ADPKD Annual Report

BC RENAL ADPKD NETWORK

2023

ADPKD Annual Report

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1   Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is the most common inherited renal disorder and is the fourth leading cause of end-stage renal disease in Canada. A lifelong disease, patients develop clusters of cysts -- noncancerous round sacs containing fluid. The disease is quite variable, from minimal impact on kidney function to rapidly progressive disease that results in kidney failure at a young age. There is no cure for ADPKD at this time, but research has led to improved tools for diagnosing and predicting the renal outcome of the disease, as well as treatments that may slow the progression of ADPKD in some people.

As part of BC Renal's overarching priorities of optimizing patient experience and outcomes as well as innovation and research in renal care, we have developed and implemented a provincial ADPKD strategy in BC's renal programs that supports equitable and sustainable care for patients and families living with ADPKD.

As a first in kind comprehensive provincial registry, the BC ADPKD registry was established to enhance the ability to identify ADPKD patients in BC and gain a better understanding of the burden of disease, current management, and outcomes of these patients. The granular data in the registry informs continuous quality improvement initiatives including facilitation of practice audits wherein individual clinicians can use the data to get a better understanding of their current ADPKD management and identify areas for improvement.

2   Data Sources

Early identification, assessment of renal progression and implementation of appropriate treatments are key components of modern ADPKD care.

The ADPKD patient registry was created to enhance identification and understanding of ADPKD in BC including basic demographics, management patterns and clinical outcomes of this specific group of patients. A main focus in the registration efforts has been identification and registration of patients seen in nephrologists' private offices, prior to enrollment in other BC Renal administered services. Over time there is a noted increase in the number of ADPKD patients registered in PROMIS which is most prominent in early-stage patients not on dialysis or transplant.

The ADPKD registry includes data that will capture patterns of treatment use, specifically tolvaptan which has been funded by BC Renal since January 2020. Cost of BCR-funded tolvaptan is being captured through a separate tolvaptan utilization report.

Further enhancements in PROMIS are pending including information such as tolvaptan approval criteria, total kidney volume and classification.

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3   ADPKD Overview

3.1   ADPKD in BC

Total number of active patients with ADPKD diagnosis registered as of March 31 each year (2015 to 2021) regardless of setting (KCC, MD office, dialysis, transplant).

Report Date As Of	2015	2016	2017	2018	2019	2020	2021	2022	2023
Total number of registered ADPKD patients	709	772	1041	1121	1179	1292	1352	1381	1431
ADPKD patients followed in KCC as of report date	211 (30%)	214 (28%)	256 (25%)	282 (25%)	342 (29%)	452 (35%)	519 (38%)	554 (40%)	597 (42%)

Total number of active patients with ADPKD diagnosis in BC regardless of setting (KCC, MD office, dialysis, transplant) as of March 31, 2023

|	#
Total number of active patients with ADPKD diagnosis in BC as of period end	**1431**
By Health Authority:	
IHA	144
FHA	347
VCH+PHC	613
VIHA	237
NHA	53
Patients whose primary nephrologist does not belong to any HARP/unknown	37

Number of active patients with ADPKD diagnosis followed in KCC and by health authority as of 03/31/2023.

