Objectives

- List treatment options for chronic kidney disease (CKD) in children
- Outline medical evaluation
- Identify donor sources
- Describe transplant surgery and post-operative care

How Common

1 to 2 children out of 100,000
Adults are 20 times more likely

Etiology of CKD in children

- Congenital
  - Structural (obstructive defects)
  - Inherited disease processes
- Acquired
  - Inflammatory/immunologic
  - Infectious
  - Other

Causes of CKD in Children

- Obstructive Uropathies
  - PUV – posterior urethral valves
  - Most common lesion of lower urinary tract in male infants
  - Folds across the urethra obstruct urinary flow
  - Causes hypertrophy of the bladder neck and dilation of the upper urinary tract
  - Advances to CKD during infancy or early childhood

Causes of CKD in Children

Obstructive Uropathies

Ureteropelvic junction (UPJ) 
vesicoureteral junction obstruction (VUJ)

Causes backflow of urine into ureters and kidney pelvis (VUR-vesicoureteral reflux)
Causes of CKD in Children

- VUR – Vesicourethral reflux (primary or secondary)
  - Grade I-V unilat or bilat, lower grade better
- Multicystic dysplasia – Often prenatal diagnosis, typically one kidney
- Hypoplasia or aplasia – Underdeveloped kidneys

- Prune belly – agenesis of abdominal musculature
- Duplication of ureters – UTIs, surgical intervention
- Horseshoe kidney – Two kidneys fused, may function normally, potential for problems later

Causes of CKD in Children (cont’d):

- Inherited disease processes
  - PKD – Polycystic kidney disease
  - Familial nephritis
  - Congenital nephrotic syndrome
  - Cystanosis
  - Primary oxalosis
  - Alports

Causes of CKD in Children

- Acquired (inflammatory/auto-immune)
- Glomerular diseases
- FSGS/nephrotic syndrome
- HSP-Vasculitis
- Lupus
- IgA nephropathy
- Amyloidosis

- Post-infectious disorders
  - HSP
  - HUS
  - Other
    - HTN
    - Kidney stones
    - Wilms tumor – Childhood cancer, usually identified by age 2 yrs
    - NSF (Nephrogenic systemic fibrosis) aka NFD (nephrogenic fibrosing dermopathy)
Treatment Options for CKD

- Peritoneal dialysis – home therapy
- Hemodialysis – in center or at home
- Transplant – may or may not need dialysis as a bridge to transplant. Transplant is not a cure, but a surgical treatment.
- No treatment

Benefits to Transplant

- NO MORE DIALYSIS!
- More energy
- No more diet or fluid restrictions
- Opportunity for growth and development

Risks

- General anesthesia
- Immunosuppressive medications and side effects
- Organ rejection and post operative complications
- Psychosocial risks: depression, anxiety, guilt, and dependence on others
- Risk of disease transmission from the donor
- Potential for recurrence of original kidney disease

Transplant is not a cure

Trading one disease state for another!!
Who can receive a transplant?

- Patients with CKD and on some form of dialysis.
- Patients with CKD, not yet on dialysis but with kidney function low enough to benefit from transplant.
  - CKD stage 4 or 5 with steady progression (GFR 20 or below)
  - Anemia
  - Low energy
  - Poor appetite
  - Growth impairment

Who can receive a transplant?

- Timing
  - IS everything
- Age
  - Really does NOT matter
- Size
  - DOES count

Renal Transplants by Age

Barriers to Evaluation/Transplant

- Medical issues such as significant irreversible heart or lung disease, cancer
- Non-compliance with dialysis, medications, or appointments
- Current use of illegal drugs
- Morbid obesity
- Not having an appropriate caregiver/support person after transplant

Pre-Transplant evaluation

- Considerations:
  - Cause of CKD
  - Medical status (co-morbidities)
  - Current kidney function
  - Living donor vs. deceased donor
  - Adherence

Pre-Transplant evaluation

- Orientation class
- Insurance approval to begin full evaluation
- Medical evaluation includes (but is not limited to):
  - Labs, EKG/echo, Chest x-ray, Renal ultrasound
  - Social Worker – to assess support system
  - Pediatric Nephrology, Pediatric Urology (as indicated)
  - Psychology
  - Transplant Surgeon
  - Health Maintenance: dental, vaccinations, well child
  - Information from dialysis unit/nephrologist
  - Transplant specific labs: Tissue typing, PRA, Cross match
Pre-Transplant Preparation
- Surgical interventions pre transplant
  - Native nephrectomy
  - Infection
  - Protein loss/FSGS
  - Bladder augmentation
  - Continent urinary diversion
  - Intestinal conduit
  - Mitrofanoff (appendicovesicostomy)
  - Native bladder

