Refractory Anemia in a 2-year-old Peritoneal Dialysis-dependent Patient

Salar Bani Hani Supervised by Dr Smitha Vidi

- H.C is a 2 year old female, ex 36 weeker with ARPKD, Caroli's disease, portal hypertension s/p bilateral nephrectomies early in life and is on peritoneal dialysis.

- In January/2020 was found to have hemoglobin of 7.7 g/dl .
- A repeat CBC showed it to be 7 and she had a positive fecal occult blood (FOBT).

HPI

- No vomiting or diarrhea, no melena, no abdominal pain.
- No increased fatigue or change in appetite or SOB.
- Had switched from Elecare to Puramino Jr 2 weeks prior due to poor weight gain and had since then gained 0.7 kg.
- No new medications.

Past Surgical and Medical history

- Nephrectomies at age 1 week
- PD catheter insertion at age 1 week
- BL inguinal hernias, GERD and Pulmonary hypoplasia
- Discovered to have biliary duct dilatation and a cystic liver “mass” on a routine follow up ultrasound at age 15 months.
- At age 23 months she underwent a magnetic resonance cholangiopancreatography imaging.
- She was found to have splenomegaly with varices and portal hypertension. She also had intrahepatic biliary dilatation with regions of saccular dilatation in the right hepatic lobe which was consistent with her diagnosis of Caroli’s disease.

Meds

- Nephronex
- Zyrtec 1.3 mg/day
- Sevelamer 6000 mg/day
- Amlodipine 1.5 mg /day
- Clonidine 0.03 mg BID
- Calcitriol 0.4 mcg/day
- Cholecalciferol 2000 units/day
- Calcium phosphate 14 cc/day
- Cyproheptadine 2 mg BID
- Epoetin alfa 2000 units SC twice a week
- Ferrous sulfate 30 mg BID
- Miralax 4.25 g PRN/day
- Kayexalate 3.5 g /day

Vitals

- Pulse 126 bpm
- BP 103/75
- Resp 22 bpm
- Ht 80 cm (0.32 %ile)
- Wt 11.2 kg (7.76%ile)

Physical exam

- General: smiling, playing, small for age, thin hair.
- Eyes: pallor, no periorbital edema
- ENT: No lesions, moist mucous membranes
- Resp: Clear lungs, no increased work of breathing
- CVS: Normal s1, s2, no murmur
- Abd: soft and lax abdomen, hepatosplenomegaly, no guarding or tenderness. PD cath site clean.
- MSK: normal ROM, bruising present on lower limbs
- Neuro: Normal exam

Lab Results

- Repeat hg 7.0 gm/dl. Trend was from 10.4 to 7 gradually over the course of 5 months.
- MCV 89
  - MCH 29
  - MCHC 32
- Platelets 132
- Retic 4.6%
- Iron 42
- TIBC 248
- Tsat 17%
- INR 3.3
- PTT 62.9
- ALT 23
- AST 39
- Total bilirubin <0.2
- LDH 210 U/L
- Direct bilirubin <0.1

Lab Results

- Weekly KT/V 2.41
- Lead level <2
- Aluminum Level 14 (0-15 ug/L)
- Folate 15.4 ng/ml (>5.38 ng/ml)
- B 12 2165 picogram/ml (220-1125 picogram/ml)
- Methylmalonic Acid 0.85 ng/ml (0-4 umol/l)
- 25-hydroxy vit D 23 (20-80 ng/ml)
- Ca 9.2, Phos 4.1
- PTH 127 pg/ml (10-65 pg/ml)
- Hemoglobinopathies screen : normal
- FOBT positive
- BUN 25, Creatinine 5.6, K 3.8, Na 142, CO2 32, Albumin 2.2

Imaging

- Abd US: Similar intrahepatic biliary ductal dilatation compared to previous mages in the patient with history of Caroli Disease and Splenomegaly

What do you think is the cause of the anemia?

Carnitine profile

- Total carnitine 64 micromole/L (38-73 micromole/L)
- Acyl carnitine 25 micromole/L (7-24 micromole/L)
- Acyl/free carnitine 0.6 (0.1-0.8)
- These were obtained in 2017

What is the next step in your management?

- GI consult
- IV Vit K x 3
- Blood transfusion
- Plan for Upper GI endoscopy and liver biopsy as outpatient.
- Discharged on oral vit K.

- FINAL DIAGNOSIS: Vit K deficiency, Occult GI bleeding

- Two months later, hgb decreased gradually to 7 g/dl despite increasing Epoetin to 3000 units 3 days a week and receiving IV iron every two weeks.
- In addition, she had begun to lose weight despite adequate caloric intake. A fecal occult blood test was positive again, but INR was 1.1. She was suspected of having GI bleeding.

- She was admitted and transfused with the plan of upper GI endoscopy and possible sclerotherapy of varices.

What was the finding on endoscopy?

MEDLINE
non calibrated

- A hair bezoar was identified occupying 30% of the gastric lumen. It extended beyond the pyloric channel and tranversed as far as the scope could be safely advanced in the proximal jejunum. It occupied 50% of the duodenal lumen.
- It was not amenable to endoscopic therapy and the need for surgical evaluation and intervention was discussed with the family.

- An explorative celiotomy for removal of gastric duodenal jejunal tricho-bezoar was done without complications

One month later…

- Hgb 10.8 g/dl
- Iron 129
- Ferritin 103.5
- Tsat 50%

- Gaining weight steadily and adequately

Reason for hair bezoar formation?

- She had been started on amlodipine for blood pressure control in October/2019 (4 months prior to presentation).

- Amlodipine is known to cause alopecia,

Stump the Consultants

Annual Dialysis Conference, March 7th 2021

Dr. Priya Saini, MD, FRCPC
The Hospital for Sick Children
Toronto, Ontario, Canada

Case Presentation -1-

- 11 year old male
- Recent left elbow injury, receiving ibuprofren Q4 hours x 1 week
- PMHX: non-medicated ADHD, height & weight 90th %le
- Family Hx: early major vascular events in mother and maternal grandparents

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Case Presentation -2-

- 1 week after his elbow injury, presented with area of white discharge from his elbow
- Also, fever, vomiting, diffuse maculopapular rash, and petechiae
- BP 90mmHg requiring bolus

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Case Presentation -3-

Blood work

Lab	Value
WBC	24.6 x10^9/L
Hemoglobin	108 g/L (10.8 g/dL)
Platelets	494 x10^9/L
Neutrophils	15.6 x10^9/L
Eosinophils	0.62 x10^9/L
Creatinine	1900 umol/L
Urea	52.8 mmol/L
Sodium	133 mmol/L
Potassium	3.5 mmol/L

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Case Presentation -4-

Urine

Lab	Value
Urine ACR	35 mg/mmol
Urine PCR	127 mg/mmol
Urine Eosinophils	Positive

- Shortly after admission, noted to be anuric

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Initial Management

- Ceftriaxone, vancomycin, and clindamycin x 14 days
- Temporary femoral CVL insertion with renal biopsy
- Intermittent HD initiated: 2 hours, 2 mL/min clearance -> 3 hours, 4 mL/min clearance
- IV pulse methylprednisone x 3 days
- Started to void on 4th day -> HD 3 times/week

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Further Workup -1-

- Normal ANA, ANCA, C3, C4, anti-GBM.
- Kidney ultrasound: right kidney 11.5 cm, left kidney 11cm. Slightly increased echogenicity but normal renal parenchyma, resistive indices, and bladder.
- Renal biopsy: evidence of ATN. Also 11/18 globally sclerosed glomeruli at the corticomedullary junction. Mild interstitial fibrosis and tubular atrophy (20%). No arteriosclerosis. Immunofluorescence staining negative. No eosinophilic infiltrate.

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Further Workup -2-

- CKD Investigations:
  - Normal ECHO
  - Normotensive on no anti-hypertensive medications
  - Renal osteodystrophy scan: normal skeletal maturation, appropriate for age
  - Mild anemia, initiated on darbepoetin and iron supplementation

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Question #1

How long would you expect a patient with AKI to be on dialysis?

Length of Dialysis for AKI -1-

CJASN® Clinical Journal of the American Society of Nephrology

Recovery of Kidney Function in Children Treated with Maintenance Dialysis

Marjolein Bonthuis,[1] Jérôme Harambat,[2] Etienne Bérard,[3] Karlien Cransberg,[4] Ali Duzova,[5] Liliana Garneata,[6] Maria Herthelius,[7] Adrian C. Lungu,[8] Timo Jahnukainen,[9] Lukas Kaltenegger,[10] Gema Ariceta,[11] Elisabeth Maurer,[12] Runolfur Palsson,[13] Manish D. Sinha,[14] Sara Testa,[15] Jaap W. Groothoff,[16] Kitty J. Jager,[1] and on behalf of the ESPN/ERA-EDTA Registry

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Length of Dialysis for AKI -2-

What is the likelihood that children starting maintenance dialysis therapy will recover kidney function?

CJASN
Clinical Journal of American Society of Nephrology

Methods and Cohort

ESPN/ERA-EDTA Registry
36 European countries

N= 6574
Age <15 years
Maintenance dialysis initiation 2000-2014

Outcome:
Dialysis recovery = discontinuing dialysis for 30 days or more

Recovery (entire cohort)

Recovery at 2 years
2%

Median time to recovery
5 months
(IQR 2-9.6)

Recovery (by cause of kidney failure)

	CAKUT	Vasculitis	Ischemia	HUS
	0.8%	11%	12%	13%
Adjusted HR (95% CI)	ref	20.4 (9.7-42.8)	11.4 (5.6-23.1)	15.6 (8.9-27.3)

Conclusions There was a recovery rate of 2% within 2 years after initiation of maintenance dialysis in children. There is a clinically important chance of recovery in children with vasculitis, ischemic kidney failure and HUS.

Marjolein Bonthuis, Jérôme Harambat, Etienne Bérard, Karlien Cransberg, Ali Duzova, Liliana Garneata, Maria Herthelius, Adrian C. Lungu, Timo Jahnukainen, Lukas Kaltenegger, Gema Ariceta, Elisabeth Maurer, Runolfur Palsson, Manish D. Sinha, Sara Testa, Jaap W. Groothoff, and Kitty J. Jager. ***Recovery of Kidney Function in Children Treated with Maintenance Dialysis.***
doi: 10.2215/CJN.01500218

Case Continued -5-

- After ~12 months on dialysis, our patient was listed for deceased donor kidney transplant while continuing to investigate the etiology
- Eye exam: chronic papilledema despite well controlled BP on ABPM. Papilledema was asymptomatic with no headaches and 20/20 vision
- MRI/MRV: showed additional signs to support raised ICP, as well as a Chiari 1 Malformation

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Question #2

What are the causes and management of papilledema in a dialysis patient?

Papilledema -1-

- Dialysis disequilibrium syndrome
- Idiopathic Intracranial Hypertension
  - Risk factors:
    - Obesity (present in this patient)
    - Otitis media
    - Head trauma
    - Certain medications – ie tetracycline, Vitamin A
    - Refeeding after malnutrition

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Papilledema -2-

- Acetazolamide
  - Carbonic acid inhibitor
  - Contraindicated in ESRD
- Topiramate
  - Anticonvulsant with weak carbonic anhydrase inhibitor properties
  - Chosen for our patient
- Mechanical reduction of CSF
- Weight loss, low sodium diet

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Case Continued -6-

- With the constellation of ESRD, increased ICP, Chiari 1 Malformation, and a maternal family history of early major vascular events, without a unifying diagnosis, a whole exome sequence was performed.
- WES revealed *CLCN5 mutation in keeping with Dent Disease*
  - Variant p.D692LfsX7 with coding DNA c.2073_2076delTGAC, inherited from his mother

Question #3

What are the presenting features and the natural history of Dent Disease?

Dent Disease -1-

- X-linked recessive disorder of the proximal tubules
- Presenting features can include: a male patient with short stature, polyuria, microscopic hematuria, asymptomatic proteinuria, hypercalciuria , nephrocalcinosis, or nephrolithiasis

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Dent Disease -2-

- Renal biopsy in Dent disease: non-specific but can include focal global glomerulosclerosis without any basement membrane abnormalities. Also can see tubular atrophy, varying degrees of interstitial inflammation, and interstitial fibrosis (seen with our patient).

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Dent Disease -3-

- 2/3 of males patients with Dent disease -> CKD

- If have CLCN5 mutation (60%) & CKD -> 2/3 will then develop end stage kidney failure, usually around 30-50 years old

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Case Resolution -1-

- Shortly after his genetic diagnosis of Dent disease, our patient was tolerating a reduction in dialysis treatments.
- Eventually stopped dialysis altogether at ***26 months after hemodialysis was initiated.***
- His position on the transplant list was put on hold and eventually he was removed from the list.

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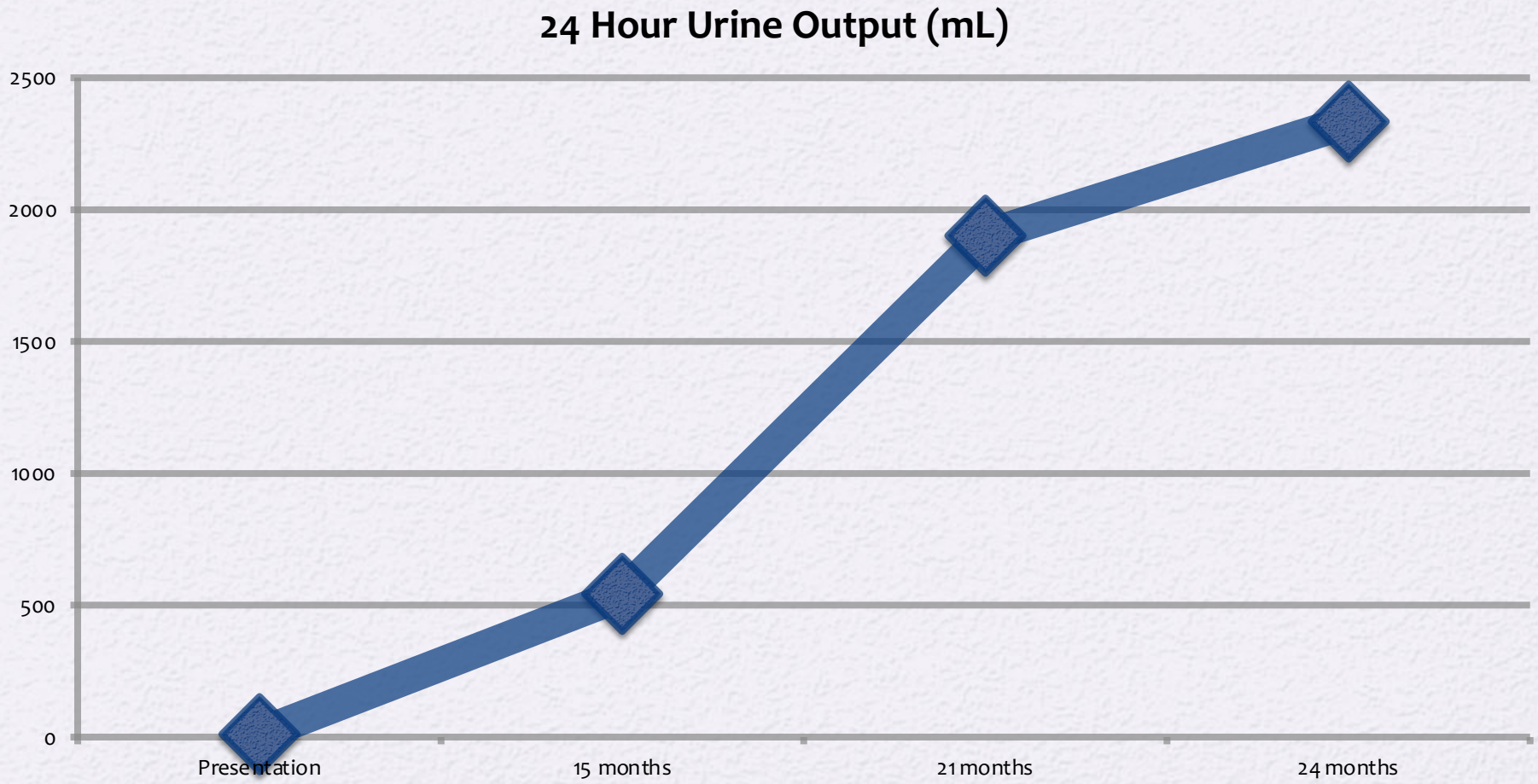
Case Resolution -2-



Case Resolution -3-

- Our patient is now 4.5 years from his initial presentation, and has remained off of dialysis for 2years without a kidney transplant or re-initiation of renal replacement therapy thus far.
- His GFR remains stable at 15 mL/min/1.73m2 and he continues to be followed in our chronic kidney disease clinic.

Thank You!

References

1. Bonthuis M, Harambat J, Bérard E, et al. Recovery of kidney function in children treated with maintenance dialysis. *Clin J Am Soc Nephrol*. 2018;13(10):1510-1516. doi:10.2215/CJN.01500218

2. Cleves-Bayon C. Idiopathic Intracranial Hypertension in Children and Adolescents: An Update. Headache. 2018;58(3). doi:10.1111/head.13236

3. Van Berkel Y, Ludwig M, Van Wijk JAE, Bökenkamp A. Proteinuria in Dent disease: a review of the literature. Pediatr Nephrol. 2017;32(10). doi:10.1007/s00467-016-3499-x

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Introduction to Resources and the *Pediatric Renal Team*

Nonnie Polderman, RD
Virtually from Vancouver, BC
March 5, 2021

BC Children's Hospital
An agency of the Provincial Health Services Authority

I have no disclosures.

Helping kids shine.

OBJECTIVES

- Discuss role of the RD as part of the interdisciplinary team
- Introduce professional resources for practitioners and educational resources for patients
- Review clinical tools to aid in nutrition assessment of pediatric renal patients

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FINDING YOUR WAY

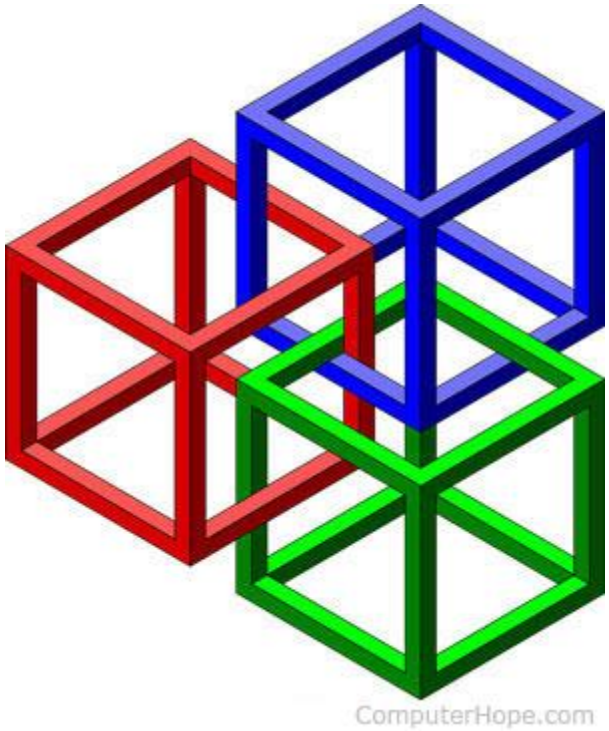
NAVIGATING PEDIATRIC NEPHROLOGY

Regulatory Guidelines

Professional Practise

Patient Resources

Knowledge/ Networking



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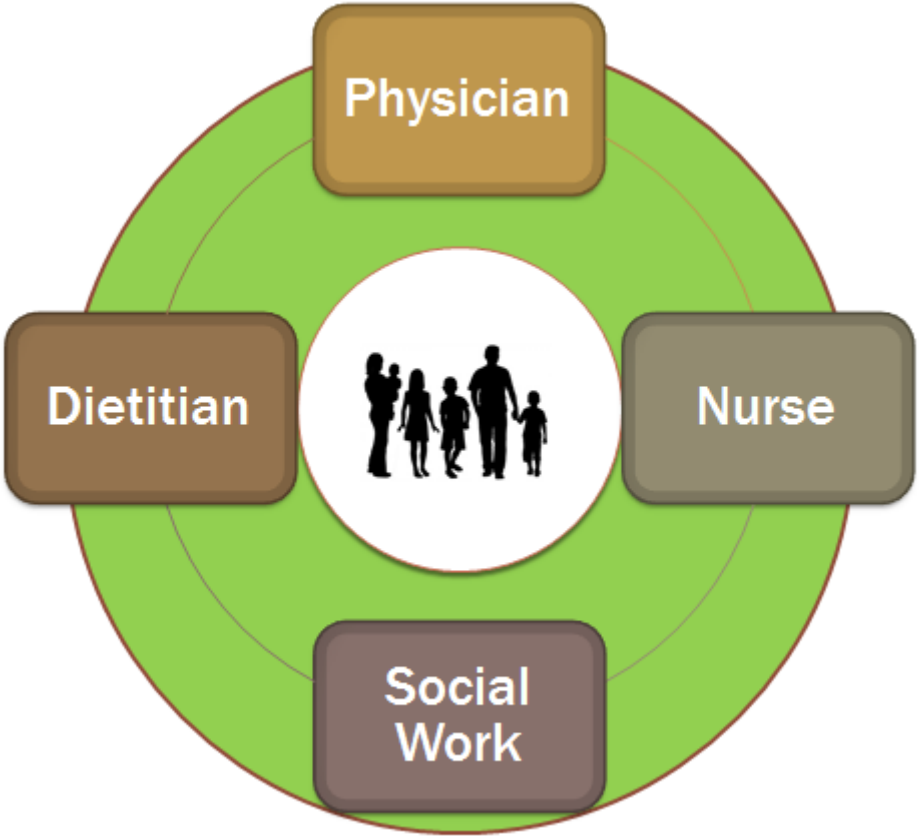
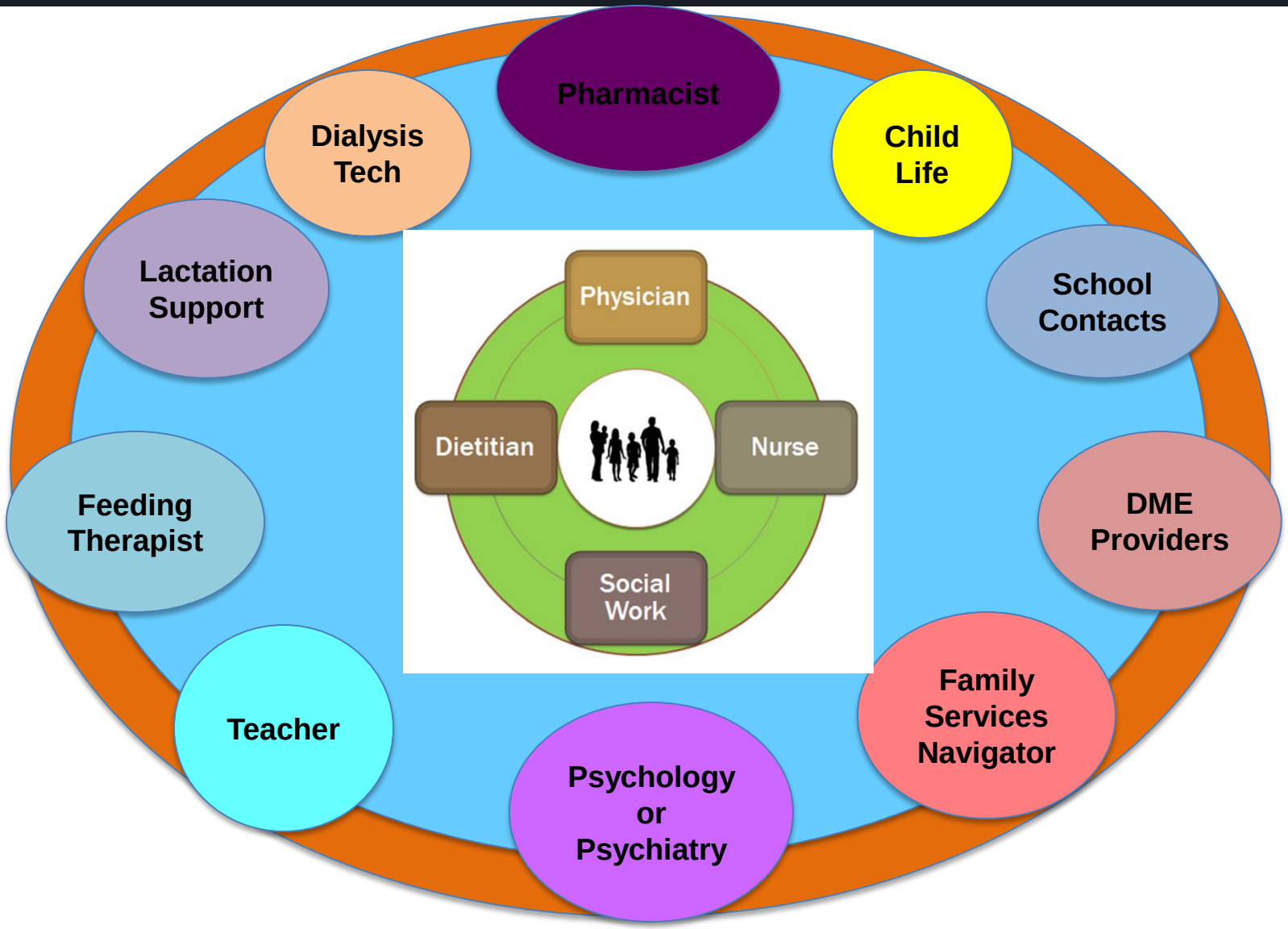
THE RENAL TEAM



