PKDCONNECT

A Community of Care

PEER MENTOR PROGRAM

Training Manual

Contents

WE GIVE HOPE.

PKD Foundation Mission

We fund researchers, advocate for patients and build a community for all affected by PKD.

No one faces PKD alone.

PKD Connect Peer to Peer Mentoring Program Vision

The PKD Foundation does not provide medical advice. Information provided to patients, family and friends through our website, educational resources, local Chapter events and peer to peer mentoring program is meant to educate and support our constituents on topics important to disease management. PKD Foundation resources are based on advice and opinion provided by PKD experts but does not constitute medical advice. We strongly recommend that care and treatment decisions related to specific health concerns be made in consultation with the patient's health professional team.

PKD Connect Peer Mentor Program overview

Welcome to PKD Connect Peer Mentor Program! The PKD Foundation is honored that you would like to share your experiences living with PKD with others. No one should feel alone as they manage living with PKD. PKD Connect Peer Mentor Program connects volunteers that are living with PKD (peer mentor) with someone that is looking for support. You, as a peer mentor, can understand what your mentee is going through and can share experiences that will help them feel that they are not alone. Your goal is to listen and help your mentee(s) become their own best health advocate.

What is mentoring?

Merriam-Webster defines a mentor as "a trusted counselor or guide." Peer mentoring is a relationship between a person (mentor) that has had an experience with a person (a mentee) that is new to the experience. At the core, mentoring means making yourself available to support someone when they need it in a way that is meaningful to the mentee. A mentoring relationship can be a short-term relationship or could last for years.

Expectations & requirements

PKD Connect staff members are here to supervise mentee relationships and provide assistance as needed. Staff members will meet regularly with peer mentors to provide support and to discuss progress. It is very important to be responsive to communications and to let PKD Connect staff members know if your availability changes. A one year commitment to the program is required.

Peer mentor expectations

- listen and understand what others are going through;
- provide resources and information, support and encouragement to assigned mentees;
- share their experiences with the people they are supporting;
- help accomplish the mission of PKD Connect Peer Mentor Program;
- report any concerns immediately to PKD Connect staff members.

Peer mentor requirements

- sign a confidentiality statement;
- provide availability for mentoring calls;
- promptly communicate availability changes;
- submit call notes for every mentor / mentee call;
- participate in ongoing training;
- Keep conversations confidential.

Respecting privacy & confidentiality

Confidentiality is extremely important. During the training sessions, as well as during your peer to peer mentoring sessions, all information shared should be kept in strict confidence. Peer mentor volunteers will be asked to sign a confidentiality agreement. We want to ensure that our volunteer mentors, as well as the people we support feel comfortable and confident sharing personal information and experiences.

How to become a PKD Connect Peer Mentor

Submitting your application

The first step in becoming a PKD Connect peer mentor is submitting the application. Applications will be reviewed by PKD Connect staff and mentors will be notified by email within 2 weeks of submission. PKD Connect is dedicated to ensuring that both mentors and mentees have a positive experience participating in the Peer Mentor Program. To ensure the program is successful and meaningful, we will work to keep a balance between the number of mentors and the number of mentees utilizing the mentoring service. If we are not accepting mentors, you will be immediately notified and your application will be kept on file.

Peer mentor training

In addition to the initial PKD Connect Peer Mentor Program application trainings, peer mentors will be required to engage in character role-play and attend ongoing training sessions. Mentors will be provided an overview of new resources available at quarterly trainings to support mentee sessions. These resources are also intended to help answer questions or concerns that peer mentors have. During quarterly sessions we will also share experiences to help all of our peer mentors support each other and the PKD community.

Peer mentor training will be an ongoing process and we thank our mentors in advance for taking the first steps in becoming a peer to peer mentor. Mentors are welcome to walk through the mentor onboarding training sessions at your own pace. Once the onboarding webinar series is complete, quarterly trainings are required. Quarterly training sessions will be recorded and archived for convenience. We are here to support the best process for you. So, let's get started.

Your knowledge about PKD counts

It isn't necessary for peer mentors to be PKD experts. We will leave that to health professionals. It is important to know basic facts. The PKD community is generally very well educated about their disease however, peer mentors will definitely be asked specific medical questions. It is our job, as mentors, to remind the people that we are supporting that we cannot provide medical advice but we can suggest resources that will address specific medical concerns.

Training will include a glossary of terms and answers to frequently asked questions to assist mentors in providing general information about PKD. It is important to be comfortable with general facts about ADPKD and ARPKD (see below) and keep your handbook or your computer accessible during your mentoring calls. You can access useful resources that address topics important to the PKD community in your handbook or online under "Resources".

General information

PKD is one of the most common life-threatening genetic diseases and there is no cure. Today, treatment options are limited to dialysis and transplantation. PKD equally affects men, women and children – regardless of age, race, geography or ethnic origin. There are two forms of PKD: autosomal dominant polycystic kidney disease (ADPKD) and autosomal recessive polycystic kidney disease (ARPKD)

Autosomal dominant polycystic kidney disease (ADPKD)

ADPKD is one of the most common, life-threatening genetic diseases. In ADPKD, fluid-filled cysts develop and enlarge in both kidneys, eventually leading to kidney failure. It is the fourth leading cause of kidney failure and more than 50 percent of people with ADPKD will develop kidney failure by age 50. Once a person has kidney failure, dialysis or a transplant are the only options.

ADPKD is a painful disease that impacts quality of life. The average size of a typical kidney is a human fist. Polycystic kidneys can get much larger, some growing as large as a football, and weighing up to 30 pounds each.

Unlike some genetic diseases, ADPKD does not skip a generation meaning it often affects many people in one family. Approximately 10 percent of the people diagnosed with ADPKD have no family history of the disease, with the disease developing as a spontaneous (new) mutation. Once a person has ADPKD, even through a spontaneous mutation, they have a 50 percent chance of passing it on to each of their children.

