Dear Sir,

Autosomal dominant polycystic kidney disease (ADPKD) is considered to be extremely rarely combined with glomerulopathies [1]. We report a case of ADPKD with chronic glomerulonephritis proved by biopsy.

In 1990 a 45-year-old man was admitted to a clinic of nephrology on account of generalized oedema all over the body, the face and the lower extremities. Physical examination was remarkable with pleural effusion (confirmed by radiography) and ascites. Enlarged kidneys were not palpated. The ultrasonography showed renal polycystosis and lack of cysts in the liver. Laboratory tests produced the following results: urinary protein excretion of 8.5-12.7 g/24 h, urinary sediment: 15-20 leucocytes, isolated erythrocytes; significance of bacteriuria, urea 14.3-18.1 mmol/l, creatinine 174^464 µmol/l; clearance of creatinine 21 ml/min; total protein 46 g/l; albumin 22%. A liver disease, diabetes mellitus, lupus and amyloidosis were excluded. In order to explain the origin of the nephrotic syndrome an open renal biopsy had been undertaken. The polycystic changes were macroscopically confirmed. Histological investigation showed the tubular changes typical for polycystosis, and chronic glomerulonephritis (membranous)-thickened glomerular basement membranes, absence of cell proliferation, subepithelial staining with IgG, IgM, IgA and C3, intense deposition of fibrin in the glomerules and in the interstitium. Therapy with prednisone, heparin, antibiotics, etc., had been administered. The oedema decreased distinctly, proteinuria fell to 2-3 g/l. The following 1 year’s outpatient observation showed no progression of the renal failure. The patient died suddenly at home in 1991.

ADPKD is diagnosed most often in the 4th to 5th decade of human life [2]. Family history remains unknown in about 25% of patients [2,3]. Our case belongs to this group, as the relatives and risk refused a screening investigation. Proteinuria in ADPKD is usually of a low degree [1-3]. During the observation of 152 patients from 51 families we found neither another case of proteinuria above 3 g/l nor a nephrotic syndrome.

Renal biopsy frequently clarifies the etiology of a nephrotic syndrome. It is generally accepted that renal biopsy is contraindicated in the case of ADPKD in view of the low reliability and danger of complications. Its usage for an early diagnosis of the cystic changes is justified only for scientific purposes [4]. This is why we did an open renal biopsy (after...
excluding diabetes mellitus, lupus and amyloidosis), at which under visual control enough material for the histomorphologic study was taken. Although the method is invasive, we assume that in our case it is justified.

Looking through more than 300 literary sources from the last 20 years we found only two similar cases. Ackerman [5] described 2 patients with ADPKD and nephrotic syndrome and without histological examination assumed that the nephrotic syndrome was caused by the main disease. However, the author pointed out that the microangiopathic hemolytic anemia showed the existence of another nephropathy. Licina et al. [6] reported a case of histologically proved crescentic nephritis without nephrotic syndrome in a patient with ADPKD. Attention is paid [5, 6] to the possibility of using a morphologic evaluation of erythrocytes in urinary sediment for the eventual presence of a glomerulopathy in such cases.

Renal function decreases very slowly in ADPKD, and it could be forecast [7]. Its rapid worsening, except for some other complication, might also be due to glomerulopathy [6]. In contrast to Licina et al. [6] we do not accept that glomerulonephritis is a complication of ADPKD, as ADPKD does not create conditions for the beginning of an immune nephropathy. Casuistry of similar statements also supports the idea of combining the two diseases in one and the same organ.

We accept that specifying the histomorphologic type of the chronic glomerulonephritis in ADPKD is necessary for a corresponding pathogenetic treatment.

In conclusion, we assume that ADPKD cannot be added to the list of the diseases that occur together with a nephrotic syndrome. Its presence in patients with ADPKD makes it necessary to look for another disease.

References