

[ CASE REPORT ]

## Autosomal Dominant Polycystic Kidney Disease in which the Polycystic Liver Volume Was Reduced by Rigorous Blood Pressure Control

Tatsuya Suwabe<sup>1,2</sup>, Yoshifumi Ubara<sup>1,2</sup>, Daisuke Ikuma<sup>1</sup>, Hiroki Mizuno<sup>1</sup>, Noriko Hayami<sup>1</sup>, Masayuki Yamanouchi<sup>1</sup> and Naoki Sawa<sup>1,2</sup>

### Abstract:

Polycystic liver disease (PLD) is the most common extrarenal manifestation of autosomal dominant polycystic kidney disease (ADPKD). However, current treatments for PLD are only supportive. We experienced a case of enlarged kidneys and liver in a 53-year-old Japanese man with ADPKD who was on hemodialysis. He underwent renal transcatheter arterial embolization (TAE) for enlarged kidneys. His blood pressure (BP) decreased after renal TAE, and his liver volume decreased from 5,259 mL to 4,647 mL (11.6% reduction) within 1 year after renal TAE. This case suggests that rigorous blood pressure control may be beneficial for ameliorating enlarged PLD.

**Key words:** ADPKD, polycystic kidney disease, polycystic liver disease, PLD, blood pressure

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### Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is a common inherited renal disorder (1, 2), and polycystic liver disease (PLD) is the most common extrarenal manifestation of ADPKD (3, 4). PLD can also arise in the absence of polycystic kidneys or in the presence of few renal cysts (5). Enlarged PLD causes abdominal fullness and severely impairs quality of life of patients (6). Enlarged liver are also related to severe cyst infection and massive enlarged liver was reported to be the strongest factor of death from hepatic cyst infection (7). Multiple studies have focused on pharmacologic approaches to slow the development of the liver cystic disease; however, the current treatment for PLD is very limited.

Recently, it was reported that rigorous blood pressure (BP) control is beneficial for slowing the progression of polycystic kidney disease, and such BP control has thus been recommended for early ADPKD (8). However, whether or not rigorous BP control is beneficial for slowing the pro-

gression of PLD is unclear.

We herein report a patient with ADPKD on hemodialysis whose polycystic liver volume decreased by 11.6% within 1 year after renal transcatheter arterial embolization (TAE). We consider his markedly decreased BP to have contributed to the reduction in his liver volume. This is the first case in which the liver volume showed a marked reduction following a decrease in the BP in a patient with ADPKD.

### Case Report

We encountered a 53-year-old Japanese man with ADPKD who had been on dialysis for 9 years with no other specific medical history. He was completely anuric. He had been hospitalized in our hospital one time due to a diagnosis of renal cyst infection the previous year. He had abdominal fullness and was admitted to our hospital to receive renal TAE in 2018.

### Physical and imaging examinations

His height was 179 cm, and his body weight was 68.8 kg.

<sup>1</sup>Department of Nephrology, Toranomon Hospital Kajigaya, Japan and <sup>2</sup>Okinaka Memorial Institute for Medical Research, Toranomon Hospital, Japan

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Correspondence to Dr. Tatsuya Suwabe, suwabetat@gmail.com



**Figure 1.** CT just before TAE reveals the marked enlargement of the bilateral kidneys and liver with numerous cysts.

His average BP in the morning over 2 non-dialysis days was 137/86 mmHg. He had no edema in his legs. Abdominal computed tomography (CT) was performed as reported previously (9). His bilateral polycystic kidneys were enlarged, and polycystic cysts were also seen diffusely throughout the enlarged liver (Fig. 1).

#### **Treatment at the hospital**

Renal TAE was performed. In total, 20 microcoils were inserted into his renal arteries as peripherally as possible (Fig. 2). He was discharged eight days after renal TAE.

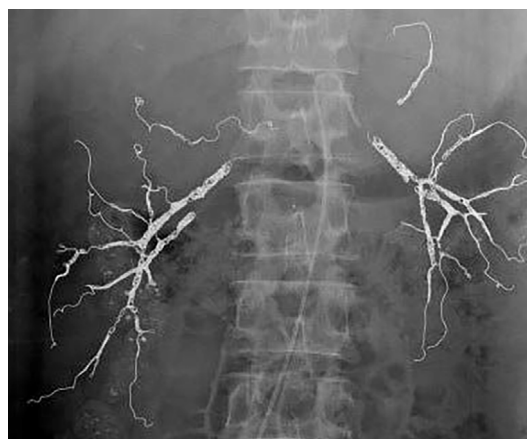
#### **Clinical course before and after renal TAE**

His renal volume and liver volume had increased gradually over the year before renal TAE (Fig. 3). However, his renal volume decreased dramatically after renal TAE, from 3,772 mL to 1,840 mL (51.2% reduction) (Fig. 3, 4). His BP also decreased from 137/86 mmHg to 107/71 mmHg after renal TAE. Surprisingly, his liver volume decreased gradually from 5,259 mL to 4,647 mL (11.6% reduction) within the year after renal TAE. His cyst volume in the liver also decreased gradually from 2,471 mL to 1,858 mL (24.8% reduction) within 1 year after renal TAE. He was administered antihypertensive medicine (Amlodipine besilate) before renal TAE and discontinued this agent seven months after the procedure.

