

Tolvaptan for Adult Polycystic Kidney Disease (ADPKD)

This leaflet is intended to provide you with information about Tolvaptan for ADPKD.

What is ADPKD?

Autosomal Dominant Polycystic Kidney Disease (ADPKD) is an inherited condition that causes fluid-filled cysts to develop within the kidneys. Over time, these cysts gradually increase in size and number. This can cause the kidneys to become enlarged and may lead to a decline in kidney function. Although the rate of progression varies between individuals, some people with ADPKD eventually develop kidney failure and require dialysis or a kidney transplant.

What is tolvaptan?

Tolvaptan (brand name Jinarc®) is currently the only licensed treatment available in the UK that can slow the progression of ADPKD. It works by blocking the action of a hormone called asopressin, which plays a role in cyst growth within the kidneys.

Clinical studies have shown that tolvaptan can:

- Slow the growth of kidney cysts
- Slow the decline in kidney function
- Delay progression to kidney failure in suitable patients
- Potentially postpone the need for dialysis or kidney transplantation

Tolvaptan is not a cure for ADPKD. However, it may help preserve kidney function for longer.

Who can take tolvaptan?

Tolvaptan is recommended for adults with ADPKD who are considered to be at risk of rapidly progressive disease. Your renal specialist will assess whether treatment is appropriate based on factors such as:

- Age
- Kidney function (eGFR)
- Kidney size and imaging findings
- Rate of kidney function decline over time
- Family and medical history

Not everyone with ADPKD will benefit from tolvaptan treatment.

What benefits can I expect?

The effects of tolvaptan on thirst and urine production begin within hours of starting treatment. The benefits on kidney disease progression occur over months and years. Studies have shown that patients taking tolvaptan experience a slower decline in kidney function compared with patients who do not receive treatment. The aim of treatment is to delay future complications and preserve kidney function for as long as possible.

Before starting tolvaptan

Before treatment begins you will:

- Meet with the renal team to discuss the benefits and risks of treatment
- Have blood tests performed, including liver function tests
- Have your kidney function assessed
- Be registered for treatment in accordance with NHS commissioning requirements

You may also be invited to participate in national renal registries that help improve understanding and treatment of ADPKD.

How do I take Tolvaptan?

Tolvaptan is taken twice daily. The first dose is taken in the morning upon waking. The second dose is taken approximately eight hours later.

Typical starting dose:

- 45 mg in the morning
- 15 mg eight hours later

Your dose may gradually increase depending on how well you tolerate treatment. The aim is to achieve the highest dose that you can comfortably tolerate. Always take tolvaptan exactly as prescribed by your renal team.

What happens if I miss a dose?

If you miss a dose:

- Take your next scheduled dose at the usual time
- Do not take an extra dose
- Do not double your next dose

Taking additional doses may increase the risk of dehydration and side effects. If you regularly miss doses, please discuss this with your renal team.

What side effects can I expect?

The most common effects of tolvaptan are related to increased urine production. You may notice:

- Increased thirst
- Passing urine more frequently
- Larger volumes of urine
- Needing to pass urine during the night
- Dry mouth

These effects are expected and show that the medicine is working. Many people find these symptoms improve as they become accustomed to treatment

Drinking fluids while taking Tolvaptan

Tolvaptan causes the kidneys to produce more dilute urine. To avoid dehydration:

- Drink regularly throughout the day
- Always have access to water
- Drink in response to thirst
- Increase fluid intake during exercise or hot weather

The amount of fluid required varies from person to person. Your renal team will advise you if you have any restrictions on fluid intake.

Liver monitoring

A small number of patients taking tolvaptan develop liver inflammation. For this reason, regular blood tests are essential. You will require:

- Blood tests before starting treatment
- Monthly blood tests for the first 18 months
- Blood tests every three months thereafter

Your renal team may advise temporarily stopping treatment if abnormal liver test results are detected.

