PKD FOUNDATION
Polycystic kidney disease

PKD Centers of Excellence
Patient Navigator Training Guide

Prepared by the PKD Foundation
Published: May 1, 2026
Version 1.0

For PKD Foundation Centers of Excellence Program Use

Authors and Contributors

Savanna Moore
Centers of Excellence Program Strategist
PKD Foundation

Nicole Torres, R.N.
Community Education Strategist
PKD Foundation

Erin Dahlen, R.N.
Mayo Clinic - Rochester, Minnesota

Gabrielle Cournoyer
Tufts Medical Center

Gauri Bhutani, M.D.
University of Wisconsin–Madison

Jennifer McFadden, R.N.
Mayo Clinic - Arizona

Rachel White, MSN, R.N.
University of Maryland Medical Center

Taylor Avrett
Mayo Clinic - Jacksonville, Florida

© 2026 - PKD. All rights reserved. PKD Foundation is a 501 (c)(3), 509 (a)(1) public charity. Federal tax ID: 43-1266906

Table of Contents

Authors and Contributors 1

Overview 3

Module 1: Understanding PKD and the Patient Navigator Role 3

Module 2: Pharmacological Treatments and Tolvaptan Therapy 8

Module 3: Blood Pressure Management in ADPKD 15

Module 4: Pain Management 21

Module 5: Nutrition and Lifestyle Counseling 26

Module 6: Advanced ADPKD Care – Kidney, Reproductive, and Extra-Renal Considerations 31

Module 7: Health Equity, Advocacy, and Emotional Support 37

Module 8: Putting It All Together – Tools for Success 40

References 44

Overview

This training guide is designed to prepare Patient Navigators to support individuals with autosomal dominant polycystic kidney disease (ADPKD) across a variety of care needs. The guide is broken into eight modules, each with clear objectives and practical tools.

Module 1: Understanding PKD and the Patient Navigator Role

Learning Objectives

- Understand the pathophysiology and clinical progression of ADPKD
- Define the scope, purpose, and responsibilities of the Patient Navigator
- Recognize how interdisciplinary coordination supports comprehensive, patient-centered care
- Become familiar with and refer to the *KDIGO 2025 ADPKD Clinical Practice Guideline for the Evaluation, Management, and Treatment of Autosomal Dominant Polycystic Kidney Disease*, for key recommendations, updates, and implications for clinical practice

Key Content

Overview of ADPKD and ARPKD

(PKD Foundation, 2015, 2025)

ADPKD is the most common inherited kidney disorder, occurring in approximately **1 in 1,000 live births**. To put this into perspective, ADPKD is more common than sickle cell anemia, cystic fibrosis, muscular dystrophy, hemophilia, Down's syndrome, and Huntington's disease combined. It's estimated to affect **over 500,000 Americans** (including pre- and post-transplant patients) and **12.4 million people worldwide**. Despite this prevalence, only about **140,000 patients in the U.S. are currently diagnosed**, giving ADPKD **orphan drug status** for emerging therapies.

What Orphan Drug Status Enables

- **Fast Track Designation:** Expedites the review of drugs that address rare and serious conditions and fulfill an unmet medical need
- **Breakthrough Therapy Designation:** Granted when preliminary clinical

evidence suggests substantial improvement over existing therapies; may also qualify for priority review
- **Accelerated Approval:** Allows approval based on surrogate endpoints that are reasonably likely to predict clinical benefits
- **Priority Review:** Shortens the FDA review timeline to expedite patient access to promising therapies

Causes and Genetics

- ADPKD is caused by mutations in the ***PKD1*** gene (≈78% of genetically defined cases) or the ***PKD2*** gene (≈15%).
- *PKD1*: Often causes more rapid disease progression
- *PKD2*: Usually progresses more slowly
- Other genes (5–10%): A small proportion of cases are caused by variants in additional genes listed in Chapter 1, Tables 1 and 4 of the *KDIGO ADPKD Guideline*, which can result in ADPKD or ADPKD-like symptoms.
- Unknown variant: pathogenic variant cannot be found in about 7% of patients
- If one parent has the mutation, the child has a 50% chance of inheriting it. However**, 20% of patients** have no known family history of the condition.

Clinical Characteristics

- ADPKD is characterized by the gradual development of bilateral renal cysts, leading to:
    - Kidney enlargement
    - Decline in kidney function
    - Early-onset hypertension
    - Pain (chronic or acute)
    - Risk of end-stage renal disease (ESRD)

Disease Impact

- PKD is the **fourth leading cause of kidney failure**, with more than **50% of patients developing kidney failure by age 60.** More than **80% of patients** develop **liver cysts** during their lifetime. ADPKD patients have a **10–15% risk of developing intracranial aneurysms**, about **five times higher than the general population.**

Autosomal Recessive Polycystic Kidney Disease (ARPKD)

- ARPKD is a **rare form of PKD**, occurring in about **1 in 20,000 children worldwide,** usually presenting in childhood. Caused by mutations in the *PKHD1*

gene, a child must receive a gene mutation from both parents to develop the disease.

- Nearly **75% of children survive the newborn stage**. About **25% of these children** may require dialysis or transplant within their first 10 years. More than **90% of children** who survive the newborn stage reach their **20th birthday**, and **half won't require dialysis or transplant**.

Disease Progression and Risk Stratification

(Otsuka America Pharmaceutical Inc., 2025)

Disease severity and progression are highly variable—even within the same family. Risk stratification helps guide treatment decisions. These include:

Total Kidney Volume (TKV)

- **Kidney volume:** Refers to a 3-dimensional measurement of kidney size.
- Total kidney volume = (left kidney volume) + (right kidney volume)
- This can be measured using imaging modalities such as MRI, CT, or ultrasound. Ultrasound is an FDA-recognized biomarker for assessing kidney size. TKV is often adjusted for patient height (height-adjusted TKV, or htTKV) and used for prognostication with the Mayo Imaging Classification described below.
- **Mayo Imaging Classification (MIC):** Categorizes patients by age and height-adjusted TKV into subclasses 1A–1E; used to identify likely rapid progressors.
- **Resource:** The Mayo Clinic's ADPKD classification tool assists clinicians in assessing disease progression by categorizing patients based on height-adjusted total kidney volume (htTKV) and age, informing prognosis and management strategies.
- There is also **Class 2 ADPKD**, which represents **atypical imaging patterns** and accounts for approximately **20% of cases**. In these patients, the **Mayo Imaging Classification (Classes 1A–1E)** may not be fully applicable, as kidney cyst distribution and morphology differ from the typical diffuse pattern used for prognostication (Irazabal et al., 2015).

Genetic Testing

- Can support diagnosis and prognosis in some cases.
- **Resource:** The "Expert" section of ADPKDSim is an educational tool for clinicians offering prognostic tools and interactive simulations to assess disease progression in ADPKD.

Common Complications of ADPKD

(PKD Foundation, 2015)

There are many **renal complications** associated with ADPKD, including:

- Hypertension: often begins in childhood or adolescence; approximately 70% of patients have hypertension by age 30 (Chapman et al., 2003; Schrier et al., 2014).
- Hematuria
- Kidney pain (rupture, infection, or mass effect)
- Cyst infection or rupture
- Kidney stones (nephrolithiasis)

Extrarenal manifestations of ADPKD include:

- Polycystic liver disease (PLD): more common in women
- Intracranial aneurysms
- Aneurysms of ascending part of the aorta
- Cysts in the pancreas and spleen
- Diverticulosis
- Cardiac valve abnormalities
- Abdominal hernias

The Role of the ADPKD Patient Navigator

The **ADPKD Patient Navigator** acts as a consistent support and point of contact throughout the patient's care journey.

