

Proceedings of the Thoracic Society

The Spring Meeting of The Thoracic Society was held at the Royal College of Physicians, London, on 6-7 March, 1970. There were eight short papers and four symposia. Summaries follow.

PRIMARY IMMUNITY DEFICIENCY SYNDROMES

Experimental Background to Immune Deficiency States

G. L. ASHERSON Immune deficiency states may affect antibody production, delayed type hypersensitivity or both types of reactions. Delayed type hypersensitivity refers to phenomena such as the Mantoux reactions, contact sensitivity, graft and tumour rejection and resistance to certain organisms which can be transferred from one animal to another by immune cells but not by immune serum. In contrast, other immunological skin reactions and resistance to organisms such as the pneumococcus and poliomyelitis virus can be transferred by immune serum. The existence of immune deficiency states which selectively affect antibody production or delayed type hypersensitivity suggests that there may be important differences in the physiology of these two types of reactions.

In the chicken, removal of the thymus early in life causes selective depression of delayed type hypersensitivity. In contrast, removal of the bursa of Fabricius leads to selective depression of antibody production and hypogammaglobulinaemia.

Similarly, in neonatal mammals, thymectomy depresses delayed type hypersensitivity and has less effect on antibody production. In mice there is a considerable reduction of the circulating lymphocytes and a larger reduction of the recirculating pool of long-lived lymphocytes which move from the blood stream through the lymph nodes and pass back to the blood stream through the thoracic duct. Neonatally thymectomized animals may waste (runt) as a result of bacterial infection. The findings in di Georgi's syndrome parallel those of thymectomized mice.

It is not certain which tissue in mammals is equivalent to the bursa of Fabricius in birds. Removal of the lymphoid tissue of the gut in neonatal rabbits causes some reduction of antibody response in the rabbit, and this tissue may be the bursal equivalent.

Primary Immunity Deficiency in Children

L. E. HILL Defects of the two fundamental immunological processes, *i.e.*, humoral and cellular immunity, are found in different combinations and degrees, and show themselves clinically in fairly well-defined disease states, such as the different degrees of antibody deficiency syndrome, di Georgi's disease and the combined immunity deficiency syndrome (Swiss type agammaglobulinaemia). Aetiological classification divides these states into those due to physiological causes (such as hypogammaglobulinaemia

of prematurity) or genetic, *e.g.*, the sex-linked or autosomal recessive immunity deficiency syndromes. Such syndromes as Aldrich's syndrome, ataxia telangiectasia and di Georgi's disease are also genetically controlled, as are those immune deficiency states with other familial immune defects of a different nature. In many cases, however, the aetiology is unknown.

The incidence of immune defects in the British Isles is discussed, as is the mortality of the different subgroups. The typical clinical history of the disease with repeated infections, most commonly of the respiratory tract, is described, and the unusual infecting organisms which may be found are mentioned. The complications of the immune deficiency states theoretically fall into two groups, which are not always easily distinguishable—those due to repeated infections, *e.g.*, bronchiectasis, and those due to some fundamental associated disease process. Of the latter, those particularly affecting children are mentioned, *e.g.*, arthritis, neutropenia and central nervous system disorders.

The question of protective immunizations and the dangers of live vaccines are discussed. Results of replacement therapy with human immunoglobulin are encouraging and, where indicated, thymus or marrow grafts should be considered. The possible disadvantages of replacement therapy are pointed out.

A brief survey of the differential diagnosis is given.

Primary Immunity Deficiency in Adults

K. M. CITRON Primary immunity deficiency in adults may be associated with hypogammaglobulinaemia, dysgammaglobulinaemia or deficiency of cellular immunity.

In 'acquired' hypogammaglobulinaemia, globulin deficiency is acquired in adult life, is occasionally transient and may be genetically determined in some patients. Bacterial infections are severe, recurrent and predominantly broncho-pulmonary, drug-resistant staphylococci being a special danger. Bronchiectasis commonly develops. About half the patients have generalized enlargement of lymph nodes with splenomegaly and some have haematological changes associated with hypersplenism. Histological features include reticuloendothelial hyperplasia and deficiency of plasma cells. Patients may also have malabsorption with steatorrhoea, polyarthritis and amyloid disease. After suffering from primary hypogammaglobulinaemia for many years, some patients develop lymphocytic malignancy.