ADPKD Patients in KCC
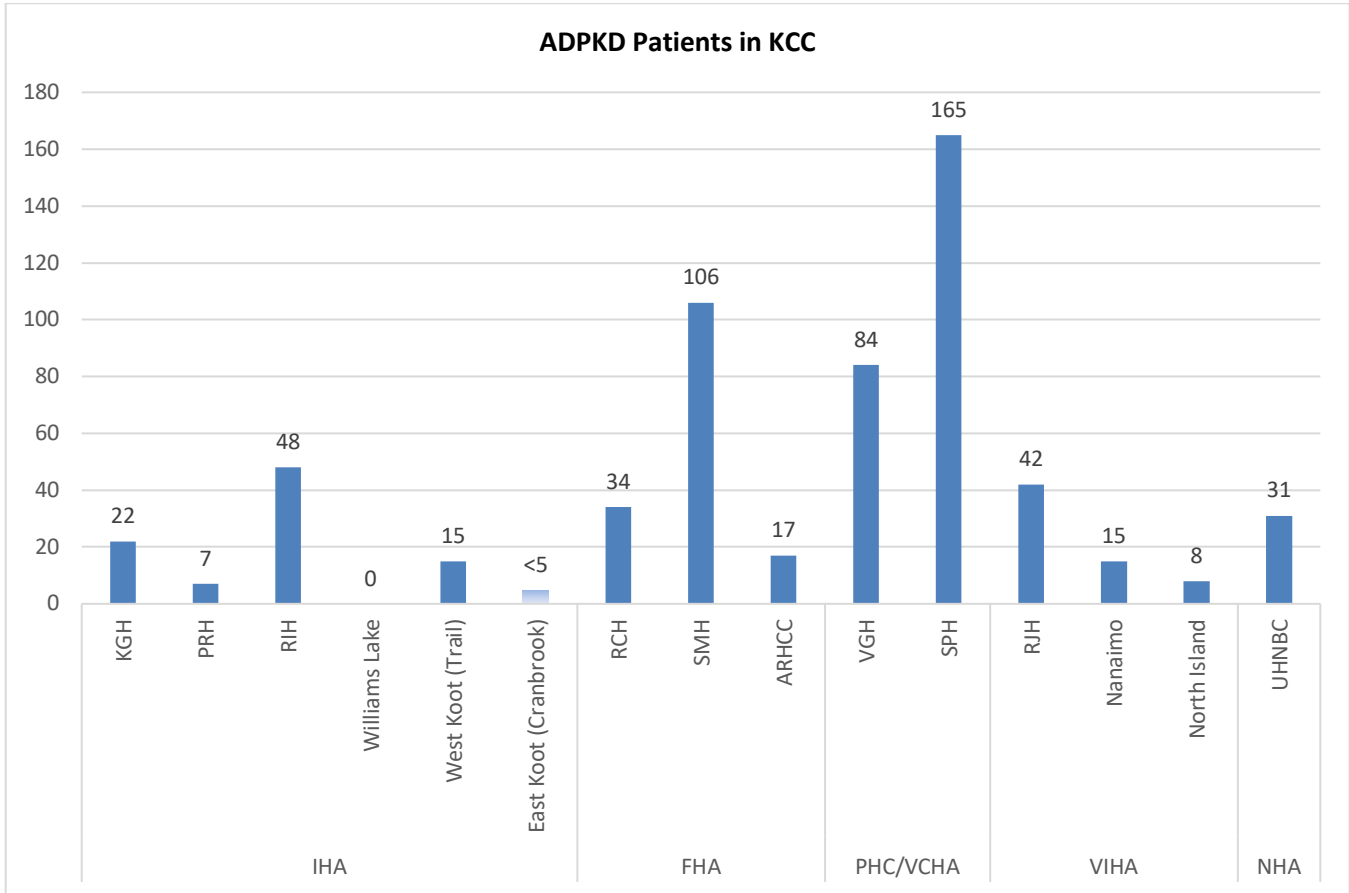
Number/proportion of active patients with ADPKD diagnosis by modality as of 03/31/2023.

	#(%)
Total number of active patients with ADPKD diagnosis in BC as of period end	**1431**
By modality:	
Active patients in pre-ESKD care	997 (70%)
Active patients in KCC care	597(60%)
Active patients whose initial registration occurred in MD office	668 (67%)
Active patients on RRT	434 (30%)