Autosomal recessive polycystic kidney disease (ARPKD)

ARPKD is a relatively rare form of PKD that affects 1 in 20,000 babies and often leads to death in the first month of life. Parents who carry the ARPKD gene have a 25 percent chance of passing the disease on to each of their children.

The immediate life-threatening issue for infants with ARPKD is lung immaturity. Lung immaturity is caused in part by insufficient amniotic fluid, produced by the kidneys, due to poor prenatal renal function. Severely enlarged kidneys caused by ARPKD also limit breathing by preventing adequate lung expansion. Death in the neonatal period can be as high as 30 to 50 percent. If an infant with ARPKD survives this critical period, kidney failure can become the most prominent life threatening issue. When the newborn's life isn't at risk, the biggest health concerns are often regulating blood pressure and the chemical balance of blood.

The improved prognosis for ARPKD may be attributed to improved prenatal sonogram technology which allows doctors to diagnose many cases of ARPKD prior to birth. Accordingly,

the birth of an affected child is better planned so the necessary specialists can be alerted. Importantly, the doctors are able to discuss with the parents what they should expect once the baby is born, advising them that the infant may need a breathing tube, may require dialysis, may have severe liver disease, and will require multiple evaluations and treatments to handle associated complications.

Mentor / mentee relationships

Matching your experience to the needs of others

The PKD Connect Resource Center will connect individuals who call us, email us, or connect with us through social media with a mentor who has shared experiences. PKD Connect staff members will introduce mentors to mentees via the email address we provide to our mentors.

The PKD Foundation protects the privacy of our volunteers. We will not share your personal information. As a volunteer peer mentor, you will be provided with a conference line number and an email address. Peer mentors are required to use the provided phone number and email address to communicate with each person that you support.

If you have any questions or concerns about confidentiality of your personal contact information, please contact the PKD Connect staff at any time.

Peer mentors will be asked to provide PKD Connect staff with your preferred times of availability.

PKD Connect members will communicate peer mentors availability up front. We want to help set you up for success so we will set the parameters up front but it will be your responsibility to manage expectations for your availability for follow up calls and email correspondence.

PKD Connect staff will introduce you to the person in need of support via your peer mentor email address.

Peer mentors should make every effort to follow up via email within 2-3 business days. If you are unable to respond due to vacations, illness, work or family needs, etc., please let PKD Connect staff members know and we will follow up or assign another peer mentor.

Agree on a time for your call upfront and log your scheduled peer support call on the shared calendar. Don't be afraid to set boundaries up front.

At the time of the call, you will need to call your mentee at the provided number. Remember: Do not share your personal phone number. Please use the Ring Central mobile phone or desktop app.

Understanding the mentor–mentee relationship

To ensure a successful mentor / mentee relationship, it is important to define exactly what the relationship is from the beginning. For mentoring to be most effective, the roles and responsibilities of both mentors and mentees should be agreed upon from the very beginning.

mentors' role should include sharing experiences with mentees as appropriate, offering encouragement and support, celebrating success, providing feedback and helping mentees find resources to help manage living with PKD. Mentors are not medical experts and cannot give advice. Mentors can and should collaborate with mentees but should not position themselves as experts. The mentors' focus should be to support the mentee and help mentees become their own best advocate. Mentees have their own role and responsibilities as well. Mentees should identify initial goals for the relationship, be open to feedback and take an active role throughout the relationship. If a mentor feels that a mentee would be better matched with another mentor, please let PKD Connect staff know and we will take an active role in assisting both the mentor and the mentee.

How commonality builds connection & trust

It would be ideal for mentors to meet face to face with their mentees however, due to logistical restrictions, most of your engagements will be by phone. Because most (if not all) of your sessions will be by phone, developing skills to help you build trust over the phone becomes critically important to your relationship. Being mindful and using responsive-listening skills are integral components to building mentee trust. Being mindful simply means to be 100% connected and present in the moment of conversation. It also important to incorporate active listening techniques with empathy, rather than judgement. Responsive-listening is also about using your ears and your heart as the barometer to help you chose the most supportive response. Everyone copes with PKD in their own way and in their own time. You may or may not agree with everything you hear and you may or may not have shared the same experiences.

It is important for mentors to understand and recognize when conversations with mentees create strong emotions for the mentors. Discussing some issues might create uncomfortable feelings for you. It is important for you as mentors to be aware of your own personal feelings and points of view and that you keep them from interfering with the support you are offering. If there are times when you are feeling overwhelmed as a mentor, this is the time to reach out to PKD Connect staff for support.

Clarifying top concerns

As mentors and mentees get to know one another, it is important to go over expectations for the relationship and clarify the mentees top concerns. It may not be possible to address all of the mentees' concerns during the first call. Understanding priorities and clear communication between mentor and mentee will set the relationship up for success right from the beginning.

Empathetic active listening

Tips for good listening skills:

- Show that you are interested in what your mentee is saying;
- Offer encouragement;
- Be present in the conversation;
- Spend more time listening than talking;

- Pay attention and jot down notes to help you stay focused on the conversation;
- Ask open ended questions to clarify what your mentee is sharing;
- Don't interrupt. Wait until the mentee finishes talking before you respond;
- Try to put yourself in your mentee's shoes;
- Paraphrase using your own words to clarify understanding;
- If you don't understand, be honest and let them know that you do not understand;
- Do not judge or show bias.

Self-care tips

There are many rewards that come with volunteering and mentoring others. It is very gratifying to know that you have been able to help others and lighten their burden. There are also challenges that may create additional stress for mentors. It is important to make sure that self care is a priority and that mentors have a support system to turn to when in need of a listener. Self care is not just something to do when on vacation. When in need of self care, try the following tips:

Take a break Remember to give yourself time to rest;

Set goals Break down your obligations to small, more attainable goals;

Say no Don't take on more than you can handle;

Create a support system Reduce stress in your life by asking others to help you;

Unplug Allow time for mindfulness by taking a break from technology;

Above all, contact PKD Connect Resource Center staff when in need of support. We are here to help and support our mentors and our mentees.

