GUIDELINE RECOMMENDATIONS*

a. We recommend that psychosocial issues are reviewed during clinical assessment and that patients should be offered multidisciplinary psychosocial support to address pain management, self-management, social challenges, psychological issues, and education and information (1C).

*Criteria used for recommendations and levels of evidence (1,2;A-D) are described in more detail in Tables 1 and 2 of Rangan G, Savige J. Introduction to the KHA-CARI Guidelines on ADPKD. Semin Nephrol.2015;35:521-3 in this issue.

UNGRADED SUGGESTIONS FOR CLINICAL CARE

Management of Chronic Renal Pain

- Explore and validate patient concerns and frustrations about the unpredictability, volatility, and intensity of chronic renal pain.
- Provide strategies and counseling on how to manage the psychological, emotional, and lifestyle impact of pain; and to limit its interference with lifestyle. Behavioral therapy, peer support, emotional disclosure, and online programs have been found to be effective for managing chronic pain, but no studies of these interventions have been conducted in patients with autosomal dominant polycystic kidney disease (ADPKD).

Self-Management

Refer to the article on Management: Diet and Lifestyle for more detail.

- Develop programs, resources, and individualized plans to equip patients with the capacity and confidence for their own self-care (including medicine-taking, lifestyle changes [diet, fluid intake, physical activity], self-monitoring, access to health care services), and to reduce the perceived treatment burden. Systematic reviews of self-management interventions in other chronic diseases suggest that they can improve knowledge and self-management behavior.

Social Work

- Provide support to address potential or actual financial discrimination related to disclosure of the genetic test results and/or patient’s diagnosis of ADPKD. Specific financial issues that have been identified in the literature and observed in clinical practice include: employment, obtaining personal insurance, applying for loans.
including mortgages, and additional expenses for medications.

**Psychological Support**
- Refer patients who may be at high risk for or have indicators for depression and anxiety to psychological services.
- Identify ways to alleviate prognostic uncertainty regarding renal disease and extrarenal complications.
- Address body image and self-esteem.
- Patient support groups are also suggested for patients to reduce their sense of isolation and to learn practical and coping strategies from other patients with ADPKD.

**Education and Information for Patients and Providers**
Refer to the article on Management: Diet and Lifestyle for more detail.

- Provide patients and their families with comprehensive, comprehensible, and practical information about ADPKD, disease complications and prognosis, self-management and monitoring, medications, and dietary and fluid intake.
- Provide succinct information (eg, printed leaflet) about ADPKD that is endorsed by experts and suitable for members of the public (eg, employers, insurers, educational institutions).

**Genetic Screening and Testing**
Refer to the article on Genetics and Genetic Counseling and Screening for Polycystic Kidney Disease for more detail.

- Provide counseling to address self-blame and guilt because of genetic transmission.
- Address family planning.
- Discuss issues around genetic testing and disclosure.

**IMPLEMENTATION AND AUDIT**
Patients with ADPKD should have access to multidisciplinary services to receive psychosocial assessment and management.

**BACKGROUND**
Autosomal dominant polycystic kidney disease is a life-threatening genetic disease with serious complications and symptoms including enlarged kidneys, end-stage kidney disease, intracranial aneurysm, pain, and infection. Constant medical monitoring, medications, and dependence on kidney replacement therapy impose a significant treatment burden. Consequently, these can have a detrimental impact on the quality of life and psychosocial and social outcomes in patients with ADPKD.13–15

A systematic review on patient perspectives of living with ADPKD found that the erratic onset and intensity of pain disrupted daily living and prevented patients from developing long-term career and family goals. They experienced persisting uncertainties including perceived ambiguities surrounding the meaning and implications of their diagnosis, disempowerment in self-management, inability to plan ahead, and financial discrimination.16 A recent KDIGO Controversies conference identified additional issues such as impaired body image, relationship strain, and limited participation in recreation and sport.17 Patients with ADPKD may also experience fatigue.18

Depression and anxiety have also been reported in patients with ADPKD. In one study, 38 patients with ADPKD completed the Beck Depression Index and 61% were found to have depression.19 Anxiety and depression were associated with lower education, female gender, and unmarried individuals.19

The aim of this guideline was to assess and summarize the evidence on psychosocial care for patients with ADPKD. This includes psychosocial assessment, psychological support, cognitive and behavioral training, social work, information and educational programs or resources, and support groups. The psychosocial aspects relating to genetic counseling are covered in the guideline subtopic Genetics and Genetic Counseling and Screening for Polycystic Kidney Disease.