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Number/proportion of active patients with ADPKD diagnosis in pre-ESKD care (KCC or MD offices) and renal replacement therapy (RRT) = dialysis modalities and transplant. Historical data shown from 2015 to 2023:
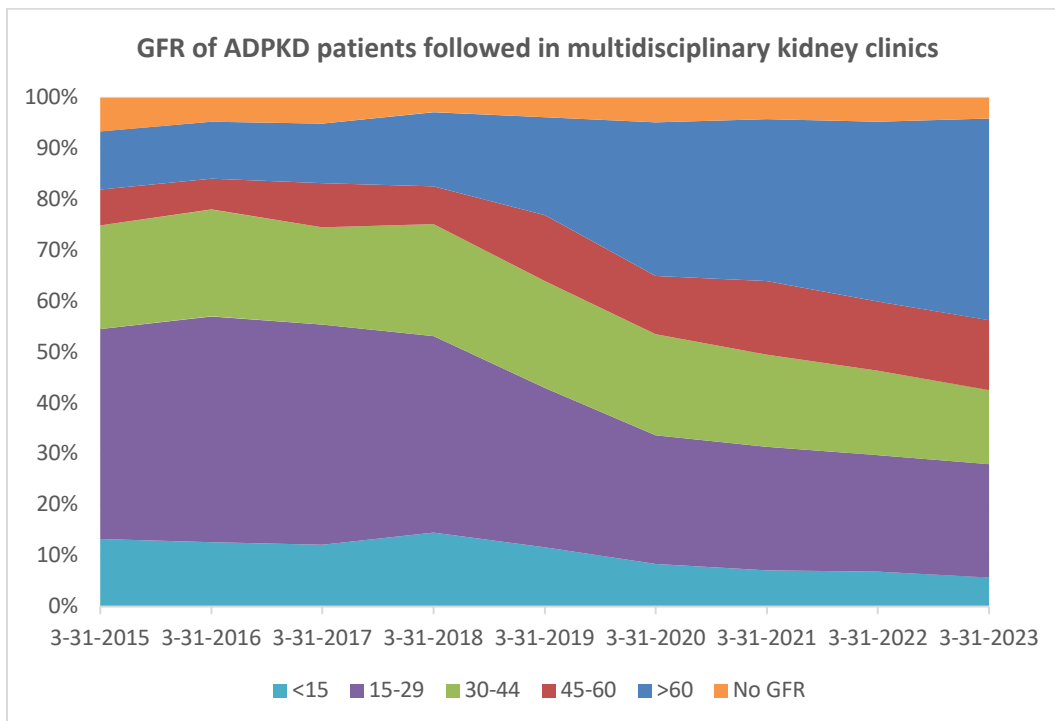

Prevalent Patients with ADPKD Diagnosis by Program and year
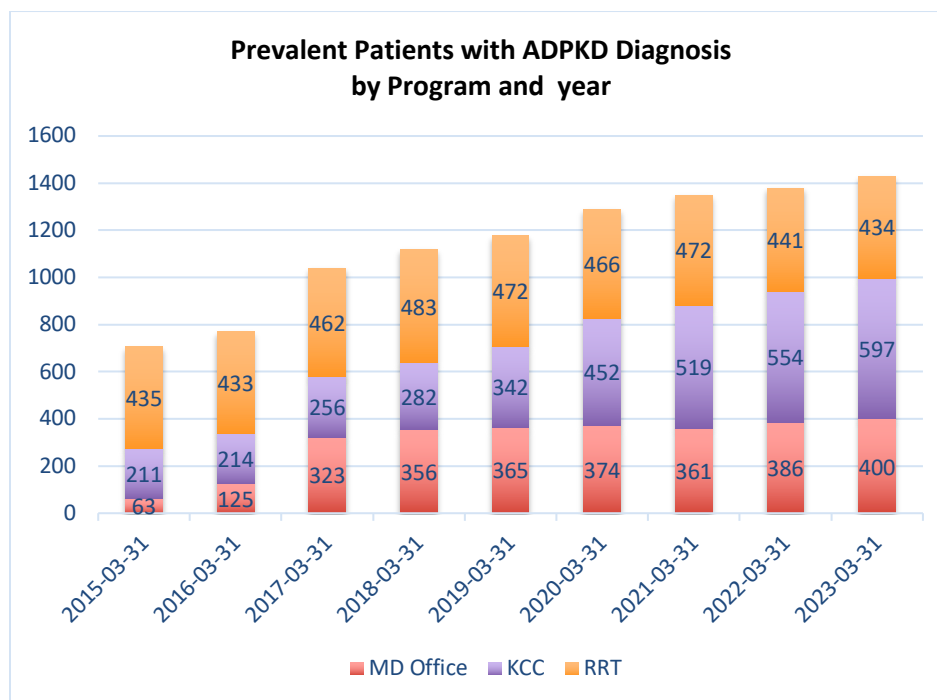
3.2   Clinical Characteristics of KCC patients with ADPKD Diagnosis

	#(%)
Total number of KCC patients with ADPKD diagnosis	**597**
Sex:	
Female	275 (46%)
Male	322 (54%)
Age (years):	
<40	138(23%)
40-54	189(32%)
55-64	130(22%)
65-74	89(15%)
75-84	36(6%)
>=85	15(3%)
Latest GFR (mL/min) during last 12 months:	
>60	237(40%)
45-60	82(14%)
30-44	87(15%)
15-29	133(22%)
<15	34(6%)
No value	24(4%)

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Number/proportion of active patients with ADPKD diagnosis in BC as of March 31, 2023