MANDATED TEAM MEMBERS (4)

- Patient and family – most important team member
- Physician – attending, fellow → **ultimate responsibility for plan**
- Nurse – most frequent contact with patient
- Social Worker – quality of life & social needs of child
- Dietitian – nutrition expert & advocate

THE RENAL TEAM… EXTENDED



REGULATORY INFORMATION

Center for Medicare & Medicaid Services:

MEASURES ASSESSMENT TOOL (MAT)

Tag	Condition/Standard ➡	Measure ➡	Values ➡	Reference ➡	Source
484.40 Water and dialysate quality:					
V196 V178 V180	Water quality; test for total chlorine Water & dialysate quality/test for microbiological contamination	Max. total chlorine (includes chloramines) Action / Max. bacteria – product water / dialysate Action / Max. endotoxin – product water / dialysate	≤0.1 mg/L daily/shift 50 CFU/mL / <200 CFU/mL 1 EU/mL / <2 EU/mL (endotoxin units)	AAMI RD52	Records
484.50 Reuse of hemodialyzers and blood lines (only applies to facilities that reuse dialyzers &/or bloodlines)					
V336	Dialyzer effectiveness	Total cell volume (TCV) of (hollow fiber) dialyzers	Measure original volume/TCV Discard if after reuse <80% of original TCV	KDOQI HD Adequacy 2006 AAMI RD47	Records Interview
484.60 Patient assessment: The interdisciplinary team (IDT), patient/designee, RN, MSW, RD, physician must provide each patient with an individualized & comprehensive assessment of needs					
V502 V503 V504 V505 V506 V507 V508 V509 V510 V511 V512 V513 V514 V515	- Health status/comorbidities - Dialysis prescription - BP & fluid management - Lab profile - Immunization & meds history - Anemia (Hgb, Hct, iron stores, ESA need) - Renal bone disease - Nutritional status - Psychosocial needs - Dialysis access type & maintenance - Abilities, interests, preferences, goals, desired participation in care, preferred modality & setting, expectations for outcomes - Suitability for transplant referral - Family & other support systems - Current physical activity level & referral to vocational & physical rehabilitation	- Medical/nursing history, physical exam findings - Evaluate: HD every mo; PD first mo & q 4 mo - Interdialytic BP & wt gain, target wt, symptoms - Monitor labs monthly & as needed - Pneumococcal, hepatitis, influenza; med allergies - Volume, bleeding, infection; ESA hypo-response - Calcium, phosphorus, PTH & medications - Multiple elements listed - Multiple elements listed - Access efficacy, fistula candidacy - Reason why patient does not participate in care, reason why patient is not a home dialysis candidate - Reason why patient is not a transplant candidate - Composition, history, availability, level of support - Abilities & barriers to independent living; achieving physical activity, education & work goals	Refer to Plan of care & QAPI sections (below) for values	Conditions for Coverage KDOQI Guidelines (see POC)	Chart Interview
484.60 Plan of care The IDT must develop & implement a written, individualized comprehensive plan of care that specifies the services necessary to address the patient's needs as identified by the comprehensive assessment & changes in the patient's condition, & must include measurable & expected outcomes & estimated timetables to achieve outcomes. Outcome goals must be consistent with current professionally accepted clinical practice standards.					
V543	(1) Dose of dialysis/volume status Monitor each treatment	Management of volume status	Euvolemic & pre-BP <140/90; post-BP <130/80 (adult); lower of 90% of normal for age/ht/wt or 130/80 (pediatric)	KDOQI HD Adequacy 2006 KDOQI Cardiovascular 2005	Chart Interview
V544	(1) Dose of dialysis (HD adequacy) Monitor adequacy monthly	Adult HD >5 hours 3x/week, minimum spKt/V Adult HD 2x/week, RKF <2 mL/min HD 2, 4-6x/week, minimum stdKt/V	≥1.2 (or URR≥65); Min. 3 hours/tx if RKF <2ml/min Inadequate treatment frequency ≥2.0/week	NQF #0249 (adult) NQF #1423 (peds) KDOQI HD Adequacy 2006	Chart Interview
V544	(1) Dose of dialysis (PD adequacy – adult) Monitor 1st month & every 4 months	Minimum delivered Kt/V_{urea}	≥1.7/week	NQF #0318 KDOQI PD Adequacy 2006	Chart Interview
V544	(1) Dose of dialysis (PD adequacy – pediatric) Monitor 1st month & every 6 months	Minimum delivered Kt/V_{urea}	≥1.8/week	KDOQI PD Adequacy 2006	Chart Interview
V545	(2) Nutritional status - Monitor albumin & body wt monthly; monitor other parameters at V509 as needed	Albumin Body weight & other parameters listed at V509	≥4.0 g/dL BCG preferred; if BCP: lab normal % usual wt, % standard wt, BMI, est. % body fat	KDOQI Nutrition 2000 KDOQI CKD 2002	Chart Interview
V545	(2) Nutritional status (pediatric) monitor monthly	Length/ht-for-age % or SD, dry wt & wt-for-age % or SD, BMI-for-ht/age % or SD, head circ/age % (age <3), nPCR	nPCR normalized HD teen (nPCR and albumin are not predictive of wt loss/nutritional status in younger children)	KDOQI Pediatric Nutrition 2008	Chart Interview
V546	(3) Mineral metabolism & renal bone disease Monitor calcium & phosphorus monthly Monitor intact PTH every 3 months	Calcium corrected for albumin (BCG) Phosphorus Intact PTH (consider with other MBD labs, not in isolation)	Normal for lab; preferred upper level <10.2 mg/dL[1] All: 3.5-5.5 mg/dL[2] Under review	NQF #1454 KDIGO CKD-MBD 2009	Chart Interview
V547	(4) Anemia – Hgb non-ESA - monitor monthly	Hemoglobin (Adult & pediatric)	No upper level established[3] See Hgb on ESA (below) for management of anemia[3]	FDA 8/24/11 for more info re CKD 5D recommendation	Chart Interview
V547 V548	(4) Anemia – Hgb on ESA – monitor weekly until stable; then monitor monthly; evaluate other anemia causes; educate patients about risks/benefits	Hemoglobin (Adult & pediatric) Blood transfusion	Initiate ESAs <10 g/dL; interrupt or ↓ dose near or >11 g/dL[3] Give lowest dose of ESAs to avoid transfusion (especially in transplant candidates); consider patient preference	FDA 8/24/11 for more info re CKD 5D recommendation	Chart Interview
V549	(4) Anemia - Monitor iron stores routinely	Adult & pediatric: transferrin saturation Adult & pediatric: serum ferritin	<20% (HD, PD), or CHr >29 pg/cell HD: >200 ng/mL; PD: >100 ng/mL HD/PD: <500 ng/mL or evaluate if indicated	KDOQI Anemia 2006	Chart Interview

Sources: DFR=Dialysis Facility Reports; OM=CROWNWeb; Chart=Patient Chart; Records=Facility Records; Interview=Patient/Staff Interview; Abbreviations: BCG/BCP=bromcresol green/purple; BMI=Body mass index; CAHPS=Consumer Assessment of Healthcare Providers & Services; CFU=colony forming units; CHr=reticulocyte hemoglobin; CMS CPM=CMS Clinical Performance Measure; DOPPS=Dialysis Outcomes & Practice Patterns Study; ESA=erythropoiesis stimulating agent; KDIGO=Kidney Disease Improving Global Outcomes; KDOQI=Kidney Disease Outcomes Quality Initiative; nPCR=normalized protein catabolic rate; NQF=National Quality Forum; RKF=residual kidney function; SD=standard deviation; spKt/V=single pool Kt/V

Measures Assessment Tool:

What standard is measured?
How it is measured?
Specific goal of the measure
Source reference for that measure

Measures Assessment Tool- Dietitian

494.80 Patient Assessment	• V508 – Renal bone disease • V509 – Nutrition status
494.90 Plan of Care	• V545 – Nutrition status • V546 – Mineral & bone disease
494.110 Quality Assessment & Performance Improvement	• V630 – Nutrition status • V631 – Mineral & bone disease

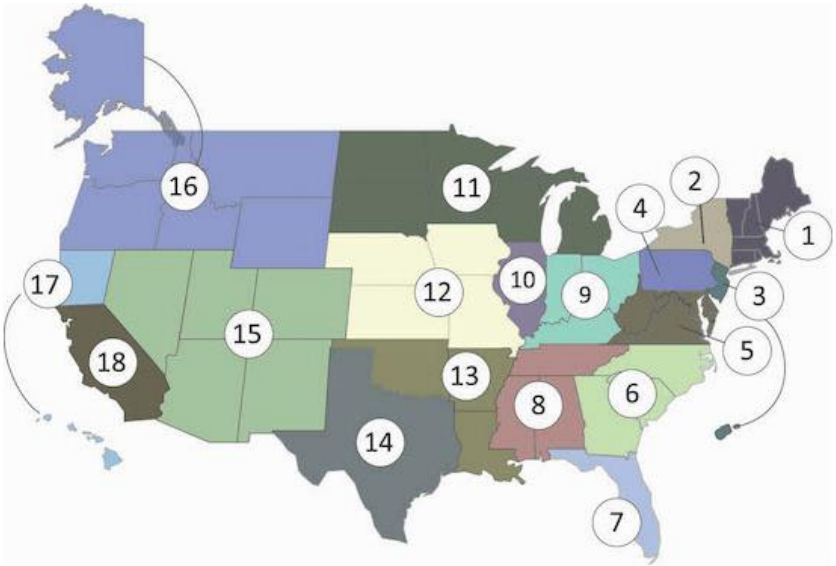
3 major phases of care

2 repeating items that are addressed

GUIDANCE FROM CMS – DIALYSIS

- V501 establishes an *interdisciplinary team* that includes a dietitian
- V509 requires the *evaluation of nutritional status* by a dietitian as part of the patient's comprehensive assessment
- V520 outlines the requirements for a patient's care plan and defines classification and follow-up for stable vs. unstable patients
- V545 states that a patient's albumin level and body weight must be measured monthly in an effort to achieve and sustain an effective nutritional status
- V546 (*pediatric*) states nPCR be monitored monthly in HD teens
- V630 requires an ongoing QA program that tracks indicators of nutrition status
- V689 and V690 state that the dietitian must be registered with CDR and have 1 year professional work experience in clinical nutrition
- V758 states that the dietitian must be available to meet patient needs

REGULATORY - USA

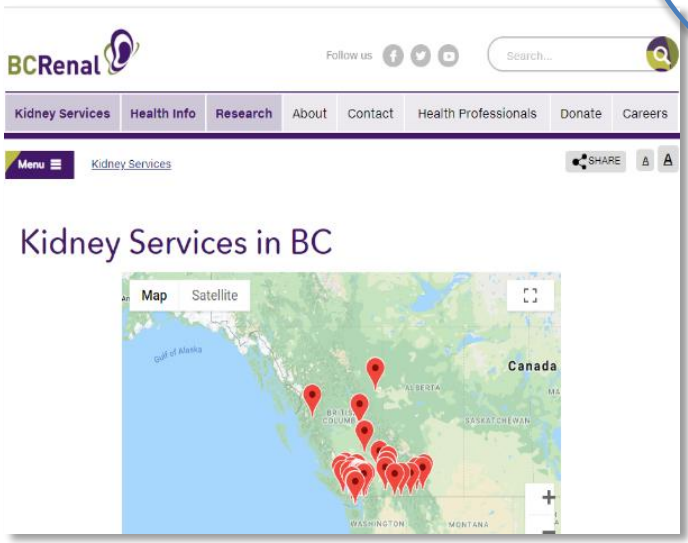


THE NATIONAL FORUM OF ESRD NETWORKS

The Forum of ESRD Networks is a not-for-profit organization that advocates on behalf of its membership and coordinates projects and activities of mutual interest to ESRD Networks. All 18 Networks are members of the Forum which facilitates the flow of information and advances a national quality agenda with CMS and other renal organizations.

http://esrdnetworks.org

REGULATORY - OTHER



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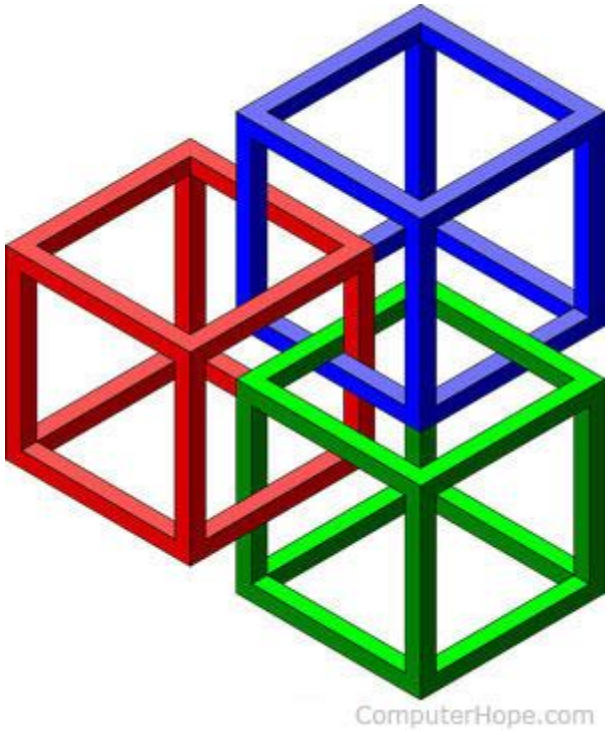

Standards of Practice & Professional Performance

Academy of Nutrition and Dietetics and National Kidney Foundation: Revised 2014 Standards of Practice and Standards of Professional Performance for Registered Dietitian Nutritionists (Competent, Proficient, and Expert) in Nephrology Nutrition

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Standards of Practice & Professional Performance

Academy of Nutrition and Dietetics: Revised 2015 Standards of Practice and Standards of Professional Performance for Registered Dietitian Nutritionists (Competent, Proficient, and Expert) in Pediatric Nutrition

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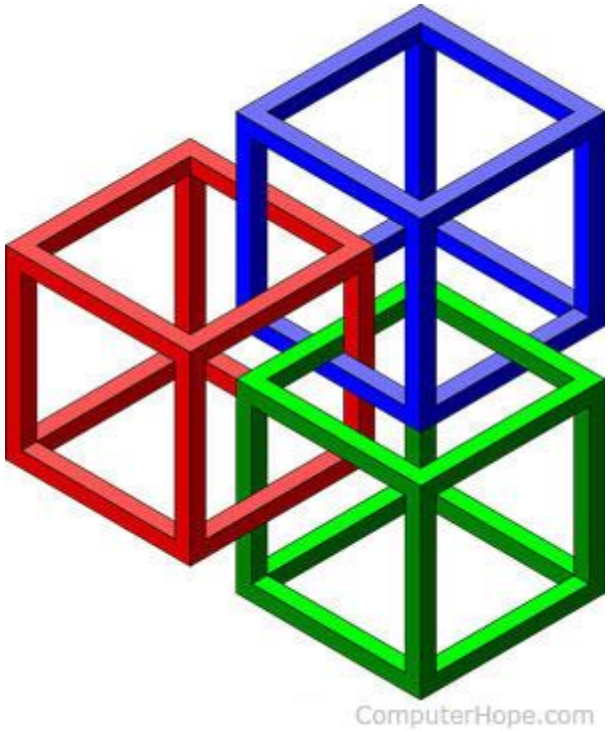
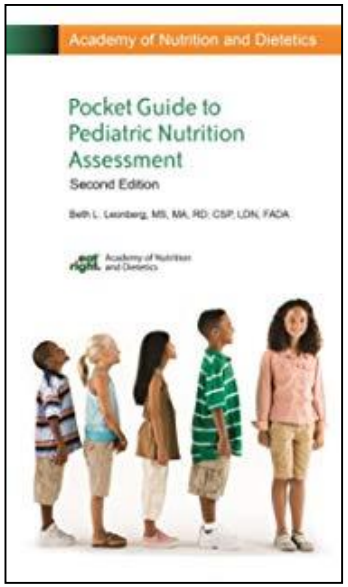
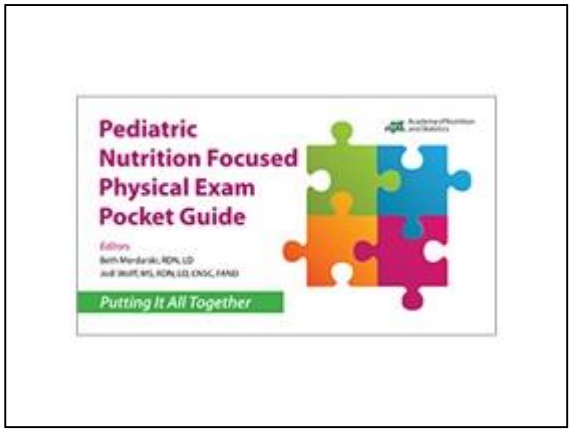
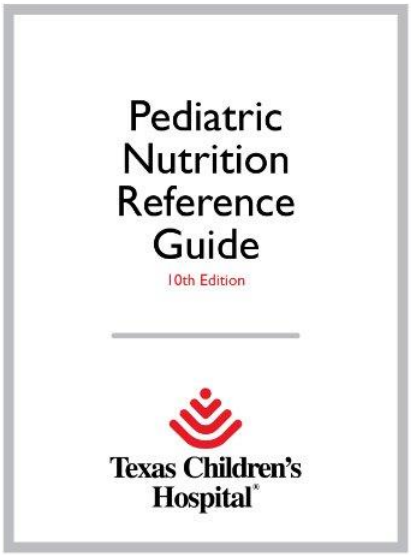


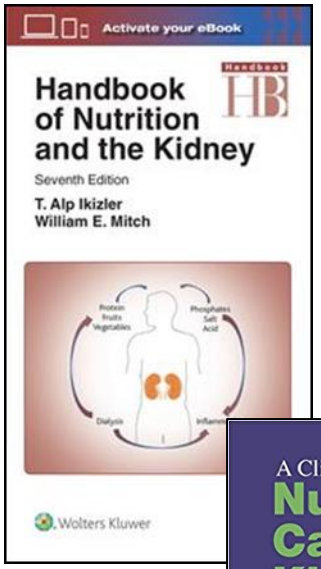
Handbooks and Pocket Guides- Pediatrics

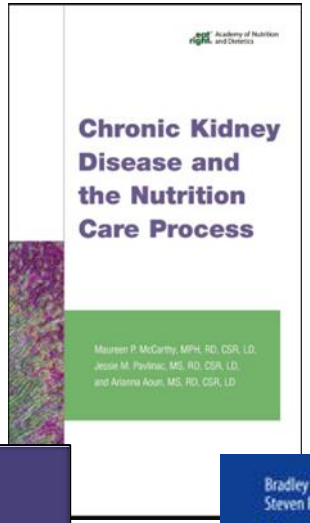


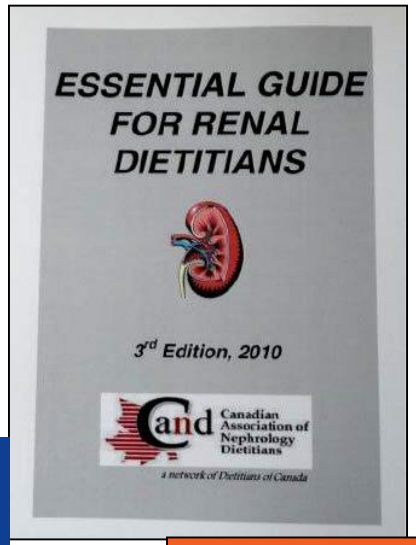


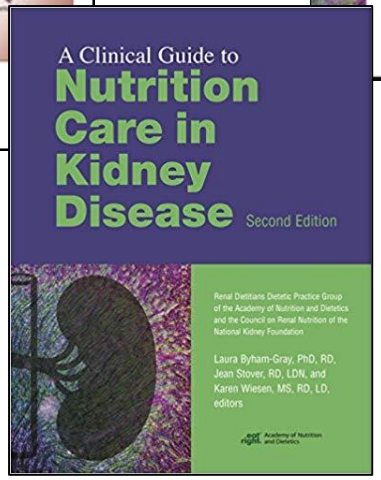


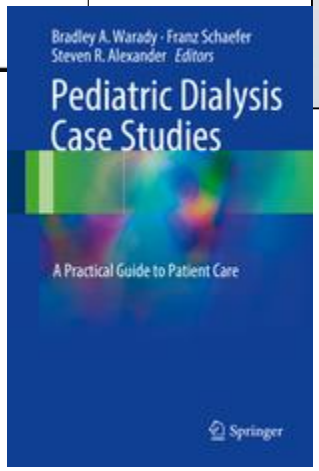
Textbooks, Handbooks, Guides: *Renal + Pediatrics*