Donor source
Type of transplant
Living donor kidney
- Donor can be related or unrelated
- *Blood type compatibility may/may not matter
- Surgery can be planned, no long waiting time for recipient

Deceased donor kidney
- Comes from someone who has died
- *Must be blood type compatible
- Unplanned, can be anytime, long waiting list

PRA
Panel Reactive Antibodies
- PRA - level of ANTIBODIES present in the blood
- Antibodies - previous transplant, pregnancy, blood transfusions
- PRA - fluctuate, measured every 1-3 months while on the waiting list
- A high PRA (>30%) may make finding a suitable kidney more difficult.

HLA Tissue Typing
Human Leukocyte Antigen
- Blood test to identify antigens
  - Protein markers on cells
- Six (6) primary ANTIGENS important in transplant. Inherit half from each parent.
  - A2, A30 (class I)
  - B8, B30 (class I)
  - DR3, DR8 (class II)
- None of the antigens HAVE to match the donors antigens.
  - Identical twins
  - 1 in 100,00 if unrelated

HLA CROSS MATCH
- Blood test done between donor (living or deceased) and recipient
- Mixes donor and recipient blood; looks for reaction
- We want NO REACTION, or NEGATIVE cross match
- POSITIVE cross match means there is a reaction, transplant from this donor would fail immediately

Live Donor evaluation
- Similar to that of the recipient: labs, x-rays, EKG, Nephrology, Transplant Surgeon, health maintenance update
- Donors over age 50 will be require more testing ie cardiac stress test.
- Contraindications to donating: diabetes, hypertension, cancer, obesity, kidney stones, current substance abuse
- Donor/kidney size can be consideration for pediatric recipients
Live Donor Evaluation

- Donor evaluation and surgery is covered for by the recipient’s insurance provider.
- Donor and recipient evaluations may be done simultaneously, if insurance allows, but only 1 donor at a time.
- Donor surgery may be laparoscopic or open, average length of stay in hospital 2-5 days.
- Donors must independently come forward and communicate directly with donor coordinator.

What if there is no living donor?

- Waiting list for a deceased donor kidney
- Average waiting time in this area is 5-6 years
- There are approx. 620 people on the UNC kidney transplant waiting list, approx. 90,000 people waiting nationwide
- UNC currently performs 70-80 kidney transplants per year
- List is managed nationally by UNOS (United Network for Organ Sharing), managed locally by CDS (Carolina Donor Services)

The Waitlist

- National list
- 11 regions
  - Kentucky, **North Carolina**, South Carolina, Tennessee, Virginia are region 11
- UNOS contracted by federal government
- Allocation point system
  - Every point = 4yr wait time
- Children are matched to donors less that 35yo first

When an kidney becomes available

- First, there is a nationwide search for a “6 antigen match”.
- If there is no “6 antigen match”, the kidney stays in the local area/region
- UNOS generates a list of patients in the area/region that best matches that specific organ. The list is called a match run list.
- Antigen match and amount of time on the waiting list are used to develop this match run list.

In North Carolina

- Five kidney transplant centers.
- Two organ procurement groups or regions: Carolina donor Services (CDS) manages organ retrieval for UNC, Duke, ECU, and Wake Forest hospitals
  Charlotte does the same for CMC.
In North Carolina

- UNC, Duke, ECU, and Baptist hospitals share a region and are essentially considered one list.
- The match run list that is generated when there is a kidney offer in the UNC area can include patients from all four of these hospitals.
- Also, there is no benefit to being listed on more than one list within the same region (OPO). May list at multiple centers in separate regions. (UNC and Charlotte)

Getting THE Call

- Nurse coordinator will call the patient, ask how they’ve been, recent illnesses, hospitalized, on antibiotics?
- We want patients at optimum health at the time of transplant.
- If there’s been a recent illness or need for antibiotics, patient not be eligible for the kidney transplant.
- Patients have the right to refuse a transplant at any time.

Getting THE Call

- When coordinator informs patient of kidney offer, will identify where their name is on the match run list, and develop a plan.
- This plan may involve waiting at home for a period of time, or we may have them come to the hospital right away.

Who gets the kidney?

- The first person on the match run list who receives medical/surgical clearance and has a negative cross match...gets the kidney.
- Unfortunately, this means some people are brought into the hospital, wait several hours, and then are sent home without a transplant.

