From Childhood to Adulthood: Guidance of Transition and Recommendations

Banafsheh Arad

Pediatric Nephrologist

- Cystinosis is a chronic childhood disease characterized by chronic renal failure and multi-organ involvement.
- Children with chronic illness need special care during the transition to adolescence and adulthood.
- With advances in treatment and kidney transplantation methods, the life expectancy of these patients has been prolonged.

The importance of the transition:

1) The inherent difficulty in caring for an adolescent/young adult with a chronic disease who has usually received a kidney transplant and thus has a high risk of graft loss
2) Extra-renal involvement progression
3) Non-compliance with treatment
4) The lack of experts close to the patient’s place of residence

What is transition?

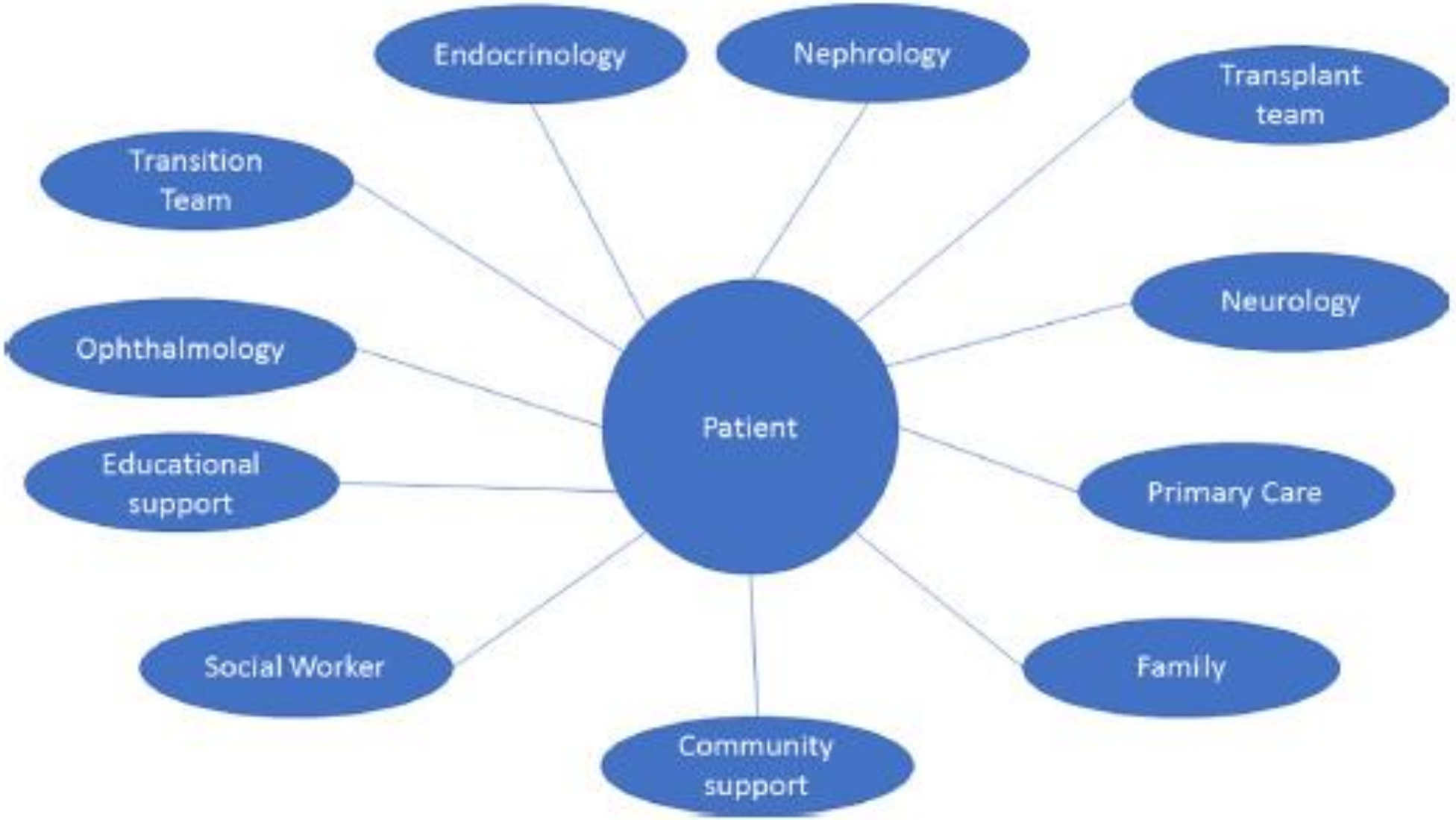
Transition is a planned and coordinated process that aims to prepare and transfer the pediatric patient from guardian-care to an adult unit where they will become responsible for their own self-care.