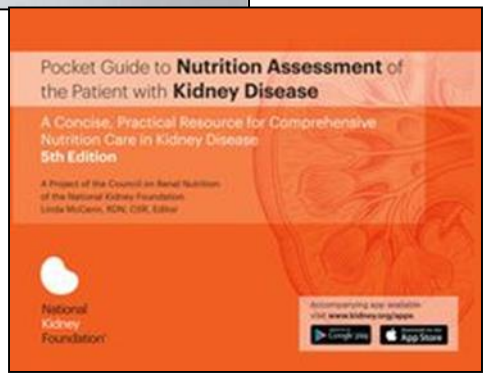




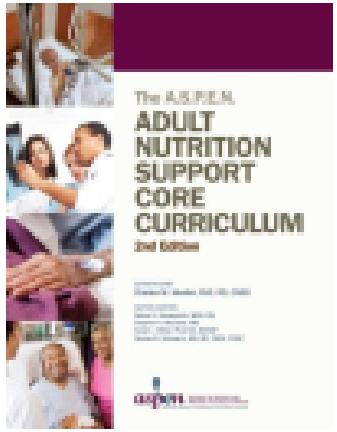


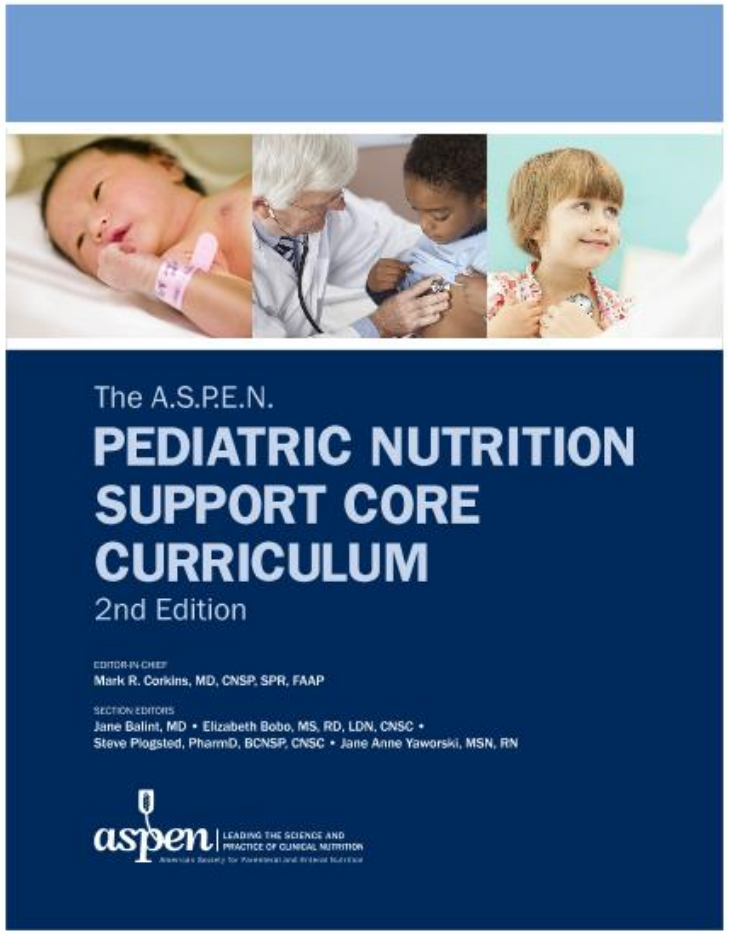




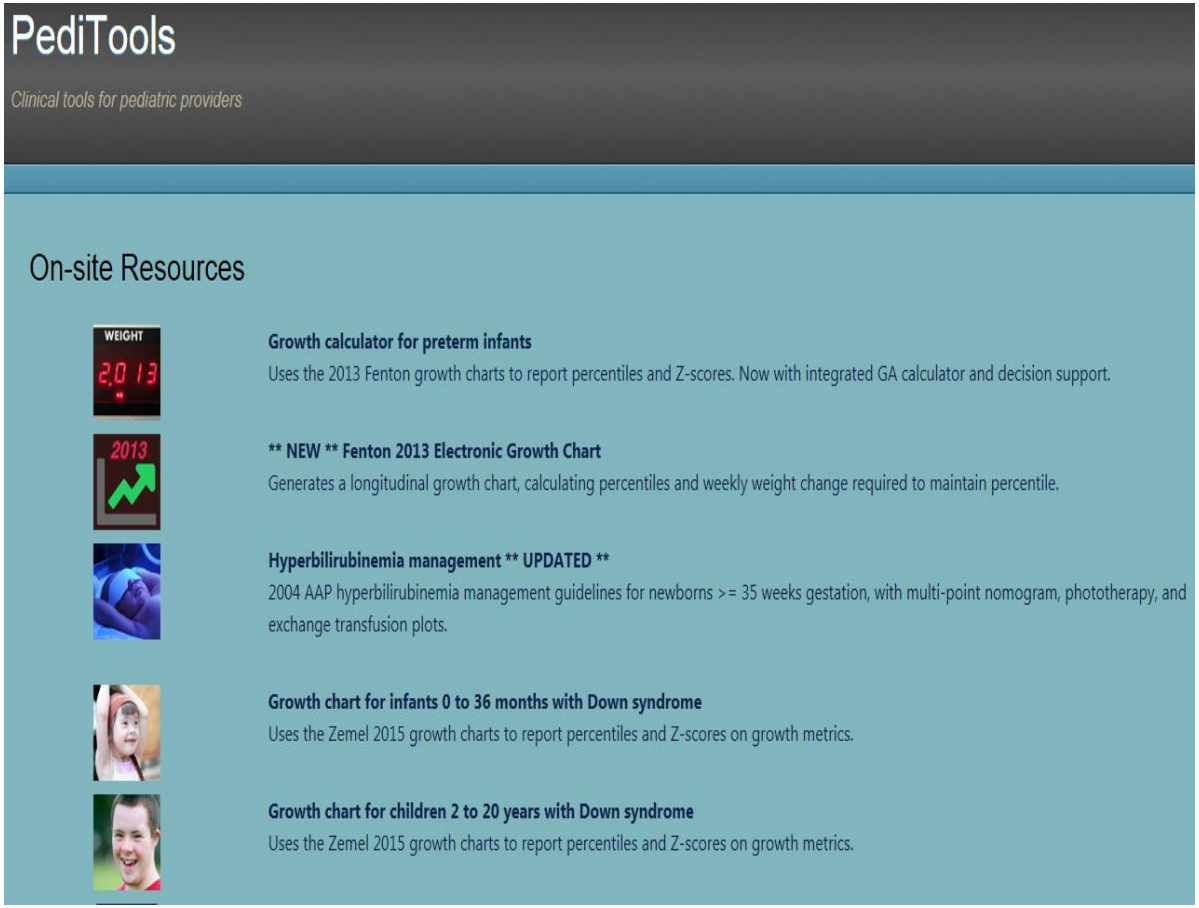


- **Renal Disease**
  Christina L. Nelms, MS, RD, CSP, LMNT
  Marisa Juarez, MPH, RD, LD
  Bradley A. Warady, MD





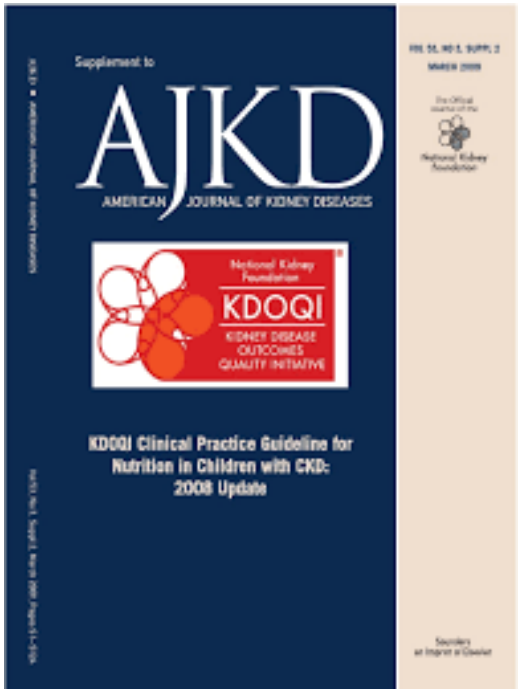
ASSESSMENT - Peditools



- Fenton – preterm infants
- Down Syndrome
- WHO
- CDC
- MUAC
- WHO arm circumference
- Skinfolds (triceps, subscapular)
- Parenteral nutrition

Kidney Disease Outcomes Quality Initiative (KDOQI)

KDOQI Clinical Practice Guidelines for Nutrition in Children with CKD: 2008 Update



RECOMMENDATIONS

NEW Clinical Practise Recommendations (CPRs)

Pediatric Renal Nutrition Taskforce (PRNT):

Dietitians and Nephrologists – Europe and North America

Since 2017….reviewing literature and publishing clinical practise recommendations.

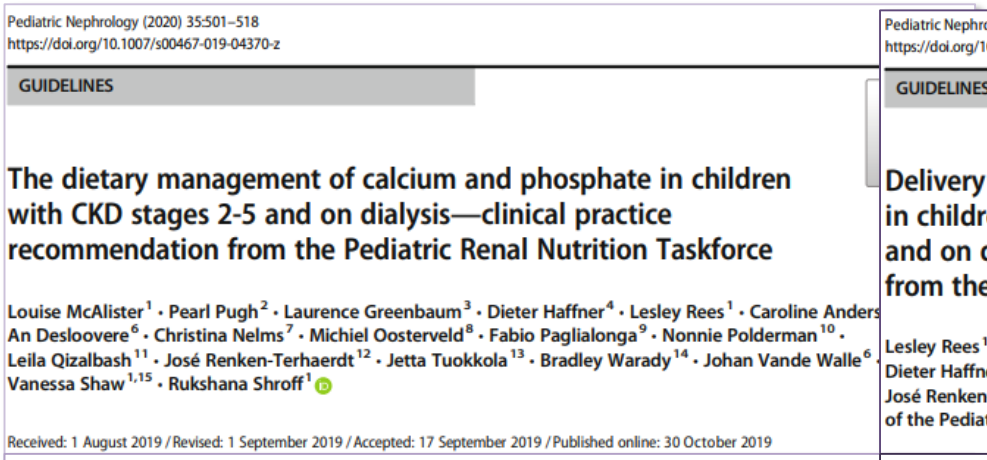
Published:

- Calcium and Phosphate
- Energy and Protein
- Assessment
- Delivery of Nutrition prescription
- Potassium management

Additional CPRs Coming soon!

- Obesity and Metabolic Syndrome
- Acute Kidney Injury
- Transplant

Clinical Practise Recommendations (CPRs)



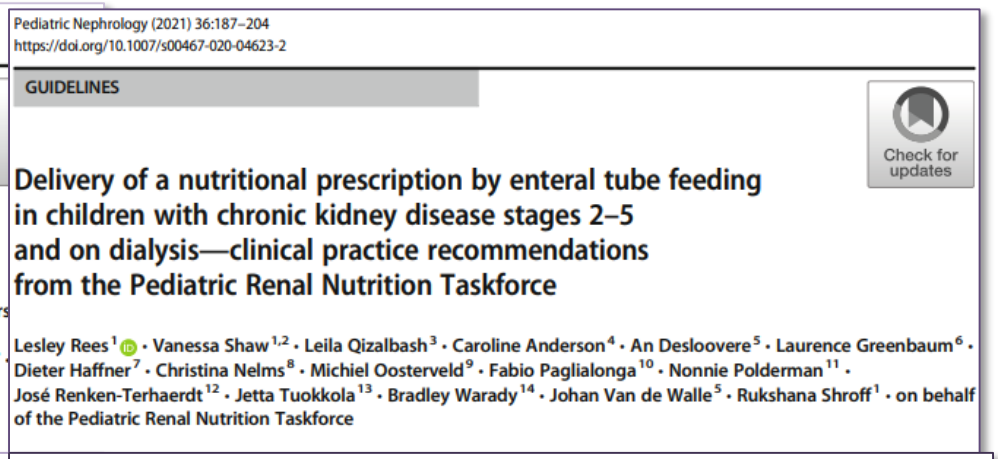
Pediatric Nephrology (2020) 35:501–518
https://doi.org/10.1007/s00467-019-04370-z

GUIDELINES

The dietary management of calcium and phosphate in children with CKD stages 2-5 and on dialysis—clinical practice recommendation from the Pediatric Renal Nutrition Taskforce

Louise McAlister[1] · Pearl Pugh[2] · Laurence Greenbaum[3] · Dieter Haffner[4] · Lesley Rees[1] · Caroline Anders
An Desloovere[6] · Christina Nelms[7] · Michiel Oosterveld[8] · Fabio Paglialonga[9] · Nonnie Polderman[10] ·
Leila Qizalbash[11] · José Renken-Terhaerdt[12] · Jetta Tuokkola[13] · Bradley Warady[14] · Johan Vande Walle[6] ·
Vanessa Shaw[1,15] · Rukshana Shroff[1]

Received: 1 August 2019 / Revised: 1 September 2019 / Accepted: 17 September 2019 / Published online: 30 October 2019



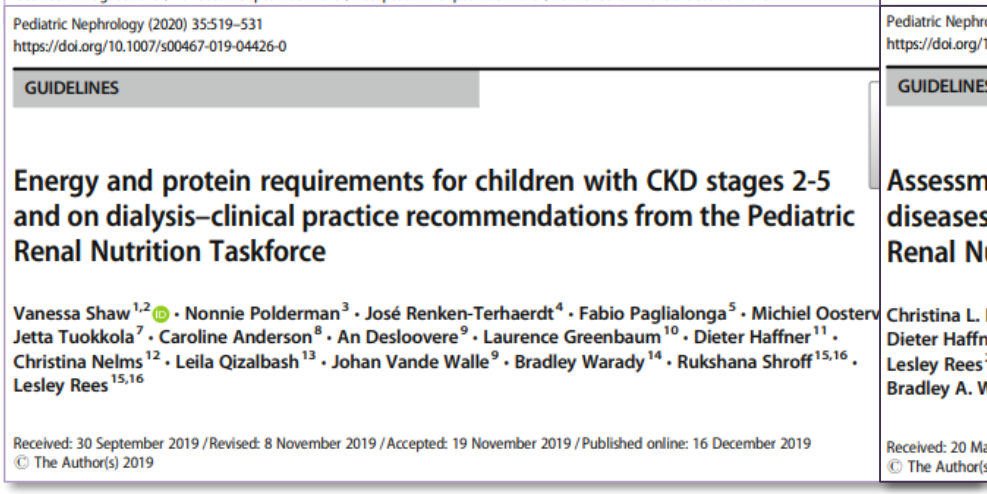
Pediatric Nephrology (2021) 36:187–204
https://doi.org/10.1007/s00467-020-04623-2

GUIDELINES

Check for updates

Delivery of a nutritional prescription by enteral tube feeding in children with chronic kidney disease stages 2–5 and on dialysis—clinical practice recommendations from the Pediatric Renal Nutrition Taskforce

Lesley Rees[1] · Vanessa Shaw[1,2] · Leila Qizalbash[3] · Caroline Anderson[4] · An Desloovere[5] · Laurence Greenbaum[6] ·
Dieter Haffner[7] · Christina Nelms[8] · Michiel Oosterveld[9] · Fabio Paglialonga[10] · Nonnie Polderman[11] ·
José Renken-Terhaerdt[12] · Jetta Tuokkola[13] · Bradley Warady[14] · Johan Van de Walle[5] · Rukshana Shroff[1] · on behalf of the Pediatric Renal Nutrition Taskforce



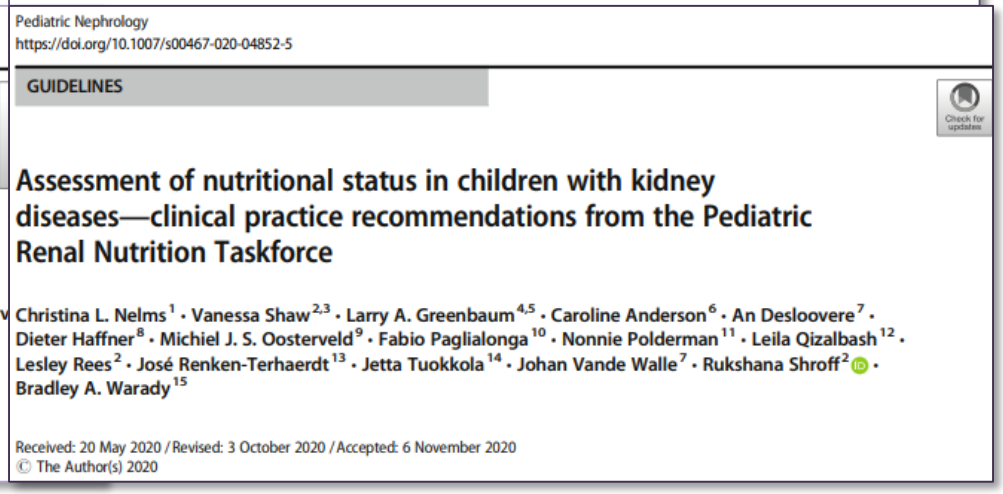
Pediatric Nephrology (2020) 35:519–531
https://doi.org/10.1007/s00467-019-04426-0

GUIDELINES

Energy and protein requirements for children with CKD stages 2-5 and on dialysis–clinical practice recommendations from the Pediatric Renal Nutrition Taskforce

Vanessa Shaw[1,2] · Nonnie Polderman[3] · José Renken-Terhaerdt[4] · Fabio Paglialonga[5] · Michiel Oosterv
Jetta Tuokkola[7] · Caroline Anderson[8] · An Desloovere[9] · Laurence Greenbaum[10] · Dieter Haffner[11] ·
Christina Nelms[12] · Leila Qizalbash[13] · Johan Vande Walle[9] · Bradley Warady[14] · Rukshana Shroff[15,16] ·
Lesley Rees[15,16]

Received: 30 September 2019 / Revised: 8 November 2019 / Accepted: 19 November 2019 / Published online: 16 December 2019



Pediatric Nephrology
https://doi.org/10.1007/s00467-020-04852-5

GUIDELINES

Check for updates

Assessment of nutritional status in children with kidney diseases—clinical practice recommendations from the Pediatric Renal Nutrition Taskforce

Christina L. Nelms[1] · Vanessa Shaw[2,3] · Larry A. Greenbaum[4,5] · Caroline Anderson[6] · An Desloovere[7] ·
Dieter Haffner[8] · Michiel J. S. Oosterveld[9] · Fabio Paglialonga[10] · Nonnie Polderman[11] · Leila Qizalbash[12] ·
Lesley Rees[2] · José Renken-Terhaerdt[13] · Jetta Tuokkola[14] · Johan Vande Walle[7] · Rukshana Shroff[2] ·
Bradley A. Warady[15]

Received: 20 May 2020 / Revised: 3 October 2020 / Accepted: 6 November 2020

Published with Open Access

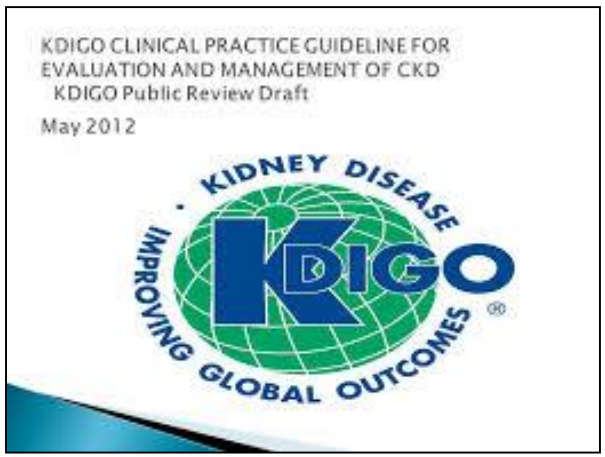
PRONA Pocket Guide

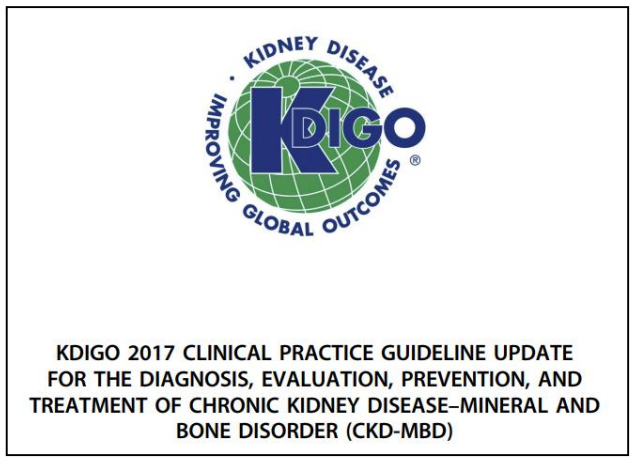
Pediatric **R**enal Dietitians **O**f **N**orth **A**merica

https://pixabay.com/illustrations/under-construction-construction-sign-2408066/

Kidney Disease Improving ***Global*** Outcomes (KDIGO)

Kidney Disease: Improving Global Outcomes (KDIGO) was originally established in 2003 by the National Kidney Foundation, a U.S. foundation experienced in developing and implementing guidelines. In 2013 KDIGO became an independently incorporated non-profit foundation, and is governed by an international volunteer Executive Committee.



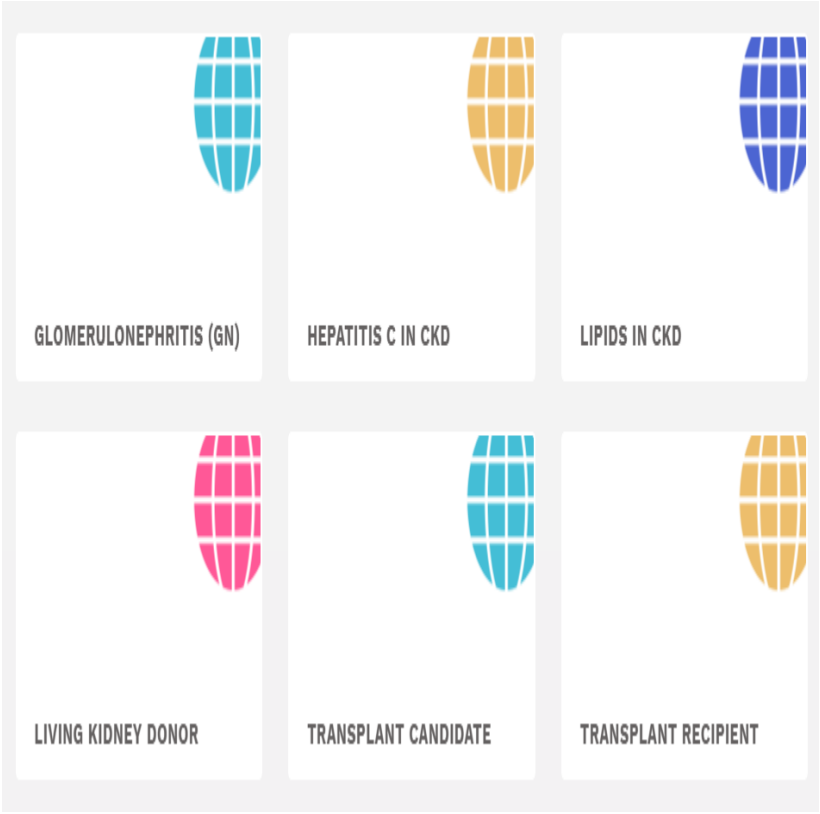


KDIGO guidelines translate the best worldwide scientific evidence into practical recommendations for clinicians and patients.

KDIGO

Eg.:
*KDIGO Clinical Practice Guideline for Evaluation and Management of CKD: 2012

*KDIGO 2017 Clinical Practice Guideline Update for the diagnosis, evaluation, prevention and treatment of CKD – (CKD-BMD)



TRIALS AND STUDIES

- Large studies that are relevant to practice…..

CKiD Study

Chronic Kidney Disease in Children

Home | Study Information | Study Administration | Investigator Resources | Coordinator's Corner | Psychologist's Corner | Family Corner

About CKiD

The CKiD Study is a NIH-funded, multicenter, prospective cohort study of children aged 6 months to 16 years with mild to moderate impaired kidney function. The primary goals of CKiD are to determine the risk factors for decline in renal function and to define how progressive decline in renal function impacts biomarkers of risk factors for cardiovascular disease; neurocognitive function and behavior; and growth failure and its associated morbidity. Two clinical coordinating centers (CCCs) (at Children's Hospital of Philadelphia and at Children's Mercy Hospital in Kansas City), a central biochemistry laboratory (at the University of Rochester), and a data coordinating center (at Johns Hopkins School of Public Health) formed a cooperative agreement to conduct the CKiD Study.

Study Aims

The specific aims are to:

- Identify novel and traditional renal disease risk factors for the progression of CKD (e.g. decline of GFR) in children
- Characterize the impact of a decline in kidney function on neurodevelopment, cognitive abilities, and behavior
- Identify the prevalence and evolution of traditional and novel cardiovascular disease risk factors in progressive CKD
- Examine the effects of declining GFR on growth and the treatment of growth failure, and to assess the consequences of growth failure on morbidity in children with CKD

CKiD Publications by Year

2006 2007 2008 2009 2010 2011 2012 2013 2014 2015 2016 2017 2018

2018 Publications

Barletta G, Pierce C, Mitsnefes M, Samuels J, Warady B, Furth S, Flynn J. Is blood pressure improving in children with chronic kidney disease?: a period analysis. Hypertension 2018;71:444-450. PMCID: PMC5812788

Brooks ER, Haymond S, Rademaker A, Pierce C, Helenowski I, Passman R, Vicente F, Warady BA, Furth SL, Langman CB. Contribution of symmetric dimethylarginine to GFR decline in pediatric chronic kidney disease. Pediatr Nephrol 2018;33:697-704. PMID: 29214443

Furth SL, Pierce C, Hui WF, White CA, Wong CS, Schaefer F, Wuehl E, Abraham A, Warady BA. Estimating time to ESRD in children with CKD. Am J Kidney Dis 2018; 71:783-792. PMCID: PMC5970998 (Letter to the Editor by KW Choy pii:S0272-6386(18)30758-3; Response on pii: S0272-6386(18)30757-1).

Ku E, McCulloch CE, Warady BA, Furth SL, Grimes BA, Mitsnefes M. Twenty-four-hour Ambulatory Blood Pressure Versus Clinic Blood Pressure Measurements and Risk of Adverse Outcomes in Children with CKD. CJASN 2018;13:422-428. PMID: 29440119

North American Pediatric Renal Trials and Collaborative Studies

NAPRTCS Online

Resources

- Home
- Announcements
- Resources
- Study Documents
- Directory
- Study Login
- Training System
- Coordinators
- Benchmark Project
- CHA-QI
- Other Information

Advantage eClinical Training Video

Advantage eClinical Quick Reference Guide

Data System Access Specification Form

Frequently Asked Questions (FAQ)

Growth Chart Calculator

HIPPA Sample Authorization Form

NAPRTCS Center in Good Standing Criteria

NAPRTCS PCC Newsletter

NAPRTCS PCC Operations Manual

Organizational Chart

Presentations from Annual Meeting

Special Studies Application with Instructions

Tax Payer ID Form

Training Manual

https://naprtcs.org/study-details/bibliography

https://web.emmes.com/study/ped/resources.htm

International Pediatric Dialysis Network

LOGIN
[Registration Form]
About IPDN
Network Participants
Links
IPDN Sponsors
Contact
Privacy Policy

About IPDN

The International Pediatric Dialysis Network is a global consortium of pediatric nephrology centers dedicated to the care of children on chronic dialysis. The IPDN aims to

- **improve the quality of pediatric dialysis care worldwide**
- **collect basic information regarding pediatric dialysis practices and outcomes**
- **provide useful tools and management algorithms for daily dialysis practice**
- **provide global benchmarking of pediatric dialysis outcomes**
- **perform prospective observational studies on important clinical issues in pediatric dialysis**

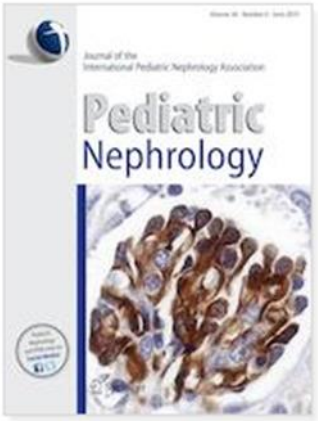
IPDN entertains two registries:
The IPPN registry for children on chronic peritoneal dialysis, and the IPHN registry for children on hemodialysis.If you would like to join the IPDN, please fill out the registration form. IPDN membership is free of charge. We grant institutional and individual memberships: With your institutional membership you have access to all information pages of the website.

