More than a kidney disease: a patient-centred approach to improving care in autosomal dominant polycystic kidney disease

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Quality of life surveys in large observational studies such as DOPPS [1] have contributed significantly to our understanding of the impact of living with end-stage renal disease (ESRD). However, the impact of chronic kidney disease (CKD) on quality of life is less well understood. As the commonest hereditary renal disease, with clinical manifestations ranging from asymptomatic to severe progressive CKD, ESRD and death, autosomal dominant polycystic kidney disease (ADPKD) presents a complex but not unique challenge to clinicians. The systematic review by Tong and colleagues highlights the impact of symptoms on quality of life in ADPKD and the all too common gap between the experience of people living with the disease and the services and support available to them. This is compounded by the, until now, lack of a disease-specific treatment for ADPKD. The publication of the TEMPO 3:4 trial analysing tolvaptan treatment presents a promising development in research in the management of ADPKD. However, the continued imperative to develop treatments to retard disease progression should not make us lose sight of the need to better understand its clinical, psychological and social consequences.

As a symptom, pain is frequently reported in ADPKD, yet as this and other surveys conducted by patient organizations show, whilst a significant proportion of patients suffer pain, many feel management of that pain is inadequate. A recent review of pain in ADPKD suggested a stepwise approach towards management [2], outlining the evidence and rationale for pharmacological, non-pharmacological and more invasive methods of pain control. This is both a timely reminder of the importance of symptom relief in a condition with no specific therapies, and a valuable starting point for a more holistic approach in which clinicians, patients and carers can act in partnership in decision-making about care. Whilst the evidence base on the ‘if’ and ‘how’ of self-management of pain is mixed, there is some evidence from other conditions that empowering patients to move from being passive recipients to active participants in the management of their condition permits a more successful outcome [3]. Self-management interventions in arthritis care have been shown to improve pain and quality of life [4], whilst a meta-analysis of self-management education in children with asthma found improved lung function and fewer attendances to the emergency department [5].

In other areas of renal care, as well as in other chronic conditions, it has long been recognized that holistic care requires a broad, inclusive and multidisciplinary approach. The diversity of health beliefs, experiences, symptoms and consequences for those with ADPKD requires care approaches tailored to individual needs. Learning from what we already know is the first step towards a more patient-centred approach to care. Self-care and self-management are central to the management of diabetes, and have also been used effectively in conditions as diverse as hypertension, heart failure and inflammatory bowel disease. A review of self-management strategies in 2011 identified provision of written information, support including care planning and follow-up, behaviour change coaching, and self-monitoring as having evidence to support their use in a range of settings [6]. There are models of specialist multidisciplinary services in renal care—of note, conservative care for ESRD, transition clinics at the interface of paediatric and adult nephrology, multidisciplinary care in pregnancy and multisystem disorders such as lupus. The development of peer support programmes has also been shown to be a useful adjunct to traditional models of care [7]. In the UK, the new national guidelines for health emphasize the importance of patient-reported outcomes, quality of life and the experience of care as being at the heart of outcomes measurement [8]. National Voices, a coalition of health and social care charities in England, has launched ‘Person-centred Care 2010’, calling on the UK government to make person-centred care the central ambition for health reform [9]. In the USA, national standards exist for diabetes self-management, and the chronic disease
self-management programme (CDSMP), devised at Stanford University, has shown improvements in self-reported health and hospitalizations in a range of settings [10, 11]. This is particularly relevant in long-term conditions and chronic pain management, where a complete cure may not be possible, and patient health and well-being are key.

Along with pain, other themes identified by the authors in their analysis relate to prognostic uncertainties, genetic ‘guilt’, genetic testing, disclosure and family planning. The authors acknowledged that this is a complex area. Several experiences and expectations described by patients—such as disempowerment in self-care and financial uncertainty—are consistent, whilst others highlight that individual needs, expectations and opinions about the role of healthcare professionals may vary. This is particularly true in decision-making around pursuing parenthood. Some described having children as a personal decision, whilst others sought directive counselling from their physician on whether to do so. This clearly requires a nuanced approach, where careful exploration of individual needs by healthcare professionals and agenda-setting by patients can enable truly personalized care. It also emphasizes that maintaining a patient-centred approach requires access to continuous education and resources for patients, to enable fully informed decision-making at every stage. Genetic testing of children and disclosure causes distress. Research has shown that families who share a family narrative about their genetic condition appear to be more resilient and adaptable [12], and the authors suggest that decision support tools should be a priority to improve patient-centred outcomes. Learning from experience in and research done into other hereditary conditions such as Huntington’s disease, cystic fibrosis, and hereditary breast and ovarian cancers, the process of genetic counselling is a multistep one. It should be nondirective, with attention paid to individual and family value systems. It encompasses not only diagnosis, but also education, support and follow-up.

