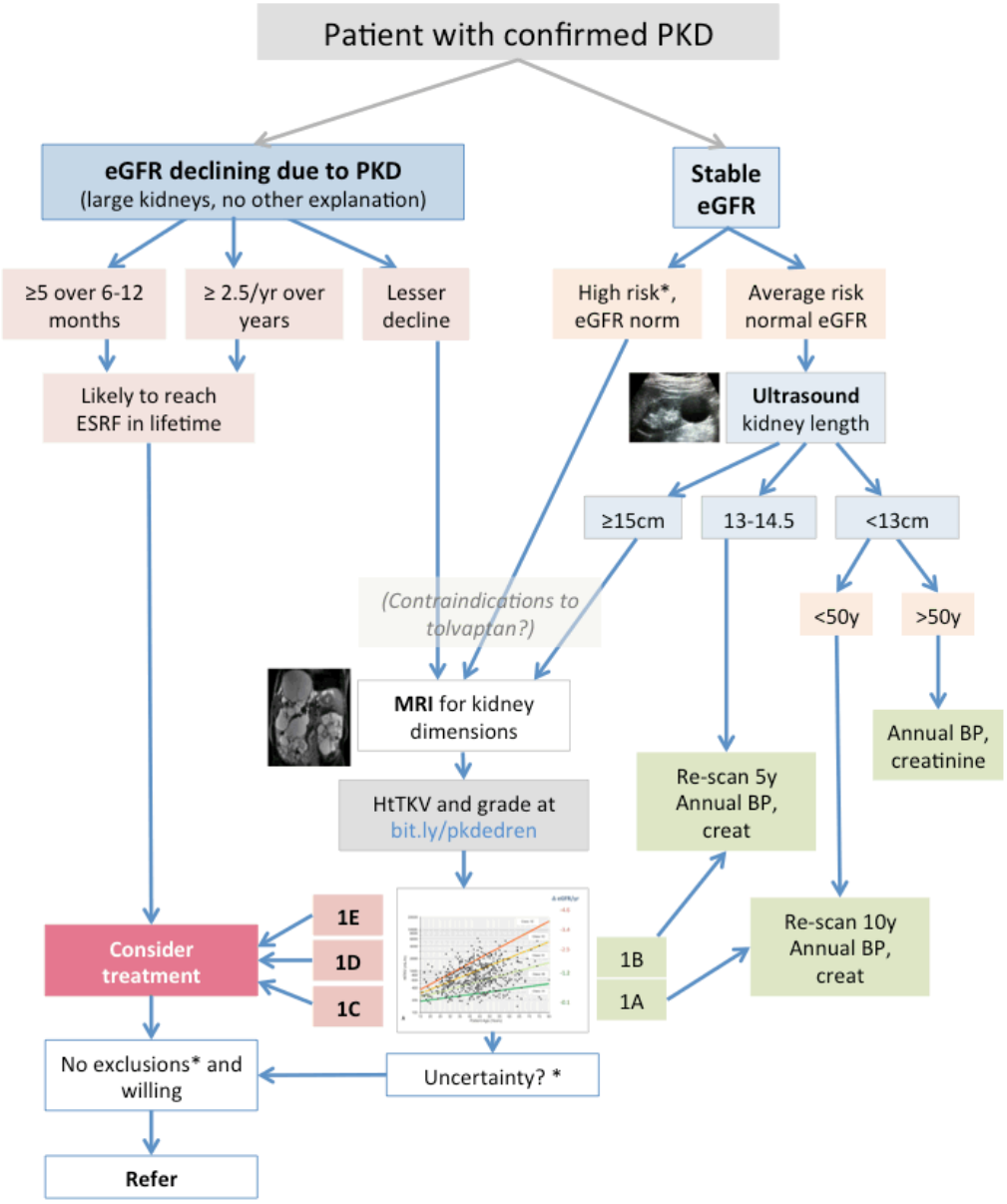
Tolvaptan for PKD

Edinburgh management pathways

Tolvaptan is approved for use in adult patients with PKD 'at high risk' of progression to ESRF. In Edinburgh we will manage this through a High Risk PKD clinic, which will initiate Tolvaptan therapy, prescribe and vary dose, and monitor therapy during the first 18 months. Most patients will later return to, or share care with, their regional or other appropriate clinic. The criteria for therapy are mainly based on (1) change in eGFR, and (2) kidney size vs. age.



Eligible patients for Tolvaptan therapy

- A definite diagnosis of ADPKD, e.g. by Levine criteria. Genetic confirmation is not required.
- Over 18 years old
- CKD stages 1-3

Exclusions/ contraindications include

- requirement for diuretics
- not able to drink freely
- at high risk of AKI from other etiologies. ACEi therapy is not a contraindication in an otherwise healthy patient who can follow Sick Day Rules.

- women wanting a family – discuss. Earlier pregnancies likely to be lower risk in PKD. >3 pregnancies is associated with earlier ESRF in some studies.
- liver disease
- not willing to comply with monitoring (monthly LFTs for the first 18 months)

High risk markers – clinical and other markers of early ESRF

- Truncating PKD1 mutation (other PKD1 mutations regarded as intermediate risk)
- Family history of early onset disease
- Early symptoms or signs (e.g. hypertension in teens/ 20s; kidney pain)
- Big kidneys – e.g. clinically palpable in young patients

Uncertainty after imaging – may arise because kidneys are not regular shape; for example distorted by a few very large cysts. Discuss.

Ultrasound size criteria; re-scan intervals – several criteria cited here are experimental, including size limits to refer for MRI; use of ultrasound vs. MRI; and re-scan intervals. Progress will be monitored and recommendations modified accordingly. Equally it may be justifiable to vary these criteria in particular circumstances.

Other aspects of management

Coding – Please code EDTA diagnosis on SERPR at Patient Review – Primary Renal Diagnosis.. Type in *Autosomal Domi …* and pick ADPKD (no need to bother with the PKD1/2 codes), then click *Apply*. Code 2718 should appear. This will make it easy to pull records of all patients in the future.

Radar and PatientView – ADPKD is now a Radar group. Please ask all patients to sign for **both** – this will give us an easy way to contact them, and to learn more about the condition nationally.

Risk category – Please record kidney size and other risk markers (above) in problem list with date, e.g.

- Kidney length 12.5, 13.5 cm Jun 2016
- Mother, uncle ESRF in 40s

Statins, CKD management – prescribe a statin if GFR is reduced, or for high-risk patients. In addition to value for cardiovascular events, there may be a modest additional effect on cyst growth. Other aspects of CKD care, including control of BP, remain as for CKD of other causes.

Genetic testing – is becoming more useful, but is still expensive, and in most cases will not affect management. Currently MRI is less expensive and more informative. Circumstances where genetic testing may be justified include

- A relative wants to be a kidney donor. Test the index patient first.
- The patient is asking about antenatal testing to prevent offspring inheriting the disease. Discuss with Clinical Genetics.
- The diagnosis is probable but not certain, and the differential diagnosis matters.

Children – It is still rarely appropriate to screen children before they are old enough to consent to it themselves. Even a few small cysts in a child of an affected parent, including in utero, makes the diagnosis likely. However absence of cysts by ultrasound cannot completely exclude the diagnosis until 30s in some individuals (but you'd expect a good prognosis if cysts are this late in appearing).

Monitoring children: Where the diagnosis is possible, occasional checking of BP is the only monitoring usually justified. Early onset hypertension sometimes occurs, in which case an ultrasound scan is indicated. Advanced ADPKD can occur in childhood, but it is very unusual.

Please send questions about this document to Madeleine Vernon or Neil Turner
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