When?

- Planning: to start the process in preadolescence (12–14 years).
- At a time of clinical and emotional stability.

How?

- Personalized for each patient
- Gradual.
- Consensus with the patient and family, and in agreement with the pediatric and adult healthcare teams
- After completion of compulsory education
- Introduce the patient to the new medical team
- Take economic factors into account


Endocrinology
Nephrology
Transplant team
Transition Team
Neurology
Ophthalmology
Patient
Educational support
Primary Care
Social Worker
Family
Community support

Prior to initiating transition, the pediatric team will ensure the patient is competent in the following areas: Milestone checklist (pre-transition stage).

Each competency consists of four to five skills the patient should acquire prior to transition. In order to pass onto active transition, the pediatric team must confirm the patients have achieved proficiency in at least 50% of the skills in each proficiency area

- ***Recognition***
- ***Insight***
- ***Self-Reliance***
- ***Establishment of health habits***

Recognition

- ☐ Disease manifestations
- ☐ Treatment regimen (Medications and Frequency and route of administration)
- ☐ Their role in disease management

Insight

- ☐ Short- and long-term health outcomes
- ☐ Medication side effects
- ☐ Consequences of non-adherence
- ☐ How cystinosis will affect social aspects of their lives

Self-Reliance

- ☐ Be able to consistently take medications
- ☐ Attend, participate, and act independently during appointments
- ☐ Call in for refills and appointments
- ☐ Recognize urgent/emergent changes in their health

Establishment of health habits

- ☐ Lifelong healthy choices
- ☐ pursuit of educational vocational goals
- ☐ acceptance of responsibilities as he/she enters adulthood

In general, there are three chronologic stages

- Preadolescence (12–15 years) Planning/introduction to the transition
- Adolescence (16–18 years) Patient education
- Young adulthood (19–25 years) Transfer

Patient competencies and skills required for transition

- **Preadolescent (12–15 years)**

Disease:

Briefly explain cystinosis

Medications:

Know the names of drugs, daily dose and schedule

Lifestyle habits:

Understand health lifestyle habits (diet, physical exercise)

Relationship with the care team:

Start to get involved at medical appointments

Patient competencies and skills required for transition

- **Adolescent (16–18 years)**

Disease:

explain the disease in greater detail

Medication:

Indication, preparation, storage, side effects, interactions

and risks of non-adherence

Lifestyle habits:

Healthy lifestyle habits, and high-risk behavior (drugs and alcohol)

Relationship with the care team:

medical appointments and communications

Patient competencies and skills required for transition:

Young adult (19–25 years)

Disease:

Understand the disease in depth

Medications:

Autonomy in the administration of medication

Lifestyle habits:

Fully understand the healthy lifestyle habits

Relationship with the care team:

Know the reference adult care team and the different specialists

Recommendations for the transition to adult nephrology

- **Kidney transplant**
- Date and type of transplant (premptive or post-dialysis)
- Donor characteristics (alive or deceased, HLA serologic tests)
- Receiver characteristics (HLA, serologic tests, other risks)
- Surgical report (graft characteristics, initial transplant evolution)
- Graft biopsy (protocol or elective, date and findings)
- Anti-donor antibodies

➢ Kidney transplant

Events

- Episodes of rejection, acute renal failure, hypertension, cardiovascular involvement, dyslipidemia, diabetes, bone disease