- They coordinate with the care team to:
    - Schedule and follow up on labs, imaging, and appointments
    - Assist with medication access and insurance processes (e.g., tolvaptan, BP meds, REMS)
    - Educate patients about ADPKD, lifestyle changes, and treatment options, as well as providing educational materials on ADPKD, local/online PKD communities, and the ADPKD Registry
    - Support care transitions (e.g., referral to transplant, dialysis education)
- They advocate for patients by:
    - Identifying barriers (e.g., transportation, cost, understanding of disease)
    - Connecting patients with resources like financial assistance and social services

Interdisciplinary Coordination

ADPKD care is best managed through a team-based, collaborative approach. Patient navigators serve as liaisons between patients and the entire care team. This team often includes:

- **Nephrologists:** Lead clinical decision-making and monitor disease progression
- **Radiologists:** Perform and interpret imaging for TKV and cyst monitoring
- **Dietitians:** Counsel on sodium restriction, hydration, and CKD nutrition
- **Social workers and case managers:** Support psychosocial and logistical needs
- **Behavioral health providers:** Help patients manage chronic disease stress and anxiety
- **Transplant coordinators:** Engage when kidney function declines toward ESRD, during and post-transplant

KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of Autosomal Dominant Polycystic Kidney Disease

The *KDIGO ADPKD Clinical Practice Guideline* offers evidence-based, expert-endorsed recommendations that support clinicians and patient navigators in optimizing disease management, standardizing care delivery, and guiding informed, patient-centered clinical decision-making.

Interactive Checklist

- ☐ Review the pathophysiology and inheritance pattern of ADPKD (*PKD1* vs *PKD2*)
- ☐ Practice explaining ADPKD progression and complications in clear, patient-friendly language
- ☐ Understand the role of TKV and Mayo Imaging Classification in identifying rapid progression
- ☐ Meet key team members (e.g., nephrology, radiology, pharmacy, social work, behavioral health)
- ☐ Shadow an ADPKD-focused clinic visit to observe team-based care in action
- ☐ Learn to use EMR tools for patient tracking, communication, and follow-up coordination
- ☐ Understand the navigator's role in REMS (Risk Evaluation and Mitigation Strategy) enrollment, insurance pre-authorization, and medication access support
- ☐ Draft a short introduction script to explain your role to new ADPKD patients

Module 2: Pharmacological Treatments and Tolvaptan Therapy

Learning Objectives:

- Understand the mechanism of action, efficacy, and side effect profile of tolvaptan in ADPKD
- Learn how to navigate the REMS (Risk Evaluation and Mitigation Strategy) program and support patient enrollment
- Recognize supportive pharmacologic and non-pharmacologic treatments for ADPKD-related symptoms and disease progression
- Educate and support patients in managing medication adherence and monitoring requirements

Key Content

Overview of Pharmacologic Treatment in ADPKD

(PKD Foundation, 2015)

There is currently no cure for ADPKD, but several medications are used to slow disease progression and manage complications.

Tolvaptan is the only FDA-approved disease-modifying therapy for adults at risk of rapid progression. It is available as generic tolvaptan. With recent generic availability, formulary placement, prior authorization requirements, and patient cost-sharing may vary by payer. Healthcare teams should verify coverage details and assess affordability before prescribing or transitioning patients between products.

Additional medications address hypertension, pain, kidney stones, urinary tract and cyst infections, and metabolic complications of kidney disease.

Tolvaptan: Mechanism of Action and Indications

In a 2012 study published in the *New England Journal of Medicine*, Torres and colleagues provided the following information:

- **Drug class:** Vasopressin V2 receptor antagonist
- **Mechanism of action:**
    - Blocks vasopressin → reduces cAMP → decreases cyst fluid accumulation and cyst growth

- **Indications:** Adults with rapidly progressing ADPKD, typically identified using:
  - Mayo Imaging Classification (Classes 1C–1E) which uses TKV, height and age as data points
  - Pro-PKD score (which includes genetic testing information and clinical information about biologic sex, hypertension, and urologic symptoms such as kidney pain and hematuria)
  - Rapid decline in eGFR
- **Clinical considerations:**
  - Helps slow kidney enlargement and preserve kidney function
  - Requires monitoring for liver safety and adherence to REMS program

Tolvaptan: Clinical Efficacy

Trial	Study Focus	**Outcomes**
TEMPO 3:4 (Torres et al., 2012)	Evaluated tolvaptan in early- to mid-stage ADPKD patients over 3 years	• Slower total kidney volume (TKV) growth and slower eGFR decline compared to placebo • Cyst growth slowed by slightly more than half in tolvaptan-treated patients
REPRISE (Torres et al., 2017)	Assessed tolvaptan efficacy in later-stage ADPKD (eGFR 25-65)	• Tolvaptan slowed the annual rate of eGFR decline by approximately 35% compared to placebo
HALT (Schrier et al., 2014)	Investigated the use of ACE inhibitors and ARBs for blood pressure control in ADPKD	• Demonstrated benefits of intensive blood pressure control on TKV growth and cardiovascular outcomes
CRISP Chapman et al., 2003)	Examined imaging biomarkers and disease progression in ADPKD	• Established the correlation between htTKV and eGFR decline, supporting the use of htTKV for prognosis and clinical trial design

Tolvaptan: Side Effects and Monitoring

Patients taking tolvaptan may experience **common side effects** such as increased urination, nighttime urination (nocturia), excessive thirst, dry mouth, and fatigue. **Rare but serious effects** include hepatoxicity, which requires regular monitoring of liver function tests as part of the REMS program.

To support treatment safety and adherence, patients are advised to maintain adequate hydration and may need to modify daily routines. Patient navigators play a critical role in helping patients understand these effects, manage expectations, and stay consistent with monitoring and medication schedules (Otsuka America Pharmaceutical, Inc., 2025; Torres et al., 2012).

Tolvaptan Counseling Script

Introduction

- "Thank you for taking time to discuss your treatment options. Today, I'll walk you through information about tolvaptan, a medication designed to slow the progression of ADPKD."

What It Does

- "Tolvaptan works by blocking the effect of vasopressin, a hormone that promotes cyst growth in the kidneys. By blocking this signal, the medication helps slow kidney cyst growth and kidney enlargement, which can help preserve kidney function over time."

Who It's For

- "This medication is generally considered for adults with ADPKD who are at risk for more rapid disease progression, based on factors such as imaging results, kidney size, and trends in kidney function tests."

Medication Options

- "Tolvaptan has been available for several years under a brand name. With the recent loss of brand exclusivity, generic tolvaptan may now also be available. This can change medication access, insurance coverage, and cost depending on your plan. Your care team and Patient Navigator can help review coverage and assistance options."

What to Expect

- "This is a long-term therapy. It does not cure ADPKD, but studies have shown

that it can slow disease progression and help delay the need for dialysis or transplant in some patients."

Safety Monitoring

- "Tolvaptan requires regular blood tests to monitor liver function due to the risk of potential liver injury. These tests are done more frequently during the first 18 months and then less often afterward. Our team will help coordinate this schedule and make sure labs are completed on time."

REMS Safety Program

- "To receive tolvaptan, patients and prescribers must participate in a safety monitoring program required by the FDA. This program is designed to ensure that liver monitoring is done regularly. We'll guide you through enrollment and explain how the process works."

Pregnancy Considerations

- "If you're able to become pregnant, effective birth control is required while taking tolvaptan. The medication should be stopped before a planned pregnancy, and you should discuss timing with your care team."

