilst discussing the role of radiology in diagnosis and surveillance.

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Introduction

The primary immunodeficiency disorders (PIDs) are a broad spectrum of genetically determined diseases which result in impaired immunity and increased susceptibility to infection. Since Ogden Bruton first described X-linked agammaglobulinaemia (XLA) in 1952, over 100 additional genetically determined PIDs have been described.[1]

Humoral immunodeficiencies, i.e., those characterized by defective B cells and antibody production, are most common, accounting for about 70% of all primary immunodeficiencies.[2,3] Respiratory disease is a significant cause of morbidity and mortality amongst affected patients. The thoracic complications of the PIDs include: respiratory tract infections, airways disease, interstitial lung disease (ILD), and malignancy.

An understanding of the PIDs, their functional consequences, and histopathological sequelae enables greater understanding of the spectrum of radiological abnormalities encountered and, in turn, permits a more focused differential diagnosis to be offered. Computed tomography (CT) plays an important role in the multidisciplinary approach to these conditions, in detecting, characterizing, and quantifying the extent of lung damage, and in directing and monitoring treatment.

The aim of this review is to provide the classification and clinical manifestations of the PIDs, describe the thoracic complications and the associated imaging findings, and discuss the role of radiology in diagnosis and surveillance.

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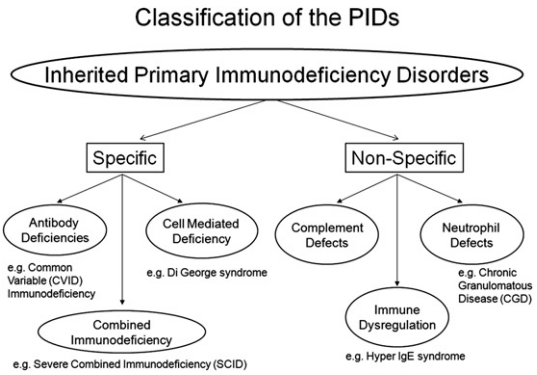

Figure 1 Classification of the PIDs.

Classification and clinical manifestations of the PIDs

PID classification is summarized in Fig 1, and the various clinical manifestations are summarized in Table 1. Common variable immunodeficiency (CVID) is the most commonly encountered PID in our centre (Papworth and Addenbrooke's Hospitals). It is an immunologically heterogeneous group of disorders characterized primarily by a generalized failure of antibody production. T-cell-mediated immunity is often intact; however, T-cell abnormalities have been found in up to 60% of individuals.[4] CVID has an estimated incidence of up to 1 in 10,000 people in the general population.[2,5] The onset of symptoms may occur in early or late childhood or adulthood. Recurrent respiratory tract infections with subsequent bronchiectasis are prominent features.

Thoracic complications and imaging findings in the PIDs

Recurrent respiratory tract infections

Recurrent pneumonia is one of the most frequent clinical manifestations of primary humoral immunodeficiency. The most common respiratory pathogens are encapsulated pyogenic bacteria, such as *Haemophilus influenzae*, *Streptococcus pneumoniae*, and *Staphylococcus*.[6] *Bordetella pertussis* is also an important cause of respiratory illness in children with CVID.[7] Recurrent infection with encapsulated bacteria is the direct result of failure to produce antigen-specific IgG antibodies.

Although there are several potential causes of slowly resolving or recurrent pneumonia, including aspiration and secondary immunocompromise, and structural causes such as bronchiectasis, obstructive lesions, and chronic obstructive pulmonary disease (COPD), radiologists should be aware of and suggest the possibility of primary immunodeficiency, particularly in the younger patient population.

The imaging features of respiratory tract infection in PIDs are similar to those in the immunocompetent patients though often more extensive and the sequelae of previous infections may be evidenced by the presence of scarring or bronchiectasis. Bacterial pneumonia most often manifests as foci of consolidation, which may be lobar or segmental. Cavitation may occur. Infective bronchiolitis may also be seen and manifest on CT as poorly defined centrilobular nodules and tree-in-bud opacities (Fig 2).

Patients with defects involving humoral immunity also have increased susceptibility to respiratory tract viral infections.[8] Rhinoviral infections are frequent and prolonged.[8] Other viral infections may also be encountered. Severe infections caused by varicella,[9] herpes simplex,[10] herpes zoster,[9] and cytomegalovirus[11] have been reported, as have cases of biphasic or fatal measles. [7,12]

Rarely, opportunistic organisms such as *Pneumocystis jirovecii*, *Mycobacterium tuberculosis*, and a variety of fungi (Fig 3) can also be encountered in primary humoral immunodeficiency disorders. This may be caused by clinically important abnormalities of cell-mediated as well as humoral immunity.

Patients with hyper-IgE syndrome are at risk from recurrent infections with *Staphylococcus aureus.* Recurrent pneumonias may result in bronchiectasis (Fig 4) and pneumatocoele formation.[13,14] Pneumatocoeles serve as foci for further infections especially with fungi.[14–16] Certain subgroups of patients also have a predisposition to infection with intracellular organisms.

Table 1
Clinical manifestations of primary immunodeficiency disorders (PIDs).

Humoral (antibody) immunodeficiency disorders, e.g., CVID, XLA	Recurrent pneumonia, otitis media, sinusitis, and septicaemia are the most common clinical manifestations of primary humoral immunodeficiency. In patients with Good's syndrome the presence of a thymoma is associated with hypogammaglobulinaemia.
Cellular and combined immunodeficiency disorders, e.g., SCID	Highly susceptible to opportunistic infections. Often have progressive pneumonia secondary to viral infection or *Pneumocystis jirovecii.* B-cell function is T-cell dependent so there may be accompanying abnormalities in antibody production.
Disorders of phagocytic cells and adhesion molecules	Predisposed to recurrent pyogenic and fungal infections.
Complement deficiencies	Increased incidence of autoimmune disease and pyogenic infections with deficiencies of early components (C1 to C4) of the classical pathway. Increased susceptibility to infections from *Neisseria* species associated with deficiencies of the terminal complement components (C5 to C9). C3 deficiency results in complications such as recurrent pneumonia.
Immune dysregulation, e.g., hyper-IgE syndrome	Highly variable phenotype as dependent upon immunological defect.

CVID, common variable immunodeficiency; XLA, X-linked agammaglobulinaemia; SCID, severe combined immunodeficiency.

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