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P397 -Clinical Practice Guideline: Monitoring children and young people with, and at risk of developing Autosomal Dominant Polycystic Kidney Disease (ADPKD)

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AIMS

There is significant variation in clinical practice across the UK with regard to the management of autosomal dominant polycystic kidney disease (ADPKD) in children and young people. We developed recommendations for monitoring children and young people with, or at risk of developing ADPKD.

METHODS

This guideline was developed on behalf of the UK Renal Association (RA) according to the clinical practice guideline development policy manual (NICE accredited). Evidence reviews were undertaken to identify relevant literature focusing on the management of ADPKD in children and young people, according to an agreed search strategy. Medline, EMBASE and PsycINFO databases were search from 1980-2017, abstracts were screened for inclusion by two authors. Included studies were critically appraised. Where evidence was lacking, formal Delphi consensus methodology was employed with representation from all specialty areas covered by the guideline including patient representation. 80% of Delphi panel agreement was taken as consensus, an iterative process was employed with three rounds.

RESULTS

Sufficient research evidence was obtained to make one evidence-based recommendation, a further 5 guideline points were made on the basis of Delphi consensus and panel discussion. Key recommendations include:

1. Information on ADPKD inheritance and the potential benefits and harms of testing should be provided by health professionals with specialist knowledge.
2. Measure blood pressure at least every 2 years in all children aged over 5 years with or at risk of ADPKD.
3. The decision to test for ADPKD in asymptomatic children and young people (CYP) at risk of developing ADPKD, should be undertaken jointly between health professionals and parents or carers and, wherever possible, the young person.
4. Either kidney ultrasound or genetic testing may be offered to asymptomatic children and young people at risk of ADPKD, where testing has been agreed by parents or carers (and, wherever possible, the young person) and health professionals.
5. If asymptomatic children at risk of ADPKD do not have cysts on ultrasound, further ultrasound testing should be deferred until adolescence (15-18 years), or later if preferred by the young person.

6. If genetic testing is planned in children and young people at risk of ADPKD, identification of the mutation in the affected adult family member (if not already known) should be undertaken prior to testing the child or young person.

Three further research recommendations were made, due to the absence of evidence and lack of consensus in the Delphi process. These covered the quantification of proteinuria and screening for intracranial aneurysms.

CONCLUSIONS

Through literature review and Delphi consensus methodology we have developed a clinical practice guideline for children and young people with ADPKD that will assist clinicians and families in making decisions about this condition and improve the consistency of clinical management across the UK.