His BP was measured with an automatic device in the sitting position, and the average of two BP readings (measured in the morning on a non-dialysis day) was used for the analysis. The kidney and liver volumes and liver cyst volume were determined using the Vincente software program, ver. 4 (Fujifilm, Tokyo, Japan). Liver cysts were defined as lesions with a density <25 HU on CT (9).

#### **Mechanism by which renal TAE reduces the renal volume**

After renal TAE, the kidney volume decreases. Histologically, some necrosis, fibrosis, or regeneration of the renal cyst may occur, as in acute renal infarction, because the renal arteries are occluded as completely as possible (10).



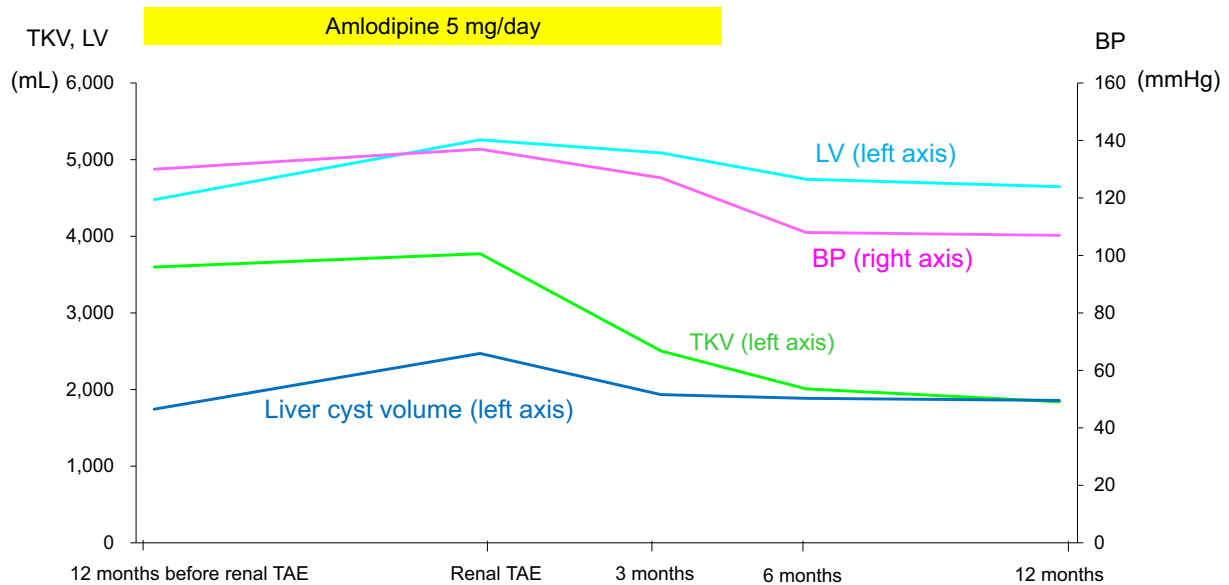
**Figure 2.** Abdominal X-ray obtained just after TAE shows that the renal arteries are occupied by microcoils from the peripheral branches to the main branches.

However, there is actually a small amount of remaining renal blood flow or re-canalization of the bilateral renal arterial branches in some cases after renal TAE (11). In other words, renal TAE may not shut down the renal artery completely but rather simply reduce the renal blood flow. However, the kidney volume always decreases after renal TAE (12), possibly for the same reason kidney shrinkage occurs following atherosclerotic renal artery stenosis. Generally speaking, progressive renal shrinkage occurs in kidneys supplied by significant renal artery stenosis (13). Thus, the kidney volume decreases as the renal blood flow decreases, even though the renal blood flow is not completely shut down. For this reason, the kidney volume may decrease after renal TAE.