At present,
245 institutions participate in the network
and
573 individual members actively contribute data to the network.

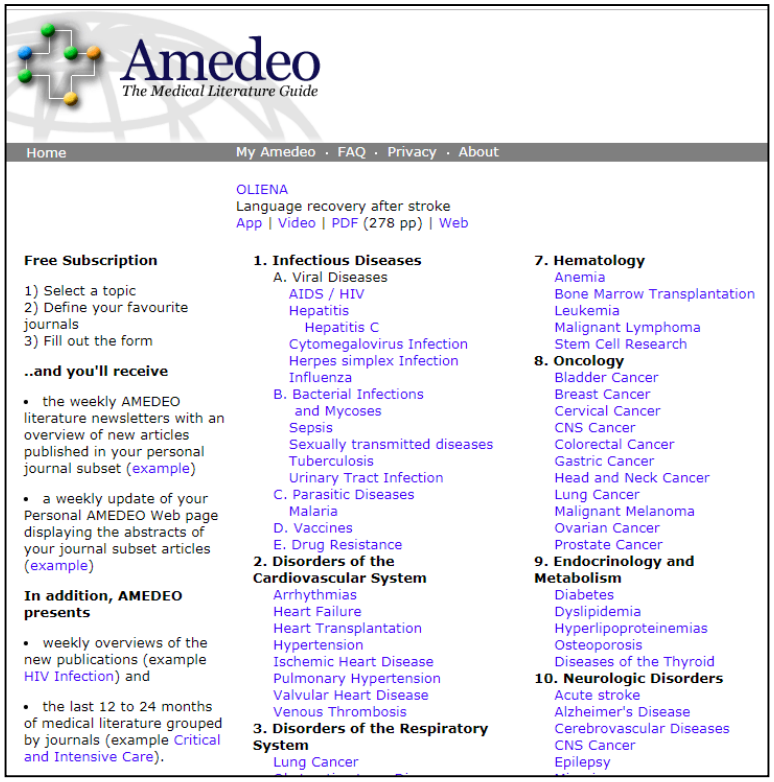
To date,
3662 patients have been enrolled in the IPPN Registry at 128 contributing centers in 43 countries
and
917 patients have been enrolled in the IPHN Registry at 84 contributing centers in 36 countries.

Relevant Journals

- Journal of Renal Nutrition
- Pediatric Nephrology
- Peritoneal Dialysis International
- American Journal of Kidney Disease
- Kidney International
- Clinical Journal of the American Society of Nephrology
- Nephrology News & Issues
- Nephrology, Dialysis, and Transpla



Literature Search Services – e.g.Amedeo



Renal
Nutrition
Pediatrics
Other interests

Weekly emails with bibliographic lists about new publications

Literature Search Services – e.g.Amedeo

We have screened the following journals for you:
Am J Kidney Dis
Am J Nephrol
BMJ
J Am Soc Nephrol
J Pediatr
J Ren Nutr
Kidney Int
Lancet
N Engl J Med
Nephrol Dial Transplant
Nephron
Pediatr Nephrol

11. LUBBE K, Nusken E, Rascher K, von Gersdorff G, et al.
Glomerular disease patients have higher odds not to reach quality targets in chronic dialysis compared with CAKUT patients: analyses from a nationwide German paediatric dialysis registry.
Pediatr Nephrol. 2019 Mar 6. pii: 10.1007/s00467-019-04218.
PubMed: www.amedeo.com/p2.php?id=30843113&s=crf&pm=a1b7681ac819d1b
ABSTRACT available
Share: http://m.amedeo.com/30843113

12. WONG VEGA M, Juarez Calderon M, Tufan Pekkucuksen N, Srivaths P, et al.
Feeding modality is a barrier to adequate protein provision in children receiving continuous renal replacement therapy (CRRT).
Pediatr Nephrol. 2019 Mar 6. pii: 10.1007/s00467-019-04211.
PubMed: www.amedeo.com/p2.php?id=30843114&s=crf&pm=a1b7681ac819d1b
ABSTRACT available
Share: http://m.amedeo.com/30843114

13. HWANG SH, Lee DH, Min J, Jeon JY, et al.
Handgrip Strength as a Predictor of All-Cause Mortality in Patients With Chronic Kidney Disease Undergoing Dialysis: A Meta-Analysis of Prospective Cohort Studies.
J Ren Nutr. 2019 Feb 28. pii: S1051-2276(19)30002.
PubMed: www.amedeo.com/p2.php?id=30827839&s=crf&pm=a1b7681ac819d1b
ABSTRACT available
Share: http://m.amedeo.com/30827839

Professional Networking

NKF: Council on Renal Nutrition
AND: Renal Practice Group
www.renalNutrition.org

AND: Pediatric Nutrition Practice Group

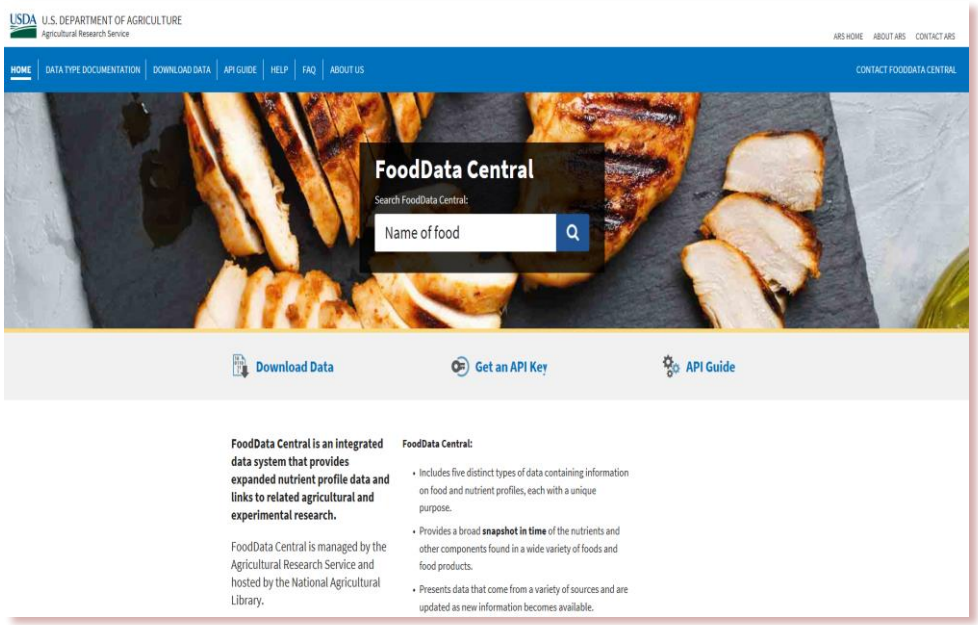
PRONA: Pediatric Renal Dietitians of North America
website is currently under development
www.prona.online

Adult Renal Listserv:
renalrd-request@mailman.srv.ualberta.ca

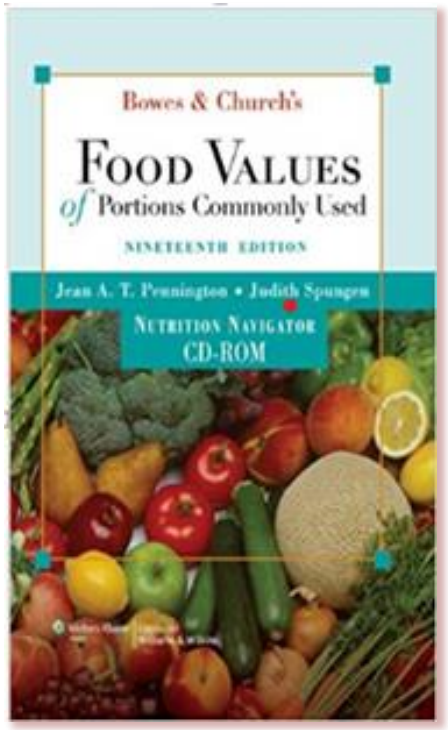
Peds Renal Listserv:
PedsRenalRD@mailman.srv.ualberta.ca

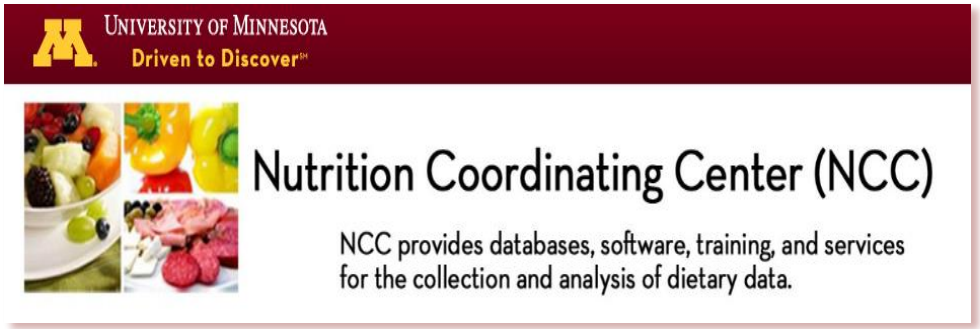
Peds Nutrition Listserv:
PEDI-RD@LIST.UIOWA.EDU

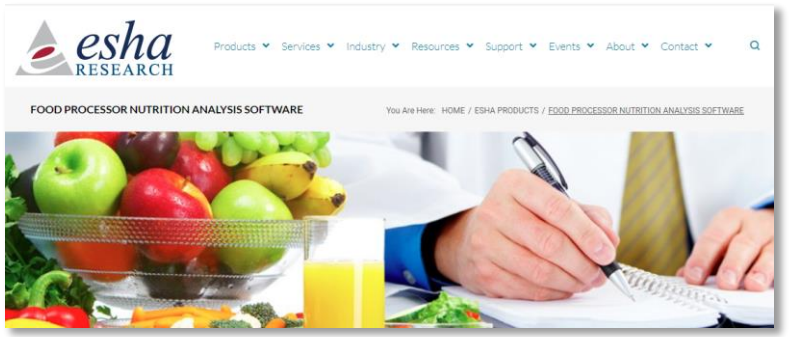
Diet Analysis



https://fdc.nal.usda.gov







BC Children's Hospital
An agency of the Provincial Health Services Authority

CALCULATORS

Conferences to Attend

Specific to Pediatric Renal Nutrition:

NKF – National Kidney Foundation – Spring Clinical Meeting

www.kidney.org/spring-clinical

WSPN – Western Society of Pediatric Nephrology

**does not always have a nutrition program*.

ASPN – American Society of Pediatric Nephrology –
Allied Health Symposium

1.5 day meeting - Las Vegas 2019

ADC – Annual Dialysis Conference
with pre-conference workshop: Fundamentals of Dialysis in Children

PRNA – Pediatric Renal Nutrition Academy

1-2 day events solely dedicated to Pediatric Renal Nutrition

NATCO – North American Transplant Coordinators Organization
www.natco1.org/education/nutrition-conference.asp

Learning Modules / Webinars

Modules:

eg. Abbott

Webinars:

eg. Vitaflo

https://anhi.org/education/course-catalog
https://education.kidney.org/
https://www.nestlehealthscience.us/vitaflo-usa/via/pediatric%20renal%20disease/pediatricrenal