Closing

- "It's completely normal to have questions about this therapy. Would you like to review how the medication access process works, or talk about common side effects and tips for managing them?"

Patient Education Tips

- Provide consistent education via PowerPoint or handouts
- Share educational videos for reinforcement
- **Example:** FDA Approves the First Treatment for ADPKD, PKD Foundation

Tolvaptan REMS Program Overview

(Otsuka America Pharmaceutical Inc., 2025)

Tolvaptan therapy requires participation in a REMS program due to the risk of serious liver injury. Patient Navigators play a key role in supporting safe treatment by coordinating care, providing education, and ensuring compliance with monitoring requirements.

Patient Eligibility

- The healthcare provider evaluates whether the patient meets clinical criteria for tolvaptan therapy before initiating treatment.

Patient Education and Counseling

- The prescriber and/or patient navigator educates the patient about therapy expectations, potential risks, and the REMS monitoring obligations.

REMS Enrollment

- Both the prescriber and patient must complete and sign the REMS enrollment forms before the medication can be dispensed.

Pharmacy Verification and Insurance Authorization

- The specialty pharmacy confirms that all REMS enrollment requirements are fulfilled. Usually, a prior authorization request is submitted to the patient's insurance to ensure coverage. The prior authorization will often be completed at your local institution and once approved the prescription can be processed by the specialty pharmacy.
- Prior authorization (PA) for tolvaptan is approved in most cases. Once the PA is approved, the specialty pharmacy determines the patient's copay (the direct out-of-pocket cost). It's important for patients to confirm this cost directly with the specialty pharmacy before starting therapy.
- If the PA is denied (meaning the patient's insurance will not cover the medication), the out-of-pocket cost for tolvaptan may exceed **$12,000 per month**, which is typically cost prohibitive. In these situations, patient assistance programs such as the Otsuka Patient Assistance Foundation (OPAF) may help eligible patients obtain coverage. In some cases, providers may also submit an appeal of the denial, or patients may explore alternative insurance coverage options during enrollment periods.

Medication Delivery

- Once enrollment and insurance steps are completed, the specialty pharmacy contacts the patient to coordinate shipment or delivery of tolvaptan.

Baseline Laboratory Testing

- Baseline liver function tests must be completed within three months prior to starting therapy.

Ongoing Safety Monitoring

- Patient navigators coordinate ongoing laboratory monitoring to ensure liver safety. The required schedule includes:
  - Day 14 after initiation
  - Day 28 after initiation
  - Monthly for the first 18 months
  - Every three months thereafter

Patient Navigator Roles

- Educate patients on potential risks and monitoring requirements.
- Schedule and coordinate lab draws according to the REMS monitoring timeline.
- Confirm that REMS consent forms are signed.
- Facilitate communication between the patient, specialty pharmacy, prior authorization team (as applicable) and prescribing team.

Lab Monitoring Template (Example)

Patient	Start Date	Baseline Labs	Day 14	Day 28	Months 1–18	Quarterly (>18 months)	Notes
John Smith	2025-05-01	2025-04-15	Complete	Complete	May, Jun, …	Not yet applicable	Mild thirst
Jane Doe	2025-03-10	2025-02-28	Complete	Complete	Mar–Aug	Dec 2026	No side effects

Additional Pharmacologic Management

(PKD Foundation, 2015)

Hypertension

- Most common early manifestation
- First-line treatment options: **ACE inhibitors or ARBs**

Pain

- Avoid NSAIDs and prefer non-opioid strategies (acetaminophen, behavioral therapy).

- Chronic pain may warrant referral to pain/palliative care.

Infection

- **Urinary tract infections (UTIs)** are relatively common in patients with PKD. Once urine culture results are available, treatment typically involves antibiotics targeted to the identified bacteria. Initial (empiric) antibiotic therapy is usually selected based on the provider's clinical judgment, local resistance patterns, and the patient's prior infection history. If a kidney infection (pyelonephritis) is suspected, treatment duration may be extended, often for approximately two weeks.
- **Cyst infections:** Lipid-soluble antibiotics, such as fluoroquinolones, penetrate kidney cyst fluid more effectively and are commonly used when a cyst infection is suspected. Treatment courses for cyst infections are typically longer, often lasting four to six weeks.
- Patients should also be monitored for **hematuria, recurrent UTIs, and kidney stones**, as these complications can occur in individuals with PKD.

Other

- Uric acid management if hyperuricemia is present
- Nephrolithiasis may require additional support

Navigator Role

- Reinforce rationale for treatment
- Encourage adherence and help manage side effects
- Support REMS navigation, lab tracking, and insurance processes
- Communicate regularly with clinical pharmacy, lab, and prescribing teams

Interactive Checklist

- ☐ Review mechanism of action and key clinical trials (TEMPO 3:4, REPRISE)
- ☐ Understand Mayo Classification and eGFR trends for patient eligibility
- ☐ Watch a tolvaptan-focused educational video (clinic or manufacturer resource)
- ☐ Read through REMS packet and patient safety guide
- ☐ Role-play a patient counseling session using the sample script
- ☐ Identify and list supportive medications and their typical uses in ADPKD
- ☐ Locate and test EMR templates for tracking lab draws and tolvaptan milestones
- ☐ Create a quick-reference handout for counseling reminders

Module 3: Blood Pressure Management in ADPKD

Learning Objectives

- Understand the relationship between hypertension and ADPKD progression
- Identify appropriate blood pressure targets for patients with ADPKD
- Recognize the role of pharmacologic and non-pharmacologic strategies in managing hypertension
- Learn how to support patients in effective blood pressure control, addressing adherence and barriers

Key Content

Hypertension and ADPKD

(Chapman et al., 2003; Schrier et al., 2014)

- 70% of ADPKD patients develop hypertension by age 30
- Hypertension may precede noticeable kidney function decline and is linked to total kidney volume (TKV)
- Larger kidneys = higher BP = faster disease progression

Impact on Disease Progression

(Chapman et al., 2003; Schrier et al., 2014)

Uncontrolled hypertension accelerates the decline in **glomerular filtration rate (GFR)** and significantly increases the risk of **cardiovascular complications**, including myocardial infarction (MI), stroke, and heart failure.

Early and sustained BP control is essential to slow disease progression.

KDIGO 2025 ADPKD Guideline: Blood Pressure Recommendations

Monitoring (Devuyst et al., 2025)

- Prefer standardized office BP measurement over routine office checks
- Home blood pressure monitoring (HBPM) is recommended as more reliable than office-only measurements

- Ambulatory BP monitoring (ABPM) should be considered for patients—both children and adults—who have:
    - Difficult-to-control BP
    - Left ventricular hypertrophy (LVH)
    - Proteinuria
    - Declining kidney function with normal office BP readings
    - Evaluate for secondary causes of hypertension when three or more antihypertensive medications are required despite adherence to diet and medication

Non-Pharmacologic Interventions (Devuyst et al., 2025)

- Limit dietary sodium, including processed foods
- Encourage maintaining a healthy weight with balanced nutrition and regular physical activity
- Optimize pain management, which can contribute to BP control

Medical Management (Devuyst et al., 2025)

- Adults aged **18–49 years** with **CKD stages G1–G2** and elevated BP should aim for a home **BP of ≤110/75 mm Hg**, if tolerated.
- Adults aged **50 years or older**, at **any CKD stage**, should target a mean systolic BP **below 120 mm Hg**, measured using standardized office methods, if tolerated.
- First-line therapy for hypertension should include renin-angiotensin system inhibitors (ACE inhibitors or ARBs) to reach recommended BP goals.
- Combination therapy with **ACE inhibitors, ARBs, and direct renin inhibitors** should generally be avoided, regardless of diabetes status. However, in patients with ADPKD—even those with advanced CKD—this combination may be safe, as suggested by the findings of the **HALT-PKD trial** (Torres et al., 2012).
- Additional antihypertensive agents may be added as needed, and therapy should be individualized based on patient age, kidney function, comorbidities, and tolerance.