When patient comes in....

- ED or 24 hr observation unit
- Blood drawn for cross match (4-5 hours to complete)
- Assessed by a member of the surgical team for surgical clearance
- Assessment by pediatric nephrologist for medical clearance
- Wait for cross match results (our patient and others)
- Ideally no more than 12-24 hr cold time

Wait listed or transplanted

- Wait listed or transplanted within year
- Percent of patients
- 91 93 95 97 99 01 03 05
- 0-17 18-34 35-49
- 50-64 65-69

USRDS 2011 ADR
Transplant Surgery

Following Surgery
- PICU for first 1-2 days
- IV, oxygen, cardiac/BP monitoring, urinary catheter, JP drain
- Out of bed as soon as possible
- Education begins as soon as possible
- Families learn about:
  - Diet
  - Medications
  - Activity level
  - Wound care
  - Avoiding infections
  - Assessing problems
- Average length of stay in hospital is 5-7 days

Post-Operative Management
- General - fever, pain, discomfort
- Renal - urinary output, pain over graft site, edema, hypertension, BUN, creatinine, electrolytes
- Urinary tract - urine characteristics (blood, cloudy, etc.), bladder spasms
- Pulmonary - decreased breath sounds, Increased respiratory rate, O₂ requirement

Post-Operative Continued
- GI - Bowel sounds, distention, N/V, appetite
- Immune - Wound site, elevated WBC
  - Anti-virals and antibiotics
  - Protective isolation precautions secondary to immunosuppression

Post Transplant Meds
- Steroids
- Calcineurin Inhibitors
  - Tacrolimus/Prograf/FK 506
  - Cyclosporin, Gengraf/Neoral/Cyclosporin modified
- Mycophenolate Mofetil (Cellcept/MMF)
  - Azathioprine/Imuran
- Bactrim (PCP prophylaxis)
  - Pentamidine aerosol
- Ganciclovir/Vålyce (CMV prophylaxis)
- Electrolyte Replacements (neutraPhos, KCl, magnesium oxide)

Nurses are Key
- Help identify knowledge deficits (patient, family, doctors)
- Aid with patient teaching (medications, symptoms of rejection, infection, etc)
- Address issues of altered body image
Outpatient post transplant
- 3x/week labs, M/W/F, 4-6 weeks
- 2x wk, M/T, 4 weeks
  - CBC with diff, Chem 7, ca, mag, phos
  - urinalysis
  - 12 hr trough to check tac or cya
- Monthly
  - Viral monitoring (CMV, EBV, urine decoys/BK)
  - UA with urine culture
- Daily home monitoring of BP, I/O, fever, weight
- Weekly clinic visits 4-6 weeks post op
  - Every 2 weeks x4
  - Monthly
  - Every 3 mo

Why We Biopsy
- Diagnostic
- Disease Staging
- Directs therapy

Post-operative complications
- INFECTION
  - High risk due to immunosuppression
- REJECTION
  - May be chronic or acute, has varying degrees
  - Can be treated in most cases if caught early
  - May require hospitalization for biopsy, IV medication
  - Signs/symptoms difficult to recognize by patient
  - Noted on labs before symptoms occur

Post Transplant Challenges
- Primary care, medical home
- Most pediatric practices will not take Medicare patients
- Adherence
  - Medications
  - Lab work
  - Clinic appointments
- Urological
- Communication
- Transitioning to adult care

UNC vs National
(1/2009-6/2011)

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<th>Deceased Donor Transplant</th>
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<td></td>
<td>Graft Survival UNC</td>
<td>National Average</td>
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<tr>
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<td>1 year (children)</td>
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<td>3 year (children)</td>
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Other options for transplant
- Expanded Criteria Donors (ECD)
  - Donors > 50 yrs old with HTN, diabetes, elevated creatinine and/or death from a stroke.
  - Optional
  - Special consent form required in advance
**Other options for transplant.....**

- Centers for Disease Control (CDC) - High Risk Donors
- Donors with high risk social behaviors such as IV drug use, time in jail, or other risk factors.
- Many tests are done to determine if donor has infectious disease prior to donation.
- Optional
- Consent form signed at time of transplant

**Final Thoughts**

Chronic Kidney Disease in children is not very common

Transplant is the best short and long term treatment option

Transplant is the most cost efficient therapy

Availability of organs is the most limiting factor for access to transplant

Children are precious, amazing and innocent. Our charge and goal is to keep them that way as long as they are children!