**SEARCH STRATEGY**

**Databases Searched**
Medical subject headings and text words for ADPKD were combined with medical subject headings terms and text words relating to psychosocial assessment and management (psychological, social, cognitive and/or behavioral training, patient education and information, support groups). The search was carried out in Ovid MEDLINE (1946 to November 2014), Embase (1974 to November 2014), PsycINFO (1806 to November 2014), and the Cochrane Database of Systematic Reviews and the Cochrane Registry of Clinical Trials (inception to November 2014).

**Date of search:** November 2014.

**WHAT IS THE EVIDENCE?**
Supplementary evidence Tables are available online.
Interventional Studies

There are no randomized controlled trials that assess the effectiveness of psychosocial interventions specifically for patients with ADPKD.

Quality-of-Life Studies

Only a few studies have assessed quality of life (QoL) in patients with ADPKD. Abdominal distension, sleep disturbances, and pain were found to impair overall QoL. Miskulin et al, in 2014, conducted a cross-sectional study involving 1,043 patients with ADPKD. Patients with lower estimated glomerular filtration rates (GFRs) of 20 to 44 mL/min/1.73 m² were significantly more likely to report that pain impacted on their daily lives and had lower QoL (Short-Form 36 [SF-36]) scores than patients with estimated GFRs of 45 to 60 and 60 or more mL/min/1.73 m².

Rizk et al, in 2009, prospectively assessed QoL in a cohort of non–dialysis-dependent patients with ADPKD (n = 152) using the SF-36. Patients with GFRs less than 80 mL/min/1.73 m² had lower physical component summary scores compared with patients with GFRs of 80 mL/min/1.73 m² or greater.

Suwabe et al, in 2013, assessed quality of life in a cross-sectional study (n = 219) and found that SF-36 scores were significant lower compared with the general population in Japan. Abdominal distension, pain, sleep disturbance, heartburn, fever, gross hematuria, and anorexia were identified to influence QoL.

Qualitative Studies

A recent systematic review identified 21 qualitative studies that reported the perspectives and experiences of 247 patients living with ADPKD. Five themes were identified: unvalidated pain (medical trivialization, inadequacy of pain management); persisting uncertainties and ambiguities (lacking diagnostic clarity, disempowerment in self-care, unpredictable daily disruptions, inability to plan ahead, financial discrimination); genetic guilt and resentment (blaming parents, self-blame, constant burden of guilt); precariousness in pursuing parenthood (prognostic uncertainty, owning the decision, needing directive counseling); and defining parental responsibility for genetic testing and disclosure (preserving normality, doubting necessity of genetic testing, respecting the child’s autonomy, hope in future technologies, facilitating preparedness).

KHA-CARI Consumer Workshop

In August 2014, patients and caregivers (n = 18) contributed to a KHA-CARI consumer consultation workshop. Participants were asked to discuss their experiences of living with ADPKD, and to identify topics, interventions, and outcomes they considered important and relevant for guidelines on ADPKD. The participants reiterated the importance of mental health assessments (to diagnose and treat depression and anxiety), supportive clinician-patient communication, patient cognition and ability to process information, and peer support. They emphasized the need for comprehensive, comprehensible, and practical information, particularly to address the following questions:

- What is the definition of ADPKD?
- What is the prognosis for an individual patient with ADPKD?
- How can patients recognize the signs and symptoms of ADPKD?
- How can patients self-manage the disease (eg, patient-friendly tool using algorithms or flowcharts)?
- What are the links between ADPKD and other extrarenal complications?
- What are the side effects of medication?
- What are the dietary and fluid intake recommendations for patients with ADPKD?