Managing conversation challenges

It is natural to want to "fix" the problem. You may want to give advice based on your own experience or use your experiences to guide your mentee to the "right" answer. This can derail your conversation and may cause your mentee to feel that you are not respectful of the feelings being shared. It is not your job to advise, direct, provide answers or solutions. It is appropriate to share your own experiences so the mentee can evaluate if what worked for you might be something they should consider for themselves. Be sure to always remind your mentee that you are not a medical professional and that you cannot evaluate your mentee's health or provide recommendations for treatment or disease management. It is always the mentees responsibility to follow up with their health professionals and discuss all questions, concerns or changes to disease management.

Be careful that your questions do not feel like you are prying. It is okay to ask questions to make sure you understand but be careful not to ask too many questions at once or make your mentee feel defensive.

Keep the conversation focused on your mentee. There are times when sharing your experiences can be beneficial. It is always your decision how much you would like to share.

SAMPLE SCRIPT

I can't answer specific medical questions.

"I'm afraid I'm unable to answer that question. You see I'm not a trained medical professional and the Foundation prohibits us from giving medical advice. I'm sure you can understand why these rules are in place. We do however, strongly recommend that you make an appointment to discuss these questions and all of your medical concerns with your healthcare professionals as soon as possible."

Mentor / mentee engagement

You now have your first mentee assignment. You have communicated with your mentee and have determined the time for your call. Let's get ready for the call!

- Be on time! This builds trust and shows your new mentee that you value them.
- Explain your role as a peer mentor. Let your mentee know that you are there to listen but you are not there to provide medical advice or counseling services. You are there to share their journey and provide a support and encouragement.
- Listen and learn about the concerns your mentee brings to the call. Develop some specific goals that your mentee wants to discuss with you.
- Show interest in the conversation.
- Focus on helping your mentee be their own best advocate.
- Provide the mentee with resources to address the concerns that they bring up during the call.
- If your mentee asks you questions that you do not know the answer to, write it down and let your mentee know you will follow up on this question with PKD Connect staff members.
- After the call, fill out the call log form to document the call and the resources provided.
- Be prepared to follow up with your mentee after a few days to see if the information was helpful and if you can connect him / her with additional information.

SAMPLE SCRIPT

Starting the call

"Hello, my name is _____. I'm the Peer Mentor that the PKD Foundation assigned to you. Is this still a good time to talk? Excellent. It's my understanding that you requested a mentor to help you with _____. Is that correct? Do you mind if I ask you a few more questions to ensure I understand your situation? Wonderful. . . ."

Call structure outline

Each call will be different but you can plan to organize your calls in a basic format that will help keep things on track.

Goal setting At the beginning of each call, make sure you identify the goals for the call. Clarify what you hope to accomplish. You may not be able to cover all of the mentee's concerns in one call so set the expectation up front.

Sharing Listen to the mentee's concerns and share experiences that may help support the mentee's goals for the mentoring session.

Reflecting As the end of the scheduled time approaches, it is time to reflect on the session, determine if goals were met and schedule the next call.

Clarifying boundaries

Set boundaries up front.

Ask your mentee what his / her goals are for the first call.

Clarify the top concern(s). You may need to choose one or two concerns to address during a call session.

Let your mentee know if any of the concerns listed need to be addressed by a health professional and be honest that you cannot provide medical advice.

SAMPLE SCRIPT

Setting boundaries

"So there are no surprises, I'd like to give you an idea of what to expect, timewise on these calls. Typically, the first call (today's call) is a bit longer than the calls to follow. Today's call usually runs about 40 – 50 minutes. On subsequent calls, however, our schedules are blocked for a 30-minute time frame. I find the best way to maximize call time is to work from a list of questions that you've prepared in advance of the call. It may also be helpful to know that while my availability varies, when I am available, I block calls between the hours of 9 – 2 PST, Mon. – Fri."

Ending the call & post-call reflection

When the end of the scheduled time for the call is approaching, begin to think about how to end the call. It is important to make sure that the mentee feels success from the mentoring session. How the call ends will reinforce a positive experience and set up future mentoring sessions for success as well.

- Ask the mentee if he/she feels that immediate concerns have been addressed.
- Schedule a follow up call.
- Review resources that were discussed to address mentee's concerns.
- Determine what topics are of concern for follow up calls.

Encourage mentees to spend some time reflecting on the conversation before the next call and to document thoughts and actions based on the conversation. Keep a list of new concerns or questions in preparation for the next call.

SAMPLE SCRIPT

Call completion

"I see we're nearing the end of our call, so let's see how we did. While I know you may have more questions, tell me, did I address your immediate area of concern? Just so you know, this doesn't have to be the last time we discuss this topic. We can continue to discuss this area of concern during future calls, if you'd like?

Preparing for next call

Would you like to schedule another call, or would you prefer to process what we discussed first? If you choose to continue, I'd like to encourage you to keep a list of future topics, so we can discuss them on future calls. Either way, keeping a list will help you drive your best path forward."

Record keeping and follow-up

PKD Connect volunteer mentors are required to provide documentation of every call. This is important to the success of the program and will help PKD Connect staff evaluate the program and provide the resources necessary to ensure that our mentors have the training and resources to support their mentees.

- Please fill out the call log form within 48 hours of your call.
- Report any concerns you may have regarding the call immediately.
- Mentees will receive a survey via email within 24 hours of receipt of call documentation.

What to do if you need help

If you do not know how to best support a mentee, be honest with your mentee. If you feel uncomfortable, it is okay to let your mentee know that you need to consult with PKD Foundation staff members. Sometimes, the people that reach out to us for support need more support than a volunteer is qualified to provide. If you feel that is the case, it is your responsibility to kindly end the call and let your mentee know that a staff member will follow up.

