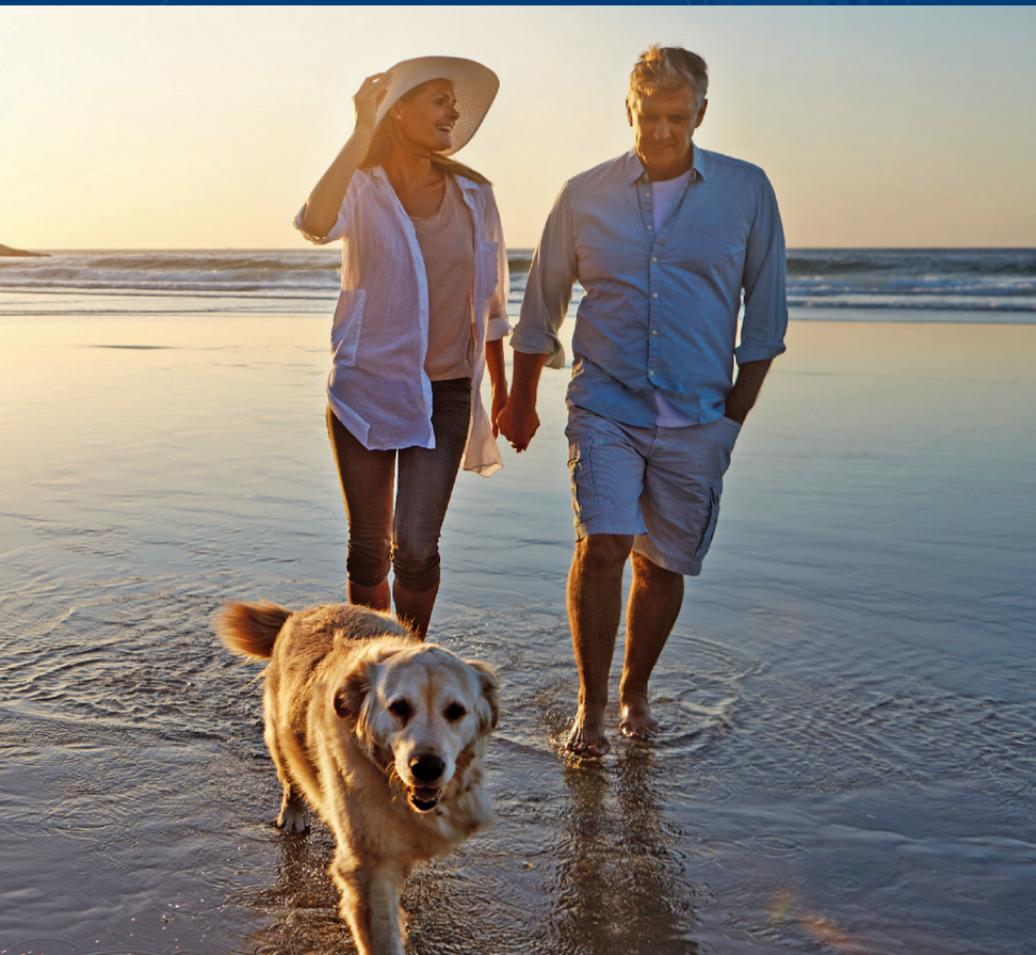


ORIJIN<sup>®</sup>:  
BEGINNING YOUR JOURNEY  
WITH Pr JINARC<sup>®</sup>



Your Treatment Companion Guide



# INSIDE YOUR COMPANION GUIDE

## **Beginning Your Journey with PrJINARC®**

is a companion guide created to help you learn more about your treatment and support you in understanding your ADPKD condition.

You can also refer to [Orijinsupport.ca](https://www.rijnsupport.ca) for additional tips and information.

Throughout your treatment with JINARC, you can use this guide to help:

- Prepare for appointments with your doctor,
- Track test results,
- Make changes in your life such as to your diet and physical activities,
- Keep all the information related to JINARC and your care in one place for easy access at any time.