Pharmacologic Management of Hypertension in ADPKD

First-Line Agents

- **ACE inhibitors (ACEi)** or **Angiotensin II Receptor Blockers (ARBs)** are often the first choice in ADPKD. These medications block the effects of the hormone **renin**, which is released from the kidneys due to the presence of cysts and is one of the main factors resulting in hypertension in ADPKD.

- **ACEi/ARBs** also offer additional benefits for patients with **proteinuria**.

Other Options (Second line medications)

- **Mineralocorticoid receptor antagonists** may be considered if BP remains uncontrolled as these target aldosterone, a hormone downstream of renin (in the renin-aldosterone pathway).
- **Beta-blockers are also good options as these can lower renin levels.**
- **Calcium channel blockers** may be used if needed, depending on individual patient response and tolerability.

Avoidance of Certain Medications

- **Traditional diuretics** (loop diuretics, thiazides) should be avoided unless clear signs of volume overload are present, such as lower extremity swelling. They may be needed in later stages of ADPKD, if at all.
- **SGLT2 inhibitors** should be avoided due to their potential concerns for safety in ADPKD as these medications cause glycosuria which can exacerbate cyst growth. PKD patients have been excluded from recent large clinical trials evaluating these agents. Clinical trials in PKD are ongoing.

Non-Pharmacologic Interventions

(PKD Foundation, 2015)

Lifestyle Counseling

- Sodium restriction
- DASH (Dietary Approaches to Stop Hypertension) diet
- Physical activity: Encouraging regular exercise (e.g., walking, swimming) can help lower BP and improve overall cardiovascular health
- Weight management
- Stress reduction

Patient Education

- Teach patients how to **monitor their own blood pressure** at home using an automated cuff. Ensure they understand how to track readings and when to report changes.
    - Resource for recommended BP monitors: https://www.validatebp.org/
- Provide resources on **stress management**, **adequate sleep**, and **avoiding excessive alcohol or caffeine intake**, all of which can contribute to elevated BP.

Resources

- *Cooking Well*, PKD Foundation
- ADPKD Diet | What is Known and What is Safe webinar: https://www.youtube.com/watch?v=J6w8C4-xAEY

Barriers to BP Control

Structural Barriers

- Limited access to healthcare, medication, or health education in underserved populations
- **Transportation**, **financial limitations**, or **language barriers** may prevent patients from attending regular appointments or filling prescriptions

Interpersonal Barriers

- Patients may face **mistrust** in the healthcare system due to past experiences, particularly in underserved communities
- **Cultural differences** and **health literacy** can affect a patient's understanding of the importance of BP management

Personal Barriers

- **Medication adherence**: Patients may struggle with remembering or managing multiple medications or may experience side effects
- **Competing priorities**: Daily life challenges, such as work, family, and personal health issues, can interfere with treatment adherence

Social Determinants of Health

- **Food deserts**, low socioeconomic status, and lack of access to healthy foods or exercise facilities are key contributors to poor BP control

Navigator Role

Providing Education and Support

- Help patients **understand** the importance of BP control and how it impacts their kidney health
- Educate on **lifestyle changes**, such as dietary adjustments, exercise, and weight management
- Provide information on **self-monitoring** and make sure the patient has access to a BP cuff

Medication Adherence

- **Remind patients** about the importance of adherence, help manage refills, and assist with prior authorization if necessary
- **Track BP readings** in the EMR or via a shared tool and use that data to help adjust treatment as needed

Encouraging Regular Check-ins

- Ensure that patients attend **follow-up visits** regularly for BP and kidney function assessments
- Use **EMR reminders** for lab tests, medication refills, and appointments.

Coordinate with Providers

- Work closely with **nephrologists** and other members of the care team—such as **primary care physicians, cardiologists, dietitians, and social workers**—to ensure a coordinated and comprehensive approach to blood pressure management
- **Assess** the need for adjustments to the medication regimen, including **ACEi/ARB dosing** or the addition of other agents

Monitor for Side Effects

- Help track **side effects** of BP medications, such as dizziness, fatigue, or electrolyte imbalances, and communicate any concerns to the provider

Interactive Checklist

Understanding Blood Pressure and ADPKD

- ☐ Review *KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of Autosomal Dominant Polycystic Kidney Disease* (ADPKD) BP targets and rationale in the context of ADPKD progression
- ☐ Understand indications and side effects of ACEi/ARBs and other agents
- ☐ Learn contraindicated therapies and why they are avoided in ADPKD

Patient Education

- ☐ Demonstrate how to use a home BP monitor and log readings
- ☐ Practice explaining the link between BP and kidney health in lay terms
- ☐ Review DASH diet and provide educational materials or referrals

Team Coordination and Follow-Up

- ☐ Track BP trends and assist in communication with managing provider

- ☐ Set reminders for labs, imaging, or medication renewals
- ☐ Assist with medication access, including prior authorizations

Addressing Barriers

- ☐ Identify social or logistical obstacles
- ☐ Offer strategies and resources tailored to the patient's needs (e.g., medication delivery services, community health programs)
- ☐ Interdisciplinary care coordination among the patient's healthcare team—including the nephrologist, primary care physician, cardiologist (if needed), dietitian, social worker, and other relevant providers—to support comprehensive management

Module 4: Pain Management

Learning Objectives

- Understand the types and causes of pain experienced by patients with ADPKD
- Recognize the importance of individualized pain management strategies
- Identify pharmacological and non-pharmacological options for pain relief
- Develop skills to support patients in effectively managing chronic pain and improving quality of life

Key Content

Types and Causes of Pain in ADPKD

(PKD Foundation, 2015, 2024)

Abdominal Pain

- Most common type, often due to cyst growth and kidney enlargement, causing tissue compression
- Can be intermittent or persistent, sometimes sharp
- May indicate cyst rupture, hemorrhage, or infection in severe cases

Back and Flank Pain

- Caused by enlarged kidneys pressing on surrounding tissues, and often worsens with movement and daily activities

Kidney Stones

- More common in ADPKD due to altered kidney function and fluid imbalance
- Causes acute, severe colicky pain, often radiating to the groin or abdomen

Cyst Infection and Hemorrhage

- Infection causes severe pain plus systemic symptoms like fever (Devuyst et al., 2025)
- Hemorrhage causes sudden, intense pain and may require urgent care (Devuyst et al., 2025)

Musculoskeletal Pain

- CKD and enlarged kidneys contribute to muscle and joint pain, often worsened by poor posture or inactivity

Importance of Pain Management

Chronic pain impacts quality of life, causing fatigue, sleep issues, anxiety, and depression. However, using proper pain control improves function, supports physical activity, and reduces unnecessary hospital visits. Pain is best managed with the help of multi-disciplinary team that may need to include a nephrologist, interventional radiologist. pain specialist, urologist, primary care, and mental health provider.

