NEUROENDOVASCULAR COILING OF BERRY ANEURYSMS IN A PATIENT WITH AUTOSOMAL - DOMINANT POLYCYSTIC KIDNEY DISEASE

Mark Joseph P. Sibal[1], Francis Angelo D. Basilio[2*], Marita B. Dantes[3], Rudolf V. Kuhn[1,4*]

[1]Section of Vascular and Interventional Radiology, Department of Medical Imaging and Therapeutic Radiology, National Kidney and Transplant Institute, Philippines
[2]Department of Radiology, University of the Philippines - Philippine General Hospital, Philippines
[3]Section of Neurology, Division of Internal Medicine, National Kidney and Transplant Institute, Philippines
[4]Faculty of Medicine, Siriraj Hospital, Mahidol University, Thailand

***Corresponding author:**
Rudolf V. Kuhn, Section of Vascular and Interventional Radiology, Department of Medical Imaging and Therapeutic Radiology, National Kidney and Transplant Institute, Philippines
Email: rudolfvkuhn@yahoo.com

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ABSTRACT

Introduction: Autosomal Dominant Polycystic Kidney Disease (ADPKD) is a prevalent cause of end-stage renal disease, significantly complicated by intracranial aneurysms. With a rupture mortality rate of 10–20%, prompt identification and management are critical.

Case Report: A 50-year-old female with ADPKD-related renal failure presented with recurrent headaches. Imaging identified a distal right middle cerebral artery (MCA) aneurysm. Although initially lost to follow-up, she eventually underwent urgent endovascular coil embolization for multifocal lesions, including the MCA and a distal anterior cerebral artery aneurysm. The procedure was successful, and 1-year follow-up imaging confirmed complete obliteration with symptom resolution.

Discussion: ADPKD-associated aneurysms often rupture at younger ages and smaller sizes than sporadic cases. While current guidelines suggest targeted screening based on family history, many ruptures occur without such history. Non-contrast MR Angiography (MRA-TOF) offers a safe, non-invasive screening tool to detect these lesions early.

Conclusion: Intracranial aneurysms pose a severe threat in ADPKD. This case demonstrates endovascular coiling as a safe, effective strategy for multifocal disease. Given the aggressive natural history, lowering the intervention threshold and considering baseline MRA screening at age 30 for high-risk individuals may optimize outcomes.

Keywords: Autosomal Dominant Polycystic Kidney Disease, Intracranial Aneurysm, Endovascular Coil Embolization, Magnetic Resonance Angiography

INTRODUCTION:

Autosomal Dominant Polycystic Kidney Disease (ADPKD) is the most prevalent inherited cause of end-stage renal disease (ESRD), with an estimated incidence of 4–10% [1]. This condition is pathologically characterized by the development of numerous, variably-sized cysts that progressively replace the renal parenchyma. By the age of 30, over half of ADPKD patients are reported to have visible renal cysts on imaging [1].

ADPKD is associated with several significant extra-renal manifestations. The most critical of these is the development of intracranial aneurysms. The reported prevalence of intracranial aneurysms in the ADPKD population is estimated to range from 9–12% [2,3]. Rupture of an intracranial aneurysms is the most devastating complication, leading to a mortality rate of 10–20% and a morbidity rate of up to 50%, often resulting in a severe decline in the patient's quality of life [4].

Given the high risk of catastrophic rupture, prompt intervention is indicated for high-risk individuals, especially those presenting with signs of aneurysm growth or neurological symptoms suggesting impending rupture.

CASE REPORT:

A 50-year-old female presented with a history of end-stage renal disease secondary to ADPKD, diagnosed in 2006 (Figure 1). She maintained a good functional status and independence in activities of daily living while undergoing thrice-weekly hemodialysis. In 2017, she presented with recurrent headaches, which prompted a neurological work-up. A CT angiogram at that time revealed a narrow-necked, saccular aneurysm, measuring 2.8×3.3 mm (height × width), located in the distal right middle cerebral artery (MCA) (Figure 2A). Surgical clipping was advised, but the patient was subsequently lost to follow-up.