This is just the starting point: we have gathered the following resources to support you.

**BEGIN HERE AND KEEP GOING.**

PrJINARC® (tolvaptan) is used in adults to treat autosomal dominant polycystic kidney disease (ADPKD).

## WHY WOULD YOUR DOCTOR PRESCRIBE PrJINARC® FOR YOU?

In autosomal dominant polycystic kidney disease (ADPKD), “cysts” or fluid-filled sacs develop on the kidneys. When cysts develop on the kidneys, they can block parts of the kidneys and interfere with the kidneys’ ability to function properly.

Over time, more cysts develop, cysts get bigger, and some may eventually burst. This is known as “disease progression”. As the cysts continue to grow in number and size, the kidneys get larger and have a harder time doing their job. Eventually, the kidneys may stop working altogether.

It has been identified that a naturally occurring hormone known as vasopressin may be involved in ADPKD. Vasopressin makes the kidneys retain water. In people with ADPKD, vasopressin levels are higher than normal. This high level of vasopressin promotes the growth of cysts in the kidneys.

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**JINARC works by blocking the effects of vasopressin. By blocking vasopressin, JINARC is able to reduce the growth of the cysts in the kidneys and slow the decline of kidney function. This should help protect your kidneys from damage and failure.**

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Because of the way that JINARC works, you will produce more urine and you may need to urinate more frequently during the day and at night. This may become less pronounced over time but talk to your doctor or your nurse at the ORIJIN® Patient Support Program if you need help coping with this effect of the medication.

## HOW TO TAKE <sup>Pr</sup>JINARC<sup>®</sup>

### Warnings and precautions

Before you start JINARC, talk to your healthcare team if you:

- have difficulty urinating or have an enlarged prostate
- are dehydrated or suffer from excessive vomiting, diarrhea or sweating
- have low levels of sodium in your blood
- have high potassium levels in your blood
- have gout or high levels of uric acid in your blood
- are taking medication to treat high blood pressure
- are less than 18 years old
- have been asked to permanently discontinue tolvaptan in the past

JINARC is to be taken twice a day in two different doses. There are five possible dose combinations that your doctor may prescribe. The recommended split doses are:

	MORNING	AFTERNOON
<b>1</b> <b>45 + 15 mg,</b> for a total daily dose of 60 mg	45 mg 	15 mg 
<b>2</b> <b>60 + 30 mg,</b> for a total daily dose of 90 mg	60 mg 	30 mg 
<b>3</b> <b>90 + 30 mg,</b> for a total daily dose of 120 mg	90 mg 	30 mg 

JINARC comes in combination blister packs of either 45 + 15 mg tablets together; 60 + 30 mg together; or 90 + 30 mg together. Each blister pack contains a week's worth of medication.

For patients taking moderate inhibitors of CYP-3A alongside JINARC, split-dose regimens should be down adjusted:

	MORNING	AFTERNOON
<b>1</b> <b>15 + 15 mg,</b> for a total daily dose of 30 mg	15 mg 	15 mg 
<b>2</b> <b>30 + 15 mg,</b> for a total daily dose of 45 mg	30 mg 	15 mg 

A schedule of taking a higher dose in the morning and a lower dose in the afternoon is called “**split-dose timing**”. Split-dose timing gives your body the best balance between the amount of medicine in your body and possible side effects (especially night-time urination).

The table below can help you select the timing you feel would work best for you and try to stick to it.

<b>Higher dose tablet</b> <b>Dose 1 – Waking dose</b> (i.e. the time you wake up)	<b>Lower dose tablet</b> <b>Dose 2 – Eight hours later</b>
5 am	1 pm
6 am	2 pm
7 am	3 pm
8 am	4 pm
9 am	5 pm
10 am	6 pm
11 am	7 pm
12 Noon	8 pm
1 pm	9 pm
2 pm	10 pm
3 pm	11 pm
4 pm	12 Midnight
5 pm	1 am
6 pm	2 am
7 pm	3 am
8 pm	4 am
9 pm	5 am
10 pm	6 am
11 pm	7 am
12 Midnight	8 am
1 am	9 am
2 am	10 am
3 am	11 am
4 am	12 Noon

## Pr JINARC® CAN BE TAKEN WITH OR WITHOUT FOOD

**Do not consume grapefruit or grapefruit juice** during treatment with JINARC. Eating grapefruit or drinking grapefruit juice (fresh or frozen) may increase the effect of JINARC. This could cause you to have too great of a drug effect, while taking JINARC.



