Pain in ADPKD: Results from a National Patient-Powered Registry

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Background

- Kidney growth and gradual function decline impacts health related quality of life (HRQoL) in people with autosomal dominant polycystic kidney disease (ADPKD). To encourage the utilization of patient-reported outcomes in both clinical practice and trial design, the PKD Foundation designed an ADPKD Registry of patient-reported data in the USA.
- **The ADPKD Registry** is a national, online collection of U.S. patient-reported data related to ADPKD, launched in September of 2019. The purpose of the Registry is to **simultaneously facilitate research discoveries while supporting patient-centered outcomes research to learn more about the patient journey and discover unmet needs.**
- The significance and relevance of the Registry design, implementation, and management are maintained through the **activities of Advisory Committees and Working Groups** consisting of clinician scientists, researchers and patient stakeholders. The Registry is hosted on a secure, online platform; participants are registered and consented through the online system.

Methods, IS and PDS

The Registry collects data on individual disease characteristics, demographics, and diagnosis. Participants also periodically report their latest kidney function lab values and complete validated HRQoL standardized instruments (the ADPKD-Pain and Discomfort Scale (PDS)[1] and the ADPKD-Impact Scale (IS[2]) (Table 1). Patient-reported pain is then tracked longitudinally with eGFR-estimated chronic kidney disease (CKD) stage and other characteristics.

Table 1

ADPKD Pain and Discomfort Scale	ADPKD Impact Scale
Measures severity and impact of 3 distinct types of ADPKD-related pain over the last 7 days.	Measures physical, emotional, and fatigue impact of ADPKD on a patient's daily life over the last 2 weeks.
Dull pain – chronic ache Sharp pain – acute, rare Discomfort – chronic fullness and pressure	Physical domain Emotional domain Fatigue domain
Response categories are: 1 = none 2 = mild 3 = moderate 4 = substantial, and 5 = extreme.	Response categories are: 1 = not difficult/bothered 2 = a little difficult/bothered 3 = somewhat difficult/bothered 4 = very difficult/bothered, and 5 = extremely difficult/bothered

ADPKD-IS and ADPKD-PDS contact information and permission to use: Mapi Research Trust, Lyon, France, https://eprovide.mapi-trust.org

Results

Figure 1

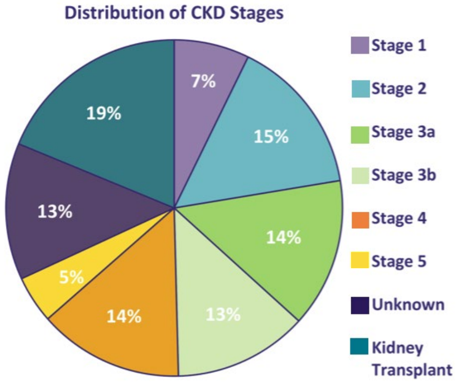
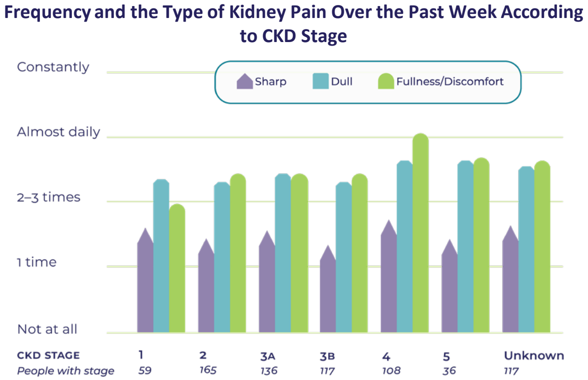

Distribution of CKD Stages

Figure 2


Frequency and the Type of Kidney Pain Over the Past Week According to CKD Stage

By November 2021, 768 ADPKD participants completed at least one PDS. The cohort (median age = 53 years, 72% female) encompasses all stages of CKD, with 19% post-kidney transplant (Figure 1).
On average, CKD Stage 4 participants reported the most-dull kidney pain (28% moderate, 12% substantial pain). Sharp kidney pain was reported more evenly across disease stages but less frequently across the entire cohort (33% sharp pain vs 57% dull pain) (Figure 2).

CKD Stage 5 participants were more likely to report an impact of dull kidney pain on sleep (21%) than other pre-transplant participants (Figure 3).
Pain/discomfort scores (from the PDS) and pain-related questions (from the IS) showed a strong positive correlation (n=763, $p<0.0001$). Participants who report being bothered more by PKD-related pain also report higher pain severity (Figure4).

Figure 3

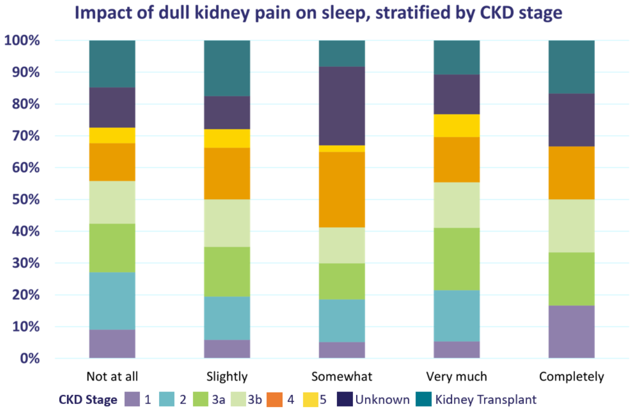
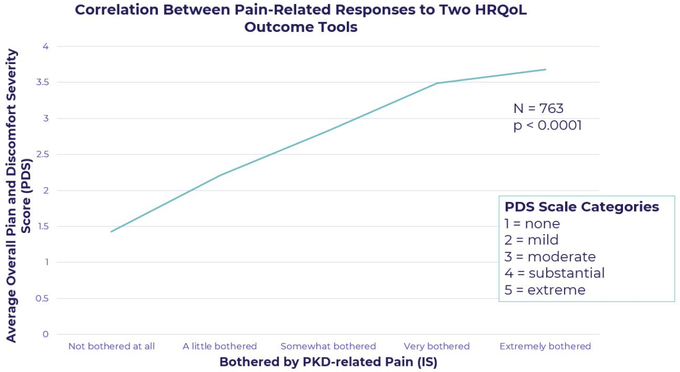

Impact of dull kidney pain on sleep, stratified by CKD stage

Figure 4


Correlation Between Pain-Related Responses to Two HRQoL Outcome Tools

PDS Scale Categories
1 = none
2 = mild
3 = moderate
4 = substantial
5 = extreme

Conclusions

- In launching the Registry, the PKDF aimed to **empower patients** to share their disease experience including the impact of PKD on their HRQoL, and to **increase the opportunities** to participate in research.
- Analysis of the initial cohort **reveals that patients from all CKD stages are willing and eager to contribute** to the body of knowledge in ADPKD.
- These data will contribute to the **increased understanding of pain experience in ADPKD**, including characterization of the patients more likely to report it.

References

1. ADPKD-PDS - Autosomal Dominant Polycystic Kidney Disease Pain and Discomfort Scale. https://eprovide.mapi-trust.org/instruments/autosomal-dominant-polycystic-kidney-disease-pain-and-discomfort-scale) 24 March 2020, date license acquired)
2. Oberdhan D, Cole J, Krasa HB, Cheng R, Czerwiec FS, Hays RD, et al. Development of the Autosomal Dominant Polycystic Kidney Disease Impact Scale: A New Health-Related Quality-of-Life Instrument. Am J Kidney Dis. 2018;71(2):225–35. pmid:29150246