Five years later after detection of the aneurysms, the patient sought medical clearance for a parathyroidectomy due to work-up for musculoskeletal pain. Review of systems noted the persistence of headaches, which were partially alleviated by analgesics. Due to the known intracranial aneurysms, a repeat cranial CT angiogram was ordered. This study demonstrated an increase in the size of the original right MCA aneurysm (3.3×4.0 mm, H × W, Figure 2B) and the development of a new narrow-necked, saccular aneurysm, measuring 5.0×6.0 mm (H × W) in the right distal anterior cerebral artery (ACA) (Figure 3), at the bifurcation of its pericallosal and callosomarginal branches. These findings necessitated further evaluation with digital subtraction angiography (DSA) for precise lesion characterization.

Following multidisciplinary consensus, urgent endovascular coil embolization was favoured over open surgical clipping as the management strategy due to the multifocality of the lesions. Simple coil embolization was performed successfully, using two detachable coils in the right MCA aneurysm and six detachable coils in the right distal ACA aneurysm (Figure 4). The procedure was well-tolerated without post-operative complications, and the patient was discharged on the fourth post-coiling day. A follow-up CT angiogram one year later showed complete obliteration with no signs of aneurysm recurrence. The patient reported resolution of headaches and subsequently underwent successful parathyroidectomy.

DISCUSSION:

The risk for intracranial aneurysm development must be considered in all patients diagnosed with ADPKD. Currently, a familial history of ruptured or unruptured intracranial aneurysms remains the only consistently identified risk factor for developing these lesions in ADPKD patients [1]. Emerging evidence suggests that the natural history of aneurysms in ADPKD differs significantly from those in the general population [4].

Intracranial aneurysms associated with

ADPKD are known to rupture at an earlier median age (41 years vs. 51 years) [4]. Furthermore, small aneurysms (measuring 5–9 mm) in ADPKD have a rupture rate of 20–30%, in contrast to the general population, where aneurysms <7 mm rarely rupture. The majority of ruptured cases in ADPKD involve the anterior cerebral circulation [4]. This is complicated by the difficulty of management of aneurysms in distal anterior cerebral artery, whether by clipping or coiling, which also often have higher complication rates. Screening guidelines for intracranial aneurysms in ADPKD remain a subject of debate. The 2025 Kidney Disease: Improving Global Outcomes (KDIGO) clinical practice guidelines recommended targeted screening only for patients with a family history of intracranial aneurysms or hemorrhagic stroke, as well as those with personal history of subarachnoid hemorrhage [5]. However, the efficacy of this strategy is questionable, as approximately 80% of reported ruptured intracranial aneurysms cases in the literature occur in patients without a known family history [5].

With advancements in Magnetic Resonance (MR) technology, non-contrast MR Angiography using Time-of-Flight (MRA-TOF) technique offers a safer diagnostic alternative by avoiding the use of iodine-based contrast agents [6]. Recent evidence suggests that the sensitivity and specificity of MRA-TOF for detecting intracranial aneurysms as small as 2–3 mm are comparable to contrast-enhanced CT angiography [6].

Considering intracranial aneurysm development occurs during adulthood, several authors recommend baseline MRA screening at approximately 30 years of age, with interval follow-up every 5 to 10 years until the age of 60, to maintain screening efficiency [6, 7]. Beyond screening, patient counseling on lifestyle modifications, including strict blood pressure control and smoking cessation, is paramount to mitigate the risk of aneurysm growth and rupture.

Given the inherently higher rupture risk in ADPKD patients, the threshold for intervention should be lowered. Aneurysm growth demonstrated on serial imaging along with neurologic symptoms, as observed in our patient, represent a clear and compelling indication for intervention.

The management of detected intracranial aneurysms in ADPKD requires a multidisciplinary approach. For unruptured lesions, endovascular techniques, such as coil embolization, provide a minimally invasive solution. In cases for aneurysms with challenging morphology or location, newer techniques involving endovascular stents and balloons or intrasaccular devices are also viable options.

CONCLUSION:

Intracranial aneurysms pose a critical threat to patients with ADPKD. While targeted screening remains the KDIGO guideline, a familial history of aneurysm or previous hemorrhagic stroke are still considered major risk factors. Close monitoring for neurological symptoms and targeted screening for high-risk individuals are essential for prompt diagnosis and timely intervention. Endovascular coil embolization offers a safe and minimally-invasive management approach, demonstrated effectively in this case for multifocal lesions. To optimize screening efficiency, a recommended baseline MRA screening starting at 30 years of age is prudent for high-risk individuals.

DATA AVAILABILITY:

Further information regarding this work can be obtained from the corresponding author upon reasonable request.