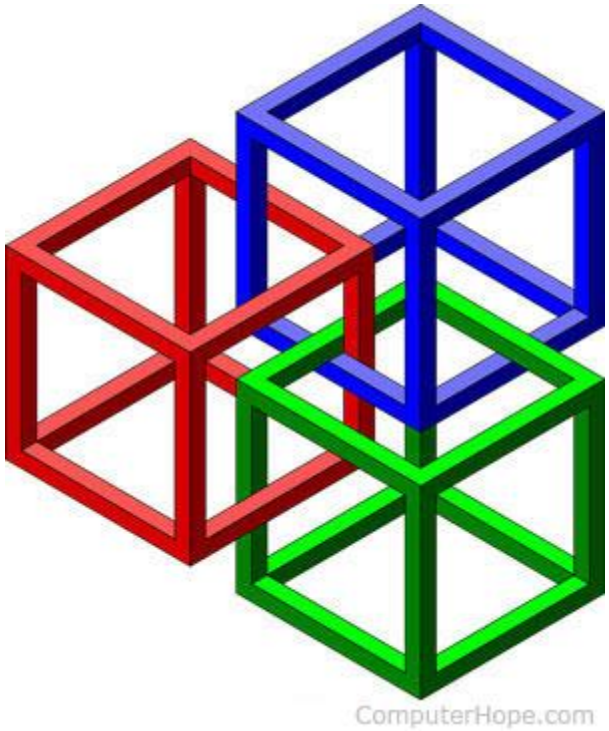
NAVIGATING PEDIATRIC NEPHROLOGY

Regulatory Guidelines

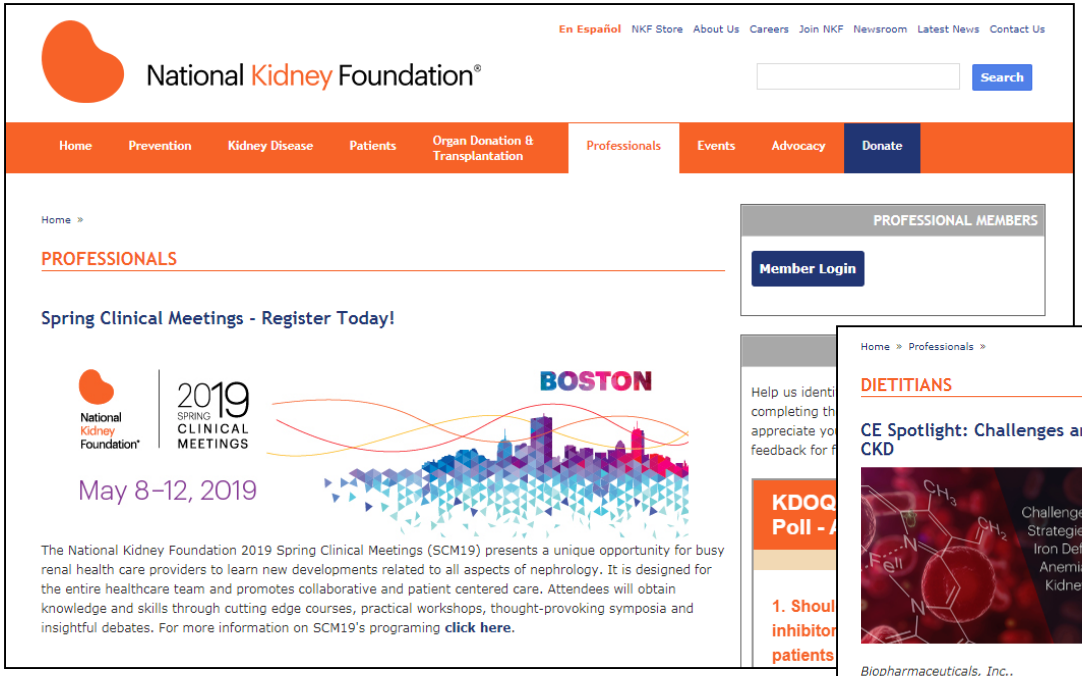
Professional Practise

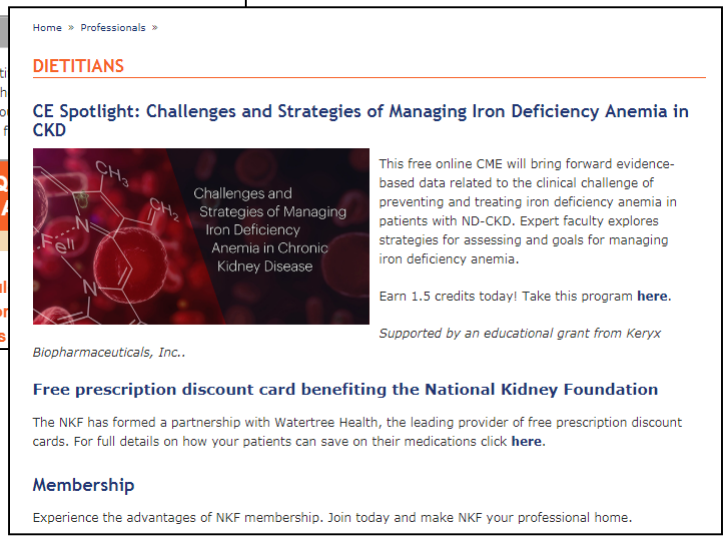
Knowledge/ Networking

Patient Resources

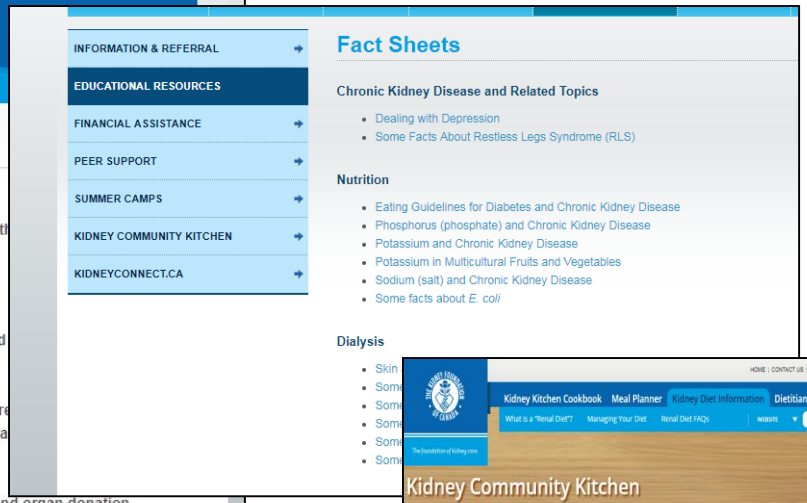


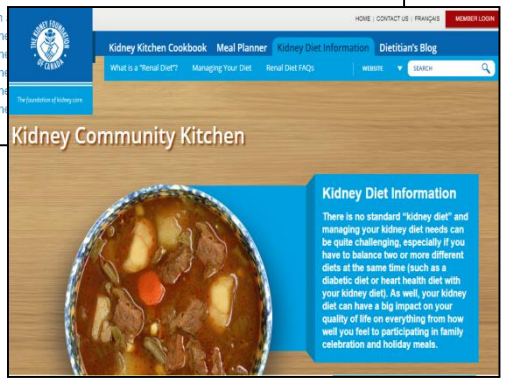
National Kidney Foundation- US



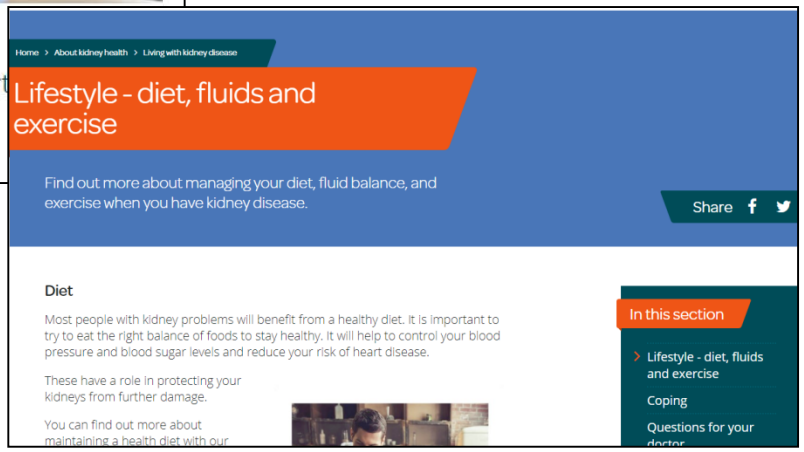


Kidney Foundation of Canada





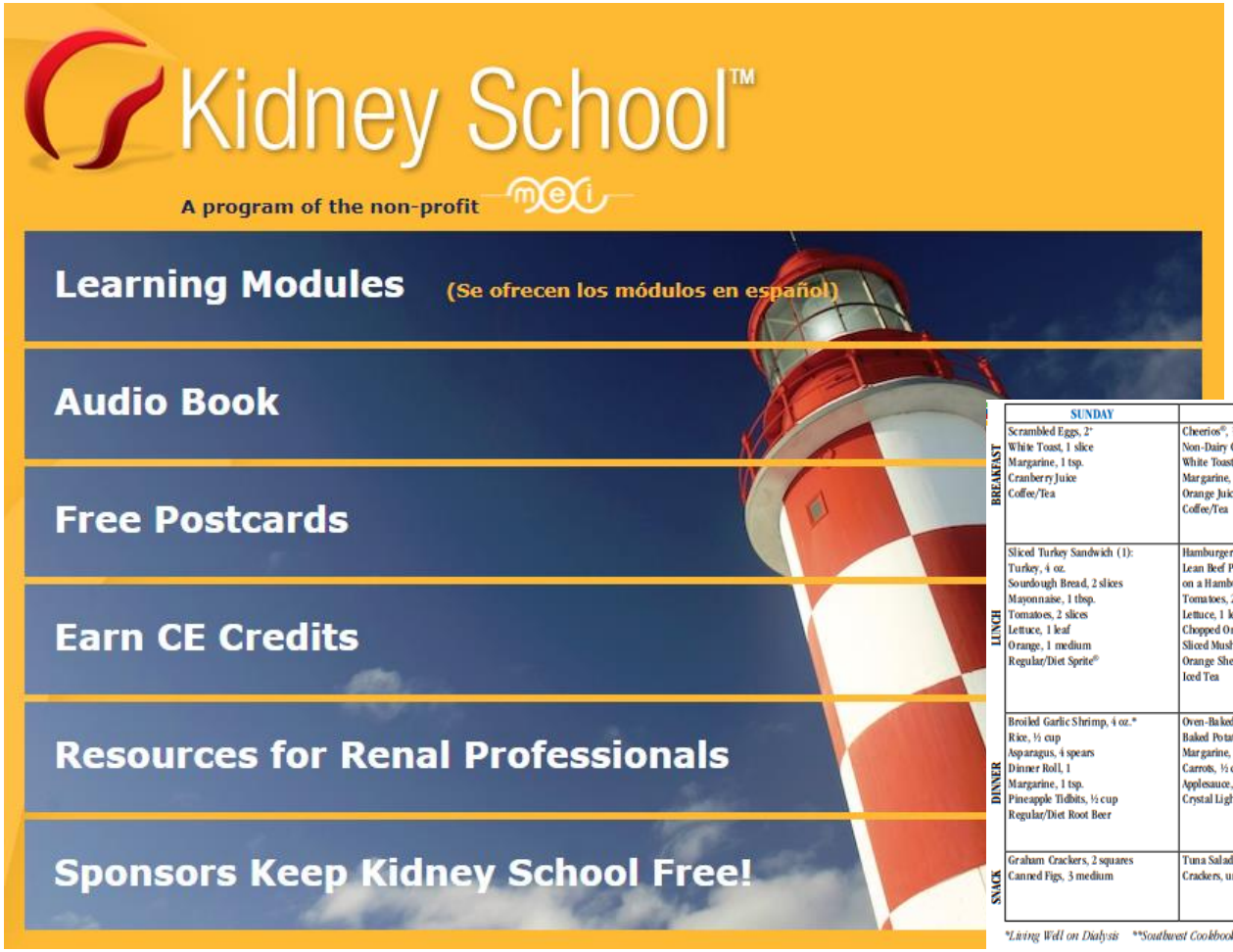
Kidney Care - UK



Kidney School

kidneyschool.org

Module on Nutrition

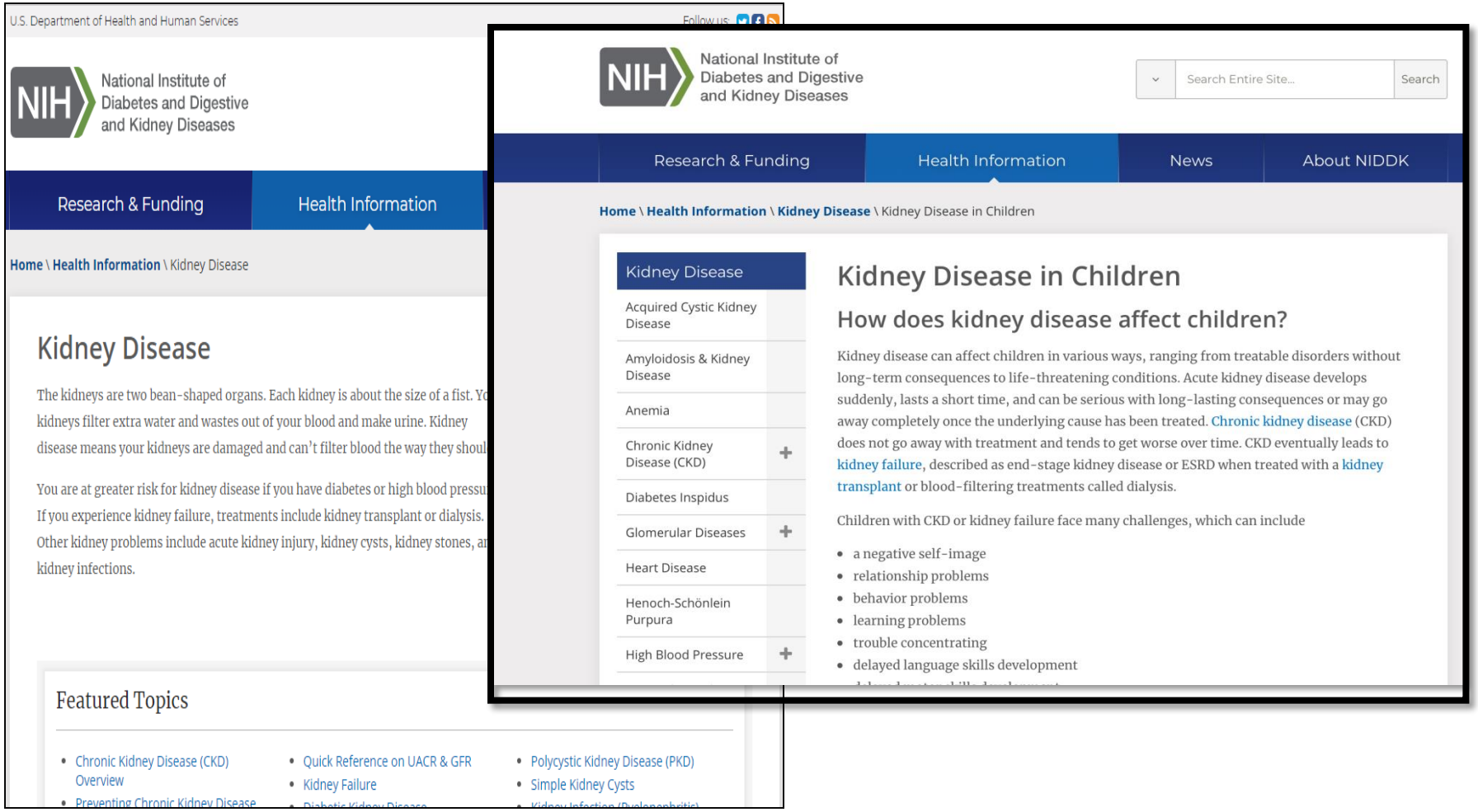


	SUNDAY	MONDAY	TUESDAY	WEDNESDAY
BREAKFAST	Scrambled Eggs, 2* White Toast, 1 slice Margarine, 1 tsp. Cranberry Juice Coffee/Tea	Cheerios®, ½ cup with Non-Dairy Creamer, ½ cup White Toast, 1 slice Margarine, 1 tsp. Orange Juice Coffee/Tea	French Toast, 2 slices: Eggs, 2* White Bread, 2 slices Margarine, 2 tsp. Maple Syrup, 4 tbsp. Strawberry/Banana Juice Coffee/Tea	Cornflakes, ½ cup with Non-Dairy Creamer, ½ cup Bagel, 1 Jam/Jelly, 2 tsp. or Cream Cheese, 2 tsp. Grapefruit, 1 whole Coffee/Tea
LUNCH	Sliced Turkey Sandwich (1): Turkey, 4 oz. Sourdough Bread, 2 slices Mayonnaise, 1 tbsp. Tomatoes, 2 slices Lettuce, 1 leaf Orange, 1 medium Regular/Diet Sprite®	Hamburger (1): Lean Beef Patty, 4 oz., on a Hamburger Bun Tomatoes, 2 slices Lettuce, 1 leaf Chopped Onion, 1 tbsp. Sliced Mushrooms, ¼ cup Orange Sherbet, ¼ cup Iced Tea	Grilled Salmon, 4 oz. Mexican Pasta, 1 cup** Corn Bread Roll, 1 Margarine, 1 tsp. Mixed Green Salad, ½ cup Oil and Vinegar Dressing: Salad/Olive Oil, 2 tsp. Vinegar, 1 tsp. Kiwi, 1 Regular/Diet Sprite®	Tuna Sandwich (1): Low Sodium/Water Packed Tu flaked, 4 oz. Mayonnaise, 1 tbsp. Chopped Onions, 1 tbsp. Chopped Celery, 1 tbsp. Swiss Cheese, 1 oz. Hard Bread Roll, 1 Apple, 1 medium Regular/Diet Root Beer
DINNER	Broiled Garlic Shrimp, 4 oz.* Rice, ½ cup Asparagus, 4 spears Dinner Roll, 1 Margarine, 1 tsp. Pineapple Tidbits, ½ cup Regular/Diet Root Beer	Oven-Baked Chicken, 4 oz. Baked Potato, 1 small Margarine, 2 tsp. Carrots, ½ cup Applesauce, ½ cup Crystal Light®	Salisbury Steak, 4 oz., with: Sliced Mushrooms, ½ cup Chopped Onions, ¼ cup Dinner Roll, 1 Margarine, 1 tsp. Artichokes, ½ cup Jell-O®, ½ cup Lemonade	Baked Pork Chop, 4 oz. Rice, ½ cup Steamed Broccoli, ½ cup Margarine, 1 tsp. Fresh Apricots, 4 Iced Tea
SNACK	Graham Crackers, 2 squares Canned Figs, 3 medium	Tuna Salad, ¼ cup Crackers, unsalted tops 6	Baked Apple with: Sugar, 2 tsp. Margarine, 2 tsp. Cinnamon, 1 tsp.	Chili Wheat Treats, ½ cup*

*Living Well on Dialysis **Southwest Cookbook *Egg substitute/egg whites can be used in place of whole eggs

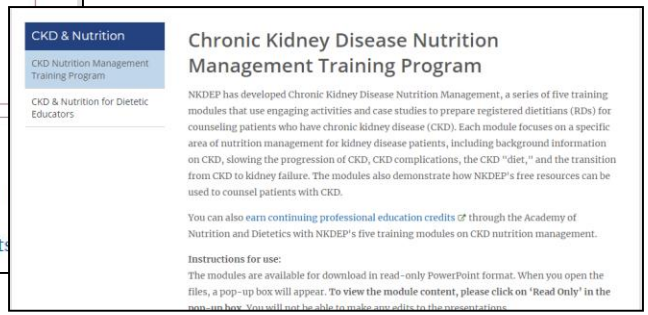
National Institute of Diabetes and Digestive and Kidney Disease

www.niddk.nih.gov



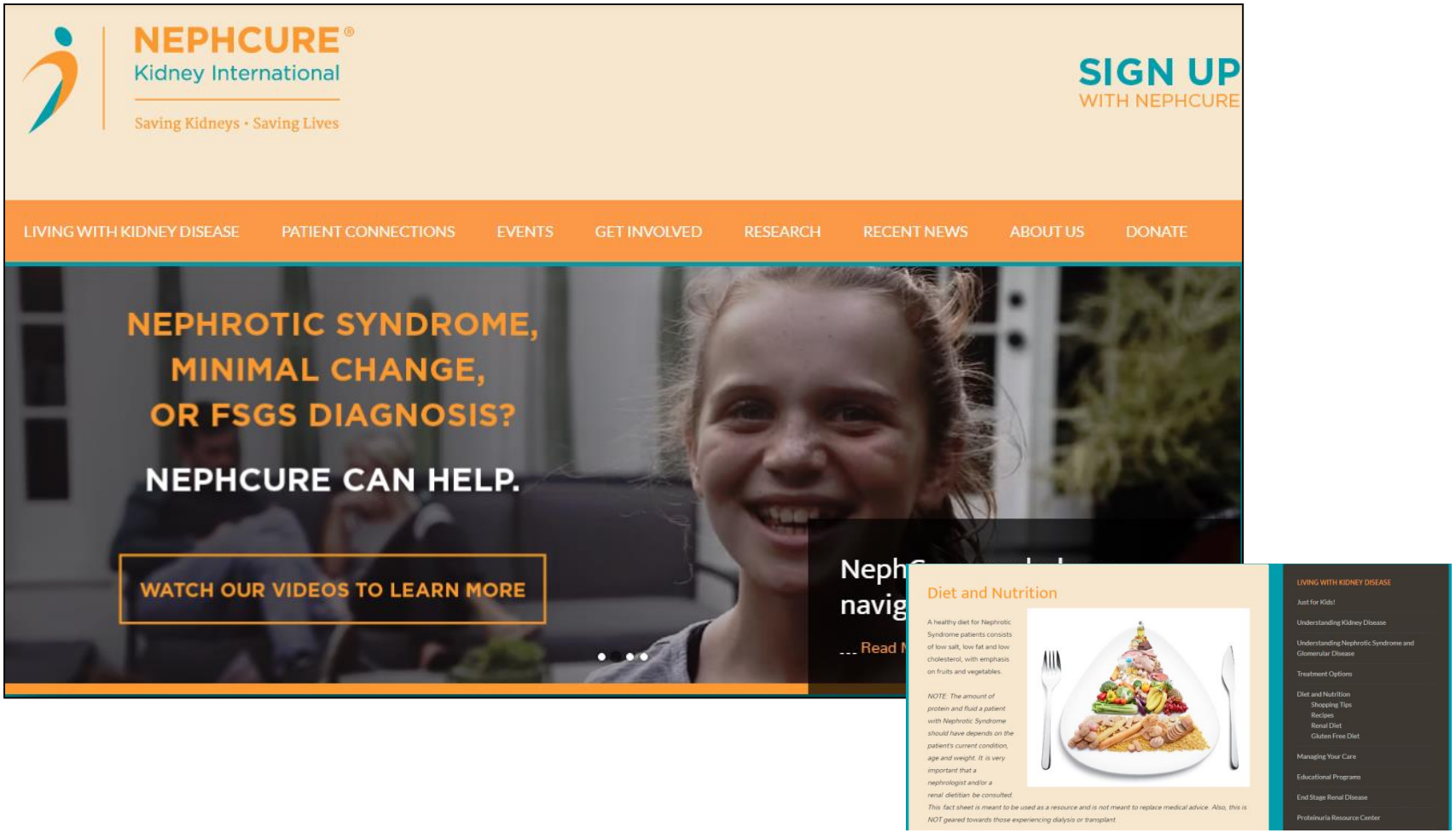
National Kidney Disease Education Program

www.niddk.nih.gov/health-information/health-communication-programs/nkdep



NephCure

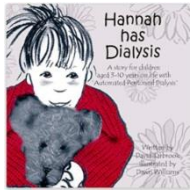
nephcure.org

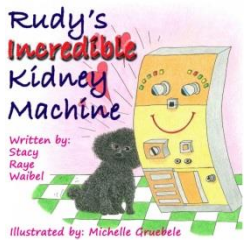


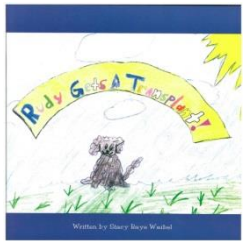
Other Patient Education

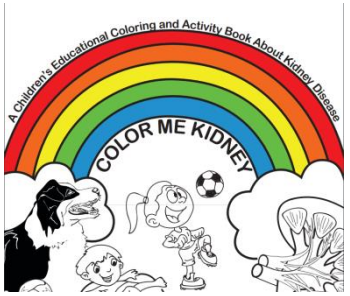
Patient Resources/Sites:
Kidney.org – NKF- patient tab
Rsnhope.org – Renal Support Network

Books/Booklets:
Rudy's Incredible Kidney Machine
Rudy Gets a Transplant
Melsy takes Dialysis to show and tell
Hannah has Dialysis
Color Me Kidney



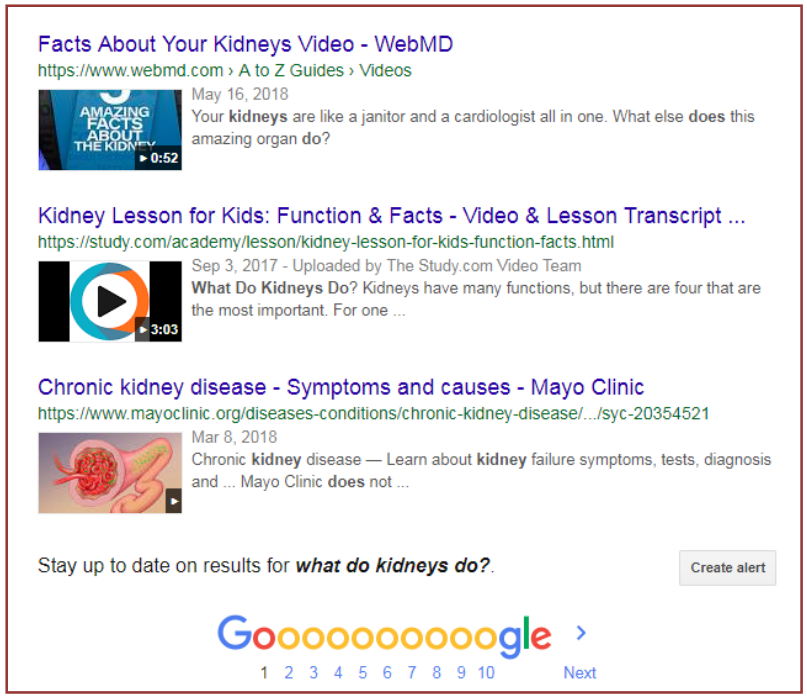






Online videos

Google and Youtube: what do kidneys do?



CHOOSE MY PLATE with CHRONIC KIDNEY DISEASE

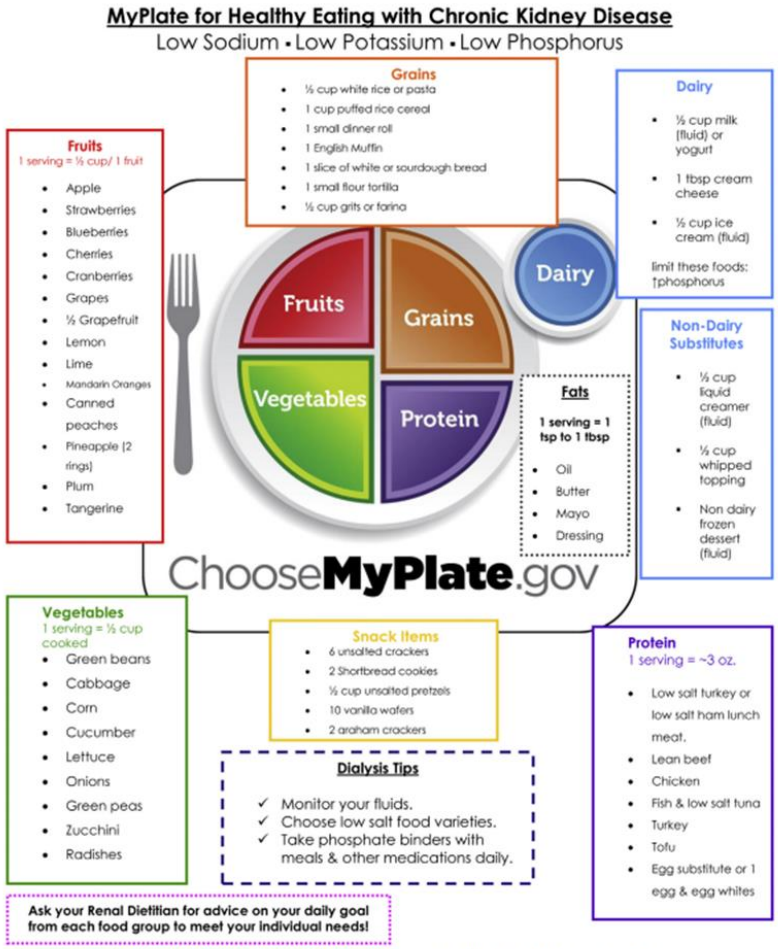


Figure 1. MyPlate handout for healthy eating with chronic kidney disease.

RECIPES!!

- Recipes
  - www.Kidneygrub.com (patient tab)
  - www.kidneyrd.com (patient tab)
  - www.kidneycommunitykitchen.ca
  - www.ultracare-dialysis.com/RecipeCenter.aspx (RD and chef)
  - www.davita.com (600 recipes with nutrient profiles)
  - www.myspiceitup.ca (great visuals with nutrient info)

- Free Downloads
  - www.dciinc.org/recipes/
  - www.kidney.org/sites/default/files/docs/kidney_cookbook_lr.pdf
  - www.kidney.org.uk/documentlibrary/food_with_thought.pdf

- Videos
  - https://www.youtube.com/user/BCRenalAgency
  - http://www.bcrenalagency.ca/health-info/managing-my-care/diet

Apps

Nutrition Apps for Managing Chronic Kidney Disease

MyFoodCoach®
Available for Free: Apple, Android
Summary: Created by the National Kidney Foundation to help you manage personalized nutritional goals. It offers nutrition information, recipes, ingredients, and full meal plans designated for patients with diabetes, CKD, and hypertension.
Link: https://www.kidney.org/apps/patients/my-food-coach-app

Fooducate®
Available for Free: Apple, Android
Summary: Records food intake, activity, sleep, and mood. This app keeps track of calories, protein, sodium, fat, and more. View your progress and stay motivated by connecting with friends and community for support.
Link: https://www.fooducate.com/

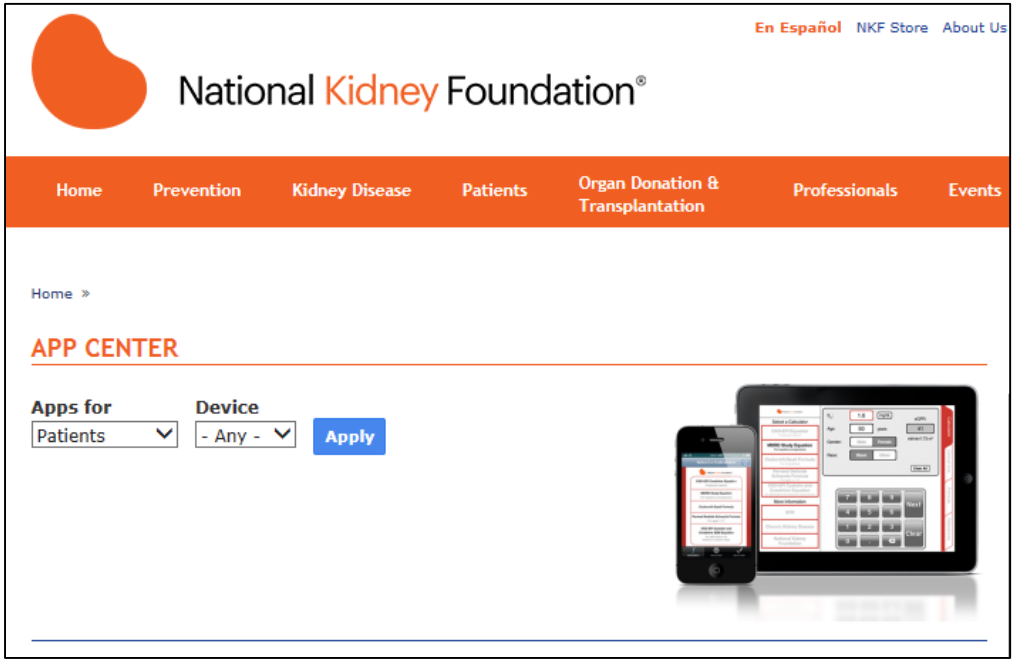
ShopWell®
Available for Free: Apple, Android
Summary: YottaMark, Inc. provides the ability to scan items to tell you ingredients of the foods you are purchasing. This app also offers suggestions of new items to try, helps identify food for certain diets or allergies, and includes an "Ask the RD" section. This can help you track sodium and other nutrients of concern while shopping.
Link: http://www.shopwell.com/mobileapp

H2Overload®
Available for Free: Apple
Summary: National Kidney Foundation's app for management of fluid intake, weight, and blood pressure and provides education on these medical conditions. The app also contains an option to send your results to healthcare providers.
Link: https://www.kidney.org/apps/H2Overload-app

Mango Health®
Available for Free: Apple, Android
Summary: This app allows you to manage your medications and create a schedule of healthy habits. It provides medication information such as food or other drug interactions. You can earn points for compliance and potentially earn rewards.
Link: https://www.mangohealth.com/

Klein,J. Utilization of Mobile Nutrition Applications by Patients With Chronic Kidney Disease. J Ren Nutr Vol 29, 1:e5-e7 (January 2019)

FINDING APPS



Evidenced based?

Limited market

Tube feeding

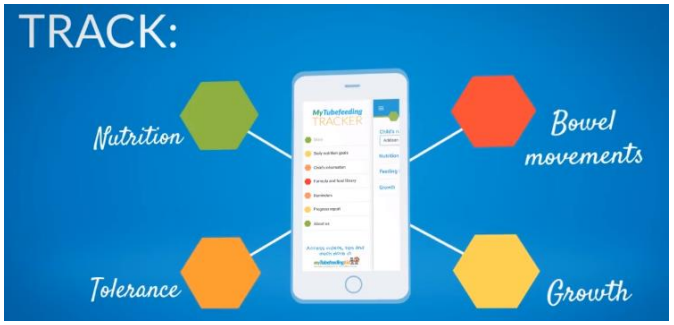
Tube Feeding Awareness Foundation

www.feedingtubeawareness.org/

Feeding pump assistance

www.infinityfeedingpump.com/virtual-pump/

App: my tube feeding tracker



IN SUMMARY…..

Delivering nutrition care in pediatric nephrology demands:

- that you have a solid understanding of pediatric nutrition as well as kidney related issues,
- that you keep an open mind and consider the many possibilities for solutions to problems,
- that you never stop learning!!!

Just the beginning!.....

…the sky's your limit!!!

Speaker Contact: Nonnie Polderman
npolderman@cw.bc.ca

UCSF Benioff Children's Hospitals

Standardizing Our Approach: Blood Pressure in Pediatric Hemodialysis Patients

Audrey Busch, MS, RN, CNN

March 5, 2021

Agenda

- Background
  - Poll this group re BP practices
  - Show results for SCOPE BP practice result
- CV disease in Peds patients (research)
- Flynn 5th report → translating into HD patient population
- Developing the bundle – what's in it?
  - in-center BP
  - Video of what to do vs what not to do
  - ABPM
- Implementing the bundle
  - Common fear → advice on how to get started
  - resources

Poll:

Does your unit have a concrete and standardized procedure for obtaining and recording blood pressure for hemodialysis patients?

- Yes
- No

Poll:

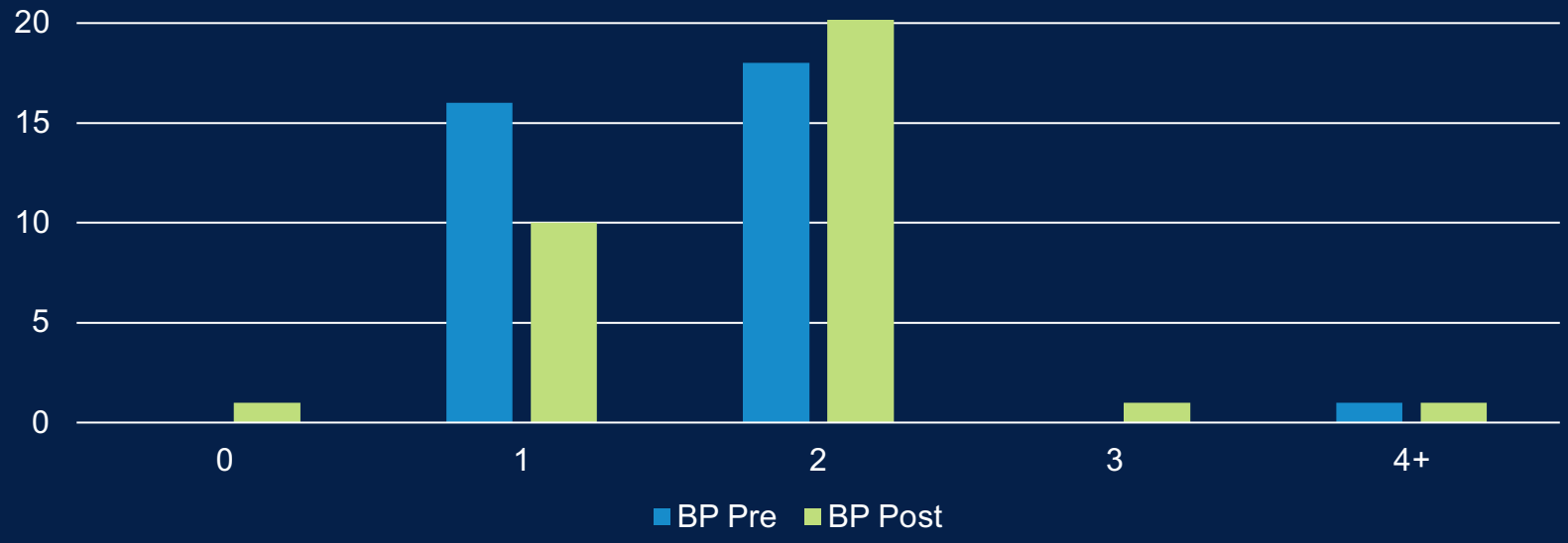
Are you confident that regardless of staff, blood pressure is being obtained and recorded the same on every patient every treatment??