**Sour oranges, such as Seville, limes, tangelos (a cross between tangerines and grapefruit), and pomelos may have an effect similar to grapefruit.**

Ask your pharmacists about other fruits that may interact with JINARC.

If you miss a dose, take your next prescribed dose at the scheduled time. Do not double up your doses. Do not take 2 doses at the same time.



Set an alarm on your smartphone to the times you selected. There are also many free medication reminder apps – just check the App Store or Google Play for options.

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## WHEN SHOULD JINARC NOT BE USED

Do you have any of the following? Check off any that apply to you. If you have checked things off, you should talk to your doctor before starting JINARC.

- you have a hard time drinking or knowing if you are thirsty
- you have elevated sodium (salt) in your blood
- you have a condition associated with a low blood volume
- your liver is not functioning properly
- you are pregnant or plan to become pregnant, or are breastfeeding
- your body is not able to make any urine
- you are allergic, hypersensitive or intolerant to any of the ingredients in JINARC (including lactose)

## Pr JINARC® AND OTHER MEDICINES

Taking JINARC with some other medicines may affect the way JINARC or the other medicines work. It is recommended to discuss your medications (prescription and non-prescription medicines, vitamins, minerals, natural supplements and alternative medicines) with your healthcare team.

## JINARC AND THE IMPORTANCE OF PROPER HYDRATION

While taking JINARC, you will produce more urine and urinate more frequently. Because you eliminate water when you urinate, it is important to ensure you are replacing what you lose. Unless your doctor tells you otherwise, drink plenty of water during the day and one or two glasses before going to bed.

Water keeps your body working by:

- maintaining your body temperature
- breaking down food so that your body can absorb the nutrients
- helping to get rid of waste products
- acting as a cushion between cells, muscles, and joints
- keeping your bowels regular

**If you suffer from vomiting, diarrhea or any other condition that might cause you to become dehydrated while taking JINARC, call your doctor.**

Some signs of dehydration include:

- Increased thirst
- Dry mouth and/or skin
- Fatigue
- Decreased amount of urine
- Headache/Dizziness
- Irregular heart beats

## WHEN SHOULD I SEEK IMMEDIATE MEDICAL HELP?



Serious angioedema and severe allergic reactions are rare and uncommon. They might occur when you take PrJINARC®.

You should stop taking JINARC and seek immediate medical help if you have any of the following:

- **Dehydration:** if you start to feel much more thirsty than normal, feel like your mouth or skin is very dry, feel really tired or fatigued, are peeing less than normal, have a headache, feel dizzy, or your heart beats are not normal.
- **Severe allergic reactions:** if your face, eyes or tongue starts to swell, if you have a hard time swallowing, if your breathing sounds like wheezing, if your skin is itchy or if you have itchy bumps on your skin (hives), a rash, if you have a fever, have cramps in your abdomen, if your chest feels tight, if you have trouble breathing, or if you pass out.



# WHAT ELSE WILL YOU NEED TO DO WHILE TAKING PrJINARC®?

## LABORATORY TESTS AND MONITORING

An important part of your treatment with JINARC is monitoring your **liver and kidney function**.

Prior to starting, and during treatment, you will have blood drawn to determine your liver function, specifically your alanine transaminase (ALT), aspartate aminotransferase (AST), total bilirubin and alkaline phosphatase (ALP) levels.

If abnormal ALT or AST increases are detected, your doctor will immediately interrupt JINARC and repeat the liver tests.