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CONFLICT OF INTEREST:

The authors have no conflicts of interest to declare and is in agreement with the contents of the manuscript.

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FIGURE LEGEND:

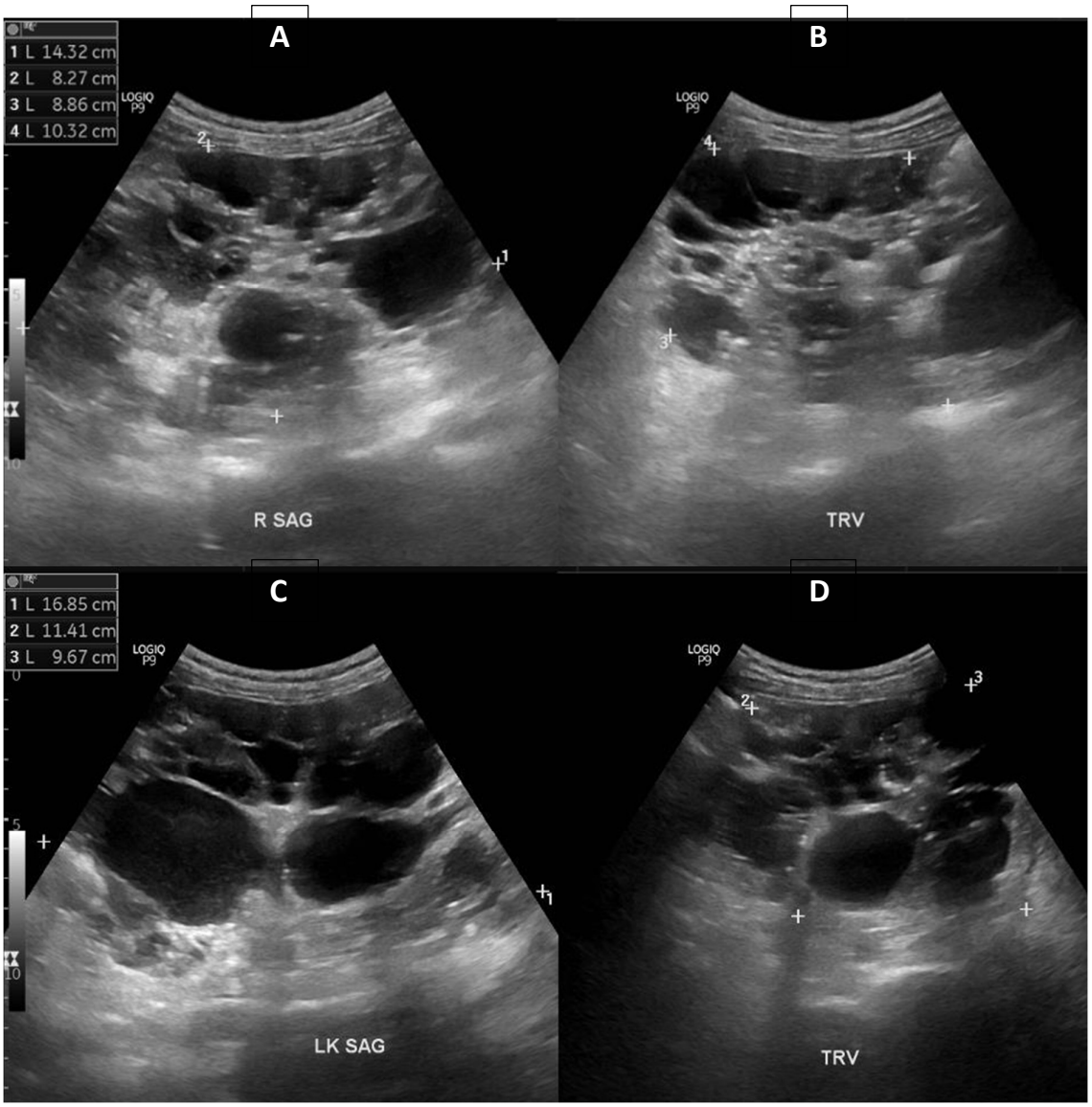

Figure 1: Ultrasound of the kidneys in sagittal (A,C) and transverse (B,D) planes showing multiple varisized cysts replacing the entire renal parenchyma, consistent with autosomal-dominant polycystic kidney disease.