- Not confident at all
- Slightly confident
- Fairly confident
- Completely confident

And the survey says….

- In early 2017, we surveyed pediatric 40+ dialysis clinics across the country.
- We queries them on the *current* and *routine* blood pressure practices in their dialysis center.
- Here is what we found:

How many BP measurements are ***routinely*** obtained on each patient pre/post-HD in your unit?



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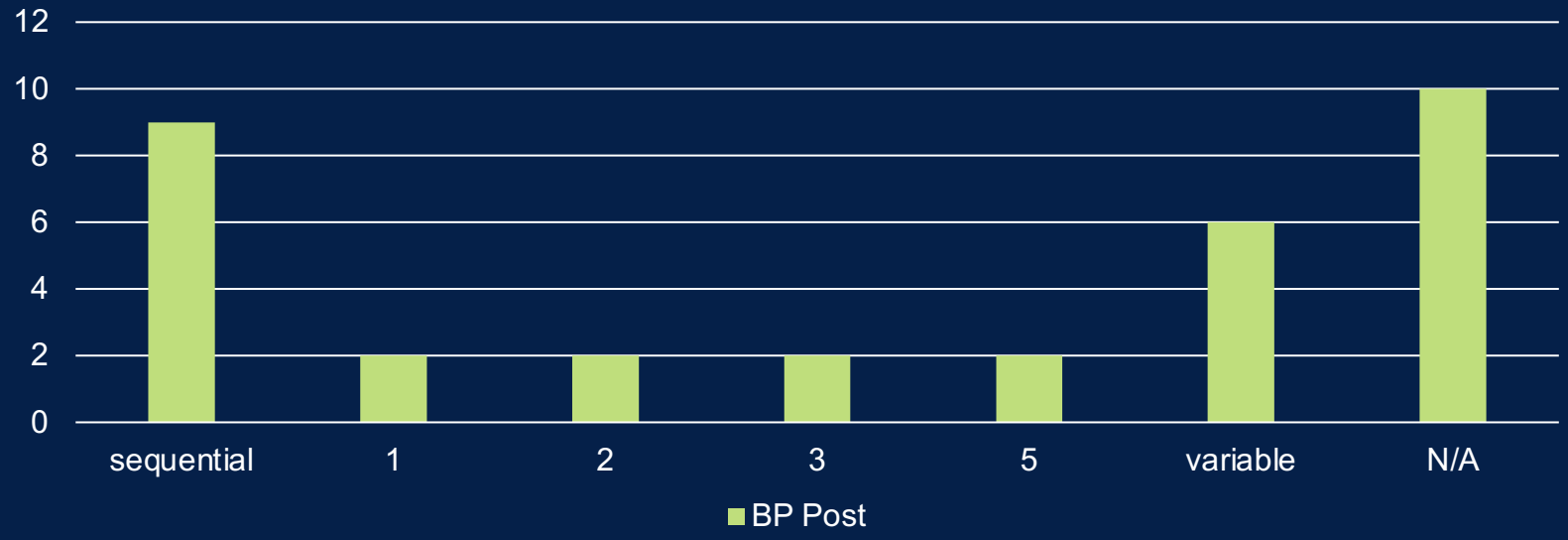
If two or more BP measurements are ***routinely*** obtained post-HD, how many minutes apart are they taken?



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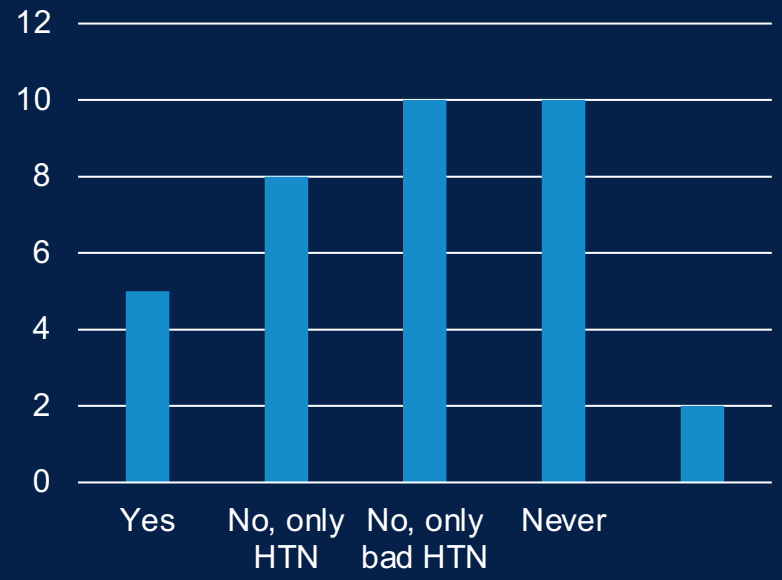
ABPM – Ambulatory Blood Pressure Monitoring

Is your unit ***routinely*** performing ABPM?



If ***routinely*** performing ABPM, how often?

Background

- Why is BP important in these patients
  - Research related to CV outcomes in pediatric dialysis patients
  - So we know that dialysis patients have ^ risk for CVD and we take a lot of BPs but how do we know if they are good quality

- Goals of standardizing BP
  - Eventually we would like to know how to treat all of these cases but *FIRST we have to know if we are even measuring correctly and uniformly.*
  - *Comparing apples and oranges*
  - The aim is STANDARDIZATION, we are not advising how to treat, simply elaborating on how to measure and when to confirm. to do. We just want to make sure we are at least gathering the right information.

How it works

- Adapted from Joseph Flynns 5th reports.
- Two parts
  - In-Center BP Measurement –
    - Standardized measurement to be performed before and after every dialysis
  - Home BP Measurement
    - ABPM every 6 months

      OR
    - Twice daily home BPs for 4 consecutive days

In Center Blood Pressures

- Describe the criteria
- (site if/when possible)

Home Blood Pressures (ABPM or Home)

- Describe the criteria (site if/when possible)
- Home BP parent training documet

How it looks

The GOOD

Common Concerns…And solutions

State clinic hesitations

Describe implementation tactic/strategies

resources

Restate the goal of implementing this into practice

Thank you!

Contact information

Citations (where otherwise not cited)

Picture of UCSF and Ped Neph team.

LIVE FROM THE WATER TREATMENT ROOM

Pam Heise, MSN RN CPN CNN

Assistant Director, Clinical Practice– Renal & Pheresis Department

Texas Children's Hospital

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OBJECTIVES

- Describe why water purification is important in dialysis
- Identify the contaminants of water that are toxic to dialysis patients
- Identify the components of the water treatment room
- Outline required water testing and documentation
- Review surveyor questions
- Describe CMS conditional level findings related to water

WHY IS WATER PURIFICATION IMPORTANT IN DIALYSIS

- Chemicals added water to make it safe for consumption
- Drink about 2 L water each day
- HD patients exposed up to 200 L water each treatment
- Many published instances where water has caused harm in HD patients

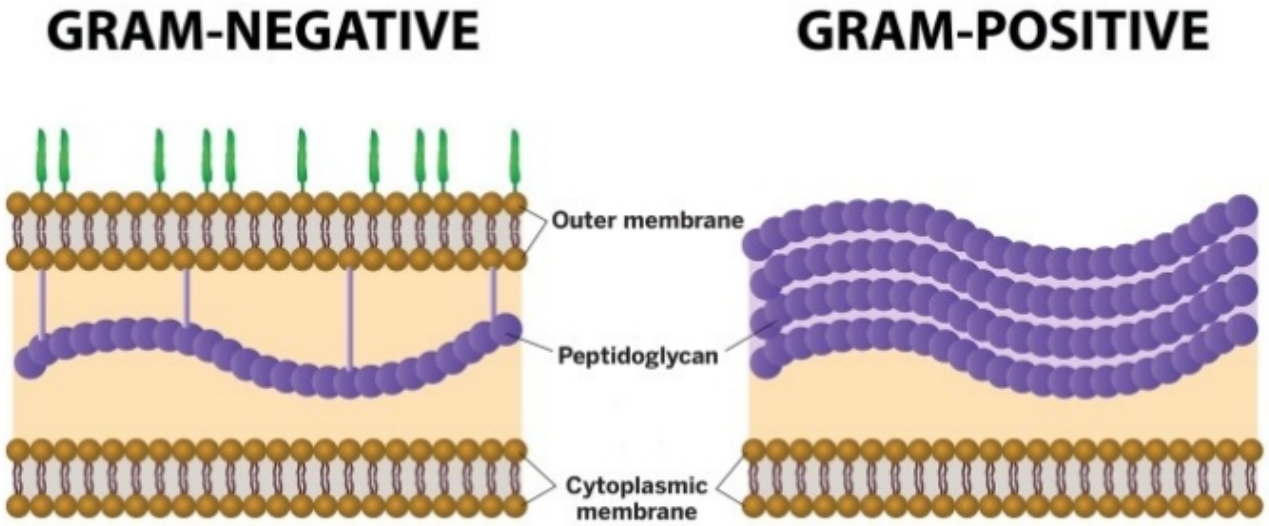
vs

WHAT ARE THE WATER STANDARDS

- Environmental Protection Agency (EPA) – minimum standards for drinking water
- Association for the Advancement of Medical Instrumentation (AAMI) – sets thresholds for acceptable levels of inorganic chemical contaminants in water used for dialysis treatments

TYPES OF CONTAMINANTS

- Microorganisms
  - Gram Negative Bacteria
    - Grow rapidly
    - Produce endotoxins
    - Quickly killed with chemicals
  - Nontuberculous mycobacteria
    - Grow slowly
    - Do not produce endotoxins
    - Take longer to kill



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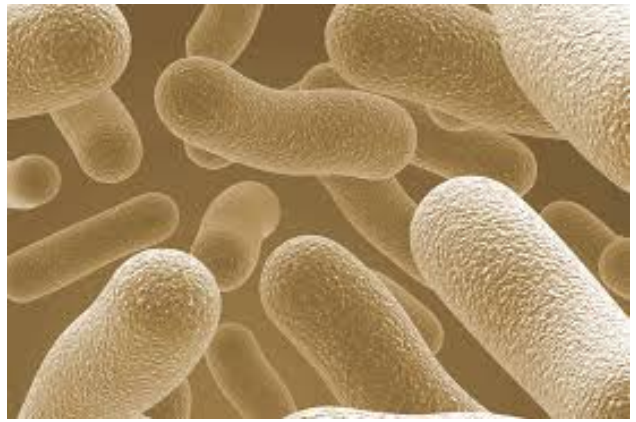
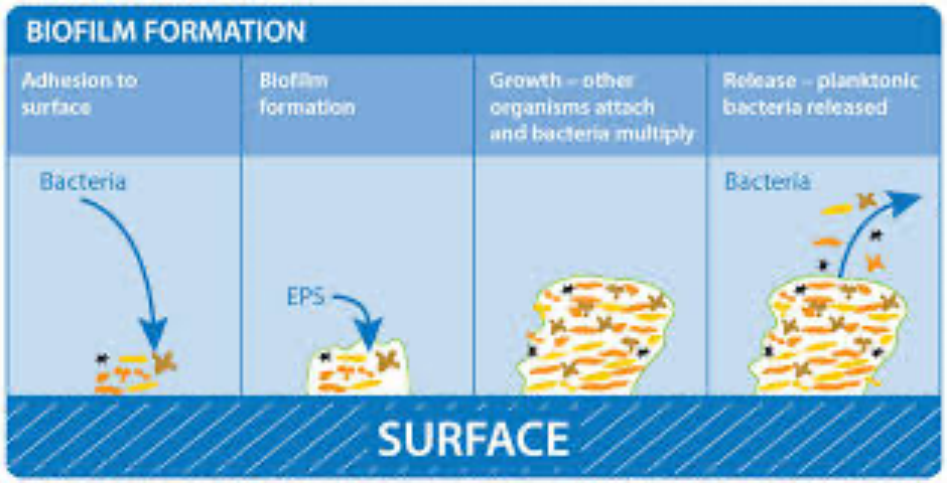
TYPES OF CONTAMINANTS

- Endotoxins
  - Part of bacterial cell wall
  - Small, so they can pass through dialyzer membrane
  - If bacteria level is kept low, less chance for endotoxins
- Biofilm
  - A gelatinous substance that is secreted by bacteria that adhere to surfaces
  - Difficult to remove
  - Bacterial cultures may not catch
  - Grows in layers, then releases toxins





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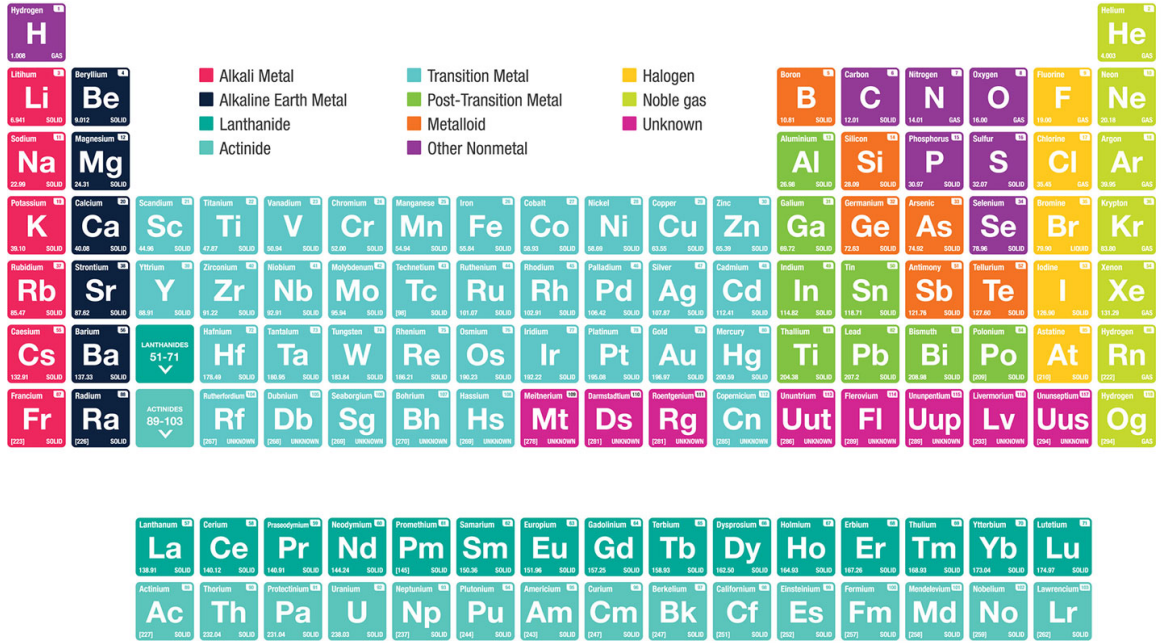
TYPES OF CONTAMINANTS

- Sediment
- Salts, metals and other chemicals
  - Sodium
  - Potassium
  - Calcium
  - Magnesium
  - Fluoride
  - Aluminum
  - Copper
  - Mercury



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CONTAMINANTS TOXIC TO PATIENTS ON DIALYSIS

CONTAMINANT	ADVERSE EVENT
Aluminum	Encephalopathy, bone disease, anemia
Calcium/ magnesium	Nausea, vomiting, muscle weakness
Chlorine/ chloramine	Hemolysis
Copper	Hemolysis, nausea, vomiting
Endotoxin	Pyrogenic reaction, chronic inflammation
Fluoride	Nausea, abdominal pain, pruritus, arrhythmia
Nitrates	Anemia
Zinc	Hemolysis, nausea, vomiting
Endotoxins	Pyrogenic reaction, fever and chills, hypotension
Bacteria	Hypotension

TOUR THE WATER ROOM

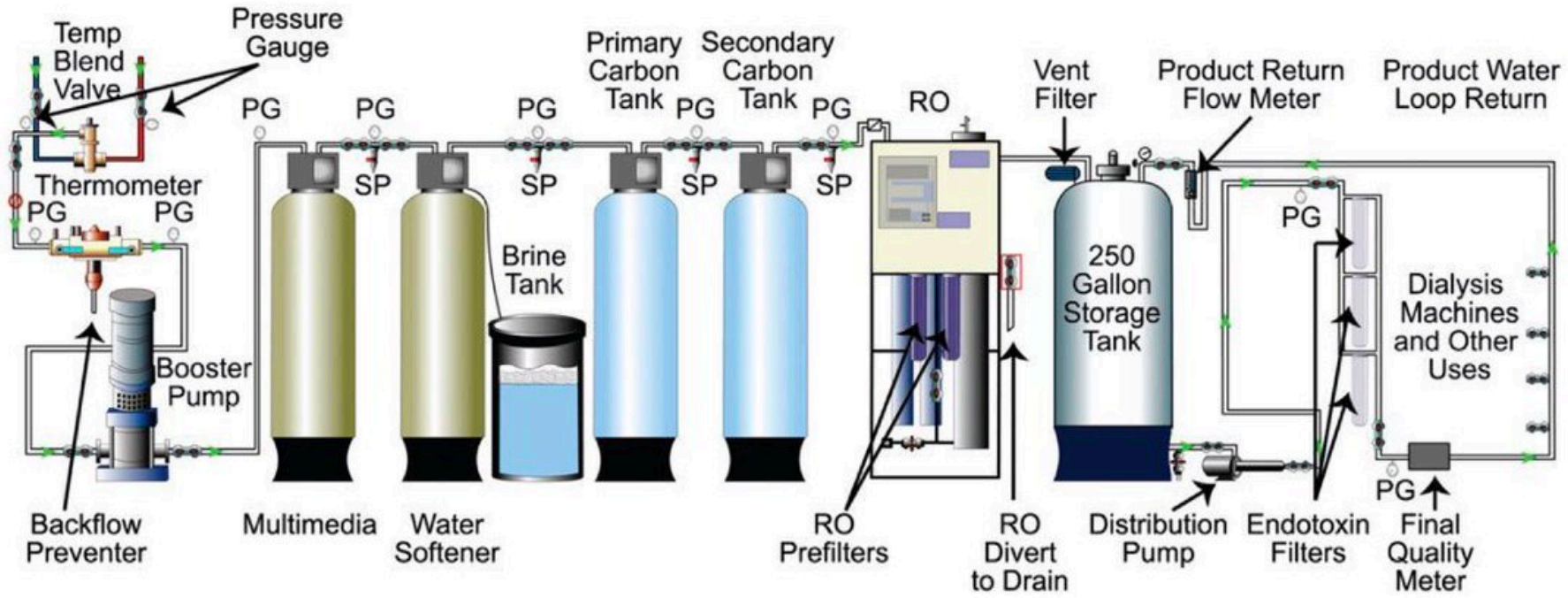

Figure 2. | The water treatment system. This schematic delineates a water treatment system with indirect product water distribution (*i.e.*, a holding tank). PG, pressure gauge; RO, reverse osmosis; SP, sampling port.

Reproduced from Ted Kasparek, and Oscar E. Rodriguez CJASN 2015;10:1061-1071

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DIFFERENT TYPES OF WATER ROOMS

LABELING REQUIREMENTS

- Schematic diagrams that identify components, valve, sample ports, and flow direction
- Function/purpose of the devices and action if out of acceptable range

A

B

BLENDING VALVE

PURPOSE:
The mixing valve blends hot and cold water to establish a preset water temperature at the valve outlet;
77° +/- 5° F for 23G RO systems
70° +/- 5° F for CWP RO systems
The performance of downstream components, in particular the carbon filters and the RO system will be affected if the water temperature varies outside of this range.

HOW DOES IT WORK:
The mixing valve is equipped with a thermometer and an adjustment knob, which allows the operator to set the desired temperature. Water must be flowing through the mixing valve when making temperature adjustments. When the operator has set the desired temperature, temperature sensitive springs will automatically adjust to make minor changes and keep the temperature at or near the desired setting.

QUALITY CHECKS:
Daily observation and recording of the blended water temperature.

NOTIFY FA AND BMT IF WATER TEMPERATURE IS NOT BETWEEN ____ AND ____ DEGREES FAHRENHEIT.

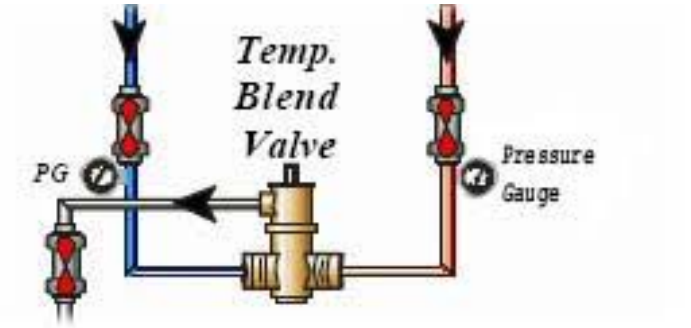
FACTORS AFFECTING OPERATION:
Improper operation of the mixing valve is usually denoted by an inability to adjust the valve to a proper temperature, leaking or dripping from the valve, or a large pressure drop across the mixing valve. The inability to adjust the mixing valve to the desired temperature may be caused by a lack of hot water entering the valve, or an incoming cold water temperature, which exceeds the operators desired temperature setting.

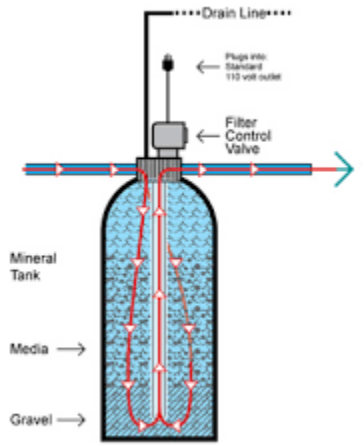
Leaks or excessive pressure drop (delta pressure) across the valve may cause shutdown or improper operation of the downstream water treatment components.

For detailed information on operating this component of the water system, refer to the system operator manual and/or clinic policies.