	#(%)
Total number of active patients with ADPKD diagnosis in BC as of period end	**1431**
Patients with blood pressure measurements during last 12 months	915 (64%)
Mean number of blood pressure measurements per Patient year	3.31
Mean blood pressure systolic/diastolic	131/78
Median systolic (IQR)	130 (120-138)
Median diastolic (IQR)	78 (71-84)
<110/75	3.7%
110/75 – 120/80	15.9%
120/80 – 140/90	52.0%
>140/90	28.4%
Number of Patients with BP measurements who meet HALT-PKD trial criteria (age 15-49, GFR>60)	164 (17.9%)
Proportion with blood pressure <110/75	7 (4.3%)

The following graphs show historical data from 2015 to 2023. These data demonstrate that since inception of the ADPKD Network, there is a trend towards inclusion of greater proportions of younger and higher eGFR patients with ADPKD diagnosis in BC KCCs.


GFR of ADPKD patients followed in multidisciplinary kidney clinics

Age of ADPKD patients followed in multidisciplinary kidney clinics

Legend: <40, 40-54, 55-64, 65-74, 75-84, 85+
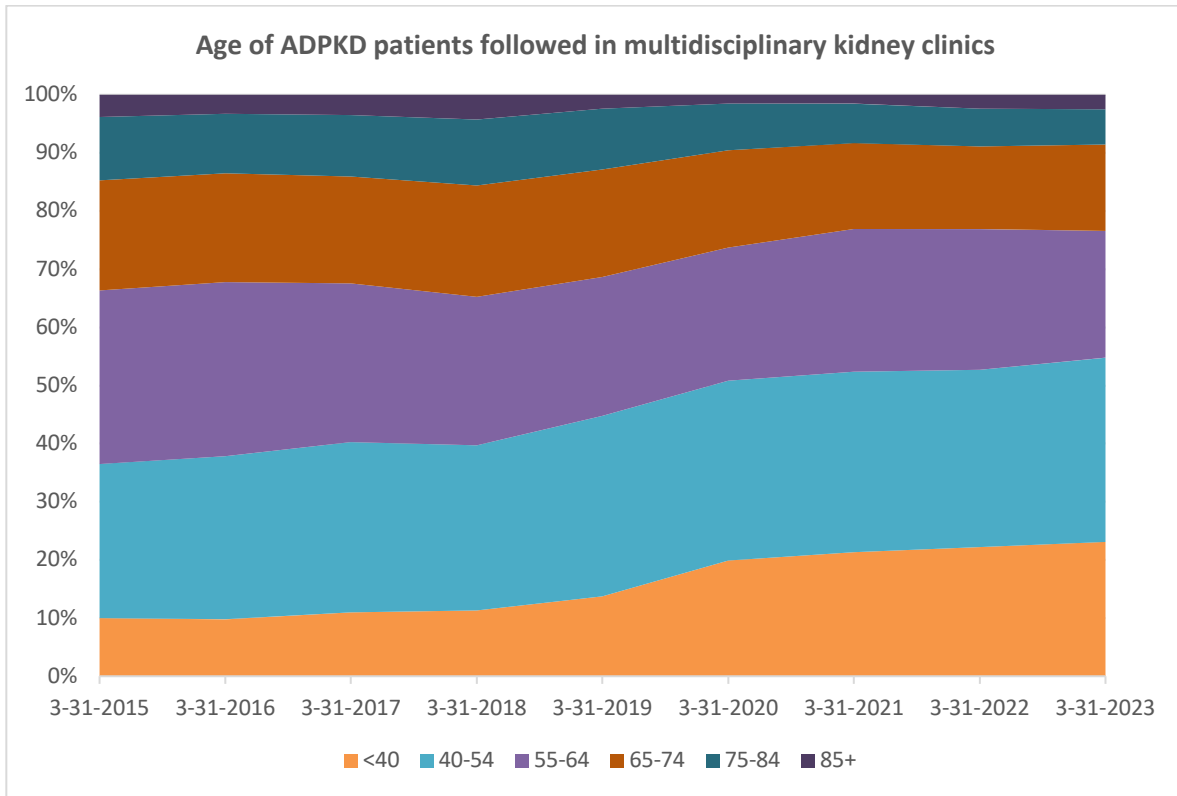
3.3 Trajectory of ADPKD patients after reaching End-Stage Kidney Disease

The following Sankey Plot shows the percentage of incident ADPKD RRT patients by treatment modality at RRT start (0 month) and status at 3 months, 1 year and 2 years post-RRT start. The same is shown for a comparator cohort of provincial renal (PR) patients. Incident RRT patients refer to patients reaching end-stage kidney disease and starting renal replacement therapy (RRT, i.e. Dialysis/transplant) for the first time. This first start is denoted by time 0 and each patient's trajectory is tracked at 3 months, 1 year, and 2 years after first RRT initiation.

