Key Takeaways for Clinicians from the KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of ADPKD: Intracranial aneurysms and other extrarenal complications

1 Increased risk awareness

Intracranial aneurysms (ICAs) constitute the most life-threatening association of ADPKD. The prevalence of ICAs and the incidence of ICA ruptures causing subarachnoid hemorrhage (SAH) are approximately 4 and 7 times higher in ADPKD than in the general population (Figure 1). In both populations, the prevalence and incidence are higher when there is a family history of ICA and especially SAH. Adults with ADPKD should be informed of their elevated risk for ICAs and SAH, with education on recognizing symptoms like thunderclap headaches that require immediate medical attention (Figure 2).

2 Comprehensive risk assessment

A detailed personal history of SAH and a family history of ICA, SAH, or unexplained sudden death are crucial in identifying people with ADPKD who are at higher risk for ICA, guiding screening decisions.

3 Screening recommendations

Screening for ICAs is recommended in people with ADPKD and a personal history of SAH, or a positive family history of ICA or SAH, or unexplained sudden death, especially in those eligible for treatment and with a reasonable life expectancy. Consider ICA screening in specific clinical contexts, such as during evaluation for kidney and/or liver transplantation or prior to major elective surgery in people with ADPKD.

4 Advantages and limitations of presymptomatic screening

Presymptomatic screening may allow for intervention if an ICA at risk of rupture is identified, which may prevent death or significant comorbidity. It may also allow adequate imaging follow-up if an ICA with low risk of rupture is identified, and it may reduce anxiety and provide reassurance when no ICA detected. However, screening may also lead to the identification of ICA with very low risk of rupture that do not require intervention but will require long term follow-up. Screening does not exclude the risk of *de novo* ICA development and rupture after screening and may also lead to procedures with possible treatment failure or complications, including death or significant morbidity, and may cause anxiety when an ICA is identified. It may also limit access to life insurance, loans, driver's licenses, or potential work opportunities.

5 Shared decision-making

Shared decision-making is essential in ICA screening. People with ADPKD not considered at increased risk who wish to be screened should be provided access after being adequately informed about the benefits and risks.

6 Imaging for ICA detection

When screening is pursued, time-of-flight (TOF) magnetic resonance angiography (MRA) without gadolinium enhancement is recommended. High-resolution computed tomography angiography (CTA), which requires contrast administration, may be used as an alternative. In high-risk people with ADPKD and negative initial ICA screening results, the timing of rescreening should be individualized, with intervals of 5–10 years, depending on age, risk factors, and life expectancy.

7 Lowering the risk of ICA development and rupture

Modifiable factors that increase the risk of ICA development and rupture include smoking, uncontrolled hypertension, and alcohol in large quantities. In the last decade, the risk of SAH in the general population has decreased worldwide, likely due to less smoking and better blood pressure control. Therefore, smoking cessation and blood pressure control are critical.

8 Other vascular and extrarenal complications

ADPKD is a systemic disease and involves almost every organ and tissue. Physicians taking care of people with ADPKD should be aware of these associations and address them appropriately when they present. Presymptomatic screening for these extrarenal associations is not indicated except in families with non-ischemic idiopathic cardiomyopathies, thoracic aortic aneurysms, or coronary artery dissections.

Figure 1

	General population	General population with family history of ICA or SAH	ADPKD population	ADPKD population with family history of ICA or SAH
Prevalence of ICA (95% CI)	2.9% (1.9–4.5)	3.4 (1.9–5.9) higher risk	12.9% (10.4–15.4)	17.1% 13.4–21.1)
Incidence rates of SAH (per 1000 person-years, 95% CI)	0.079 (0.069–0.09)	3–7 higher risk	0.57 (0.19–1.14)	Likely higher (based on data from general population)

Figure 2

Thunderclap headache

Definition:
- Strikes suddenly
- Intense pain: "worst headache in my life"
- Reaches maximal intensity within 60 seconds

May be associated with or followed by:
- Nausea or vomiting
- Seizures
- Altered mental state/loss of consciousness

What to do:
- Seek immediate medical attention
- Have evaluation in an emergency department equipped with CT scan
- Inform caregivers about the increased risk for subarachnoid hemorrhage associated with ADPKD

Figure 3

Organ	Association	Screening
Heart	Valvular heart disease, non-ischemic cardiomyopathies, congenital heart disease, atrial fibrillation, pericardial effusion	Non-ischemic cardiomyopathies (when there is a family history)
Arteries	Aortic root dilatation, thoracic aortic aneurysms (TAA), coronary artery dissections (CAD) and aneurysms, other aneurysms	TAA or CAD (when there is a family history)
CNS	Arachnoid cyst, dural diverticula	No
Bowel	Colon diverticulosis and diverticulitis, small bowel and duodenal diverticula	No
Liver	Dilatation of common bile duct, congenital hepatic fibrosis, cholangiocarcinoma	No
Pancreas	Cysts	No
Spleen	Cysts	No
Lung	Bronchiectasis, pleural effusion	No
Abdominal wall	Hernias	No
Genital	Seminal vesical cyst, sperm abnormalities	No

ADPKD, autosomal dominant polycystic kidney disease; ICA, intracranial aneurysms; SAH, subarachnoid hemorrhage