PRETREATMENT - TESTING AND DOCUMENTING WATER SYSTEM

- Blending valve
  - Monitor temperature at the start of each day
    - 65-85^{o}F Ideally 77^{o}F
- Multi media filter
  - Monitor pressure drop across the filter at the start of each day
    - Δ≤15psi





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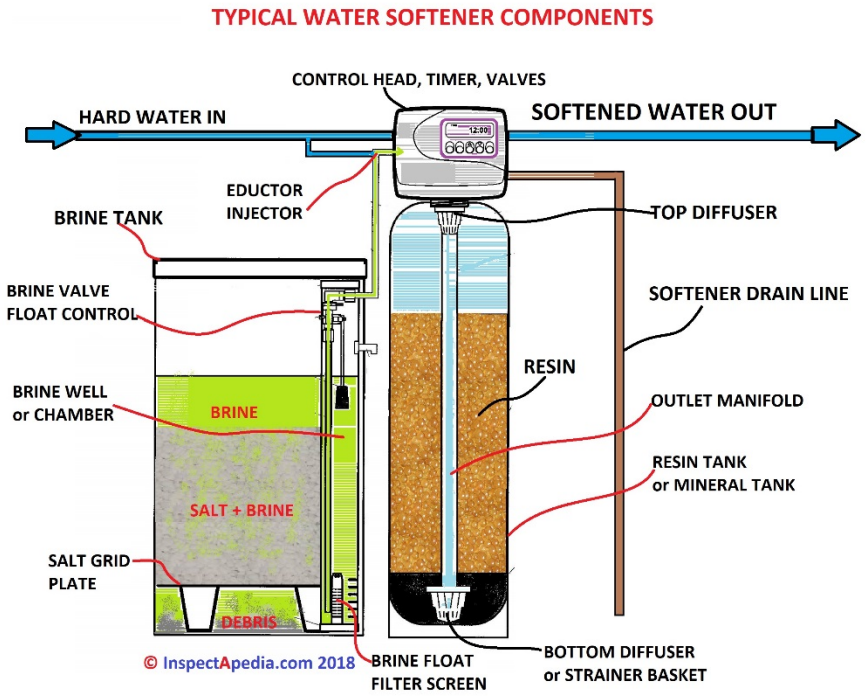
PRETREATMENT - TESTING AND DOCUMENTING WATER SYSTEM

- Water softener
  - Monitor water harness at the start and end of each day
    - <1 grain per gallon (gpg) or <1 parts per million (ppm)
  - Monitor media regeneration time monthly
    - Regenerate media with brine was after hours
- Brine Tank
  - Monitor salt level at the beginning of the day
    - Salt above water line



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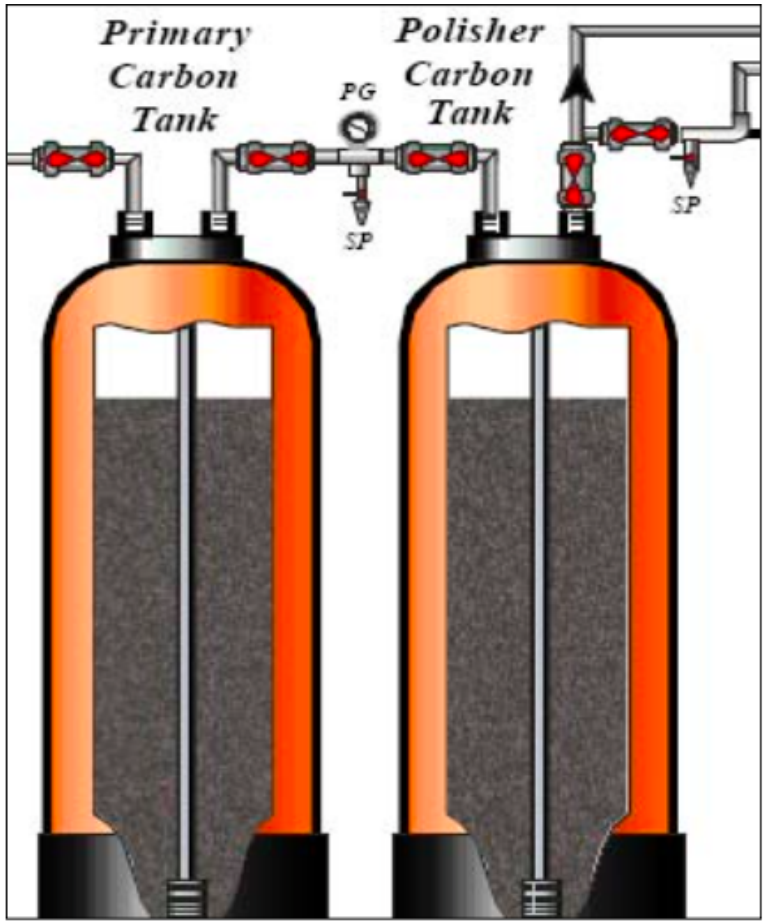
PRETREATMENT - TESTING AND DOCUMENTING WATER SYSTEM

- Carbon Tanks
  - Monitor chlorine and chloramine at the start of each day and every 4 hours
    - ≤0.1 PPM
    - Empty bed contact time (EBCT) 10 minutes
- RO prefilter
  - Monitor pressure drop across device at the start of each day
    - Δ≤20psi



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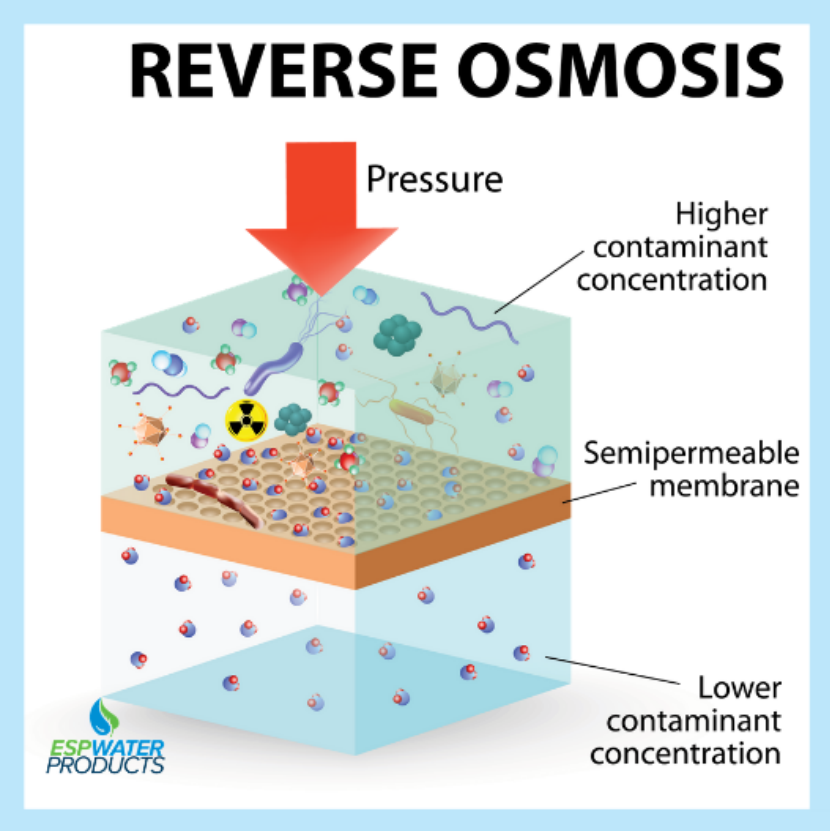
TREATMENT - TESTING AND DOCUMENTING WATER SYSTEM

- Reverse Osmosis (RO)
  - Monitor % rejection level at the start of each day
    - ≥ 90%
  - Monitor water purity
    - Conductivity and Total Dissolved Solids (TDS)



DISTRIBUTION – TESTING AND DOCUMENTING

- Distribution loop
  - Monitor flow of the water at the end of the loop periodically
    - >3ft/second (indirect loop)
    - >1.5ft/s (direct loop)

BACTERIOLOGY OF WATER AND DIALYSATE– TESTING AND DOCUMENTING

- Water cultures
  - Sample collection – Direct plate counts
    - Different parts of the water distribution system
      - i.e. After RO, after endotoxin filter, beginning and end of loop)
    - At least two machines monthly and from enough machines so each machine is tested at least once per year.
  - Established systems - Monitor no less than one time per month
  - New systems – Monitor weekly
    - Acceptable level: <50 colony forming bacteria (CFU)
    - Action level: 50 CFU/mL-199 CFU/mL (can complete treatments for the day)
    - Unacceptable level: >=200 CFU/mL (must stop treatments)

ENDOTOXINS – TESTING AND DOCUMENTING

- Endotoxins – dead bacteria
  - Monitor Limulus Amebocyte Lysate (LAL) no less than one time per month (*NEW AAMI 23500 STANDARDS*)
    - Acceptable level: <0.25 Endotoxin Units (EU)/mL
    - Action level: >=0.25 EU/mL to <0.5 EU/mL
    - Unacceptable level: >=2 EU/mL

SURVEYOR QUESTIONS – CARBON SYSTEM AND CHLORINE REMOVAL

1. What is the Empty Bed Contact Time (EBCT) of the carbon system?

   At least 10 minutes per carbon tank

2. What test is done for chlorine in the water system?

   RO must be running for 15 minutes, DPD4 or chlorine strips, after primary carbon tank

3. When is the test done?

   Before the start of the day and every 4 hours

SURVEYOR QUESTIONS – CARBON SYSTEM AND CHLORINE REMOVAL

4. What is the maximum allowable result?

   ≤ 0.1 mg/L

5. If maximum level of is exceeded, what actions are taken?

   Recheck after carbon tank 2 (primer tank).

   What if that level is ≤ 0.1 mg/L?

   Recheck post carbon tank every hour, continue treatments.

   What if that level is > 0.1 mg/L?

   Stop treatments, notify MD.

WATER TESTING FOR TOTAL CHLORINE

A surveyor will observe and look for the following:

1. Did the sample come from the sample port after the primary carbon tank
2. Testing reagents within the expiration dates
3. If digital meter is used, is it zeroed prior to testing.
4. Was correct PPE worn during the testing.

SURVEYOR QUESTIONS - REVERSE OSMOSIS & CONTINUOUS WATER QUALITY MONITOR

1. How is water quality monitored?

   Audible alarm in the unit

2. What is the set point for the water quality alarm?

   < 90% reject

3. What actions are taken if the percent rejection falls below 90% or the water quality exceeds the set point?

   Stop treatments, water is diverted down the drain, reevaluate AAMI analysis

SURVEYOR QUESTIONS – DISINFECTION, WATER & DIALYSATE MICROBIOLOGY MONITORING

1. How often is the water distribution system disinfected?

   At least monthly

2. When are the water cultures and endotoxins obtained in relation to disinfection and from what sites?

   At least monthly, from several different parts of the water distribution.

3. How often are dialysate cultures taken from each machine?

   At least 2 dialysis machines every month so that each machine is tested at least annually

SURVEYOR QUESTIONS – DISINFECTION, WATER & DIALYSATE MICROBIOLOGY MONITORING

4. How are samples of water and dialysate collected and how are cultures and LALs performed?

   Direct plate counts and measurement of endotoxins collected at least monthly on established systems. 2 machines every month and at least once annually

5. What are the action and maximum allowable microbiological levels for the product water and dialysate?

Microorganism	Standard	Acceptable Level	Action Level	Unacceptable Level
Bacteria	ANSI/AAMI	< 50 CFU/mL	50 – 199 CFU/mL	≥ 200 CFU/mL
	AAMI 23500 Updated STD	< 50 CFU/mL	50 – 99 CFU/mL	≥ 100 CFU/mL
Endotoxin	ANSI/AAMI	< 1 EU/mL	1 - <2 EU/mL	≥ 2 EU/mL
	AAMI 23500 Updated STD	< 0.25 EU/mL	0.25 - < 0.5 EU/mL	≥ 0.5 EU/mL

6. What actions are taken when those levels are exceeded?

   Action level: can complete treatments for the day

   Unacceptable level: must stop treatments

CONDITIONAL LEVEL FINDINGS

- Lack of knowledge or training of staff assigned to operate and monitor water treatment or Dialysate preparation
- Failure to perform and document tests for chlorine and chloramine
- Unsafe practices in preparation, labeling or delivery of Dialysate
- Failure to address out of range tests

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Centers for Medicare & Medicaid Services, HHS: Medicare and Medicaid programs: Conditions for coverage for end-stage renal disease facilities: Interpretive guidance 2008

Centers for Medicare & Medicaid Services. ESRD Survey Training: ESRD Core Survey Field Manual Version 1.9, Bethesda, MD, Centers.

75 years of community water fluoridation. (2020, January 15). Retrieved January 26, 2021, from https://www.cdc.gov/fluoridation/basics/anniversary.htm

Henner, D. H2knOw: What you need to know about water treatment in dialysis. Retrieved from Renal Healthcare Association member's only website.

Kasparek, T., & Rodriguez, O. (2015, June 05). What medical directors need to know About Dialysis Facility water management. Retrieved January 26, 2021, from https://cjasn.asnjournals.org/content/10/6/1061/tab-article-info

Sukumar, Ali, L., Mohamed, M., Morsi, E., Faswal, & Farooq, U. (2020, October 21). Water treatment for hemodialysis. Retrieved January 26, 2021, from https://www.renalfellow.org/2020/10/21/water-treatment-for-hemodialysis/

Texas Children's Hospital®

COMMENTS/QUESTIONS?

Quality of Life
Round Table Discussion

Kelli Scott, LCSW, LMSW

Disclosures

I have no disclosures.

Tools Used

- Core Version
- ESRD Specific

CMS Requirements

- Completed within first 30 days and at least annually there after
- Completed if patient experiences a life changing event or change in health status

Areas Assessed

- Physical
- Emotional
- Social
- School/Work

Scoring

- Will add picture of scoring scale compared to general population

Questions???

What questions do you have about the PedsQOL?

Have you come across concerns after QOL is completed?

For those that have experience with tool, how has it improved patient care?

How have you dealt with any identified concerns after the QOL is completed?

References

- Pedsql.org

Renal Anemia: The Basics

Meredith Atkinson, M.D., M.H.S.
Associate Professor of Pediatrics
Johns Hopkins School of Medicine
5 March 2021

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CHILDREN'S CENTER

No Disclosures

Learning Objectives

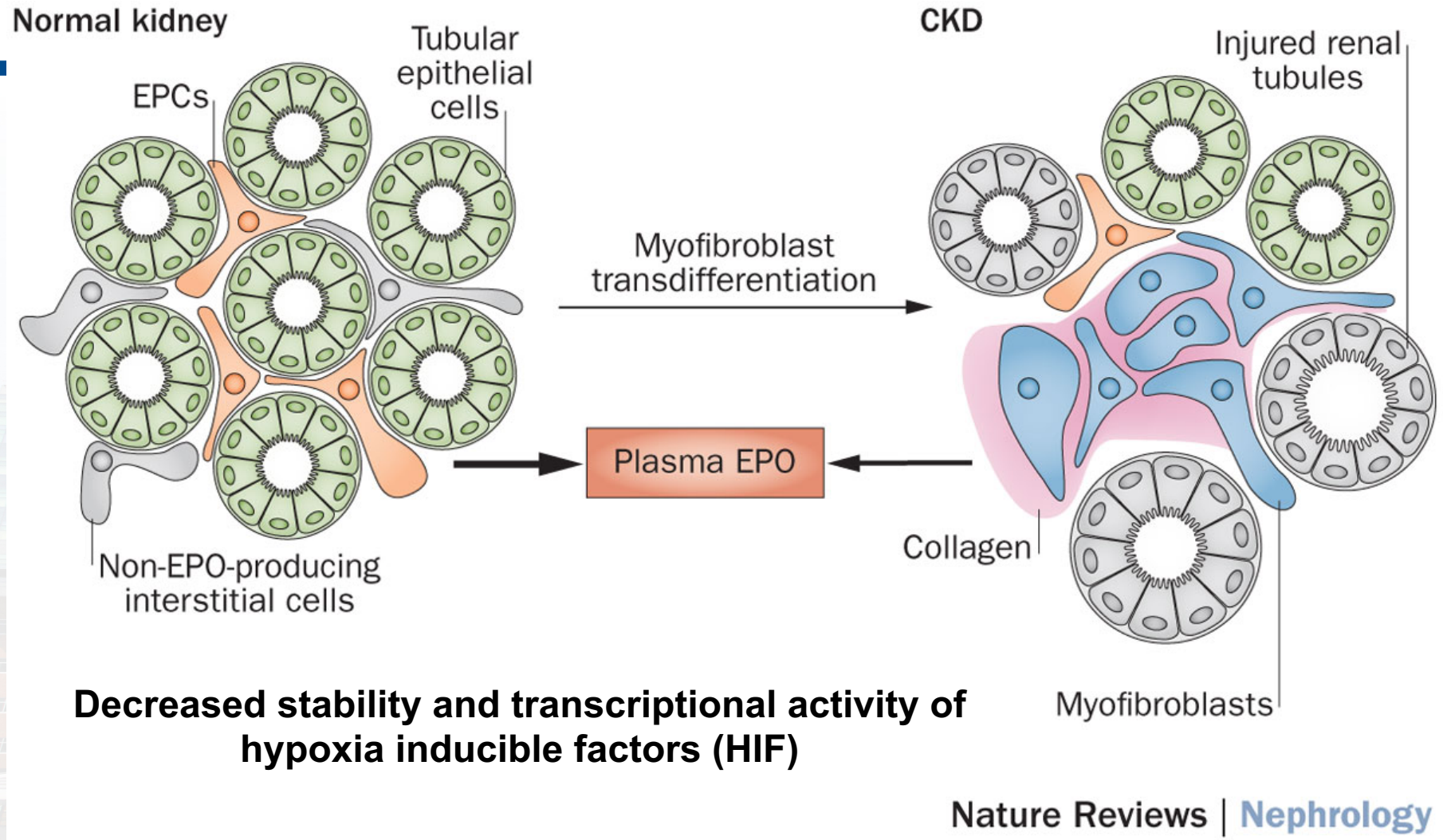
- At the end of this session the listener will be able to:
  - Describe the etiology of the anemia of CKD in children
  - Define anemia and initiate a work-up
  - Understand formulations and dosing of erythropoiesis stimulating agents (ESA)
  - Describe indications for and approach to iron supplementation
  - Recognize emerging anemia therapies
    - Newer ESAs, novel iron supplementation, HIF stabilizers

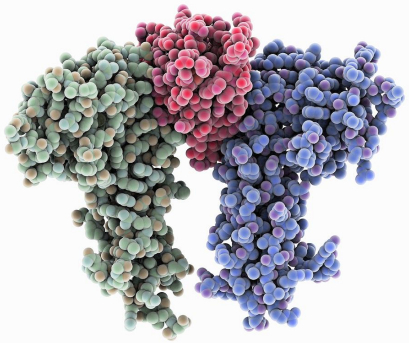
Anemia of Kidney Disease

- 1839 - "..by far the most remarkable character of the blood in the advanced stage of Bright's disease is a gradual and rapid reduction of its colouring…no other natural disease comes as close to hemorrhage for impoverishing the red particles of the blood."

Fishbane, Spinowitz. Update on Anemia in ESRD and Earlier Stages of CKD: Core Curriculum 2018. AJKD 71(3):423-435.

Figure 5 Cellular basis of erythropoietin deficiency in renal failure

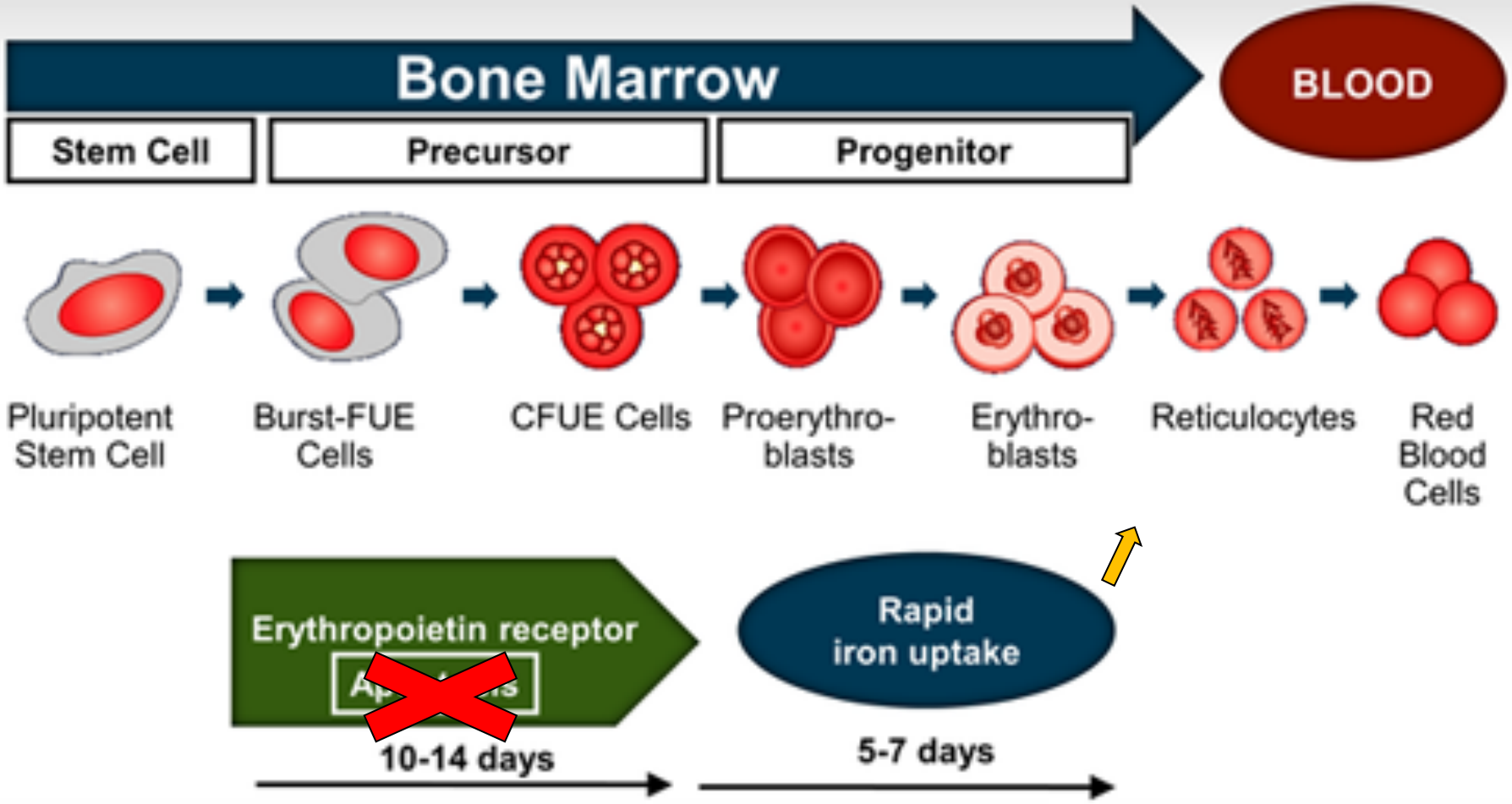




Human erythropoietin
Glycoprotein hormone

Koury, M. J. & Haase, V. H. (2015) Anaemia in kidney disease: harnessing hypoxia responses for therapy
Nat. Rev. Nephrol. doi:10.1038/nrneph.2015.82

Red Blood Cell Maturation



Requires on average a 3-week cycle for red blood cell maturation

Reprinted from Lankhorst CE, Wish JB. Anemia in renal disease: diagnosis and management. *Blood Rev*. 2010;24:39-47, with permission from Elsevier.