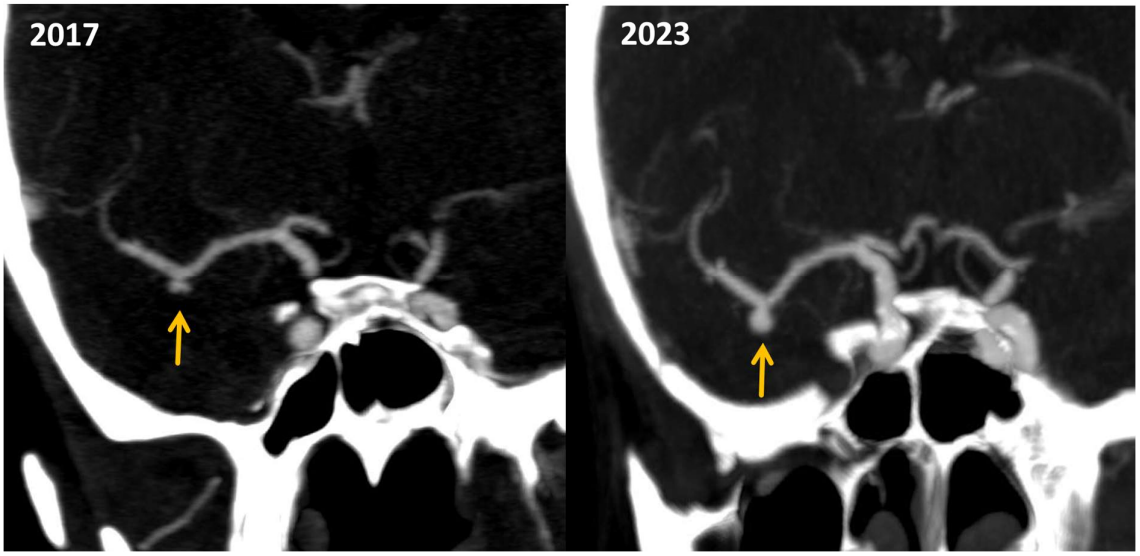

Figure 2: Comparative cerebral CT angiograms in coronal plane (5 years apart) demonstrating the interval increase in size of the right MCA aneurysm (yellow arrow): previously measuring 2.8 x 3.3 mm (HxW), now measuring 3.3 x 4.4mm (HxW).

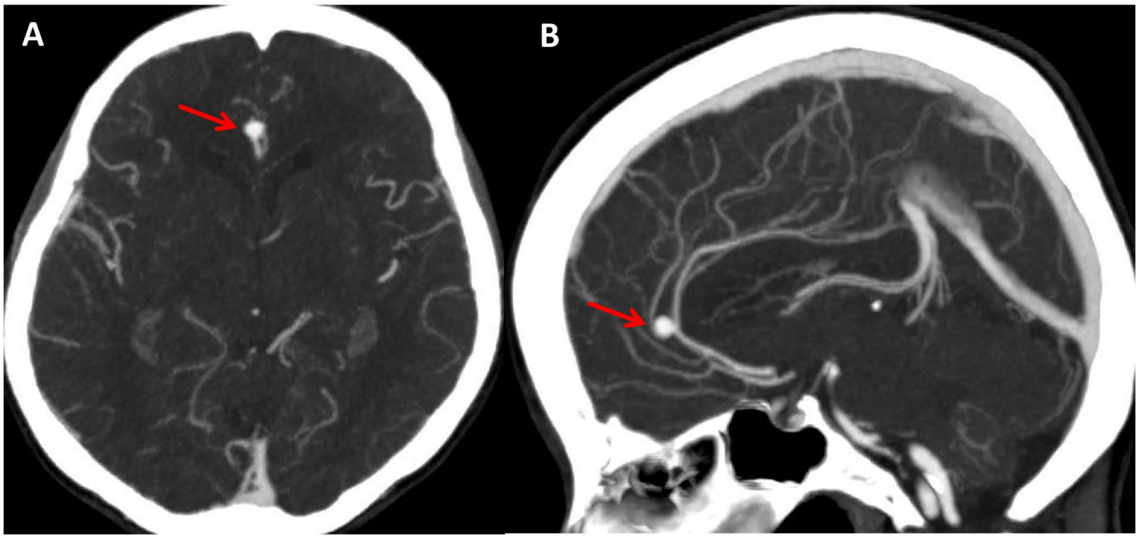

Figure 3: Cerebral CT angiogram in axial (A) and sagittal (B) planes showing the new saccular aneurysm at the right distal ACA (red arrow).