Handling a crisis

It is possible that a volunteer mentor may determine that the mentee is in crisis. The PKD Connect Peer Mentor Program is not a crisis service. It is important that our mentees offer support in the moment and follow up with resources for the appropriate services and support.

Recognising signs of crisis

- Sounding sad
- Expressing feeling anxious, agitated, or unable to sleep
- Frequent and dramatic mood changes
- Feeling that life is not worth living, having no sense of purpose in live
- Talking about feeling trapped- like there is no way out of a situation
- Withdrawing from mentoring program

Tips for handling crisis:

- Active listening: Give the mentee the opportunity to openly tell their story without interruptions or unnecessary questions. Be patient with them, show care while being a compassionate listener. Use active listening questions: “What happened next?”, “How did you react?” and “What was it like?”
- Things not to say: “It could be worse,” “Get over it,” “Why are you making such a big deal about it?” and “Does this happen to you a lot?”
- Express respect, empathy, and compassion. Try not to show your personal feelings, especially if you are amazed or frightened.
- Give them the most possible control to make decisions. This will help them decrease their sense of helplessness. It is important to keep in mind that the goal is to help

them become more independent. Be cautious of them developing an unhealthy relationship of dependency towards you.

- Use breathing techniques to help the mentee calm down.
- Help the mentee identify natural support system (family, friends, church, etc.) in case of an emergency or when you are not available to talk.
- In a timely manner, reach out to PKD Connect staff who can help you evaluate the interaction you have had with the mentee in crisis. Talk about everything you saw, heard, felt, and experienced.

If you feel that your mentee is in crisis, please follow up with the PKD Foundation immediately by phone or email.

PKD Hopeline: 844.PKD.HOPE

Crisis Text Line: 741741

National Suicide Prevention Hotline: 1-800-273-8255

Ending a peer to peer relationship

Most likely, peer to peer relationships will end naturally as mentees feel they have received the support and information needed at the time. If you feel that a relationship with a mentee is no longer productive or no longer feels positive, it is your responsibility to gently conclude the relationship. If you feel at any time that a mentee is not a good match for you as a mentor, please let PKD Connect staff know immediately. A peer to peer relationship can be ended at any time by the mentor or the mentee.

It may be that there are other resources that can be offered to assist your mentee. If you need assistance, please contact PKD Connect staff for help.

SAMPLE SCRIPT

Ending a relationship

Mentor: "Now that we've had 4 sessions, how do you feel about continuing or ending furture engagement."

Mentee: "I haven't really thought about that. I do feel like I can move forward on my own, but can we keep the door open if I feel the need to talk again sometime in the future?"

Peer mentor resource guide

Biopsy

PKD is usually not diagnosed by biopsy. Usually, patients are diagnosed by ultrasound.

Blood pressure

pkdcure.org/what-is-pkd/adpkd/what-are-the-symptoms

Blood pressure is a measurement of the force of the blood as it flows through the body.

High blood pressure or hypertension affects about 60–70 percent of people with ADPKD and begins early in the course of the disease. Many times, the increase in blood pressure will be the first sign of ADPKD and is the primary reason a person gets tested.

High blood pressure should be treated aggressively. If left untreated, hypertension causes further damage to the kidneys, enlarges and thickens the heart muscle and increases the risk for strokes and other cardiovascular events.

Resources:

Webinar — The HALT_PKD study: What do the results of this study mean for me?
pkdcure.org/resource/halt-pkd-study-results

Webinar — Hypertension and HALT PKD Study Results
pkdcure.org/resource/hypertension-halt-pkd-study-results/

Caffeine

pkdcure.org/living-with-pkd/nutrition

There is no direct evidence that caffeine will damage your polycystic kidneys. However, studies of PKD cells grown in a lab have shown that caffeine-like substances promoted cyst growth in PKD. At this time, it may be wise to limit caffeine intake to less than 200 to 250 mg per day.

Chronic Kidney Disease stages

Stage 1: Includes signs of mild kidney disease, with a normal GFR showing 90% or higher kidney function.

Stage 2: Includes signs of mild kidney disease with a GFR showing 60–89% kidney function.

Stage 3: Includes signs of moderate kidney disease and a GFR showing 30–59% kidney function.

Stage 4: Includes signs of severe kidney disease and a GFR showing 15–29% kidney function.

Stage 5: Includes signs of severe kidney disease and kidney failure, with a GFR showing less than 15% kidney function.
davita.com/kidney-disease/overview/stages-of-kidney-disease

Clinical trials

pkdcure.org/living-with-pkd/clinical-studies

Patients play a key role in the research and development process by volunteering to participate in clinical studies. From observational studies to clinical trials, you can help researchers unlock the secrets of PKD and find a treatment by participating in a study.

The link provided will direct you to a list of studies that are currently recruiting participants. Use the contact information provided to find out how to participate.

Creatinine

Creatinine is a measure of kidney function. It is a waste product of muscle metabolism (the work the muscles do). After creatinine leaves the muscles, it enters into the blood, then is filtered by the kidneys and ends up in the urine. There is always some creatinine in the blood and some in the

urine. When there is a loss in kidney function, the kidneys do not clear creatinine from the blood as efficiently as they once did. This causes an increase of creatinine in the blood, which can be measured by a simple blood test.

Diagnosing ADPKD

pkdcure.org/what-is-pkd/adpkd/how-is-adpkd-diagnosed/

Currently, there are three main clinical tests that can be used to diagnose a person with PKD: ultrasound, computed tomography (CT) or magnetic resonance imaging (MRI).

Ultrasound is the most common and least costly screening method for ADPKD.

Testing children

pkdcure.org/what-is-pkd/adpkd/how-is-adpkd-diagnosed/

The question of whether or not to have your child / children tested for PKD is a difficult and personal one. Once a diagnosis is confirmed by imaging or genetic testing, it becomes part of an individual's health records.