Cohorts: ADPKD and PR patients, respectively, who started RRT between April 1, 2015, and March 31, 2021; with follow-up data collected for 2 years on each patient.

ADPKD Patients

PR Patients

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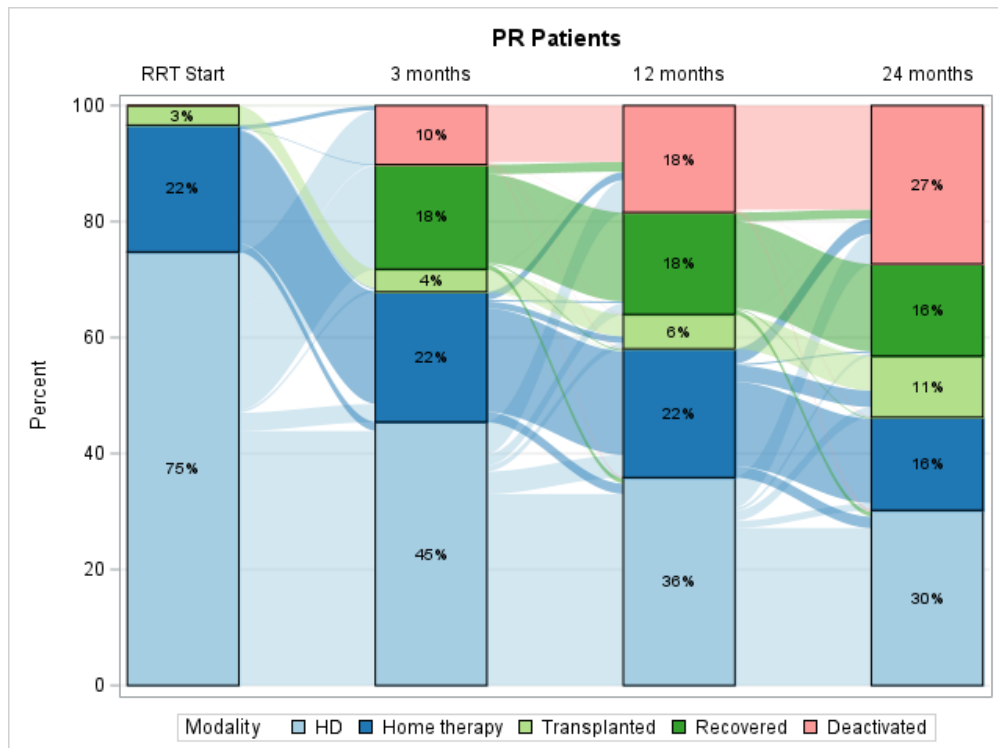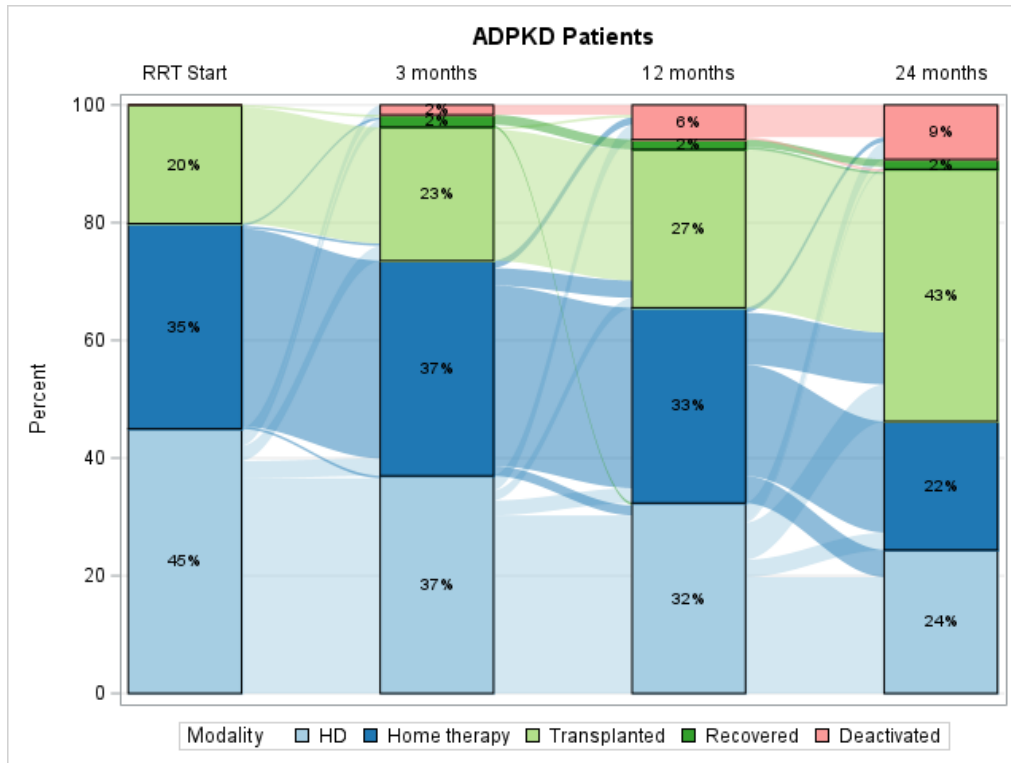
4   Tolvaptan in ADPKD