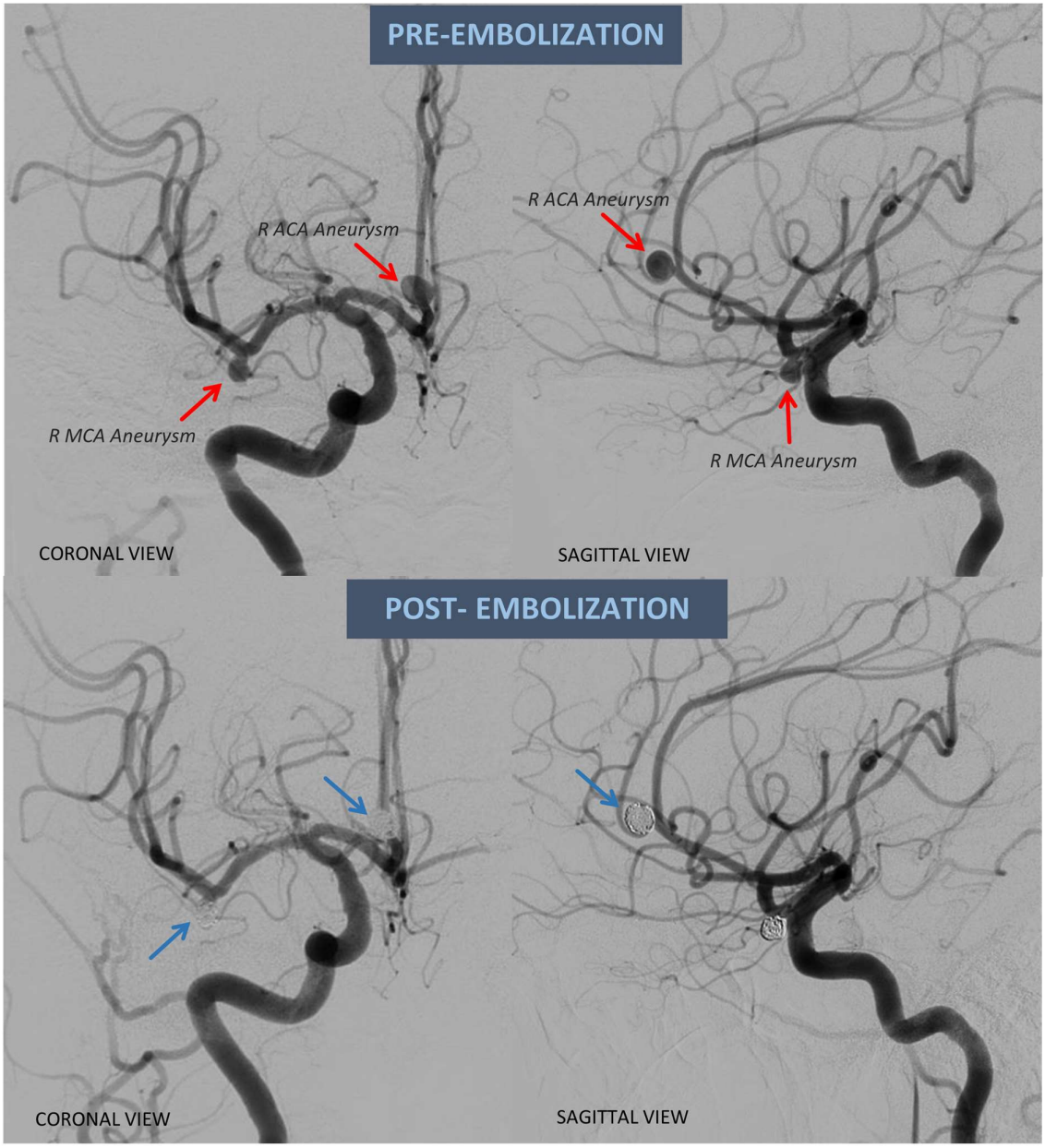

Figure 4: DSA of the right internal carotid artery in anteroposterior and lateral views. Panel above shows representative images highlighting the narrow-necked, saccular aneurysms in the right MCA and R ACA (red arrows). Panel below shows representative images after deployment of detachable coils obliterating the flow into the aneurysms. No contrast extravasation, dissection or thrombus formation.