PKD patients get diagnosed at all different ages.

Dialysis

pkdcure.org/living-with-pkd/dialysis/

How will I know when I need a transplant or dialysis?

Renal replacement in the form of dialysis is typically recommended when the GRF is about 10 percent. However, this does require planning, discussion with your nephrologist about dialysis modalities and what will work best for you and placement of dialysis access in advance. Your nephrologist will not only be looking at the creatinine level, but also how you are feeling (signs and symptoms) and other electrolyte imbalances.

How do I find a dialysis center in my area?

To find a dialysis center near you and find out how it ranks in terms of quality and care, you can go use the "Dialysis Center Comparison" tool provided by the Department of Health and Human Services. To access this tool, click here.

For a quick listing of dialysis centers with no comparison information, click here.

If you are looking to change dialysis facilities because you are not happy with your current center, keep in mind you may also need to change doctors.

Financial assistance for dialysis

The American Kidney Fund offers several programs that help qualifying dialysis patients pay for health insurance, medications and other treatment-related expenses. You can visit their website at kidneyfund.org for more information. Also, speak with the social worker at your dialysis center as he / she would be able to assist you in applying for these programs and they may have additional suggestions for you as well.

Genetics & genetic testing

pkdcure.org/what-is-pkd/adpkd/how-is-adpkd-diagnosed/

PKD is a genetic disease passed from one generation to the next. It doesn't skip generations and it affects both men and women alike. A person with PKD has a 50 percent chance of passing it on to each of his or her children. Children who do not inherit the disease would have no chance of passing it to their children.

Two genes that cause ADPKD have been identified. About 85% of people with ADPKD have mutations in the PKD1 gene, located on chromosome 16. The remaining 15% of individuals have mutations in the PKD2 gene located on chromosome 4.

The disease caused by ADPKD1 is more severe than that caused by ADPKD2. Individuals with mutations in the PKD1 gene develop cysts, hypertension and loss of kidney function at an earlier age compared to the ADPKD2 gene.

Not everyone experiences all of the common symptoms of PKD and the rate of progression for each family member can vary.

DNA testing is available for ADPKD.

Resources:

Webinar — PKD Genetics, Peter Harris, Ph.D.
pkdcure.org/resource/pkd-genetics/

Athena diagnostics
athenadiagnostics.com/

Insurance & Medicare

Employer Group Health Plan (EGHP)

If you or your spouse are working, you may be eligible for insurance through your employer. Employers with 50 full-time employees or more must offer insurance to their employees. Children up to age 26 are eligible under parents' insurance plan.

Affordable Care Act

If you do not have medical insurance through your employer, you can apply for individual insurance plans through the Affordable Care Act Marketplace. For more information, visit healthcare.gov.

Medicare

Patients that have ESRD (end stage renal disease) can apply for medicare coverage regardless of age.
medicare.gov/people-like-me/esrd/esrd.html

Medigap is extra health insurance that you buy from a private company to pay health care costs not covered by Original Medicare, such as co-payments, deductibles, and health care if you travel outside the U.S.
medicare.com/medicare-supplement/can-i-get-medicare-supplement-coverage-if-i-have-end-stage-renal-disease-esrd

Kidney donor

pkdcure.org/living-with-pkd/transplant/about-donating-your-kidney

For more information about being a kidney donor, please contact a transplant center near you and ask for the living donor coordinator.

Resources:

Webinar — Living Donation, Lisa King and Margo Vandrovec, Mayo Clinic
pkdcure.org/resource/living-donation-2

Guide to Transplantation & Living Donation (hard copy)
Guide to Transplantation (pdf)
Living Donor Insert (pdf)

Kidney failure

Signs of kidney failure are:

- Fatigue
- Poor appetite
- Nausea/vomiting
- Trouble concentrating (in severe cases, confusion)
- Dry, itchy skin, especially if phosphate is high
- Funny taste in your mouth; food tastes funny, metallic
- Muscle cramps at night
- Swelling in feet and ankles

Kidney stones

Kidney stones occur in about 20 to 30 percent of people who have ADPKD as compared to one to two percent of the general population.

The symptoms of kidney stones are severe pain in the back, side or into the groin. Often there will be blood in the urine when passing a kidney stone.

Resources:

We have information about kidney stones available online at pkdcure.org, in our PKD Patient Handbook, and also a webinar on kidney stones that can provide you information. Here are links where you can access this information.

Webinar — Kidney Stones, Dr. Arlene Chapman, Dr. Bharathi Reddy and Dr. Anna Zisman
pkdcure.org/learn/multimedia/webinars/kidney-stones

Medications to avoid

pkdcure.org/living-with-pkd/working-with-your-doctor/

In general, you should avoid any medication that could harm your kidneys or affect your blood pressure. This includes:

- Non-Steroidal Anti-inflammatory Agents, such as Advil, Motrin and Aleve: If you have pain, use can use safe doses of Tylenol or Ultram
- Cold or allergy pills with pseudafed (pseudoephedrine): These can raise your blood pressure.
- Over-the-counter diet pills: These may contain diuretics or stimulants.
- Herbal supplements — discuss any herbal supplements with your physician

As always, you should consult your physician/nephrologist before taking any over-the-counter medications and supplements. He or she can help you weigh the risks and benefits, depending on your degree of renal dysfunction.

Nephrectomy — kidney removal

pkdcure.org/living-with-pkd/donating-pkd-affected-kidneys-for-research/

Nephrectomy is a surgery to remove your kidney. The decision whether or not to have a nephrectomy should be made with your physician on a case by case basis.

Nutrition

pkdcure.org/living-with-pkd/nutrition/

Currently no specific diet has been proven to slow progression of PKD. It is, however, ideal to eat a balanced and healthy diet to maintain optimal body conditions. A healthy body is able to fight infection better, and bounce back faster.

