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

The table below illustrates the prognosis of CKD by GFR and Albuminuria Categories according to KDIGO 2012.

<table>
<thead>
<tr>
<th>GFR categories (ml/min/1.73 m²)</th>
<th>Description and range</th>
<th>Persistent albuminuria categories</th>
<th>Description and range</th>
</tr>
</thead>
<tbody>
<tr>
<td>G1</td>
<td>Normal or high</td>
<td>A1</td>
<td>Normal to mildly increased</td>
</tr>
<tr>
<td>G2</td>
<td>Mildly decreased</td>
<td>A2</td>
<td>Moderately increased</td>
</tr>
<tr>
<td>G3a</td>
<td>Mildly to moderately decreased</td>
<td>A3</td>
<td>Severely increased</td>
</tr>
<tr>
<td>G3b</td>
<td>Moderately to severely decreased</td>
<td></td>
<td></td>
</tr>
<tr>
<td>G4</td>
<td>Severely decreased</td>
<td></td>
<td></td>
</tr>
<tr>
<td>G5</td>
<td>Kidney failure</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

- **A1** (Normal to mildly increased): 
  - <30 mg/g (<3 mg/mmol)
- **A2** (Moderately increased): 
  - 30-300 mg/g (3-30 mg/mmol)
- **A3** (Severely increased): 
  - >300 mg/g (>30 mg/mmol)

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

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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 \( \geq 30 \text{mg/24 hours} \) (or equivalent*) whose office BP is consistently \( >130 \text{mm Hg systolic or } >80 \text{mm Hg diastolic} \) be treated with BP-lowering drugs to maintain a BP that is consistently \( \leq 130 \text{mm Hg systolic and } \leq 80 \text{mm Hg diastolic} \). (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 \( >300 \text{ mg/24 hours} \) (or equivalent*). (1B)
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). (IC)

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. (IC)

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. (IC)
Treatment of Other Complications

- Anemia
- Hyperuricemia
- CKD Metabolic Bone Disease
- Acidosis
- Hyperkalemia
- Cardiovascular Disease Prevention
CKD progression

- Normal
- Increased risk
- Damage
- Kidney failure
- Death

Complications

Glomerular filtration rate

Damage

- Kidney disease
  - Duration ≤3 months = acute
  - Duration >3 months = chronic

Acute kidney injury
  - Change within 1 week
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.