**ALT, AST and ALP are liver enzymes. When they are elevated, it may indicate acute and chronic liver inflammation, liver disease and/or damage.**

Sometimes these are done when you visit your doctor, or they may be done at a community laboratory. Your schedule for the liver function tests will be as follows:

JINARC treatment stage	Timing of ALT/AST test
Baseline test	Before you start JINARC
First 18 months of treatment	Every month
19-30 months	Every 3 months
After 30 months	Every 3-6 months for duration of treatment

## LIVER FUNCTION TESTS OR LIVER PANEL

These are an important part of your <sup>Pr</sup>JINARC<sup>®</sup> treatment to detect acute and chronic liver inflammation, liver disease and/or damage by measuring substances, which have a role in your body and need to be kept at a certain level.

The JINARC Hepatic Safety Monitoring and Distribution (HSMD) Programme is responsible for monitoring the liver function and minimizing the risk for all patients who have been prescribed JINARC. The HSMD Programme is managed by the ORIJIN<sup>®</sup> Patient Support Program. Therefore, your ORIJIN nurse may call you with reminders when it's time for you to go for liver function testing (ALT/AST). In addition to these calls, if you have a smartphone or similar device, you can use your calendar app to input reminders for when you need to go for another test.

Before taking JINARC, your doctor will ask that you sign a patient-prescriber agreement form (PPAF), stating you understand the benefits and risks of treatment and that you agree to take these blood tests as prescribed by your doctor to start and remain on treatment.

You can obtain support by calling the following number directly 1-844-2JINARC (1-844-254-6272).

## BLOOD PRESSURE

Another important test for people living with ADPKD is blood pressure monitoring. Blood pressure shows the force of the blood against blood vessel walls as it moves through your body. Your physician will test your blood pressure as needed during your treatment. Your reading is made up of 2 measurements: systolic pressure and diastolic pressure.

**Systolic pressure** is the measurement of pressure in your blood vessels when your heart beats (contracts). Systolic pressure is always presented as the upper or first number.

**Diastolic pressure** is the measurement of pressure in your blood vessels between beats when your heart is at rest. Diastolic pressure is always presented as the lower or second number.

The recommended target blood pressure is 110/75 mmHg for ADPKD patients who are younger than 50 years with eGFR  $> 60$  mL/min/1.73 m<sup>2</sup> and without significant cardiovascular morbidities. This indicated that in some patients, an individual target may be needed.

It is encouraged to discuss your personal target blood pressure with your physician.

eGFR: estimated Glomerular Filtration Rate



## FREQUENT URINATION (POLYURIA)



All medications have the potential for side effects, although not everyone gets them.

Two of the most common side effects seen with PrJINARC® are increased frequency of urination and increased amount of urine.

Other common side effects may include thirst, headache, constipation, diarrhea, dry mouth, indigestion, decreased appetite, fatigue, weakness, dizziness, trouble sleeping, muscle spasms, rash or itching.

We encourage you to talk with your doctor, nurse or pharmacist **if you experience any of the following:**

- **Irregular heartbeats, muscle weakness and generally feeling unwell:** could be symptoms of increased levels of potassium in the blood
- **Joint pain, commonly in the big toe, followed by redness, swelling or warmth:** could indicate increased levels of uric acid in the blood (gout)
- **Yellowing of the skin or eyes, dark urine, abdominal pain, nausea, vomiting or loss of appetite:** could be a sign that your liver is being affected by the treatment
- Any other side effects that are concerning you

## MANAGING FREQUENT URINATION

### The Way PrJINARC® Works

In this section we are going to focus on helping you manage frequent urination. The good news is that the need to urinate frequently may become less pronounced over time.

Because of the way that JINARC works, you will produce more urine. This will cause you to urinate more frequently during the day and at night and the amount of urine you pass may also increase. While this may be inconvenient, it is also a sign that the medication is working as intended. This may become less pronounced over time.

#### IMPORTANT:

Please do not cut back on how much water you drink. While taking JINARC, be sure to drink plenty of water! In fact, it is a good idea to have water available to drink with you at all times (see tab Why do I need to drink lots of water?).