Resource:

Webinar — Keeping on Track with a Healthy Diet, Jacob Taylor
pkdcure.org/resource/keeping-track-healthy-diet/

Pain Management

pkdcure.org/living-with-pkd/chronic-pain-management/

Chronic pain is one of the most common problems for patients with PKD. The pain is usually in the back or the side and occasionally in the stomach. It can be intermittent and mild requiring only occasional pain medicine such as acetaminophen (Tylenol). However, in a small number of patients with severe PKD, the pain can be constant and more severe.

If you are experiencing pain, it is important to consult your physician.

Renal denervation

Renal denervation has been used successfully to treat chronic pain in patients with PKD. Surgical procedures may be performed when more conservative treatments have failed to provide relief for chronic pain. It is imperative to consult with your physician and discuss risks and benefits associated with any surgical procedure.

Resource:

Webinar — Pain and PKD, Dr. Theodore Steinman
pkdcure.org/resource/pain-and-pkd/

Marijuana for pain management

At this time, marijuana has not been studied as a treatment to relieve pain for patients with PKD so we are unable to comment as to whether or not it would be effective or safe for PKD patients.

Advocating for PKD to be an approved condition for medical marijuana

Marijuana has not been studied as a treatment to relieve pain for patients with PKD. Until clinical studies have been conducted to understand the safety and effectiveness, we cannot advocate for marijuana to be used as a treatment until studies are completed.

Pregnancy & PKD

pkdcure.org/living-with-pkd/pregnancy-and-pkd/

The diagnosis of ADPKD is no longer only made years after a person has had a family. The use of noninvasive ultrasonography has made the testing for ADPKD safe for all ages. Thus, screening specifically for ADPKD or finding evidence of ADPKD while doing an ultrasound for other reasons has made early diagnosis a more common occurrence. As the age of diagnosis is lower, an understanding of the risks of pregnancy in women with ADPKD is important.

Generally, women with ADPKD who have normal blood pressure and normal kidney function have uneventful pregnancies and deliver healthy babies. Risk factors associated with pregnancy and ADPKD are due to increased blood pressure.

It is important for a woman with ADPKD to be closely monitored during pregnancy whether she has hypertension or not.

The decision to have children is a very personal one. It is important to discuss concerns with your doctor.

Resource:

Webinar — Family Planning and Pregnancy, Joanne Stone, Mount Sinai
pkdcure.org/resource/pkdchf-cystic-diseases-diagnosis-daily-living/

Protein

pkdcure.org/living-with-pkd/nutrition/

Diets based on plant proteins help lower blood pressure and may help slow the growth of kidney cysts.

Aim for no more than 0.8 grams of protein per kilogram of body weight, per day. To calculate your weight in kilograms, divide your weight in pounds by 2.2. Multiply that number by 0.8 to get your recommended amount of protein per day. Brightly colored fruits and vegetables, whole grains and legumes are rich sources of antioxidants which may help protect the kidneys.

Resource:

Webinar — Keeping on Track with a Healthy Diet, Jacob Taylor
pkdcure.org/resource/keeping-track-healthy-diet/

Slow or stop progress of PKD

There are several things patients can do to try to delay the onset of renal failure:

- Control blood pressure.
- Control cardiovascular risk factors by maintaining a healthy diet, maintaining healthy body weight, exercising regularly, avoiding smoking, etc.
- As with any other kind of kidney disease, avoiding medications that could possibly injure the kidneys. Discuss all medications with your physician.

Spontaneous mutation

pkdcure.org/what-is-pkd/adpkd/

Approximately 10% of patients with PKD are the result of a spontaneous mutation meaning that there is no family history of the disease and the disease is due to a spontaneous mutation of the PKD1 or PKD2 gene.

Symptoms of PKD

pkdcure.org/what-is-pkd/adpkd/what-are-the-symptoms/

Common symptoms include:

- High blood pressure
- Chronic pain or heaviness in the back, sides or abdomen
- Blood in the urine
- Urinary tract infection (UTI)
- Kidney stones

Transplantation

pkdcure.org/living-with-pkd/transplant/pre-transplant-preparation/

Insurance coverage for a transplant

All transplant candidates are entitled to Medicare benefits. Your social worker or your financial coordinator at the transplant center should be providing you with information about how to sign up for Medicare benefits.

Your local Social Security Medicare office can also provide detailed information about Medicare and payment of costs associated with transplant. Out of pocket expenses are different with each insurance plan so you will need to contact your insurance provider for information about your specific benefits.

Glossary

0–9

24-hour urine collection A test done in combination with the blood creatinine test to determine kidney function, called creatinine clearance and is an approximation of glomerular filtration rate

A

ACE inhibitors Angiotensin converting enzyme (ACE) inhibitors; a group of drugs commonly used to treat hypertension in PKD patients

ADPKD Autosomal dominant polycystic kidney disease; the more common form of PKD, it occurs in approx. 1 in 500 live births

Aldosteron A hormone that causes the body to retain salt and lose potassium

Amniocentesis A test used in prenatal diagnosis of chromosomal abnormalities in which a small amount of amniotic fluid, which contains fetal tissues, is sampled from the amniotic sac surrounding a developing fetus, and the fetal DNA is examined for genetic abnormalities

Amniotic fluid The protective fluid contained in the amniotic sac of a pregnant female; the fluid is partially supplied by fetal urine which is produced by the fetal kidneys; in ARPKD, poor prenatal renal function causes a reduction in this fluid

Aneurysm An outpouching in a blood vessel which can leak or rupture

Angiogram Procedures that utilize contrast dye injected into the blood vessels in order to clearly visualize them; it is typically used when an aneurysm is suspected or to look for blockages in heart vessels

Angiotensin A powerful constrictor of blood vessels; it stimulates the production of aldosterone

Angiotensinogen A substance in the blood that forms a hormone called angiotensin

ARBs Angiotensin receptor blockers (ARBs); a group of drugs commonly used to treat hypertension in PKD patients

ARPKD Autosomal recessive polycystic kidney disease; occurs in approx. 1 in 20,000 live births.

arteriovenous (AV) graft A looped, plastic tube that connects an artery to a vein

arteriovenus (AV) fistula A surgically-created connection from an artery to a vein

Aspirate To draw fluid by suction

Autosomes A chromsome that is not a sex chromosome; most cells in our body have 22 sets of autosomes

B

Bladder A muscular sac in the pelvis that collects urine

Blood pressure A measurement of the force of the blood as it flows through the body.

