

NEPHROTIC PROTEINURIA IN ADPKD - A CLINICAL CHALLENGE. Prabodh Ranjan, H Magoo, EH Anjum, C Desai, D Baumstein, A Chaudhari, AM Tannenber, Metropolitan Hospital Center, NewYork Medical College,NY.

In autosomal polycystic kidney disease (ADPKD) the protein excretion is generally less than 1g/24h and the association with the nephrotic range proteinuria is considered rare. We describe a patient of ADPKD with nephrotic range proteinuria, who exhibited rapid progression to ESRD.

A 52 years old nonsmoker male with ADPKD and controlled hypertension developed nephrotic range proteinuria with rapid progression to ESRD. The patient was first found to have nephrotic range proteinuria in October 2002, with BUN of 35mg/dl and serum creatinine (SCr) of 2.5mg/dl. His 24hour urine protein and creatinine were 3.4gms and 1.8gms respectively. The serological workup including ANA, Hep C antibody, HBsAg and HBsAb were negative with normal complements. His renal ultrasound revealed multiple cysts in both the kidneys with right and left renal size of 24.9 x10.9cms and 21.5 x 10.5 cms respectively. Noticeably, five of his brothers have ADPKD and two already had renal transplant. The patient was started on maximum dose of ACE-inhibitor. Despite the control of hypertension with ACE-inhibitor and use of statins, the renal function (BUN/SCr - 66/10 mg/dl) deteriorated rapidly over five years to present need for renal replacement therapy.

Nephrotic range proteinuria is rare in ADPKD and its presence is considered mostly coincidental. Almost every form of glomerulopathy has been reported in ADPKD with the higher frequency of FSGS. In ADPKD, the renal function does not decrease at a constant rate between birth and ESRD but remain well preserved for many years; however, nephrotic range proteinuria accelerates its course, as it was observed in our case. Higher complications and technical difficulty precludes renal biopsy in ADPKD. Under these circumstances, Can a trial of steroid be helpful in delaying the progression to ESRD? The question is yet to be answered.