Treatment plan

- Immunosuppression regimen
- Specific cysteamine therapy

➢ Chronic dialysis

- Start date and type of dialysis
- Place of treatment
- Vascular access
- Kidney transplant waiting list
- Dialysis prescription and efficacy (KT/V)
- Residual renal function, Fanconi Syndrome
- Episodes of superimposed kidney failure, HTN, cardiovascular involvement, dyslipidemia, diabetes, bone disease
- Prescribed drugs

Chronic kidney disease

- Stage and rate of progression
- Kidney transplant waiting list
- HLA, serologic tests
- Residual renal function, Fanconi Syndrome
- Superimposed kidney failure, HTN, cardiovascular involvement, dyslipidemia, diabetes, bone disease
- Prescribed drugs

Recommendations for the transition to adult ophthalmology

- ***Information about the disease***
- ***Patient's medical record***
- ***Patient monitoring***

In adolescence (16–18)	Tests every 6 months
In adults	Tests every year

- ***Optimal periodic follow-up***

Corneal crystal distribution

Measure intraocular pressure

Examination of the ocular fundus

Recommendations for the transition to adult ophthalmology

- ***Recommendations in exceptional circumstances:***

Ocular fundus examination 2–3 months after starting treatment with certain drugs (growth hormone, oral contraceptives, etc.)

- ***Topical ophthalmic treatment:***

Cysteamine saline solution 0.55%

Cysteamine viscous solution 0.55%

Ophthalmologic emergency:

Severe and/or acute loss of vision are not typical of cystinosis

Recommendations for the transition to adult endocrinology

- ***Age***
- ***Pubescent stage at transfer:*** Tanner IV or below, or a history of delayed puberty (onset of puberty: ♂ > 14 years old; ♀ > 12 years old)
- **Hormone replacement therapy*:*** Thyroxine, insulin, GH or testosterone
- ***Endocrine parameters:*** T4, TSH, glycemia, HbA1c, GH, IGF-1, testosterone, LH, FSH
- **Staturo-ponderal development:** Weight, height and weight/height percentile, bone age
- **Diabetes?**

Recommendations for the transition to adult neurology

- ***Information about the disease***
- **Neurocognitive, behavioral or school performance problems**

Attention, executive functions, language and memory, perceptive, visuospatial and visuoconstructional functions and voluntary cognitive motor control.

- **Neuropsychologic explorations**

Cognitive performance, anxiety, depression and behavioral alterations

- **Muscular strength**
- **Orofacial and deglutory motor function, respiratory function**
- **Neuroimaging tests**

Non-adherence

- Non-adherence was reported about 89% in cystinotic children younger than 11, and 56% of older than this age.
- Patients may miss the drug doses, change medication by themselves, and not take the recommended doses.
- The patients complain of unpleasant cysteamine odor, gastrointestinal upset, and numerous medications with multiple daily doses

Assessment of non-adherence

- **Monitoring of intragranulocyte cystine levels:**

(6 h post-Cystagon dosing)

- In most of cystinotic patients, leukocyte cystine level is 3.0-23.0 nmol half cytine/mg protein without treatment.
- In a mixed leukocyte augmented with pholymophonuclears with HPLC method
- Normal leukocyte cystine value is below 0.2 nmol hemi cystine/mg proteins

- **Assessment of corneal cystine deposits**

Recommendations for improving treatment adherence in transition:

- Education about the consequences of cystine accumulation in different organs and kidney transplant rejection.
- Behavioral techniques to remember the time for taking medications. (parents can remind him or he can use an alarm or a calendar)
- Psychological problems such as depression, anxiety, and attention deficit disorder
- Engage family members, friends, and medical team to improve patient's adherence to treatment
- Regular appointments
- A good relationship between the patient/carer and medical team

Recommendations for promoting self-care among adolescent and young adult patients:

- ***Cystinosis dossier***

1) Medical visit schedule
2) List of contacts
3) Medical reports by date and specialist
4) Immunization card
5) Record of allergies, if any
6) Any documents from appointments, blood tests, etc.
7) Medication monitoring


Thanks for your attention