Seek urgent medical advice if you develop:

- Yellowing of the skin or eyes
- Dark urine
- Severe nausea
- Unexplained tiredness
- Persistent abdominal pain

Sick day rules

You should temporarily stop tolvaptan if you develop:

- Vomiting
- Diarrhoea
- Significant dehydration
- Heat-related illness
- Any illness that prevents you drinking normally

Continue drinking clear fluids if possible.

Restart tolvaptan only when:

- You are eating and drinking normally
- Vomiting and diarrhoea have stopped
- You have been symptom-free for at least 24 hours

If you are unsure, contact the renal team for advice.

Hot weather and Travel

Hot weather can increase your risk of dehydration. During periods of extreme heat:

- Increase your fluid intake
- Monitor your symptoms carefully
- Contact the renal team if you are struggling to keep hydrated

If you are travelling or expect limited access to fluids or toilet facilities, discuss this with your renal team beforehand. Temporary interruption of treatment may occasionally be appropriate

Pregnancy and breastfeeding

Tolvaptan must not be taken:

- During pregnancy
- If you are trying to become pregnant
- While breastfeeding

Women of childbearing potential should use reliable contraception before, during and for four weeks after treatment.

Please inform your renal team immediately if you become pregnant or are planning pregnancy.

Medicines and foods that can interact with Tolvaptan

Some medicines can affect the way tolvaptan works. These include certain:

- Antibiotics
- Antifungal medicines
- Blood pressure medications
- Herbal remedies

Avoid:

- Grapefruit
- Grapefruit juice
- Seville oranges

Always tell your doctor, pharmacist or renal team about any medicines, supplements or herbal products you are taking.

Patient Knows Best (PKB)

Patient Knows Best (PKB) is a secure online patient portal. Through PKB you may be able to:

- View clinic letters
- Access blood test results
- Review appointment information
- Access educational resources
- Communicate with your healthcare team where available

Please ask a member of the renal team for information about registering for PKB.

National Registries

You may be invited to join the UK Kidney Association Rare Renal Disease Registry (RaDaR). This registry helps improve understanding, treatment and outcomes for people living with rare kidney diseases, including ADPKD. Participation is voluntary.

Further information

Video courtesy of University Hospitals of Leicester NHS Trust



Useful websites

PKD Charity <https://pkdcharity.org.uk>
Information and support for people living with ADPKD

Kidney Care UK www.kidneycareuk.org
Information, support and practical advice for people with kidney disease.

UK Kidney Association www.ukkidney.org
Professional and patient information about kidney disease

NHS Website www.nhs.uk
General information about ADPKD and kidney health.

Useful contact details

East and North Hertfordshire Teaching NHS Trust:

- Website www.enherts-tr.nhs.uk
- Telephone 01438 314333

Renal Pharmacy Team:

- Telephone 01438 284677
- Email renalpharmacists.enh-tr@nhs.net

Dr Jeevaratnam's Secretary:

- Telephone 01438 284230

References

1. Torres VE et al. Tolvaptan in Patients with Autosomal Dominant Polycystic Kidney Disease. New England Journal of Medicine. 2012.
2. Torres VE et al. Tolvaptan in Later-Stage Autosomal Dominant Polycystic Kidney Disease (REPRISE). New England Journal of Medicine. 2017.
3. National Institute for Health and Care Excellence (NICE). Tolvaptan for Treating Autosomal Dominant Polycystic Kidney Disease.
4. UK Kidney Association. Clinical Practice Guidance on ADPKD and Tolvaptan.
Jinarc® (Tolvaptan) Summary of Product Characteristics

Leaflet information

**You can request this information in a different format or another language;
please speak to your doctor or nurse.**

Date of publication: August 2018

Version number: 3

Author: C Morlidge

Reference: Renal Pharmacy

Review Date: August 2029

© East and North Hertfordshire Teaching NHS Trust
www.enherts-tr.nhs.uk