Non-Pharmacological Pain Management

Heat and Cold Therapy

- Warm compresses for muscle and abdominal cramps; cold for inflammation or acute injury

Physical Therapy and Stretching

- Reduces musculoskeletal pain and improves mobility
- Low- to moderate-intensity exercises for core strength and posture (e.g., Alexander Technique) are recommended (Little et al., 2008)
- Supportive garments may help manage chronic pain in patients with very enlarged kidneys

Massage Therapy

- Can help relieve muscle stiffness and improve circulation

Complementary Medicine

- TENS units and acupuncture may provide additional relief for chronic pain in some patients

Mental Health and Wellness

- **Relaxation Techniques**
  - Mindfulness, deep breathing, and progressive muscle relaxation can reduce pain perception and stress
- **Depression Screening**
  - Recommended for patients with persistent or refractory chronic pain
- **Cognitive Behavioral Therapy (CBT)**
  - Provides coping strategies and support for managing chronic pain
  - Should be delivered by a qualified mental health professional

- **Patient Education:**
    - Providing patient education on acute and chronic pain in PKD and potential treatment options

Pharmacological Pain Management

First-Line Analgesics

- **Acetaminophen:** Preferred for mild to moderate pain; safe with liver considerations
- **NSAIDs:** Avoid or use cautiously due to risk of nephrotoxicity, especially in advanced CKD or high-risk ADPKD
- **Tolvaptan:** May improve acute and chronic pain in patients with ADPKD at high risk for progression

Neuropathic Pain Treatments

- **Antidepressants** (e.g., amitriptyline) or **anticonvulsants** (e.g., gabapentin) for nerve-related pain if first-line analgesics fail
- **Clonidine:** Can help with pain and lower blood pressure; may reduce heart rate

Antispasmodics

- May relieve crampy or colicky pain from stones or cysts
- Usually not recommended for chronic use

Opioids

- Reserved for severe or acute pain (e.g., cyst rupture, stones)
- Use cautiously due to dependence risk and side effects; provide bowel regimen support
- Chronic use is a last-line treatment option for chronic kidney pain; tramadol is typically the starting agent
- Require pain contract and close monitoring with primary care provider

Interventions for Pain management

Cyst-Specific Treatments

- **Percutaneous Cyst Aspiration and Sclerotherapy**
  - Minimally invasive procedure for large dominant kidney or liver cysts causing pain
  - Uses alcohol or foam and is usually performed by an interventional radiologist
- **Cyst Deroofing**
  - Surgical option performed by a urologist
  - More invasive than percutaneous approaches

Nerve Blocks and Interventional Pain Procedures

- Considered for refractory chronic pain requiring significant opioid use
- May include celiac plexus block, splanchnic nerve block with radiofrequency ablation, renal denervation, or spinal cord stimulator
- Usually performed by a pain team or interventional radiology
- Typically considered last-line interventions before nephrectomy

Nephrectomy

- Reserved for severe, unmanageable pain
- Usually performed in patients with end-stage kidney disease, on dialysis, or post-transplant

Managing Acute Pain Episodes

(PKD Foundation, 2024)

Kidney Stones

- Confirm with imaging, usually **non-contrast CT of the abdomen**
- Management includes **hydration, pain control**, and **alpha-1 blocker** (e.g., tamsulosin) to help pass stones naturally
- Short courses of opioids may be needed for severe pain
- **Larger stones** may require urologic intervention

Cyst Rupture/Hemorrhage

- Can cause severe, acute kidney pain and blood in the urine, similar in severity to stone pain
- Most episodes **resolve on their own** within 1–2 weeks
- Rarely, severe hemorrhage may require **hospitalization, acute pain management, or surgery**

Barriers to Effective Pain Management

- Concerns about medication side effects and addiction
- Access to care limitations (remote areas, insurance)
- Psychological factors influencing pain perception and reporting
- Cultural and communication barriers

Navigator Role

- Conduct regular pain assessments with validated scales
- Educate and provide resources on pain management options
- Coordinate with specialists (pain, nephrology, interventional radiology, urology, primary care)
- Monitor medication adherence and side effects
- Advocate for patient needs and connect with support services

Interactive Checklist

- ☐ Review common pain types in ADPKD and their causes
- ☐ Assess patient pain severity regularly using standard scales (follow provider protocols as needed)
- ☐ Educate patients on medication options, benefits, and side effects
- ☐ Understand safe use of acetaminophen, NSAIDs, and opioids
- ☐ Support careful monitoring of opioid use and adherence according to provider protocols
- ☐ Provide resources for complementary or integrative therapies, such as stretching, nutrition and dietary support, and mental health support

Module 5: Nutrition and Lifestyle Counseling

Learning Objectives

- Understand the nutritional needs of patients with ADPKD, including how diet can influence disease progression
- Recognize the importance of individualized nutrition plans tailored to kidney function, comorbidities, and patient preferences
- Identify key lifestyle modifications—physical activity, weight management, fluid intake—to support kidney health
- Develop skills to counsel patients on sustainable lifestyle changes that improve health and quality of life

Key Content

Nutritional Needs in ADPKD

Nutrition guidance for people with ADPKD and CKD G1-G4

(Devuyst et al., 2025)

Fluids / Water

- Encourage intake of **2–3 liters of fluid daily** to maintain dilute urine and reduce kidney stone risk
- Target **morning urine osmolality <280 mOsm/kg,** if measured
- Adjust fluid intake for **medications or impaired urine-dilution** to prevent hyponatremia
- In some cases, providers may recommend **>3 liters per day** based on individual needs

Salt / Sodium

- Aim for **<2 g sodium (~5 g salt) per day**
- Avoid added table salt and heavily processed foods
- High sodium intake is linked to faster kidney enlargement and decline in ADPKD

Protein Intake

- Maintain **moderate protein intake (0.8–1.0 g/kg/day)** for CKD stages G1–G4
- No strong evidence for stricter restriction in ADPKD

- Favor **plant-based proteins** over large amounts of red or processed meat
- KDIGO recommends a balanced diet; no specific diet proven to delay ADPKD progression

Energy / Calories and Weight

- General guideline: **25–35 kcal/kg/day**
- Adjust for weight management (reduce intake if overweight or obese)
- Higher BMI may accelerate disease progression in ADPKD

Fats and Lipids

- Limit fat and sugar intake as part of a **heart-healthy dietary pattern**
- Practical target: **<30% of total energy from fat**, **<10% from saturated fat**

Dietary Fiber and Whole Foods

- Encourage **fiber-rich diet (~25–38 g/day)** from fruits, vegetables, whole grains, legumes, and plant-based proteins
- Aim for **≥400 g of fruits and vegetables daily (~5 portions)**, excluding high-starch vegetables like potatoes
- Limit free sugars to **<10% of total energy** (ideally <5%)

Overall Dietary Pattern

- Emphasize **vegetables, fruits, whole grains, legumes, nuts, plant-based proteins, and unsaturated fats**
- Limit **processed meats, refined sugars, sugar-sweetened beverages, added salt, and calorie-dense foods**
- Follow general population and CKD dietary guidance for heart and kidney health

Nutrition Guidance for people with ADPKD with CKD G4-G5 from the *KDIGO 2024 Clinical Practice Guideline for the Evaluation and Management of Chronic Kidney Disease*

Individuals should be advised to continue eating healthy with special considerations for advanced CKD and metabolic complications.