#### ORIJIN TIP

There are apps that can help you find public bathrooms.

Check out the App Store or Google Play.



## WHY DO I NEED TO INCREASE MY FLUID INTAKE?

While on PrJINARC®, you will produce more urine and urinate more frequently, thus eliminating more fluid from your body. To prevent becoming dehydrated, have water available to drink while taking JINARC. Drinking water will help replace what you lose and maintain fluid balance.



Thirst isn't always a reliable early indicator of the body's need for water. Many people, particularly older adults, don't feel thirsty until they're already dehydrated. The signs and symptoms of dehydration also may differ by age.

### Adults

- Extreme thirst
- Less frequent urination
- Dark-coloured urine
- Fatigue
- Dizziness
- Confusion

## TIPS ON FLUID INTAKE

Chances are that you will find that you are much thirstier than usual while taking JINARC. This should help you to drink more fluids.

Here are some ideas that other patients have told us have been helpful in drinking more.



- Water can get boring. Try adding bits of fruit or vegetables or diluting small amounts of fruit juices (other than grapefruit juice) in the water to give it some flavour. Just make sure that water remains the main ingredient of whatever you're drinking.\*
- When you feel a craving for snacks, drink a glass of water as well.
- Always carry a water bottle with you and refill it as necessary.
- Drinking through a straw may help you drink more.

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\* If your doctor has put you on a low-potassium diet because your kidneys aren't working well, be careful not to eat fruits or vegetables that are high in potassium, like oranges, kiwis, carrots or tomatoes, among others. In this case, low potassium options such as apples, cherries or grapes are better choices.

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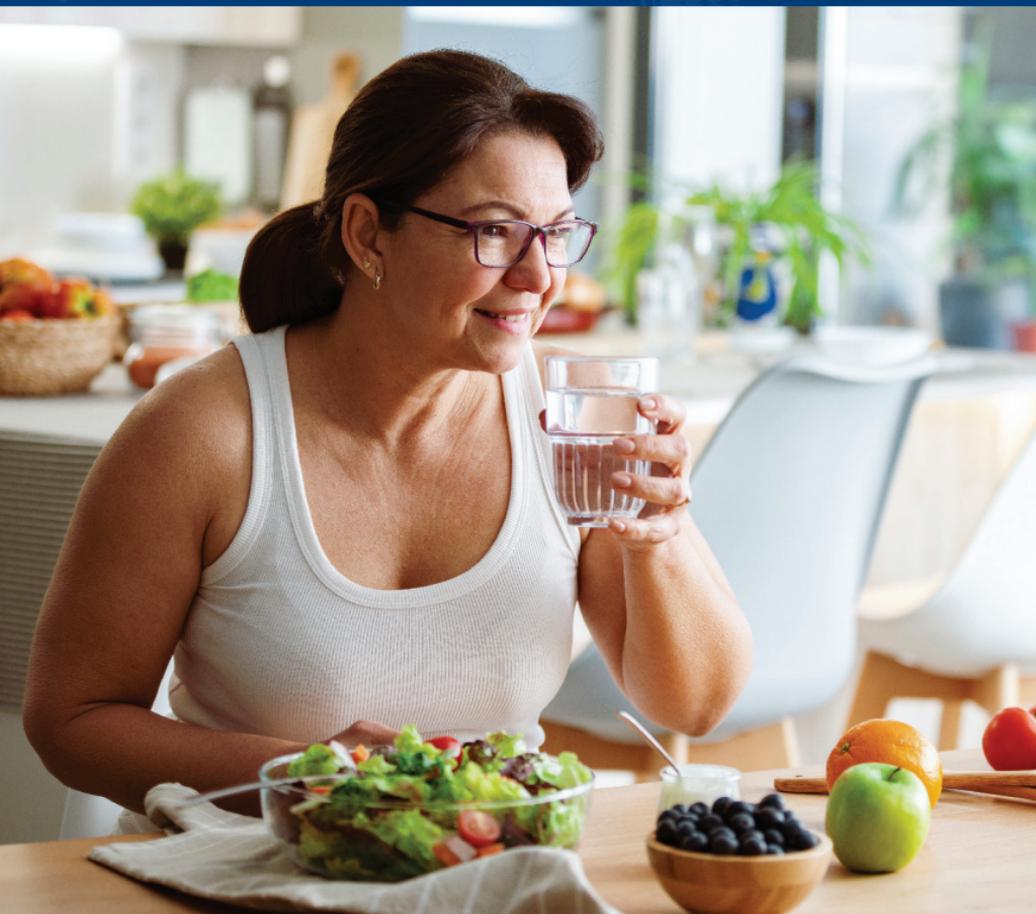
## WHEN AND **HOW** TO DRINK:

- For better hydration, it's best to continually sip water throughout the day as taking in large quantities of water generally ends up being eliminated in the urine quickly.
- Keep a large glass of water beside your bed or in the bathroom, so that you have water handy during the night.
- Much of our fluid comes from the food we eat! In fact, fluid makes up a large part of many of our foods, such as fresh fruit, yogurt and soups.

In the next sections, we have included some common health topics that are important for people living with ADPKD to consider.

The following topics include:

- Nutrition
- Physical activity (next page)



## START SMALL!

Making lifestyle changes can sometimes feel daunting. Breaking down your goals into simple, daily steps can help. Just remember to select those that are important or relevant to you.

## NUTRITION: EATING HEALTHY FOR YOUR KIDNEYS

Currently, no research has shown that any specific diet can make your kidneys better or keep them from getting worse. However, it is always ideal to eat a healthy, balanced diet to:

- fight infection
- bounce back from illness faster
- be in good health

It is a good idea to learn how certain nutrients can affect the kidneys, cysts, blood pressure, etc.



### THE TIPS BELOW MAY HELP

- Determine how much protein your body needs
- Individual requirements vary based on lab results, age, weight, gender, and health concerns
- Favour plant proteins, rich in antioxidants which help lower blood pressure and may slow the growth of kidney cysts
- Opt for brightly coloured fruits and vegetables
- **For patients on a low-potassium diet:** select lower potassium options like broccoli, eggplant or cucumber
- Choose low-sodium foods to help reduce blood pressure

## PHYSICAL ACTIVITY AND YOUR KIDNEYS

Physical activity is an important part of a healthy lifestyle. In general, most physical activity doesn't interfere with kidney function. But with ADPKD, there are some concerns and precautions you should think about. Low impact exercises are safer for people with ADPKD.

Contact sports can be risky. If your kidneys are enlarged, a strong impact to your side or lower back could damage them, or cause cysts to rupture. It's best to avoid these sports. If you do end up doing these sports, you can limit the risk by using protective gear.

### Low-impact activities include:

- Walking
- Swimming
- Tennis
- Rowing
- Golf
- Cycling

### Some high-risk contact sports include:

- Football
- Rugby
- Soccer
- Boxing or kickboxing
- Hockey
- Basketball

While not contact sports, horseback riding and cross-country cycling involve repetitive impact. They have been known to cause blood in the urine in ADPKD patients due to the possibility of a kidney cyst rupturing.

You should talk to your doctor about the kind of physical activity that would be safe for you.

**REMEMBER:** Fluid intake is important when you're taking PrJINARC®. Drink lots of fluids when you exercise and watch out for signs of dehydration.

## FREQUENTLY ASKED QUESTIONS

These are general questions often asked by people with ADPKD. For any specific questions you may have, please contact your healthcare team.

### **How did I get ADPKD?**

ADPKD is a genetic condition that is passed from an affected parent to a child and affects approximately 1 in 400 to 1 in 1000 live births. This condition can be found worldwide in all ethnic and racial groups and in both men and women. If one of your parents has this affected PKD gene, each time they produce an offspring there is a 50% chance of having a child with this disease.

