

Adult Management of Renal Function Decline

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Disclosure of Interests

Otsuka: research support, Consultant

•Sanofi-Genzyme: Consultant

Novartis: Consultant

Kadmon: Consultant

Misubishi-Tanabe: Consultant

UpToDate: Section Editor, Cystic Kidney Disease

•US Dept. of Defense: research support



Definition of Chronic Kidney Disease

CKD is defined as abnormalities of kidney structure or function, present for >3 months, with implications for health.

CKD is classified based on cause, GFR category, and albuminuria category (CGA).



Prognosis by GFR and Albuminuria

Prognosis of CKD by GFR and Albuminuria Categories: KDIGO 2012

Normal or high

Mildly decreased

decreased

Moderately to

Kidney failure

Mildly to moderately

severely decreased

Severely decreased

	Persistent albuminuria categories Description and range		
	A1	A2	АЗ
	Normal to mildly increased	Moderately increased	Severely increased
	<30 mg/g <3 mg/mmol	30-300 mg/g 3-30 mg/mmol	>300 mg/g >30 mg/mmol
≥90			
60-89			
45-59			
30-44			
15-29			
<15			



categories (ml/min/ 1.73 m^2)

GFR

Description and range

G1

G2

G3a

G3b

G4

G5

Green: low risk (if no other markers of kidney disease, no CKD); Yellow: moderately increased risk; Orange: high risk; Red, very high risk.

3.1 Prevention of CKD Progression

Treatment is focused on management of blood pressure and proteinuria, (BP goals have changed since KDIGO-CKD 2012)

- 3.1.5: We suggest that in both diabetic and non-diabetic adults with CKD and with urine albumin excretion of <a>30mg/24 hours (or equivalent*) whose office BP is consistently <a>130mm Hg systolic or <a>80mm Hg diastolic be treated with BP-lowering drugs to maintain a BP that is consistently <a>130mm Hg systolic and <a>80mm Hg diastolic. (2D)
- 3.1.7: We recommend that **an ARB or ACE-I** be used in both diabetic and non-diabetic adults with CKD and urine albumin excretion >300 mg/24 hours (or equivalent*). (1B)



3.1.19 Salt Intake

Salt intake

- 3.1.19: We recommend lowering salt intake to <90 mmol (<2 g) per day of sodium (corresponding to 5 g of sodium chloride) in adults, unless contraindicated (see rationale). (1C)
 - 3.1.19.1: We recommend restriction of sodium intake for children with CKD who have hypertension (systolic and/or diastolic blood pressure > 95th percentile) or prehypertension (systolic and/or diastolic blood pressure > 90th percentile and < 95th percentile), following the age-based Recommended Daily Intake. (1C)
 - 3.1.19.2: We recommend supplemental free water and sodium supplements for children with CKD and polyuria to avoid chronic intravascular depletion and to promote optimal growth. (1C)



Treatment of Other Complications

Anemia

Hyperuricemia

CKD Metabolic Bone Disease

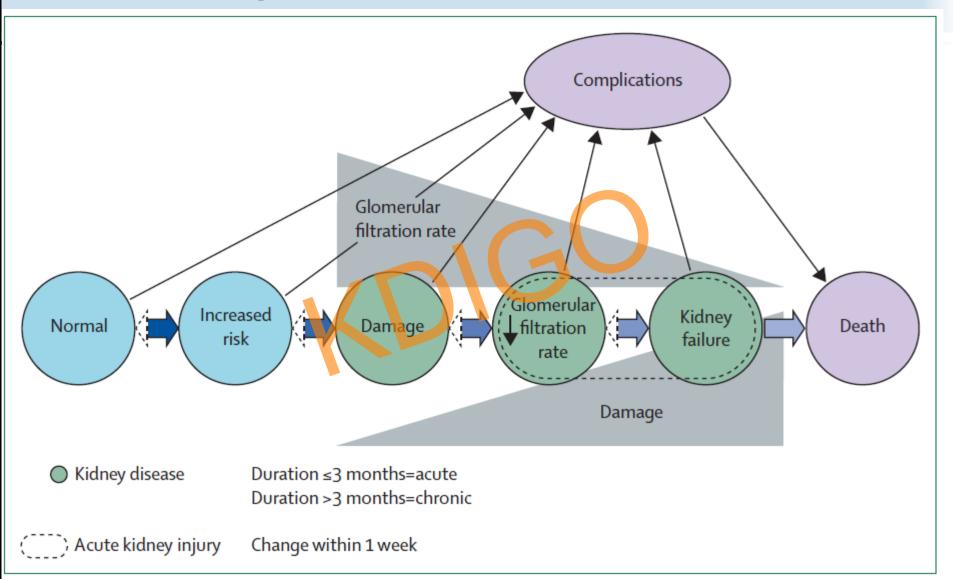
Acidosis

Hyperkalemia

Cardiovascular Disease Prevention



CKD progression





What About Uncommon Diseases?

Frequently not associated with heavy proteinuria in childhood What does the patient look like in adulthood?

- -likely reduced GFR
- -likely to have many of the metabolic complications of CKD
- -planning for renal replacement therapy will be important



Potential Framework for Consideration

Tubulopathies/channelopathies

Progression of renal injury with development of glomerulosclerosis, irrespective of initial cause

Primary glomerular involvement with albuminuria



Tubulopathies and Channelopathies

Not initially associated with hypertension or albuminuria, although proximal tubulopathy may exhibit albuminuria due to impaired reabsorption rather than glomerular injury

Focus of treatment on managing electrolyte disturbance, polyuria, salt loss, and correcting specific metabolic defect (Cystinosis)

Treatment of hypertension may not require RAAS blockade; RAAS blockade could be harmful

Management of other complications of CKD



Progression of Tubulopathy/ Channelopathy

Development of hypertension and albuminuria, consistent with glomerulosclerosis

Treatment of hypertension and albuminuria with RAAS blockade, adjusted to reflect channel dysfunction and resulting electrolyte abnormalities.

Duration of treatment to correct specific metabolic defect?

Management of other complications of CKD



Glomerular Involvement: Albuminuria

Initial manifestation with albuminuria, development of hypertension, consistent with glomerular involvement

Treatment of hypertension and albuminuria with RAAS blockade

What is the duration of treatment to correct specific metabolic defect? (Fabry)

Management of other complications of CKD



Literature is Scant

Absence of controlled trials, primarily retrospective or experience based





Transition of Care

Care of adolescent can be challenging

- -lack of compliance
- -rebellion

This is a difficult time to transition care to adult nephrology, particularly if this involves switching institutions

What is the optimal support system?



What is the Optimal Method for Supporting Patient and Family?

Managing the transition from adolescence to adult care

Managing the transition to ESRD, potential live donor transplantation



Summary

There is a lack of literature regarding the management of rare pediatric kidney diseases in adulthood

Standard clinical practices (RAAS blockade, salt restriction) may not be appropriate

