Counseling and Diagnostic Implications

Patient Working Group

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Disclosure of Interests

No relevant disclosures
ADPKD treatment and counseling challenges for doctors / nephrologists

- If diagnosed at 20, patient might reach ESRD:
  - In <10 years
  - 30-40 years (average)
  - Never

- Known impact of lifestyle choices is limited
  - Unlike with diabetes or hypertension

- Rapid increase in knowledge and potential treatments decreases certainty that future outcomes will mirror the past
  - There is a case to be made for increased hope, but how much?

When you consider these major factors, how can patients world-wide expect consistent treatment and counseling from their doctor?

One area we will explore is the potential benefit and content of checklists for doctors and patients.
The “IF / THEN / ELSE” approach
- not recommended

Tell patient they have PKD

Patient is determined to beat the disease
Provide pamphlet and PKDcure.org website

Patient is upset
Pat patient on back and say, “There, There”

Patient wants more active care than you can provide
Educate self about PKD
Refer them to PKD specialist

It works! Move on to next patient

Patient is still upset
Prescribe Zoloft
Refer to PKD Support group
Experiences at first diagnosis

• If the patient has PKD in their family, they may be prepared for the diagnosis, but it will be devastating nonetheless
  – They will relate their condition to older relatives who have suffered and perhaps passed away early from PKD
    • Feelings of hopelessness, resignation, depression are common
• If the patient does not have PKD in their family (mutation), this will likely be the first time they will confront their mortality
  – Confusion, panic, desperate seeking of advice should be expected, both immediately and later
  – Denial is common, and not in the patient’s best interest
• Patients will have a large number of informational needs
  – Family planning
  – Nutrition & Exercise
  – Medication
  – Financial (health and life insurance, career, longevity-related decisions)
  – Sexuality concerns (physical and emotional)
  – Health outcome scenarios
    • Dialysis/transplant
    • Related conditions such as aneurysms, blood pressure effects/stroke/vascular, PLD, etc
Key Questions to resolve (in order of priority)

**First Diagnosis**

1. What should a doctor tell or give to a patient at first diagnosis?
   - What reactions should the doctor be prepared for?
   - Would checklists be helpful? Content?
   - Side topic: Should the PKD patient be classified differently from the current CKD classification by eGFR stage?
2. What are all the issues related to family planning decisions?
   – Consider ethical, moral, legal, financial, and religious perspectives
   – Include pregnancy: enlarging uterus, BP control and obstetric considerations during term
3. What is the best treatment plan (in all stages) for pain management?

- Exercises
- Meditation / Relaxation techniques
- Over the Counter medications
- “Natural” remedies
- Prescription Medication
- Surgery
- etc
Children

4. How and when do I talk to my children about PKD?
   – When should they be tested?
   – What do I tell them to do lifestyle-wise?
   – What are the impacts to adolescent quality of life?
   – PKD discussions with unaffected family members?

– Side topic: Should a kidney growth chart for pediatrics be created?

Caution is advised in handling the screening of children <18 that in some countries is not advised if not illegal.
5. What are the possible PKD financial impacts?
   – Career progress
   – Potentially reduced income
   – Life and health insurance
   – Long-term care
   – etc

   – Are these impacts barriers to diagnosis and early treatment? If so, what can be done to change these?
6. What practical advice and psychological support should be given to patients regarding aneurysm screening and follow-up?

- Screening recommended at diagnosis?
  - Criteria for follow-up screening?
  - MRI or CT?
7. What should patients be told about the presence of cysts in other organs than the kidneys and liver?

– What advice should patients be given about these cysts to reassure or allay fears of their effects?
Summary

• Questions? Comments?
• We welcome your input

• We look forward to collaborating with the other groups on areas of similar focus (aneurysms, pain mgt, PLD, etc)