CARdiovascular complications in patients with adult polycystic kidney disease

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Summary

Adult polycystic kidney disease is often associated with developmental abnormalities other than in the kidney. Our study demonstrates an increased incidence of cardiovascular complications. This data and reports of other authors suggest a general defect of basement membranes. Patients with polycystic kidney disease should be regularly examined to detect cardiovascular complications, in order to initiate therapy appropriately.

Introduction

According to statistical data from the EDTA Registry for 1982 10 per cent of all patients undergoing regular dialysis treatment suffer from polycystic kidney disease [1].

It is well established, that the adult form of polycystic kidney disease is associated with developmental and acquired abnormalities. Cysts may be found not only in the kidneys, but also in liver, pancreas and in rare cases in the spleen and in the thyroid gland [2]. Clinically significant lesions associated with this disease are aneurysms of cerebral and visceral arteries [3,4]. Scheff et al reported an increased incidence of diverticulosis coli in patients with polycystic kidney disease [5] and recently several authors have described cardiovascular abnormalities [6,7].

This study investigates cardiovascular complications in patients with polycystic kidney disease undergoing regular dialysis treatment. In addition the clinical relevance of these abnormalities was evaluated.

Methods

Patients Thirty-eight patients (27 males, 11 females) undergoing regular dialysis treatment, gave informed consent to participate in this study. The mean age was
52.9±7.2 years, they had undergone regular dialysis treatment for 48.2±14.6 months.

Clinical investigations These patients underwent non-invasive and invasive examinations such as: ECG, chest X-ray, echocardiography, abdominal sonography, cardiac catheterization and angiography of the carotid arteries.

Results

Cardiovascular complications were present in 24 patients (63%). Left ventricular or septal hypertrophy occurred in 11 cases. In five patients hypertrophy was not associated with elevated blood pressure. Three patients showed atrial dilatation (Figure 1). Three patients suffered from conduction disturbances and required pacemaker therapy (Figure 2). Cardiac catheterization demonstrated an atrial septal defect in one and a ventricular septal defect in another patient. Angiography of the coronary arteries displayed multiple aneurysms in one case (Figure 3). Three patients had a cardiomyopathy (Figure 4). Sonographically in one patient an aortic aneurysm could be detected. Carotid angiography showed stenosis in one case.

During the clinical course presenting symptoms were heart failure, bradycardia, and angina pectoris. Eight patients without clinical symptoms demonstrated cardiovascular abnormalities after intensive cardiovascular examination.

Figure 1. Echocardiography of a patient demonstrating dilatation of the left atrium
Figure 2. ECG of a patient with polycystic kidney disease demonstrating SA-block II°

Figure 3. Cardiac catheterization of patient demonstrating multiple aneurysms of the coronary arteries

Discussion
Our data indicate an increased incidence of cardiovascular abnormalities in patients suffering from polycystic kidney disease. Of note is the fact that in five
patients left ventricular hypertrophy was not associated with elevated blood pressure, suggesting a developmental abnormality of the myocardium. In our study group three patients required cardiac pacemaker therapy without major electrolyte disturbances. In contrast to patients undergoing regular dialysis treatment, patients with polycystic kidney disease showed a higher incidence of atrial — and ventricular — septal defects and aneurysms of the coronary arteries. In addition other authors have reported cardiac valve abnormalities [6].

Histological analysis of available valve tissue demonstrated myxomatous degeneration with disruption of the collagen texture. Years ago malformations other than cardiovascular were reported by several authors. Cameron described a syndrome of bilateral teventodactyly of the feet in combination with polycystic kidney disease [8] and Tucker reported the peculiar oral-facial-digital syndrome [9].

Therefore we suggest, that in patients with polycystic kidney disease, the developmental disturbance is not only limited to the kidneys, but a variety of other organs. Because of these multi-organ abnormalities we suggest there is a general defect of the basement-membrane.

References

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