4.1   Tolvaptan Utilization

Report Period 10/01/2020 to 03/31/2023 for the following tables:

Report Date:	Unit	*10/01/2020-03/31/2021	04/01/2021-03/31/2022	04/01/2022-03/31/2023
Total patients on tolvaptan	#	**125**	**153**	**198**
Number of tolvaptan applications received	#	45	46	75
Number of tolvaptan applications approved	#	39	39	66
% applications approved	%	86.7	84.8	88.0
Criteria for tolvaptan use in ADPKD at initiation as per tolvaptan application form:				
Group A	#	32	38	61
Group B	#	3	1	7
Group C	#	6	5	7
On tolvaptan, but do not meet any of the criteria as per tolvaptan application form	#	0	0	0

*6 months of data available from 10/01/2020 to 03/31/2021

Report Period 04/01/2022 to 03/31/2023 for the following tables:

Number of patients on tolvaptan at any time during report period	198
FHA	69
VCH+PHC	61
VIHA	29
NHA	7
IHA	25
Patients whose primary nephrologist does not belong to any HARP	7

4.2 Clinical Parameters of ADPKD patients treated with Tolvaptan

Report Period 04/01/2022 to 03/31/2023 for the following tables:

Demographics:	Unit	
Total number of patients on tolvaptan	#	**198**
Mean age at start of treatment (SD)	years	45.3 (11.2)
Median continuous duration on treatment (IQR)	months	22.4 (7.4, 30.4)
On treatment for ≤ 3 months	#(%)	22 (11.1)
On treatment for 3-6 months	#(%)	21 (10.6)
On treatment for 6-12 months	#(%)	34 (17.3)
On treatment for 12-18 months	#(%)	12 (6.1)
On treatment for 18+ months	#(%)	109 (55.3)
Height	cm	172.6 (12.8)
eGFR, closest to start of therapy (median, IQR)	mL/min	51.5 (37,76)
Blood pressure within 12 months	mean systolic/diastolic	126/79
% followed by KCC (at time of latest dispense)	#(%)	157 (79)
Dosage (at time of latest dispense):		
15 + 15	#(%)	12(6)
30 + 15	#(%)	5(3)
45 + 15	#(%)	105(53)
60 + 30	#(%)	32(16)
90 + 30	#(%)	42(21)

Monitoring – All patients on tolvaptan within April 2021 to March 2022	Unit	
Total number of patients	#	153
Treatment persistence from time of first start/refill:		
3 months	#(%)	150(98)
6 months		143(93)
12 months		133(87)
Patients who had urine osmolarity test done during report period	#	140
Proportion of patients who had had urine osmolarity result less than 250	#(%)	130(93)
Patients who did not have any AST or ALT labs during monitoring period	#	5
Patients who have had an AST or ALT above normal during report period	#(%)	19(12)
Patients with labs that have higher AST/ALT		
LFTs > 1-3 x ULN	#	18
LFTs > 3-5 x ULN	#	0
LFTs > 5 x ULN	#	1
Out of patients who had lab results >3x ULN, how many were rechallenged and stayed on treatment until report end	#	0
Out of patients who were rechallenged, how many had another >3x abnormal lab result	N/A	N/A

5 Future Topics

Total Kidney Volume (TKV) is currently in the process of being updated; future reports will describe based on that in terms of management outcomes and treatment utilization patterns.