Blood type A classification of blood based on the presence or absence of antigens on the surface of red blood cells; there are four major blood types - A, B, AB, and O; your blood type must be compatible with a potential kidney donor

Blood type incompatible A transplant option that allows you to receive a kidney from a living donor who has an incompatible blood type; specialized medical treatment is required before and after transplant to prevent rejection

Blood urea nitrogen (BUN) A measure of kidney function; urea nitrogen is the waste product of dietary protein, so if the urea nitrogen builds up in the blood, it is a sign of decreased kidney function

C

Caffeine A substance found in coffee, tea, soft-drinks, etc.; it is generally considered best for PKD patients to limit caffeine intake to less than 200–250 mg daily

Calcium A mineral that the body needs for strong bones and teeth. Calcium may form stones in the kidney.

Calcium oxalate A common type of crystal that can lead to kidney stones

Carrier An individual who carries one copy of a recessive gene like that for ARPKD; they do not have the disease but can pass the mutation on to their offspring.

Cell proliferation Cell growth

Chorionic villus sampling A test used in prenatal diagnosis of chromosomal abnormalities in which a sample of chorionic villi is removed from the placenta for testing; also called CVS

Chronic pain Pain that is constant over a long time; long-term pain

Computed tomography (CT) A screening test that may involve radiation or iodinated contrast dye, which can be toxic to kidneys.

Congenital hepatic fibrosis A liver abnormality common in children with ARPKD; it may lead eventually lead to enlargement of the liver and spleen

Creatinine A waste product of muscle metabolism; the level of creatinine in the blood is a measure of kidney function

Creatinine clearance A test to calculate approximately how much actual kidney function you have

Crossmatch A bloodtest that tells you what antibodies you have in your body

Cyclic AMP (cAMP) Signalling molecule in cells that form tubules in the kidneys; abnormalities can lead to cyst formation

Cystitis A UTI with infection in the bladder

D

Diastolic pressure The bottom/second number of the blood pressure reading; it measures the pressure when the heart is relaxing between beats

Direct mutation analysis/ DNA sequencing A type of DNA testing requires only a single sample from the person being tested. An analysis of the DNA sequences of the PKD1 and PKD2 genes is performed.

Directed donation The most common type of living donation; when a living donor (see living donation) names the person who will receive their organ

Diverticula outpouchings on the large intestine

Diverticulitis Can occur when diverticuli rupture or become infected

DNA testing A way to find out if you have a PKD gene. See also gene linkage testing and direct mutation analysis

E

Echocardiogram An ultrasound of the heart

End-stage renal disease (ESRD) When normal kidney function declines and needs to be replaced by dialysis or transplantation; also known as kidney failure; typically considered to occur when GFR is at 10 or less

Erythropoietin Also called EPO; a hormone made in the kidney that tells the bone marrow to make red blood cells; if your kidneys fail or are removed, you must be given EPO via blood transfusions or a synthetic supplement

Estimated post-transplant survival score (EPTS)
A percentile score that ranges from zero to 100 and is assigned to each potential transplant recipient; the score is based on how long you will need a functioning kidney as compared to all other transplant candidates on the list

G

Gadolinium A special dye used to improve visualization in MRIs

Gene linkage testing A type of DNA testing that can determine if you have PKD with 99 percent probability in those with a family history. It requires samples from several family members and looks for "markers" in the DNA. A detailed family history is also required.

Glomerular filtration rate (GFR) The test used to check how well the kidneys are working; it estimates how much blood passes through the glomeruli each minute. Glomeruli are the tiny filters in the kidneys that filter waste from the blood.

Glomerulus A small tuft of blood capillaries in the kidney, responsible for filtering out waste products.

H

Hematuria Blood in the urine

Hemodialysis (Hemo) A procedure that removes extra fluid, electrolytes and waste from blood using a dialysis machine

Hernia Occurs when the contents of a body cavity buldge out of the area where it is normally contained. Two types of hernia, inguinal and umbilical, are more common in those with PKD

Human leuocyte antigens (HLA) Markers that let your immune system know which cells belong to your body and which do not; used in tissue typing

Hypertension High blood pressure; it affects about 60–70 percent of PKD patients and begins early in the course of the disease

I

Inactivating mutation A change in the DNA that leads to a reduced or complete loss of function of a protein

Intercranial aneurysm An aneurysm that occurs in the blood vessels of the brain

K

Kidney Allocation System (KAS) The system that allocates deceased kidney donations to waiting recipients

Kidney donor profile index (KDPI) A percentile score that ranges from zero to 100 and is assigned to each available deceased kidney; the score is associated with how long the kidney is likely to function as compared to other kidneys, based on information about the donor

Kidney stones Small, hard deposits made of minerals and acid salts that form inside the kidneys

L

Lithotripter A machine that uses ultrasound waves to treat large kidney stones

Liver function tests Blood tests that help determine how well the liver is functioning

Living donation When a living person choses to donate their kidney (or other organ) to someone who needs a transplant

M

Magnesium A mineral in the body that is important for metabolism; a deficiency has been associated with high blood pressure

Magnetic resonance arteriogram (MRA) A type of MRI used to visualize the blood vessels in the brain to screen for aneurysms; it is similar to an MRI scan but does not use contrast dye or radiation

Magnetic resonance imaging (MRI) A screening test that uses a powerful magnetic field, radio frequency pulses and a computer to produce detailed pictures of the inside of the body.

