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Advances in Nephrology®

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Advances in Nephrology® From the Necker Hospital

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Preface

The 1994 *Advances in Nephrology* once again attests to the wide spectrum of nephrology. Clinical nephrology is not restricted to primary renal diseases. Nephrologists are exposed to the management of patients with multisystem diseases. This year, emphasis is put on little-known phakomatoses, such as von Hippel-Lindau disease and tuberous sclerosis, which are often "neglected." Correct identification of the renal manifestations is crucial in these diseases. In addition, these diseases (most particularly, von Hippel-Lindau disease) are fascinating examples for those interested in oncogenesis. von Hippel-Lindau disease is a probable example of how the loss of a tumor suppressor gene, or more exactly, two copies of such a gene on the two homologous chromosomes, leads to tumor formation.

Among the diseases with systemic manifestations, sickle cell disease deserves special comment with regard to its global prevalence. Renal involvement has been known for many decades, but the mechanisms and mode of progression of the glomerular lesions have not been elucidated. The role of glomerular hypertrophy and glomerulosclerosis is given special emphasis in the chapter devoted to this topic. The authors also suggest that angiotensin-converting enzyme inhibition might be of long-term value in patients, creating new hope for preventing the progression of glomerular changes.

Nephrologists also should be aware of the emergence of inherited "ion channel diseases," such as cystic fibrosis, involving chloride channels and dyskalemic periodic paralysis. In the latter group, progress in pathophysiology and molecular genetics has been crucial for identifying the defects involving skeletal muscle sodium and chloride channels in hyperkalemic periodic paralysis (Gamstorp's disease) and paramyotonia congenita, and in mytonia congenita, respectively.

This volume contains information on physiology and pathophysiology. Three chapters in this volume focus on the parathyroid hormone receptor and its mode of action, and also on the parathyroid hormone-related peptide. This peptide, whose hyperproduction is responsible for most cases of humoral malignant hypercalcemia, is also involved in fetus physiology and lactation. Another pathophysiological topic deals with the nitric oxide system, which has been recognized as central in cardiovascular control. Pharmacologic inhibition of nitric oxide synthesis provided evidence of the permanent vasodilatory effect exerted by this system in humans. The question is raised as to the presence of endogenous inhibitors of this system, some of which may accumulate in renal failure and trigger hypertension.

Finally, this volume, like the previous ones in *Advances in Nephrology*, comprises four contributions on dialysis and transplantation: commencement of dialysis, cardiovascular mortality and morbidity in dialysis patients

(cardiovascular complications are still the leading cause of death in these patients, and good data on epidemiology and prevention are scarce), prevention of cytomegalovirus infection in the transplant patient, and molecular basis of graft rejection and tolerance.

Jean-Pierre Grünfeld, M.D.

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