Application for Tolvaptan in ADPKD

Fax Cover Sheet

BCRenal
Provincial Health Services Authority

To:	Brenda Lee, Coordinator Tolvaptan Adjudication Team	From:
Fax:	604-875-7366	Fax:
Phone:	604-875-7340	Phone:
Subject:	Application for Tolvaptan in ADPKD	Date:

Please include the following forms. This is a mandatory requirement for review and approval of this application:

- ☐ Tolvaptan Application Form
- ☐ Patient-Prescriber Agreement Form (PPAF)
- ☐ Copy of Imaging Report
- ☐ Copy of GFR Report
- ☐ Any Other Supporting Documentation

Ensure all items listed as mandatory on the application form are included.

Comments: _____

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PATIENT INFORMATION LABEL

Name: _____

Address: _____

Phone: _____

Date of Birth (MM/DD/YYYY): _____

PHN: _____

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Rev: March 2021

Instructions:

- Ensure the patient is registered in PROMIS, with a diagnosis of ADPKD
- Complete the information below, fax this form along with the PPAF/Tolvaptan consent for monitoring and any supporting documentation (e.g., imaging reports) to BC Renal at 604-875-7366

***Indicates Mandatory field**

The following information is required for approval:

*Confirmed diagnosis of ADPKD: ☐ Yes ☐ No

Tolvaptan is only indicated for use in ADPKD and not in any other renal cystic disease.

Current patient characteristics:

*Current age: _____ years DOB: _____/_____/_____ *Most recent BP: _____/_____ mmHg
*Patient height: _____ cm Mayo Class (if known) _____

Imaging:

To interpret these results please provide confirmation of typical morphology of ADPKD ***and*** renal sizes. 'Typical morphology' is defined as diffuse, bilateral cystic involvement of the kidneys (i.e., not atypical morphology which includes asymmetric, unilateral or segmental cystic involvement)

*Typical morphology? Yes _____ No _____ Unknown _____

***Current renal size** - At least one of these measurements must be included. If both are available, include TKV. Ultrasound is not sufficiently accurate to assess TKV and should be used for kidney length only, if TKV is not available.

TKV _____mL ☐ CT ☐ MRI Date _____
Ultrasound kidney length: R _____ cm L _____ cm Date _____

☐ ***Attach a copy of the imaging report along with this application.**

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Evidence of disease progression

Demonstration of disease progression may assist with determining candidacy; this would include evidence of GFR decline or rapidly increasing kidney volume over the course of the last ≥2 years.

GFR - Please provide all available GFR results over the last 3 years. If unavailable, please enter GFR results in the table below; provide values approximately 1 year apart to determine annual decline.

GFR*			**Date**
Current		mL/min/1.73m^2	
Previous		mL/min/1.73m^2	
Previous		mL/min/1.73m^2	

Historical renal size – Provide values at least 1 year apart to determine rate of growth.

Previous TKV	**CT**	**MRI**	**Date**
1.	☐	☐	
2.	☐	☐	
3.	☐	☐	

Please list any other clinical criteria not listed above that may impact patient's candidacy:

Please attach all supporting documents along with this application.

Criteria for tolvaptan use in ADPKD

Potential candidates for treatment with tolvaptan are those with ADPKD and more rapidly progressing disease. These criteria are based on evidence from clinical trials[1,2], and the Updated Canadian Expert Consensus on Assessing Risk of Disease Progression and Pharmacological Management of Autosomal Dominant Polycystic Kidney Disease[3].

Please indicate which group (**A, B or C**) best reflects your patient's characteristics, as well as which criteria within that group are met.

☐ **Group A**: Patients 18-55 years old who are similar to those in clinical trials[1,2]:

eGFR >25 mL/min/1.73 m^2 **AND** Evidence of renal enlargement
Renal enlargement can be documented as any of:

- ☐ TKV >750 mL in those with eGFR >45 mL/min/1.73m^2
- ☐ Class 1C, 1D or 1E on the Mayo Clinic Classification

Although not a criterion in the REPRISE trials, documentation of renal enlargement has been included here as a criterion. In those patients with advanced or rapidly progressive CKD without enlarged kidneys, an alternate diagnosis for CKD should be investigated.

☐ **Group B**: Patients 55-65 years old who would have met criteria for the REPRISE trial[2], and who also have evidence of rapid disease progression. **All three of these criteria must be met:**

- ☐ eGFR of 25 to 44 mL/min/1.73 m^2

AND

- ☐ Historical evidence of a decline in eGFR >2.0 mL/min/1.73 m^2/year

AND

- ☐ Class 1D or 1E on the Mayo Clinic Classification

Although not a criterion in the REPRISE trial, documentation of renal enlargement has been included here as a criterion to ensure that only the more rapidly progressing patients are chosen for tolvaptan treatment. In those patients with advanced or rapidly progressive CKD without enlarged kidneys, an alternate diagnosis for CKD should be investigated.

☐ **Group C**: Patients 18-65 years of age with eGFR ≥25 mL/min/1.73 m^2 who do not otherwise fit into the trial criteria may also be considered if they display other markers of rapid disease progression, including those listed below. Not all factors in this group in isolation would meet criteria for tolvaptan treatment. Therefore, all or many of these factors should be considered together:

- ☐ Annual decrease in eGFR of >2.5 mL/min/1.73 m^2 and alternate CKD diagnoses have been excluded
- ☐ Increase in TKV of >5% per year
- ☐ Mayo Clinic Classification groups 1D or 1E
- ☐ PKD 1 protein truncating mutation
- ☐ Classified as high risk via the PROPKD risk score (7-9 points)
- ☐ MRI or CT cannot be done to assess TKV and patient has large kidney lengths (e.g. >20cm) on ultrasound. Ultrasounds are not as strong of a predictor of progression as total kidney volume and should only be considered if TKV is not possible to obtain
- ☐ Consideration can be given to patients with high symptom burden related to renal expansion, but this alone is not generally an indication for tolvaptan treatment. Symptom burden should be considered in addition to other clinical criteria.