Microscopic hematuria Small amounts of blood in the urine

Mitral valve prolapse (MVP) Occurs when the valve between your heart's left upper chamber (left atrium) and the left lower chamber (left ventricle) doesn't close properly.

Mutation An unintended change or typo in a person's genetic code

N

National Institutes of Health (NIH) The agency of the U.S. government primarily responsible for biomedical and health-related research

National Organ Procurement and Transplantation Network (OPTN) The national registry where donor organs are matched to waiting recipients

Neonatal period The first month of life of a newborn

Nephrectomy A surgical procedure to remove one or both kidneys

Nephrogenic Systemic Fibrosis (NSF) A rare but serious complication that can arise from the use of gadolinium

Nephrologist A doctor who specializes in kidneys

Nephrons Tiny filters in the kidney made of think blood vessels; each kidney has about one million nephrons

Neuroradiologist The type of surgeon who repairs aneurysms

Non-directed donation When a living donor (see living donation) does not name a specific person to receive their organ; also called altruistic donation

Non-inactivating mutation A change in the DNA that does not lead to a loss of function of a protein

Non-truncating mutation A change in the DNA that does not truncate or shorten the protein

NSAIDs Non-steroidal anti-inflammatory drugs like aspirin or ibuprofen; these are not advisable for PKD patients to take

P

Pain clinic A clinic or office that uses biofeedback and supports groups to help manage pain.

Paired donation A transplant option for candidates who have a living donor who is medically able, but cannot donate a kidney to their intended candidate because they are incompatible (i.e. poorly matched); consists of two or more kidney donor/ recipient pairs whose blood types are not compatible; the two recipients trade donors so that each recipient can receive a kidney with a compatible blood type.

Peritoneal dialysis (PD) A type of dialysis that removes extra fluid, electrolytes and waste using the lining of the abdominal cavity

Peritoneum The abdominal cavity

PKD Polycystic kidney disease; a genetic disease which causes uncontrolled growth of cysts in the kidneys. There are two forms of PKD: ADPKD and ARPKD.

PKD1 The gene that provides instructions for the polycystin-1 protein; a mutation of the PKD1 gene will cause a person to have ADPKD

PKD2 The gene that provides instructions for the polycystin-2 protein; a mutation of the PKD2 gene will cause a person to have ADPKD

PKHD1 The gene that codes for ARPKD

Polycystic liver disease (PLD) More than 80 percent of PKD patients will develop liver cysts; severe cystic liver disease is uncommon

Polycystin A protein that is encoded by the PKD1 and PKD2 genes; regulates many important tubular cell functions

Polycystin-1 The protein that is coded by the PKD1 gene

Polycystin-2 The protein that is coded by the PKD2 gene

Portal hypertension An abnormality in the liver which can impede the return of blood from the intestine to the liver; it can lead to distention and increased pressure in the veins around the esophagus, the stomach, and the intestine. This can rupture, leading to possibly life-threatening gastro-intestinal bleeding. In addition, portal hypertension can cause spleen enlargement and hypersplenism resulting in low red blood cell, white blood cell and platelet counts.

Positive crossmatch A transplant option that allows you to receive a kidney from a living donor who does not match you due to antibodies; specialized medical treatment is required before and after transplant to prevent rejection; this type of donation is only considered when no other option is available

Potassium A substance essential to all living cells found in most foods; supplements should not be taken without consultation by your doctor or dietician

Pre-emptive transplantation Having a transplant before dialysis is required

Pre-implantation genetic diagnosis A form of early genetic diagnoses that uses vitro fertilization; eggs harvested from a mother are fertilized in a laboratory with the father's sperm then the fertilized embryos are tested for ARPKD; embryos that are diagnosed as free of the disorder are then placed in the uterus with the intent to initiate a pregnancy.

Proteinuria Protein in the urine

Pyelonephritis A UTI when the infection is in the kidney

R

Red blood cells (RBCs) RBCs in the urine is called hematuria.

Regurgitation When blood leaks back to the top part of the heart caused by MVP

Renal dietician A dietician with special knowledge and experience in kidney disease

Renin An enzyme produced in the kidneys

Renin-angiotensin-aldosterone system A hormone system in the body that regulates blood pressure and fluid balance.

S

Satiety Feeling full; a common side effect of severe PLD

Sclerose To harden

Sex chromosomes The chromosomes that contain genes that determine gender

Sonogram See ultrasound

Spontaneous mutation A mutation that arises naturally and is not inherited from parents; also called a de novo

Systolic pressure The top/first number of the blood pressure reading; it measures the pressure when the heart is pumping

T

Therapeutic coil A device surgically placed in an aneurysm to repair it

Total kidney volume (TKV) The total volume your kidney holds and is typically measured by MRI

Truncating mutation A change in the DNA that can truncate or shorten the protein

Tuberous sclerosis complex A rare, multi-system genetic disease that causes benign tumors to grow in the brain and on other vital organs such as the kidneys, heart, eyes, lungs, and skin; can affect infants who have ADPKD

Tubules The filtering part of the kidney

U

Ultrasound The most common and least costly screening method for PKD; a screening method that uses sound waves to develop images of the inside of the body.

United Network for Organ Sharing (UNOS) The organization that sets transplantation policy and who operates NOTA (see National Organ Procurement and Transplantation Network)

Ureters The tubes from the kidneys to the bladder

Urethra The tube that goes from the bladder to the outside

Uric acid A common type of crystal that can lead to kidney stones

Urinalysis An analysis of the urine to determine the type of baceria that is causing infection

Urinary tract infection Commonly called a UTI, an infection caused by bacteria in the bladder, kidneys, or cysts.

Urine Liquid by-product of the body secreted by the kidneys

PKD FOUNDATION
Polycystic Kidney Disease