body alone was associated with a better prognosis. The observation that the titre of antigen may fall during exacerbation of the illness and that clinical improvement may coincide with disappearance of antigen and appearance of antibody is compatible with the hypothesis of an immune complex mechanism for the pathogenesis of polyarteritis nodosa. Such a mechanism is also suggested by analogy with much of the symptomatology, including arthralgia, myalgia, urticaria, and glomerulonephritis, and the characteristic features of human or experimental serum sickness.

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Mechanisms of immune complex induced nephritis

J. G. P. Sissons (*Department of Medicine, Royal Postgraduate Medical School, Hammersmith*) The particular susceptibility of the kidney to involvement by circulating immune complexes (IC) results, at least in part, from its anatomical structure with high intracapillary pressure and blood flow; reduction of renal blood flow reduces IC deposition. However, the recent claim for a specific receptor for C3b in the human glomerulus suggests another potential mechanism of IC localization. Local release of vasoactive amines from platelets (Gelfand *et al*, 1975), brought about by the release of a platelet activating factor from basophils, increases glomerular basement membrane permeability and is one determinant of IC localization in experimental nephritis.

The mediation of allergic glomerular injury has been studied mainly in experimental acute IC nephritis and nephrotoxic serum nephritis. In the acute IC model, glomerular injury can occur independently of C and polymorphonuclear leucocytes (PMN) (Henson and Cochrane, 1971). In the chronic IC model defibrination or massive heparinization prevents crescent formation (Thomson *et al*, 1975) but the role of C and PMN have not been assessed. However, in the nephrotoxic serum model PMN-mediated damage can occur independently of C⁴, possibly because PMN have an Fc receptor.

The size and rate of deposition of IC

appear to determine the histological pattern of nephritis in experimental models, and morphological variations in human nephritis may occur on a similar basis.

It has been postulated that relative immunodeficiency may predispose to the development of spontaneous IC disease (Alpers *et al*, 1972; Peters and Lachmann, 1974). There is no direct evidence for this in human nephritis, except perhaps in rare patients with genetic C deficiencies. In the majority of patients with isolated nephritis of presumed IC aetiology definite antigens and circulating IC can still not be identified, making more logical approaches to therapy difficult.

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Enzyme defects in mucopolysaccharidoses

I. C. BARNES AND C. A. PENNOCK (*Department of Chemical Pathology, Bristol Royal Infirmary, Bristol*) The mucopolysaccharidoses are a group of inherited connective tissue disorders in which excessive cellular accumulation and urinary excretion of glycosaminoglycans (GAG) occur. They are now recognized as disorders of GAG degradation affecting metabolism of heparan sulphate, dermatan sulphate, and keratan sulphate. In the past three years all of the enzymes which are either absent or show diminished activity have been identified. Types I H (Hurler) and I S (Scheie) are deficient in α -L-iduronidase; type II (Hunter) in dermatan sulphate sulphatase; type III (Sanfilippo) subtype a in heparan sulphamidase, and subtype b

in N - acetyl - α - D - glucosaminidase; type IV (Morquio) in N-acetyl-hexosamine - 6 - sulphate sulphatase, type VI (Maroteaux-Lamy) in N - acetylgalactosamine-4-sulphate sulphatase, and type VII in β -glucuronidase.

The importance of these recent discoveries is that definitive diagnosis may be achieved, antenatal diagnosis is now available, and enzyme replacement therapy is possible. However, a number of practical difficulties may be encountered when the enzymes are assayed, especially on cultured amniotic fluid cells or cultured fibroblasts. Most of the enzymes require a natural substrate which may be difficult to purify, or a suitable artificial substrate which may be difficult to synthesize.

Advantages of the 'blind simulated clinical' specimen in quality control in microbiology

W. A. BLACK AND SUE DORSE (*Department of Microbiology, University Hospital, Ontario, Canada*) The use of recognizable 'specimens' either in the form of lyophilized cultures or simulated clinical specimens is suitable for measuring the best performance of a laboratory in bacterial identification and antibiotic sensitivity testing. It may also be the only technique which is feasible when the participating laboratories are spread over a large geographic area. However, the test conditions are abnormal, and this method of control gives little indication to the laboratory director of the real quality of work in his laboratory.

Our regional programme compared data retrieved from 'blind, simulated clinical' specimens and lyophilized specimens using the same organisms in each specimen type. Six surveys were carried out over two years with an average of six simulated 'specimens' per survey, usually two 'urines', two 'faeces', and two 'pus swabs'. These specimens were introduced surreptitiously into the 17-20 participating laboratories. To arrange this properly involved about 1000 miles of travel per survey. Major quantitative and qualitative differences in performance with simulated and lyophilized test specimens were noted. In particular, whereas the putative origin of the specimen did not affect performance with the lyophilized specimens, it played a major role in performance with the simulated specimens, showing for the latter deteriorating performance in the order faeces > pus > urine. Bacterial identification was noticeably better with lyophilized specimens where this depen-