SUMMARY OF THE EVIDENCE

No studies have evaluated ADPKD-specific psychosocial or educational interventions. Quality-of-life assessments in this population are sparse. Findings from qualitative studies indicate a need to develop and evaluate interventions that address pain management, empowerment for self-care, prognostic uncertainty, and financial discrimination.

WHAT DO THE OTHER GUIDELINES SAY?

Kidney Disease Outcomes Quality Initiative: No recommendation.

UK Renal Association: No recommendation.

Canadian Society of Nephrology: No recommendation.

European Best Practice Guidelines: No recommendation.

International Guidelines: No recommendation.

National Institute for Health and Care Excellence (NICE):

The UK NICE clinical guideline on early identification and management of chronic kidney disease (CKD) in adults in primary and secondary care provides general recommendations addressing psychosocial care in CKD, however, no recommendations were specific to ADPKD. The guideline provides suggestions on
patient information and education, which are summarized in the following:

- Offer patients education and information tailored to the severity and cause of CKD, the associated complications and risk of progression;
- Involve patients with CKD in the development of information and education programs;
- Offer high-quality information or education programs as appropriate to the severity of their condition to allow time for them to understand and make informed choices about treatment;
- Health care providers providing information and education should have specialist knowledge about CKD and the skills to facilitate learning;
- Health professionals need to take account of the psychological aspects of coping with CKD;
- Offer access to relevant support (support groups, counseling, specialist nurse).

**KDIGO Conference Executive Summary**\(^7\): The KDIGO Conference on ADPKD (January 2014) identified key psychosocial issues including: psychological help to address diagnosis, asymptomatic nature of disease, slow deterioration in quality of life, impact on body image, relationships, and recreation and sport. The working group identified challenges in identifying factual and unbiased support via health care professionals, internet, family members, and support groups. They suggested the provision of integrated (coordinated and interdisciplinary) care for patients with PKD.

**SUGGESTIONS FOR FUTURE RESEARCH**

- Psychosocial interventions targeting the priorities and needs in the ADPKD population need to be developed and evaluated. Patients with ADPKD should be involved in the development of psychosocial interventions, as well as to identify the outcomes that are relevant and important from their perspective.
- Generic quality-of-life instruments (SF-36) do not capture many of the issues relevant in ADPKD. Therefore, we suggest the development of quality-of-life instruments that include the specific concerns identified by patients with ADPKD. Randomized controlled trials could then be conducted to assess the effectiveness of the psychosocial intervention compared with standard care.

**APPENDIX A. SUPPORTING INFORMATION**

Supplementary data associated with this article can be found in the online version at [http://dx.doi.org/10.1016/j.semnephrol.2015.10.010](http://dx.doi.org/10.1016/j.semnephrol.2015.10.010).

**REFERENCES**


### PSYCHOSOCIAL CARE

Date written: March 2015  
Author: Allison Tong, Andrew Mallet, Pamela Lopez-Vargas, Gopala K. Rangan