Whilst there are currently no formal international standards, recently there has been progress towards developing international guidelines on ADPKD, with a conference held by KDIGO in Edinburgh in 2014 to discuss controversies and evaluate the evidence base [13]. Consequently, at present the format and availability of services for ADPKD varies. In the developed world the majority of care is provided in general nephrology, pre-dialysis and RRT clinics, whilst some is provided in specialist genetics clinics, which may also be involved in research and offer genetic counselling, with social, emotional and family support existing in the form of charitable organisations. Central to empowering ADPKD patients and carers is understanding that it is a systemic disease that may require the involvement of multiple specialists. Recognizing this, and involving patients and carers in the design of services so that these better reflect their needs, perhaps within or with access to specialized centres, may further accelerate and improve our understanding. As an example, the European ADPKD Forum Brussels Declaration calls for the development of a network of European ADPKD reference centres [14]. Rapid advances in genetics, diagnostics and technology may raise further complex ethical considerations in the future, where the existence of a framework within which healthcare professionals, patients and carers can consider these questions would be invaluable. More importantly, in a condition with such a spectrum of natural histories, developing tools to deliver personalized care—the heart of a patient-centred approach to health—should be an essential and achievable goal.

Learning to understand the needs of patients living with chronic disease is part of the evolution of the doctor–patient relationship, as healthcare evolves from a disease-focused to a patient-centred paradigm. Partnering with patients to enable them to take control of their own health and well-being requires effective communication, adequate information, access to support services and specialist advice. Perhaps most importantly, it is about reframing traditional models of communication and learning to truly listen to patients.

CONFLICTS OF INTEREST STATEMENT

None declared.

(See related article by Tong et al. A painful inheritance—patient perspectives on living with polycystic kidney disease: thematic synthesis of qualitative research. Nephrol Dial Transplant 2015; 30: 790–800.)

REFERENCES

Interarm blood pressure difference: more than an epiphenomenon

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Differences in blood pressure measurements between arms are common; typically a systolic difference ≥10 mmHg is detected in 4.4% of subjects in unselected cohorts free of vascular disease, but prevalence rises to 7.0% in diabetes and 13.6% in the presence of hypertension [1]. Current hypertension guidelines therefore advocate checking both arms, and using the higher reading arm for therapeutic decisions [2].

Inter-arm differences are associated with increased cardiovascular and all-cause mortality [3]. This association is observed in cohorts without pre-existing cardiovascular disease [4–7], and also in selected cohorts with hypertension, diabetes or established cerebrovascular disease [8–10]. Further cross-sectional studies have demonstrated associations of systolic inter-arm differences with peripheral arterial disease [11–15], increased carotid intima-media thickness [12], higher coronary artery calcium scores [12] and presence of left ventricular hypertrophy [11]. These findings have led to the recognition of inter-arm blood pressure difference as a potential risk marker for cardiovascular disease.

Chronic kidney disease (CKD) is also a recognized cardiovascular risk marker [16]. In the context of diabetes, inter-arm blood pressure differences have been associated with the presence of chronic kidney disease and albuminuria [17]. Evidence for the association of inter-arm disease and prognosis in chronic kidney disease is limited to one previous cohort study of subjects pooled from general medical and renal clinics, which found incremental increases in all-cause mortality for each 10 mmHg increase in systolic inter-arm difference, with a strong additional mortality risk conferred by chronic kidney disease at any level of inter-arm difference [18]. The linked study of Quiroga et al. in this issue provides a new insight into this relationship. They present data from 652 hypertensive subjects with relatively early (Stage 1–3) CKD followed up for a mean of 19 months. Inter-arm difference was measured more accurately by using the mean of the second and third pairs of readings from two simultaneously activated automated sphygmomanometers. Based on these readings, the authors reported high prevalences of 28 and 15% for systolic inter-arm differences ≥10 and ≥15 mmHg, respectively. After adjusting for age, gender, history of cardiovascular disease, diastolic blood pressure and use of anti-hypertensive medication, the authors found that inter-arm differences ≥10 and ≥15 mmHg conferred similar excess risks of 80 and 86%, respectively, for cardiovascular events. These findings provide the first prospective evidence for increased cardiovascular events in renal disease associated with simultaneously measured inter-arm blood pressure difference. There are, however, questions left to answer.

Simultaneous measurement methods, as used in this study, are preferred in epidemiological study as they avoid overestimation of inter-arm difference, which may in part be due to white coat effects, as well as short-term blood pressure variability [19, 20]. However, the reported prevalences of inter-arm difference in this study exceed most published estimates.