- **Water:**
    - High water intake may no longer be recommended at this stage unless on tolvaptan

- May need moderation to avoid hyponatremia

Lifestyle in ADPKD

(Devuyst et al., 2025)

Physical Activity

- Aim for **≥150 minutes/week** of moderate aerobic activity (e.g., brisk walking, cycling, swimming) as tolerated
- Include **muscle-strengthening activities 2–3 times/week**
- Minimize prolonged sitting; take frequent movement breaks
- **KDIGO note:** Regular activity benefits cardiovascular health and overall well-being in ADPKD

Body Weight and Nutrition

- Maintain **healthy BMI:** ~18.5–24.9 kg/m².
- Avoid excessive weight gain; high body weight may accelerate kidney growth and decline
- Combine healthy eating with regular exercise for sustainable weight control
- Follow a generally **healthy diet:**
  - Emphasize fresh fruits & vegetables, whole grains, fish, nuts
  - Limit salt, sugar, fats, and excessive animal protein

Sport and Exercise Safety in ADPKD

- Avoid **collision/contact sports** (e.g., football, rugby, boxing, hockey, lacrosse, wrestling, judo) if cysts are large or superficial
- Use **protective equipment** (e.g., athletic corset) if activity causes visible blood in urine or kidney pain
- Monitor **blood pressure** before, during, and after exercise if hypertension or kidney impairment is present

Tobacco and Recreational Toxins

- Avoid all **tobacco use** and secondhand smoke (linked to cardiovascular disease and faster kidney decline)
- Avoid **recreational drugs** and **limit alcohol** for general health

Alcohol and Caffeine Intake

- **Alcohol:** Limit to ~1 drink/day for women, 2 drinks/day for men; consider kidney and heart health
- **Caffeine:** Evidence limited and conflicting; expert opinion suggests **<2400 mg/day (~24 cups coffee)**, especially during pregnancy
    - Higher intake linked to faster kidney growth and pregnancy risks

Supplements and Alternative Nutrients

- No strong evidence supports supplements (β-hydroxybutyrate, curcumin, ginkgo, omega-3s, α-lipoic acid, isoflavones) for slowing ADPKD progression
- Focus on **evidence-based lifestyle and pharmacologic therapies** rather than unproven supplements

Patient-Centered Nutrition Counseling

Note: The Patient Navigator may not have a primary or proactive role in this area; however, they should maintain a strong working knowledge of these elements to support patient education and provide accurate information. The role may also include reinforcing and reviewing recommendations and educational resources shared by the nephrologist with patients.

- Understand the patient's individualized nutrition plan in collaboration with dietitians, considering disease stage, comorbidities, and preferences
- Coordinate with provider teams to answer patient questions about nutrition
- Encourage use of food journals or apps to monitor sodium, protein, potassium, and fluid intake
- Identify and address barriers such as cultural food preferences, financial constraints, and cooking skills by providing practical solutions and resources
- Provide psychosocial support for emotional challenges that affect lifestyle changes

Monitoring and Adjusting the Plan

- Regular follow-ups to assess diet and lifestyle effectiveness on BP, weight, and kidney function
- Periodic labs (renal function, electrolytes, lipids) to guide nutritional adjustments
- Coordinate interdisciplinary care with nephrologists, dietitians, and physical therapists

- Refer to specialists as needed for advanced or complex care

Barriers to Successful Lifestyle Changes

- Knowledge gaps about nutrition and kidney health
- Emotional/psychological stress affecting adherence
- Financial constraints limiting access to healthy foods
- Cultural preferences conflicting with dietary recommendations

Navigator Role

- Educate patients on nutrition and lifestyle's impact on ADPKD progression
- Support goal setting and monitor adherence
- Help overcome barriers by providing tailored resources and emotional support
- Coordinate care and referrals within the healthcare team

Interactive Checklist

- ☐ Assess patient's nutrition knowledge and kidney function stage
- ☐ Educate about protein, sodium, potassium, fluid, and fat management
- ☐ Collaborate with a dietitian to develop individualized nutrition plans
- ☐ Encourage use of tracking tools for food and fluid intake
- ☐ Support lifestyle changes: regular exercise, smoking cessation, alcohol moderation
- ☐ Monitor weight, blood pressure, and lab results regularly
- ☐ Address emotional, financial, and cultural barriers with practical solutions

Module 6: Advanced ADPKD Care – Kidney, Reproductive, and Extra-Renal Considerations

Learning Objectives

- Understand the importance of interdisciplinary care coordination to optimize patient outcomes during transition to kidney replacement therapy or end of life care
- Develop skills to facilitate patient access to specialists and support services
- Recognize the Patient Navigator's role in coordinating care and advocating for complex ADPKD patients

Key Content

Other Medical Interventions in ADPKD

(PKD Foundation, 2015)

Dialysis and Renal Replacement Therapy

- Necessary for patients with end-stage renal disease (ESRD)
- Types: Hemodialysis (blood filtered externally, usually thrice weekly, can be done in-center or at home) and peritoneal dialysis (uses abdominal lining, can be done at home)
- Patient Navigator supports education, access, and decision-making around dialysis options

Kidney Transplantation

- Preferred treatment for ESRD in ADPKD; improves quality of life over dialysis
- Eligibility criteria include absence of severe comorbidities and psychosocial stability
- Living donor transplants may offer better outcomes
- Navigator assists with referrals, pre-transplant evaluation, and post-transplant care coordination

End-of-Life and Palliative Care

- Important for advanced ADPKD/end-stage kidney disease
- Focuses on symptom relief, psychosocial support, and quality of life

- Navigator facilitates advance care planning, DNR/living wills, and hospice access

Care Coordination in ADPKD

(PKD Foundation, 2015)

Interdisciplinary Teamwork

- Advocate for referring patient for kidney transplantation once eGFR 20 or less
- Support patients in developing their preferred dialysis plan at the same time.
- Collaborate with nephrologists, vascular surgery, general surgery in referrals for dialysis access creation based on preferred dialysis modality, likelihood of transplant and progression rate of the ADPKD
- Ensure clear communication, coordinated care plans, and timely treatment adjustments

Specialty Provider Coordination

- Assist patients and their providers in coordinating referrals to dialysis centers when it's time to initiate therapy
- Monitor referral status and ensure appointments are scheduled and completed

Patient Advocacy and Access

- Address barriers such as financial constraints, transportation, and insurance coverage
- Help patients access assistance programs and community resources
- Advocate for coverage of medications, dialysis, and transplant costs

Patient Education and Self-Management

- Educate on adherence, lifestyle, and self-monitoring (BP, weight)
- Provide tailored educational materials and empower patient engagement

Genetic Testing and Reproductive Decision-Making

Fertility Considerations

- Most individuals with ADPKD have normal fertility
- Advanced CKD may reduce fertility and complicate pregnancy outcomes

Pregnancy Risks

- Increased risk of hypertension, preeclampsia, worsening kidney function, and cyst complications during pregnancy (Harrison et al., 2016; Wong et al., 2020)

Preconception Counseling

- Kidney function assessment and medication review (e.g., discontinuation of teratogenic drugs such as tolvaptan)
- Consultation with nephrology, genetic counseling, and maternal-fetal medicine specialists

Contraception

- Safe options include non-estrogen methods (e.g., progestin-only methods, intrauterine devices), particularly for patients with hypertension, CKD, or polycystic liver disease (PLD).