<table>
<thead>
<tr>
<th>Study ID (author, year)</th>
<th>N</th>
<th>Study Design</th>
<th>Setting</th>
<th>Participants</th>
<th>Intervention (experimental group)</th>
<th>Intervention (control group)</th>
<th>Primary outcomes</th>
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<tbody>
<tr>
<td>Tong et al. (2015)[16]</td>
<td>21</td>
<td>Qualitative systematic review</td>
<td>Multi-center studies</td>
<td>Review of qualitative studies involving patients with ADPKD, 18 years of age or older in any stage of CKD (1-5, 5D or 5T)</td>
<td>Thematic synthesis of qualitative studies as defined by the Enhancing Transparency of Reporting the Synthesis of Qualitative research (ENTREQ).</td>
<td>Not applicable</td>
<td>Perspectives and experiences of living with ADPKD – symptoms, complications, management and genetic testing.</td>
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<tr>
<td>Miskulin et al. (2014)[11]</td>
<td>1,043</td>
<td>Randomized, double-blind, placebo-controlled (HALT PKD trials)</td>
<td>Multicentre, USA</td>
<td>Patients with ADPKD, hypertension and baseline estimated GFR (eGFR) &gt;20 mL/min/1.73m² were included.</td>
<td>Completed self-administered questionnaires 36-Item Short Form Health Survey (SF-36) and Wisconsin Survey during baseline</td>
<td>Completed self-administered questionnaires SF-36 and Wisconsin Survey during baseline. Some were also compared to the general population.</td>
<td>Quality of life - Wisconsin Brief Pain Survey</td>
</tr>
<tr>
<td>Suwabe et al. (2013)[14]</td>
<td>219</td>
<td>Cross-sectional study</td>
<td>Single centre, Japan</td>
<td>Pre-dialysis and dialysis patients with ADPKD over 20 years of age</td>
<td>Completed the self-administered 36-Item Short Form questionnaire (SF-36) and a 12-Item questionnaire about specific symptoms which was designed for the study. Patients also underwent transcatheater arterial embolization (TAE) and abdominal computed tomography (CT) or magnetic resonance imaging (MRI) was performed.</td>
<td>Compared with the general Japanese population.</td>
<td>Quality of life (QoL) - Physical component summary score (PCS) - Mental component summary score (MCS) - Quality of life (QoL) - Role/social component summary score (RCS).</td>
</tr>
<tr>
<td>Rizk et al. (2009)[12]</td>
<td>152</td>
<td>Prospective observational cohort study</td>
<td>Multi-center, USA</td>
<td>Pre-dialysis patients older than 18 years of age with ADPKD</td>
<td>Completed the self-administered 36-Item Short Form questionnaire (SF-36)</td>
<td>Compared to the general population.</td>
<td>Physical component summary (PCS) score - Mental component summary (MCS) score.</td>
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Table 2. Risk of bias – Randomised controlled studies - quality appraisal tool EPOC

<table>
<thead>
<tr>
<th>Study ID (author, year)</th>
<th>Type of study</th>
<th>Random sequence</th>
<th>Allocation concealment</th>
<th>Blinding of participants &amp; personnel</th>
<th>Baseline similarities</th>
<th>Outcome assessment</th>
<th>Reporting bias</th>
<th>Selective reporting</th>
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</tr>
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<tbody>
<tr>
<td>Miskulin et al. (2014)[11]</td>
<td>Randomised, double-blind, placebo-controlled (HALT PKD trials)</td>
<td>Low</td>
<td>Low</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Low</td>
<td>Participants were only assessed at baseline, there was no follow-up for this study. Participants belong to related but different HALT trials.</td>
</tr>
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</table>

Table 3. Risk of bias - qualitative review – quality appraisal tool ENTREQ

<table>
<thead>
<tr>
<th>Study ID (author, year)</th>
<th>Synthesis methodology</th>
<th>Inclusion criteria, data sources, search strategy and screening methods</th>
<th>Study characteristics including list of studies</th>
<th>Rationale for appraisal, appraisal tools used, appraisal process and study quality</th>
<th>Process used, software used, number of reviewer, coding and study comparison</th>
<th>Inductive or deductive</th>
<th>Quotes provided</th>
<th>Adequateness of summary, results</th>
<th>Conflict of interest</th>
<th>Quality</th>
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</thead>
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<tr>
<td>Tong et al. (2015)[16]</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Inductive</td>
<td>Yes</td>
<td>Relevant</td>
<td>Mentioned</td>
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</table>
### Table 4. Risk of bias - Other studies

<table>
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<th>Study ID (author, year)</th>
<th>N</th>
<th>Study type</th>
<th>Quality</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Risk et al. (2009)[12]</td>
<td>152</td>
<td>Observational study</td>
<td>Very low</td>
<td>No quality appraisal tool available; small sample size, short term follow-up</td>
</tr>
<tr>
<td>Suwabe et al. (2013)[14]</td>
<td>219</td>
<td>Cross-sectional study</td>
<td>Very low</td>
<td>No quality appraisal tool available; small sample size</td>
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</table>