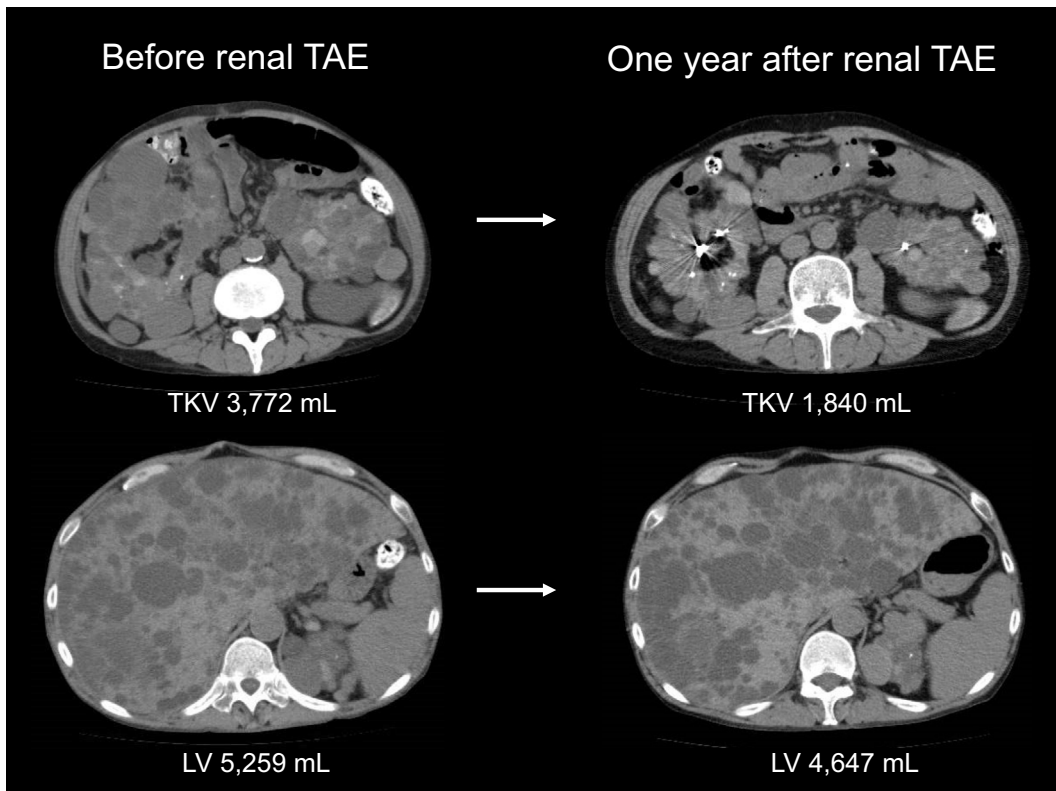
### **Discussion**

We reported a case of ADPKD in which the liver volume as well as the renal volume decreased after renal TAE. The patient's BP after renal TAE was much lower than it had been before renal TAE. A decrease in the BP is often seen after renal TAE (12). However, we reported that the least squares mean (95% confidence interval) of the systolic BP (sBP) decreased from 135 (132-137) mmHg to 123 (120-126) mmHg. The sBP at one year after renal TAE in this case was much lower than the average sBP in our patients who underwent renal TAE. This marked reduction in the BP might have influenced the liver volume decrease after renal TAE.

The renin-angiotensin-aldosterone system (RAAS) reportedly plays a central role in renal cyst growth, which is considered one of the reasons why strict BP control with RAAS blockers is effective for slowing cyst growth (8). However, the role of RAAS in hepatic cyst growth has been unclear. Of note, the patient had not been administered any RAAS inhibitors after renal TAE. Therefore, we attribute the reduction in hepatic cysts solely to the reduction in the BP itself. The hepatic blood flow is estimated to account for about



**Figure 3.** The clinical course (blood pressure, total kidney volume, and liver volume) during the year before and after renal TAE.



**Figure 4.** CT before and 1 year after renal TAE shows a reduction in the total kidney volume (TKV) from 3,772 to 1,840 mL. CT before and 1 year after renal TAE also shows a reduction in the liver volume (LV) from 5,259 to 4,647 mL.

25% of the cardiac output (14), which is known to correlate with the mean arterial pressure. Therefore, the blood flow can be reduced by lowering the BP. This reduction in the hepatic blood flow may be associated with the reduction in hepatic cyst volume. As mentioned above, the renal blood flow is shut down or reduced after renal TAE, and the kidney volume decreases. For the same reason, the hepatic vol-

ume decreases after hepatic TAE, as the hepatic blood flow is shut down or reduced (15). This reduction in the hepatic volume after hepatic TAE via a reduction in the hepatic blood flow supports our hypothesis that the hepatic volume decreases via a reduction in the hepatic blood flow following a reduction in the blood pressure.

In the present patient, the volume of hepatic cysts de-

creased more markedly than that of the liver parenchyma. This suggests that the hepatic cyst volume is more markedly affected by the blood pressure than that of the liver parenchyma.

Another possible mechanism underlying the hepatic volume reduction in this patient might be the improvement of appetite loss and the nutrition state after renal TAE, leading to the improvement of hypo-albuminuria and interstitial tissue edema, which may reduce the liver volume. Indeed, the ascites disappeared at six months after renal TAE in the present patient.

While approaches to slowing the progression of hepatic cyst growth are very limited at present, strict BP control may be a promising treatment for PLD as well as PKD. The ideal approximate BP for patients with PLD is unclear. Further studies are therefore needed to clarify the role and effectiveness of rigorous BP control in managing the progression of PLD.

In conclusion, we experienced a case of ADPKD in which the liver volume decreased by 11.6% within 1 year after renal TAE. This marked reduction in BP after renal TAE might have influenced the liver volume reduction.

Informed and voluntary consent for publication was obtained from the patient described in the article.

**The authors state that they have no Conflict of Interest (COI).**

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