Genetic Testing

- ADPKD is an autosomal dominant condition; an affected individual has a ~50% chance of passing the mutation (*PKD1* or *PKD2*) to offspring (Murphy et al., 2018)
- Genetic testing confirms the specific mutation, informs reproductive decisions, and allows at-risk relatives to consider predictive testing (Murphy et al., 2018; Campbell et al., 2025)

Indications for Testing

- Imaging is inconclusive
- At-risk relatives want early diagnosis
- Potential living kidney donors need evaluation
- Knowledge of the familial mutation can guide reproductive planning and participation in clinical trials based on genotype (Campbell et al., 2025)

Preimplantation and Prenatal Options

- **Preimplantation Genetic Testing (PGT-M):** When used with IVF, it allows selection of embryos unlikely to carry the familial ADPKD mutation, reducing the risk of affected offspring to ~1–2% (Murphy et al., 2018)
- **Prenatal Genetic Diagnosis:** Chorionic villus sampling or amniocentesis can detect fetal mutation status but carries procedural and ethical considerations (Murphy et al., 2018; Campbell et al., 2025)

Genetic Counseling

- All individuals considering testing should receive **pre- and post-test counseling** to understand inheritance patterns, test implications, reproductive options (including IVF with PGT-M), and psychosocial considerations (Murphy et al., 2018)

Extra-Renal Manifestations

- **Hypertension:** Often present early; requires ongoing monitoring and management to slow kidney disease progression
- **Hepatic cysts:** Common, usually asymptomatic, but may cause pain or discomfort
- **Intracranial aneurysms:** Risk increased in patients with family history; screening recommended for high-risk individuals
- **Cardiac manifestations:** Mitral valve prolapse and left ventricular hypertrophy may occur
- **Other manifestations:** Pancreatic cysts, colonic diverticula, and abdominal hernias can appear

Navigator Role

- Facilitate timely referrals to specialists, including nephrology, transplant, reproductive genetic counseling, cardiology, neurology, and hepatology
- Coordinate multidisciplinary care, including dialysis, transplant preparation, extra-renal monitoring, and high-risk pregnancies
- Support patient education on kidney disease management, extra-renal manifestations, reproductive options, genetic testing, and self-monitoring (BP, weight, symptoms)
- Assist with insurance coverage, financial resources, and access to medications, dialysis, transplantation, and genetic testing services
- Monitor treatment adherence, follow-up appointments, and care transitions
- Provide emotional support, advocate for patient needs, and empower patient engagement in shared decision-making

- Educate patients on warning signs that require urgent evaluation and support adherence to screening or imaging recommendations (Irazabal et al., 2015; Torres et al., 2012)

Interactive Checklist

Care Coordination and Advocacy

- ☐ Review and align interdisciplinary care plans
- ☐ Identify involved specialists (nephrology, transplant, reproductive, genetic counseling, cardiology, neurology, hepatology) and facilitate communication
- ☐ Confirm scheduling of tests, procedures, and referrals
- ☐ Assess and address barriers (financial, transport, insurance)

Patient Education and Empowerment

- ☐ Provide information about dialysis, kidney transplant, extra-renal manifestations, and reproductive planning
- ☐ Discuss medication adherence, lifestyle modifications, and self-monitoring (BP, weight, symptoms)
- ☐ Offer mental health and coping support
- ☐ Educate about warning signs requiring urgent evaluation

Dialysis, Transplant, and Genetic Support

- ☐ Assist with transplant referrals, pre-transplant evaluation, and post-transplant follow-up coordination to maintain care continuity as needed (not all navigators will be involved)
- ☐ Coordinate dialysis initiation and ongoing care
- ☐ Support access to genetic counseling, testing, and reproductive decision-making as appropriate

Module 7: Health Equity, Advocacy, and Emotional Support

Learning Objectives

- Understand health equity's importance in ADPKD care, especially for underserved populations
- Identify strategies to address health disparities and promote equitable access
- Recognize the critical role of emotional support throughout the ADPKD disease course
- Develop skills for patient-centered advocacy that respects and amplifies the patient's voice

Key Content

Health Equity in ADPKD Care

Definition

- Health equity ensures all patients can achieve their highest health potential regardless of socioeconomic, racial, geographic, or cultural differences

Barriers to Equity

- Limited healthcare access (rural, underserved areas)
- Economic hardship impacting ability to afford treatments
- Language and cultural differences affecting communication and care understanding

Health Disparities

- Delays in diagnosis and treatment in some ethnic groups
- Financial and insurance barriers to medications, dialysis, and transplantation
- Possible ethnic variations in disease progression requiring tailored approaches

Strategies to Address Equity

- Deliver culturally competent care
- Advocate for equitable resource distribution (specialty care, dialysis, transplants)
- Address social determinants of health (housing, education, employment)
- Connect patients with community and social resources

Advocacy for ADPKD Patients

Patient-Centered Advocacy

- Empower patients through education on disease and treatment options
- Assist with navigating health systems, insurance, government programs
- Support legal aspects (disability benefits, workplace rights, advance care planning)

Resource Referrals

- Provide access to financial aid, transportation, social services, and support groups
- Encourage participation in peer support communities

Policy Advocacy

- Engage with national organizations (PKD Foundation, National Kidney Foundation) to promote policies improving treatment access and affordability

Emotional Support for Patients and Families

Psychosocial Impact

- Chronic illness-related anxiety, depression, and grief
- Family stress due to genetic implications and caregiving burdens
- Social isolation and financial stress

Providing Support

- Practice active listening with empathy
- Refer to mental health professionals when needed
- Teach stress management (mindfulness, relaxation, CBT techniques)
- Facilitate support group involvement

Supporting Caregivers

- Provide resources for caregiver support groups and counseling
- Offer family counseling for emotional challenges

Encouraging Positive Coping

- Promote resilience and empowerment
- Encourage self-care activities and focus on controllable factors (diet, exercise, monitoring)

Interactive Checklist

Health Equity and Advocacy

- ☐ Identify barriers to care for underserved patients
- ☐ Ensure culturally sensitive care and access to translated materials
- ☐ Advocate for financial and insurance support
- ☐ Connect patients with community resources

Emotional Support

- ☐ Provide mental health referrals and support group info
- ☐ Teach coping and stress management strategies
- ☐ Foster safe spaces for emotional expression

Patient-Centered Advocacy

- ☐ Support navigation of healthcare and legal systems
- ☐ Facilitate advance care planning discussions
- ☐ Empower informed decision-making through accessible education

Module 8: Putting It All Together – Tools for Success

Learning Objectives

- Integrate knowledge from prior modules to deliver comprehensive, patient-centered ADPKD care
- Use effective communication skills to foster collaboration with patients, families, and the care team
- Develop and implement practical tools and strategies for care coordination and ongoing patient support
- Establish a framework for ongoing professional growth to remain informed and effective as a Patient Navigator

Key Content

Comprehensive Care Coordination

Multidisciplinary Care Approach

- ADPKD management requires collaboration across nephrologists, dietitians, pain specialists, social workers, and mental health professionals
- As a Patient Navigator, facilitate clear communication and timely referrals to appropriate specialists
- Understand each team member's role and coordinate interventions accordingly (e.g., hypertension management may involve nephrologists, cardiologists, and dietitians)

Comprehensive Care Plan

- Develop individualized care plans incorporating:
    - Pharmacologic therapies (e.g., tolvaptan, antihypertensives)
    - Lifestyle changes (exercise, nutrition)
    - Pain management (physical therapy, medications)
    - Psychosocial supports (counseling, support groups)
    - Advance care planning (dialysis, transplant discussions)
    - Follow-up timelines that keep patients engaged in their ongoing care

Patient-Centered Care

- Actively involves patients in decisions about their care, respecting their values

and preferences
- Use shared decision-making tools to help guide decisions about complex treatment options like dialysis or transplant
- Encourage patient questions and address concerns to improve adherence and satisfaction

Effective Communication Strategies

Patient Communication

- Practice active listening to understand patient concerns, fears, and goals
- Use clear, jargon-free language to explain medical concepts
- Employ teach-back methods to confirm patient understanding
- Confirm how the patient prefers to receive follow-up communication (e.g., *MyChart* message, phone call)