### Table 5. ADPKD symptoms, complications outcomes

<table>
<thead>
<tr>
<th>Study ID</th>
<th>N</th>
<th>Study type</th>
<th>Intervention</th>
<th>Outcome</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tong et al. (2015)[16] (n=247)</td>
<td>21 studies</td>
<td>Qualitative systematic review</td>
<td>Thematic synthesis of qualitative studies as defined by the Enhancing Transparency of Reporting the Synthesis of Qualitative research (ENTREQ).</td>
<td>Perspectives and experiences of living with ADPKD - symptoms, complications, management and genetic testing.</td>
<td>Five themes were identified: 1. Unvalidated pain - pain was described as intense and unpredictable and sometimes the severity and magnitude of pain was not recognized by their physician. Persistent and unresolved pain also caused frustration for patients. 2. Persisting uncertainties and ambiguities - asymptomatic patients found it difficult to understand their diagnosis and some were in denial. While others proactively searched for more information. 3. Genetic guilt and resentment - some patients blamed their parents, others blamed themselves for passing on the condition. 4. Precariousness in pursuing parenthood - some patients refused genetic testing for fear of having to terminate a pregnancy if the foetus tests positive for PKD. Others requested genetic counseling to inform their decision. 5. Defining parental responsibility for genetic testing and disclosure - patients had varying views about this. Some did not believe it was necessary to screen for PKD in their children as they felt the harms outweighed the benefits of knowing. Others saw this as an opportunity to promote awareness and as an opportunity to prepare their child to cope and plan for the future.</td>
</tr>
<tr>
<td>Study ID</td>
<td>N</td>
<td>Study type</td>
<td>Intervention</td>
<td>Control</td>
<td>Outcome</td>
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<td>Completed self-administered questionnaires SF-36 and Wisconsin Survey during baseline</td>
<td>36-item Short Form Health Survey (SF-36)</td>
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<tr>
<td>et al. (2014)[11]</td>
<td></td>
<td>placebo-controlled (HALT PKD trials)</td>
<td>(SF-36) and Wisconsin Survey during baseline</td>
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<tr>
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</table>
| Suwabe et al. (2013)[14] | 219 | Cross-sectional study              | Quality of life (QoL) - Role/social component summary score (RCS) | Quality of life (QoL) - Role/social component summary score (RCS) | - RCS was significantly influenced by serum albumin B = 0.247 P = 0.001
  Comment: several symptoms affected QoL: these included abdominal distension, pain, sleep disturbance, heartburn, fever, gross hematuria and anorexia. |                                                                  |         |
| Rüks et al. (2009)[12]    | 152 | Prospective observational cohort study | Completed the self-administered 36-item Short Form questionnaire (SF-36) | Compared to the general population | - The mean PCS score was 46.9±11.3, similar to the general population
  - Patients taking pain medication had lower PCS scores than those not on medication 39.2±10.6 versus 47.5±11.2, P<0.05 respectively
  - Physical function index and bodily pain index scores were also lower for patients taking pain medication versus patients not taking pain medication: 61.8±27.4 vs 44.8±22.5, P<0.005 and 50.6±22.2 vs 76.2±24.7, P<0.005, respectively.
  - PCS scores were greater for patients with eGFR ≥80 mL/min/1.73m² 50.3±9.7 compared to those with eGFR <80 mL/min/1.73m² 45.1±9.7, P=0.01.  
  Comment: age, BMI, education level, pulse pressure and pain medication use independently associate with PCS score and account for 32% of the variability of the measurement (R²). Strength of the association (R) between predictor variables and PCS score was 0.56. |                                                                  | Very low |

| Mental component summary (MCS) score | The mean MCS score was 51.0±9.0 similar to the general population | Very low |