### **I am the only person in my family with ADPKD. My parents are healthy so how did I get it?**

Research shows there is a chance that a new mutation or spontaneous change in a gene can occur up to 20% of the time for ADPKD. Whether it was an environmental factor or another gene (polygenic) that may have triggered it, you can be the first case in the family with ADPKD. If this is the case, your children still are at 50% risk of inheriting this condition.

### **Most of my family members who have ADPKD appear to need dialysis or kidney transplants. Does that mean that I will eventually need that too?**

ADPKD is a condition that does not really have a clear path. Some may develop kidney failure, but others may not. Because the evolution of ADPKD can be unpredictable, it is hard to know how someone with ADPKD is going to progress in the future.

### **Can I still become pregnant?**

While on treatment with PrJINARC®, **you should not become pregnant or breast-feed**. If you are already pregnant, you should not begin treatment with JINARC. To prevent pregnancy while on JINARC treatment, women of childbearing potential should have effective contraceptive measures in place before and during treatment. It is not known if JINARC will harm your unborn baby, or if JINARC will pass into your breast milk.

### **I already have children, will they get ADPKD?**

ADPKD is a genetic disease, which means that it is an inherited disease (although some cases do occur spontaneously). “Autosomal dominant” refers to the pattern of inheritance. In the case of ADPKD, the abnormal gene is dominant. Therefore, there is a 50% chance of the abnormality passing to a person’s children.

### **How will ADPKD affect my job and ability to work?**

The answer to this question varies greatly. It depends on the type of work you do, and how far advanced your ADPKD is.

If your job requires a significant amount of physical work, it may be difficult to perform certain tasks, especially as the disease progresses. If this is the case, if possible, you may want to talk to your employer about adjusting your responsibilities to those that are less physically demanding.

However, even if your job requires little physical activity, you may still find it challenging to perform your regular tasks.

Before disclosing your health status, you may want to contact your province’s Department of Labour to understand your rights as an employee. They can also advise you if disclosure of your medical condition could affect your employment status.

## WHERE TO GET HELP

ORIJIN® Patient Support Program:

[www.orijin-support.ca](http://www.orijin-support.ca)

or call **1-844-254-6272** (1-844-2JINARC)

The PrJINARC® Site:

[www.jinarc.ca](http://www.jinarc.ca)

The PKD Foundation of Canada:

[www.endpkd.ca](http://www.endpkd.ca)\*

The Kidney Foundation of Canada:

[www.kidney.ca](http://www.kidney.ca)\*

Canadian Association of Nephrology Dietitians:

[www.renalrd.ca](http://www.renalrd.ca)\*

Canadian Association of Nephrology  
Social Workers:

[www.cansw.org](http://www.cansw.org)\*

The PKD Foundation (US):

[www.pkdcure.org](http://www.pkdcure.org)\*

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## GLOSSARY OF TERMS

**Autosomal** – refers to a specific disease, condition, or trait due to a mutation in a gene on one of the first 22 non-sex chromosomes. These chromosomes are called autosomes. A single abnormal gene on one autosome from either parent can cause an autosomal disorder.

**Blister pack** – preformed plastic packaging that has several individual, self-contained pockets each designed to hold a single pill.

**Cyst** – an abnormal closed sac or pocket found in the body that contains fluid.

**Disease progression** – refers to the changes or worsening of a disease over time.

**Dominant** – when a single abnormal gene on one autosome from either parent causes an autosomal disorder. The abnormal gene is said to be dominant. For an “autosomal recessive” disorder to occur requires an abnormal gene from each parent. These 2 genes would be recessive genes.

**Polycystic** – “poly” comes from the Greek language and means “many” and “cystic” refers to the presence of cysts. Therefore “polycystic” means having many cysts.

**Urinary** – describes anything relating to the parts of the body responsible for urination or to the process of urination or urinating (peeing); for example, a ‘urinary’ diary.

**Urination** – the release of urine (pee) from the body.

**Vasopressin** – is a hormone often called an antidiuretic hormone. Its main role is to control the reabsorption of water in the kidneys.



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