Team Communication

- Keep the interdisciplinary team updated on patient status, symptom changes, and treatment plans
- Use care coordination tools such as shared electronic health records (EHR) and regular team meetings

Tools for Success – Resources for Navigators

Clinical and Educational Resources

- Provide patient-friendly educational materials (print, video, web) explaining ADPKD and management strategies
- Stay current with evidence-based clinical guidelines on treatments and supportive care

Tracking and Documentation Tools

- Use EHR systems to maintain accurate patient records and document clinical notes
- Develop and customize care plan templates for ADPKD patients
- Encourage patient use of monitoring tools (e.g., *MyChart*, a daily blood pressure and/or fluid log, or a symptom diary) to monitor blood pressure, kidney function, fluid intake, and symptoms

Care Coordination Platforms

- Explore software solutions to schedule appointments, manage referrals, and

improve team communication
- Implement patient reminder systems (text, email) to promote appointment adherence and medication compliance

Ongoing Professional Development

Continuous Education

- Keep up to date with the latest research, treatment options, and patient care strategies via conferences, webinars, and professional organizations (e.g., PKD Foundation)

Self-Reflection and Peer Learning

- Regularly evaluate your skills and knowledge to identify improvement areas
- Engage in peer review and knowledge-sharing with colleagues (e.g. request to observe a nephrology visit focused on a topic of interest)
    - Request to observe case discussions between the nephrologist and interdisciplinary teams for complex or specialized patient cases. Set professional goals focused on areas like patient communication, genetic counseling, or dialysis management.

Sustainability and Long-Term Success

Developing Patient Navigator Programs

- Advocate for formal integration of Patient Navigator roles within the PKD Foundation's Centers of Excellence
- Establish protocols for onboarding, training, and mentoring new navigators
- Promote access to affordable medications, educational tools, and patient support services
- Engage in advocacy for policy changes at local and national levels

Patient Retention and Engagement

- Create strategies to maintain patient engagement throughout disease progression (e.g., loyalty programs, care bundles)
- Implement regular patient feedback systems to assess satisfaction with care coordination and support services

Interactive Checklist

Patient Coordination

- ☐ Confirm the care plan includes pharmacological treatment, lifestyle changes, and psychosocial support
- ☐ Coordinate referrals to all necessary specialists and facilitate communication among care team members
- ☐ Develop and communicate a clear timeline for follow-up appointments, explaining their purpose to patients

Communication Strategies

- ☐ Ensure patient communication is clear, simple, and patient-friendly
- ☐ Use active listening to address patient concerns promptly
- ☐ Keep the care team regularly informed of patient progress and treatment updates

Tools and Resources

- ☐ Develop and share accessible educational materials with patients and families
- ☐ Utilize tracking tools for vital health indicators and symptom monitoring
- ☐ Maintain accurate and up-to-date patient records within the EHR system

Ongoing Learning

- ☐ Participate in continuous education on ADPKD research and best practices
- ☐ Engage in peer learning and knowledge exchange with colleagues
- ☐ Regularly assess your competencies and set goals for professional development

References

Campbell, R. E., Edelstein, C. L., & Chonchol, M. (2025). Overview of ADPKD in pregnancy. *Kidney International Reports, 10*(4), 1011–1019. https://doi.org/10.1016/j.ekir.2024.12.035

Chapman, A. B., Bost, J. E., Torres, V. E., et al. (2003). Renal structure in early autosomal-dominant polycystic kidney disease: The Consortium for Radiologic Imaging Studies of Polycystic Kidney Disease (CRISP) cohort. *Kidney International, 64*(3), 1035–1045. https://doi.org/10.1046/j.1523-1755.2003.00185.x

Chebib, F. T., et al. (Year). U.S. commentary on the KDIGO clinical practice guideline for autosomal dominant polycystic kidney disease. *American Journal of Kidney Diseases*, xx(x), xxx–xxx.

Devuyst, O., Ahn, C., Barten, T. R. M., et al.; KDIGO ADPKD Workgroup. (2025). KDIGO 2025 clinical practice guideline for the evaluation, management, and treatment of autosomal dominant polycystic kidney disease (ADPKD). *Kidney International, 107*(2 Suppl), S1–S239. https://doi.org/10.1016/j.kint.2024.11.012

Harrison, T. J., et al. (2016). Pregnancy outcomes in women with autosomal dominant polycystic kidney disease. *Kidney International, 89*(4), 923–931. https://doi.org/10.1016/j.kint.2015.12.021

Irazabal, M. V., Rangel, L. J., Bergstralh, E. J., et al. (2015). Imaging classification of autosomal dominant polycystic kidney disease: A simple model for selecting patients for clinical trials. *Journal of the American Society of Nephrology, 26*(1), 160–172. https://doi.org/10.1681/ASN.2013101111

Levin, A., Eckardt, K.-U., Agarwal, R., et al.; KDIGO CKD Work Group. (2024). KDIGO 2024 clinical practice guideline for the evaluation and management of chronic kidney disease (CKD). *Kidney International, 105*(4 Suppl), S117–S314. https://doi.org/10.1016/j.kint.2024.01.002

Little, P., Lewith, G., Webley, F., et al. (2008). Randomised controlled trial of Alexander Technique lessons, exercise, and massage (ATEAM) for chronic and recurrent back pain. *BMJ, 337*, a884. https://doi.org/10.1136/bmj.a884

Murphy, E. L., Droher, M. L., DiMaio, M. S., & Dahl, N. K. (2018). Preimplantation genetic diagnosis counseling in autosomal dominant polycystic kidney disease. *American Journal of Kidney Diseases, 72*(6), 866–872. https://doi.org/10.1053/j.ajkd.2018.01.048

Otsuka America Pharmaceutical, Inc. (2025). *Tolvaptan REMS program overview*. https://tolvaptanadpkdsharedrems.com

PKD Foundation. (2015). *ADPKD patient handbook*. PKD Foundation.

PKD Foundation. (2024). *When to seek care for a ruptured cyst*. PKD Foundation.

PKD Foundation. (2025). *ARPKD patient handbook*. PKD Foundation.

Schrier, R. W., Abebe, K. Z., Perrone, R. D., et al.; HALT-PKD Study Group. (2014). Blood pressure in early autosomal-dominant polycystic kidney disease. *The New England Journal of Medicine, 371*(24), 2255–2266. https://doi.org/10.1056/NEJMoa1402682

Torres, V. E., Chapman, A. B., Devuyst, O., Gansevoort, R. T., Grantham, J. J., Higashihara, E., Schrier, R. W.; TEMPO 3:4 Trial Investigators. (2012). Tolvaptan in patients with autosomal dominant polycystic kidney disease. *The New England Journal of Medicine, 367*(25), 2407–2418. https://doi.org/10.1056/NEJMoa1205511

Torres, V. E., et al. (2012). Effect of intensive blood-pressure control on kidney outcomes in autosomal dominant polycystic kidney disease: The HALT-PKD randomized trial. *The New England Journal of Medicine, 367*(2), 307–318. https://doi.org/10.1056/NEJMoa1109177

Torres, V. E., Chapman, A. B., Devuyst, O., et al. (2017). Tolvaptan in later-stage autosomal dominant polycystic kidney disease. *The New England Journal of Medicine, 377*(2), 193–203. https://doi.org/10.1056/NEJMoa1702900

Wong, S. W., et al. (2020). Maternal and fetal outcomes in pregnancy with ADPKD. *Clinical Kidney Journal, 13*(4), 543–551. https://doi.org/10.1093/ckj/sfaa042

PKD FOUNDATION
Polycystic kidney disease