3-4 grams of iron in the typical adult human body:

- 2-3 grams incorporated into hemoglobin (Hgb)

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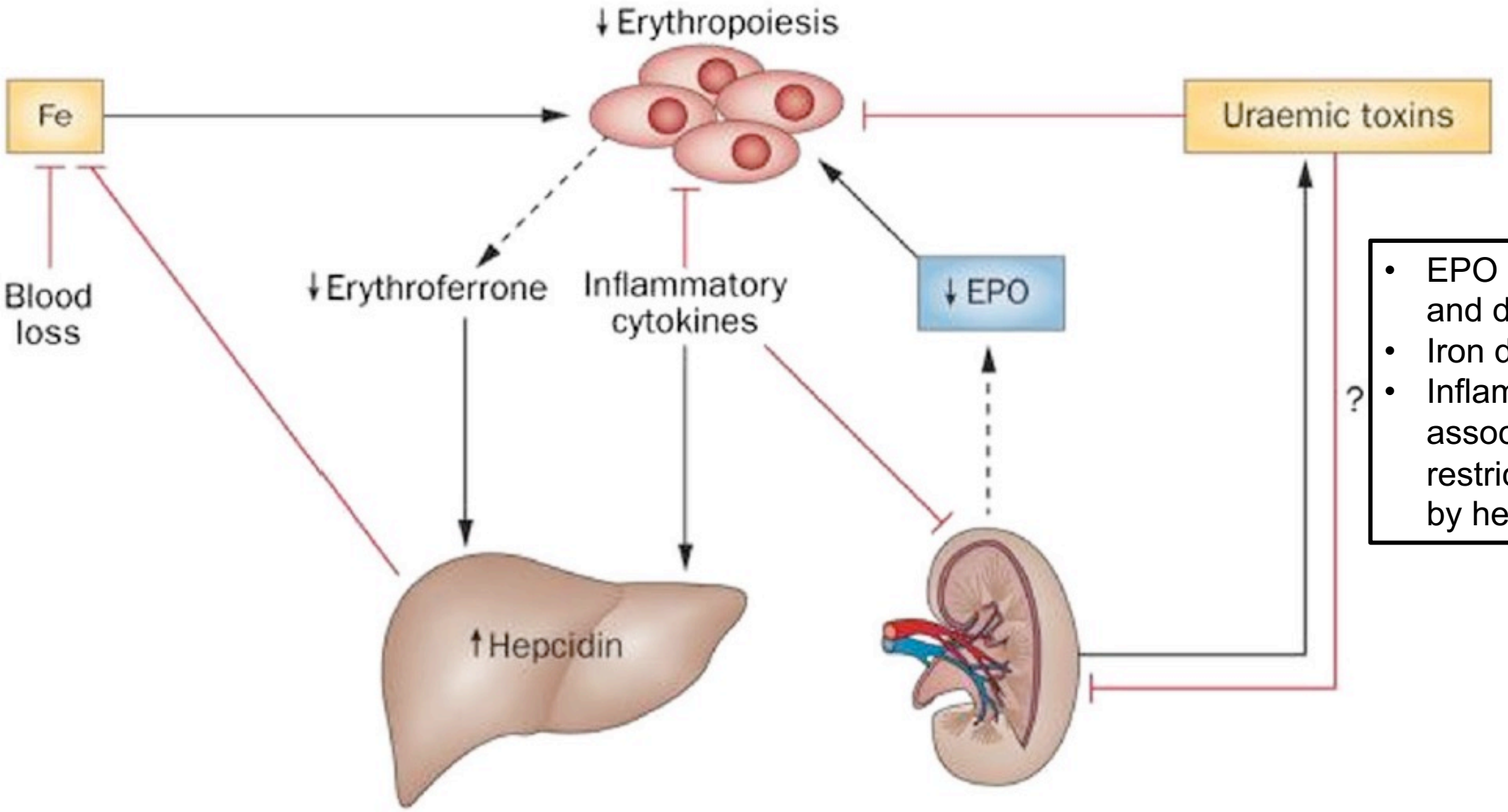
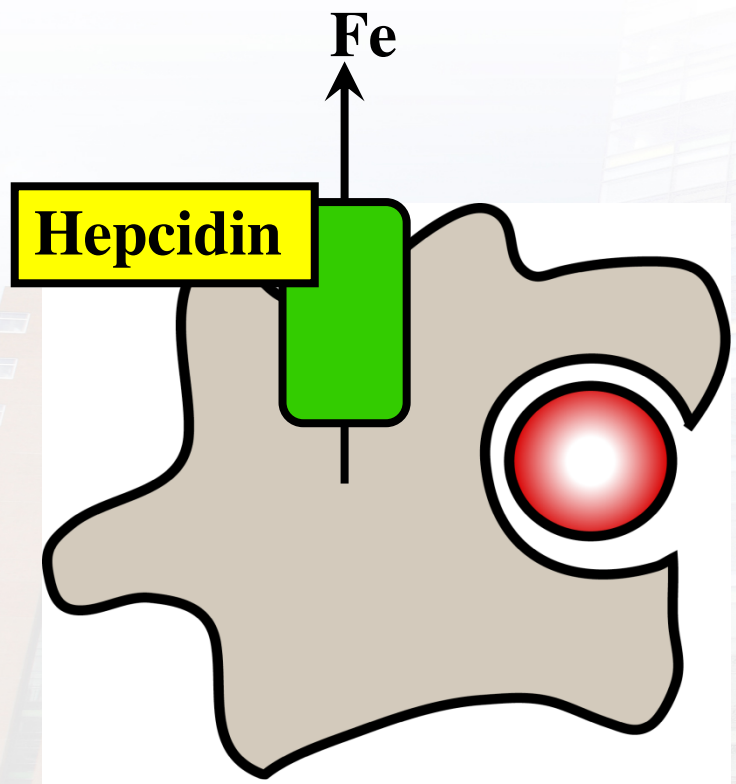
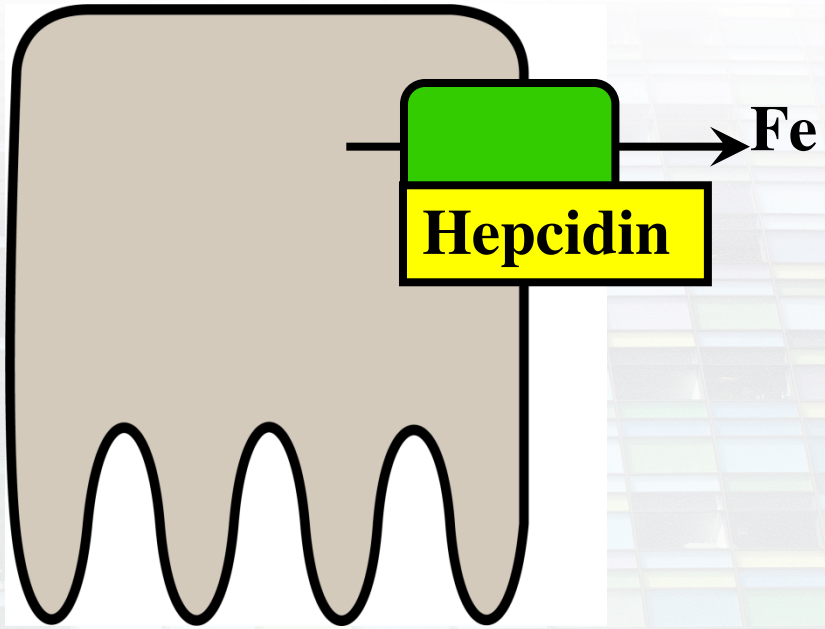


- EPO dysregulation and deficiency
- Iron deficiency
- Inflammation-associated iron restriction mediated by hepcidin

Hepcidin and Ferroportin



macrophage



enterocyte

(*Donovan et al, 2005; Nemeth et al, 2004*)

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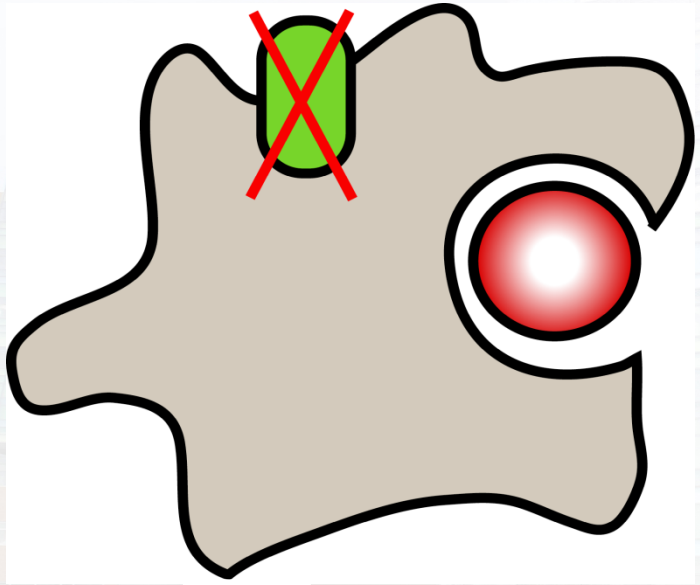
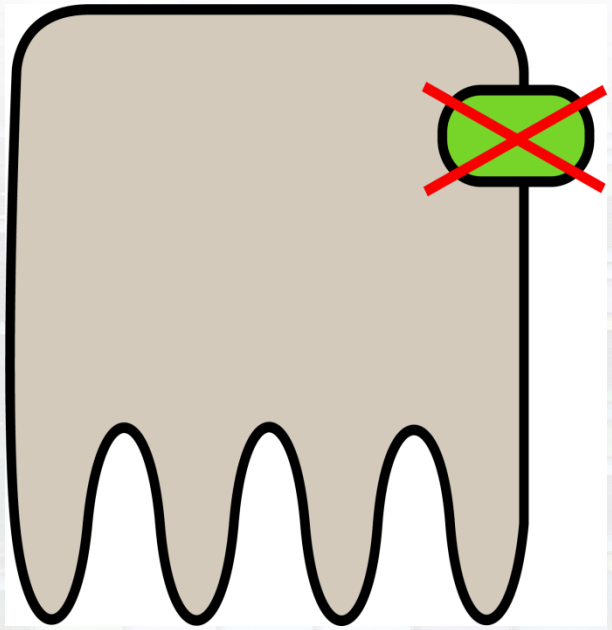
Hepcidin and Ferroportin





Decreased Fe recycling

Decreased dietary uptake

(*Donovan et al, 2005; Nemeth et al, 2004*)

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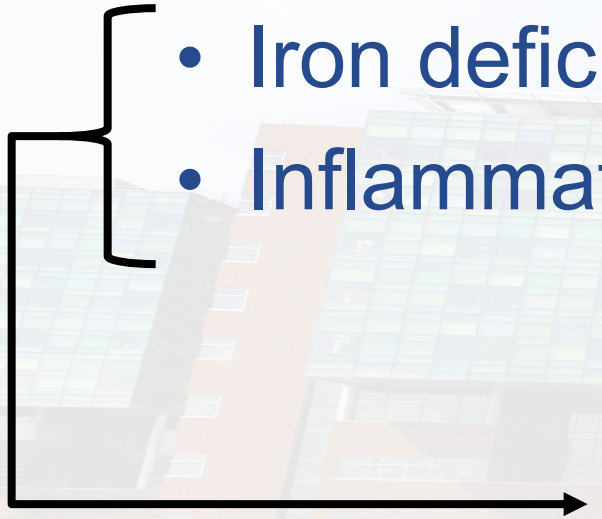
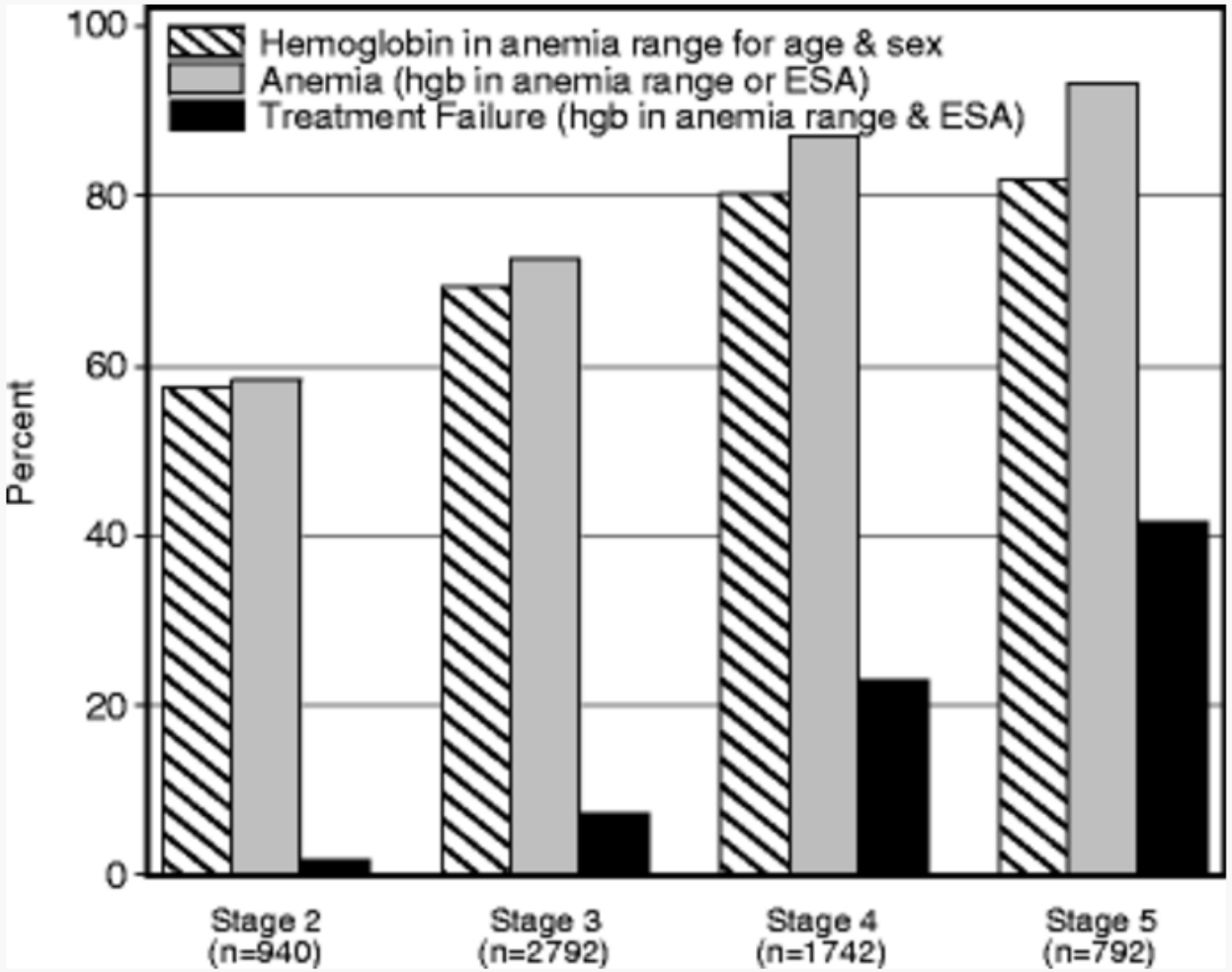
Etiology of Anemia of CKD

- Erythropoietin dysregulation and deficiency
- Iron deficiency
- Inflammation-associated iron restriction



ESA-Resistant Anemia

Epidemiology of Anemia in CKD/ESRD



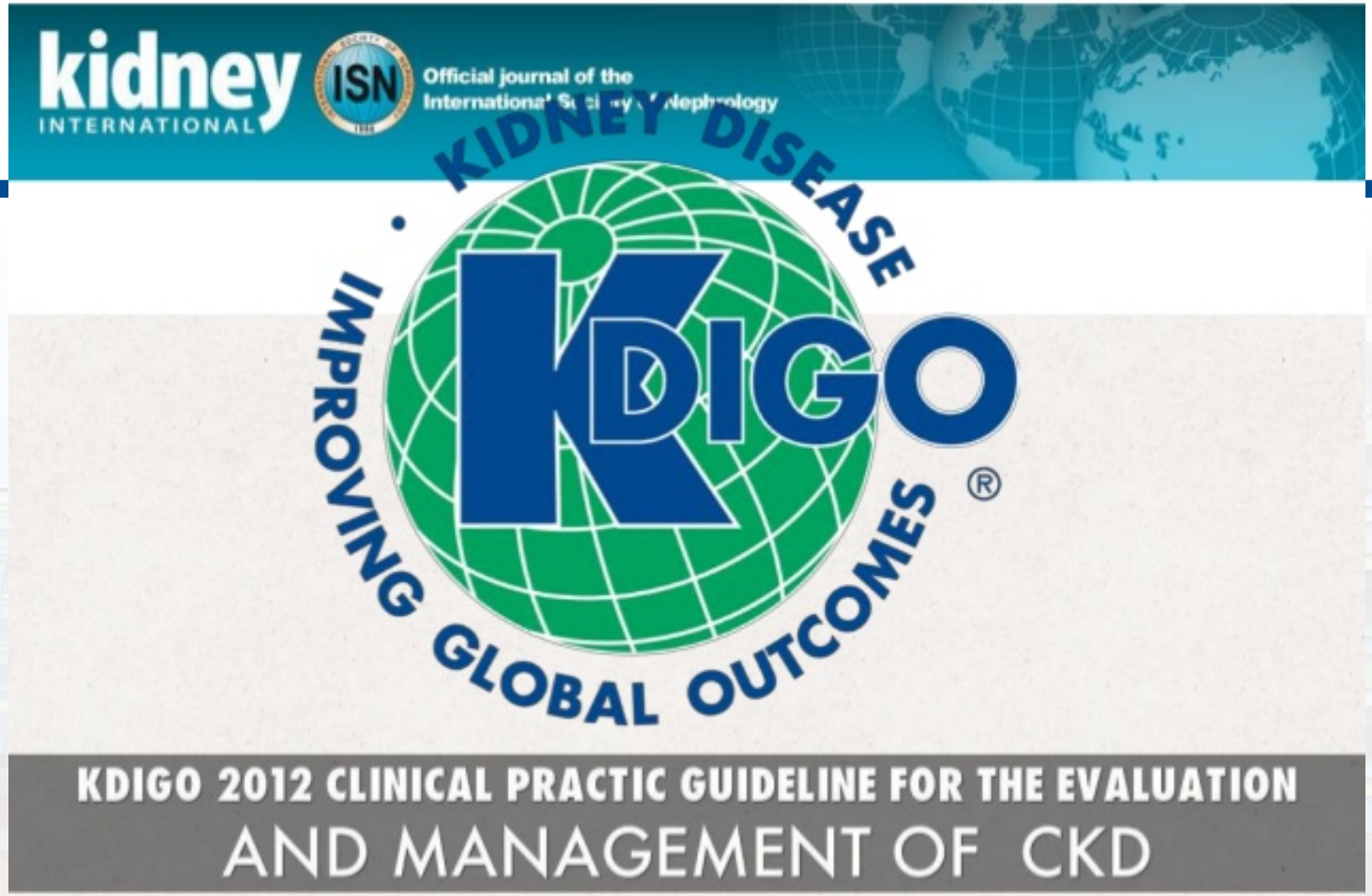
Atkinson, et. al. Pediatr Nephrol (2010) 25:1699-1709

Etiology of Anemia of CKD

- *Erythropoietin deficiency*
- *Iron deficiency*
- *Inflammation-associated iron restriction*
- Hyperparathyroidism
- “Uremic toxins”/Oxidative stress
- Other nutritional deficiencies

Adverse Associations of Anemia

- Anemia in CKD has been associated with a wide variety of adverse effects/outcomes including:
  - Hospitalization and mortality
  - Decreased quality of life
  - Increased risk for cardiovascular disease and CKD progression
  - Transfusions and allo-sensitization



http://www.kdigo.org/clinical_practice_guidelines/pdf/KDIGO-Anemia%20GL.pdf

Definition and Evaluation of Anemia

- CBC including red blood cell indices, WBC differential, and platelets
- Absolute reticulocyte count
- Serum ferritin
- Serum transferrin saturation, iron
  - % hypochromic red blood cells
  - Reticulocyte hemoglobin content
- Serum B_{12} and folate

Definition and Evaluation of Anemia

- *CBC including red blood cell indices, WBC differential, and platelets*
- *Absolute reticulocyte count*
- *Serum ferritin*
- *Serum transferrin saturation, iron*
  - *% hypochromic red blood cells*
  - *Reticulocyte hemoglobin content*
- *Serum B_{12} and folate*
- Hemoglobin electrophoresis
- Screen for hemolysis
- Screen for blood loss

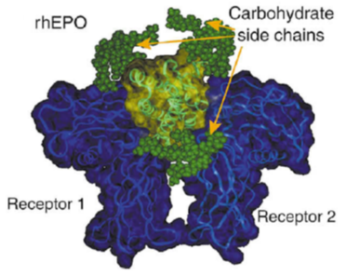
ESA for Anemia of CKD

1985

Human *EPO* gene isolated

↓

rHuEPO



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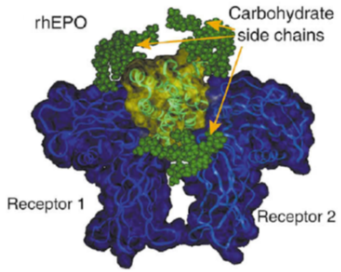
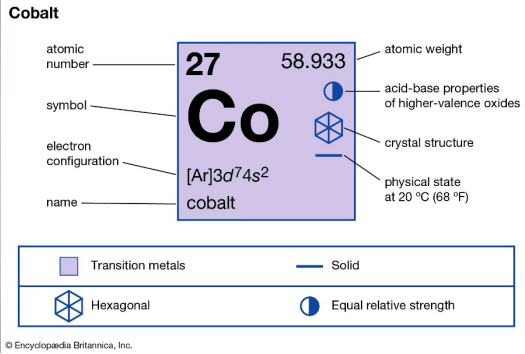


Prior to availability of rHuEPO, cobalt salts and androgens were used for treatment of anemia of CKD

Long-term cobalt ingestion caused cardiomyopathy, neuropathy, thyroid dysfunction



HIF stabilizer?

1 Epo unit = dose producing the same erythropoiesis-stimulating response as 5 µmol cobaltous chloride in experimental animals

Packed red blood cell transfusions

- Infection
- Iron overload
- Allosensitization

ESA for Anemia of CKD

1985

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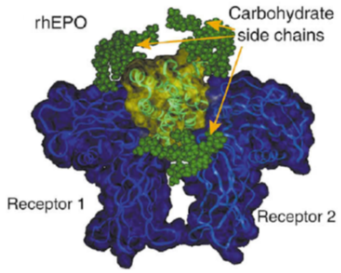
↓

rHuEPO



1987

Seminal *NEJM* paper reported that rHuEPO effectively raised Hgb and eliminated transfusions in 25 adults on HD

Eschbach et al. (1987)
NEJM 316(2):73-78

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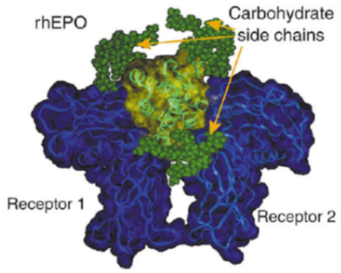

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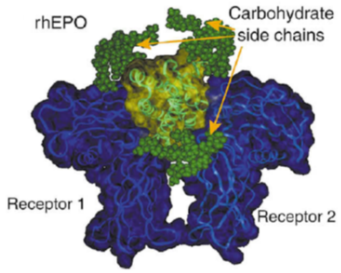

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NEJM 316(2):73-78

1989

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2001

Darbepoetin alfa approved by U.S. FDA

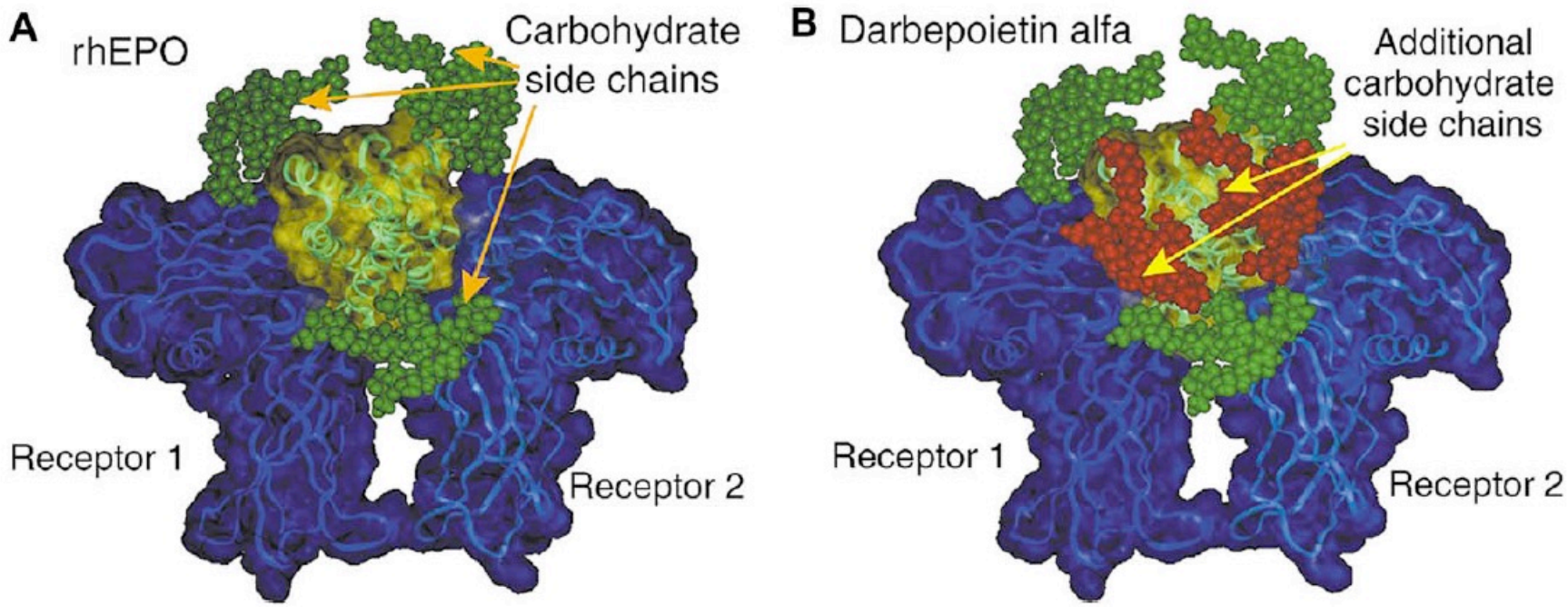


Figure 5. Molecular structures of rhEPO (**A**) and darbepoetin alfa (**B**). Reprinted by permission from Macmillan Publishers Ltd: Nature Biotechnology [7], 2003. rhEPO = recombinant human erythropoietin.

Experimental Hematology 2008;36:1573–1584

- Darbepoietin alfa – two additional sialic-acid-containing carbohydrates result in extended in vivo biologic activity

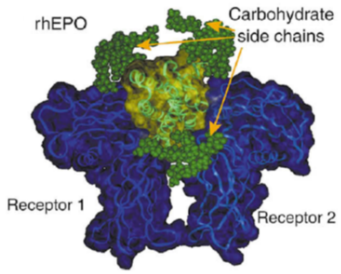
ESA for Anemia of CKD

1985

Human *EPO* gene isolated

↓

rHuEPO



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NEJM 316(2):73-78

1989

Epoetin alfa approved by U.S. FDA

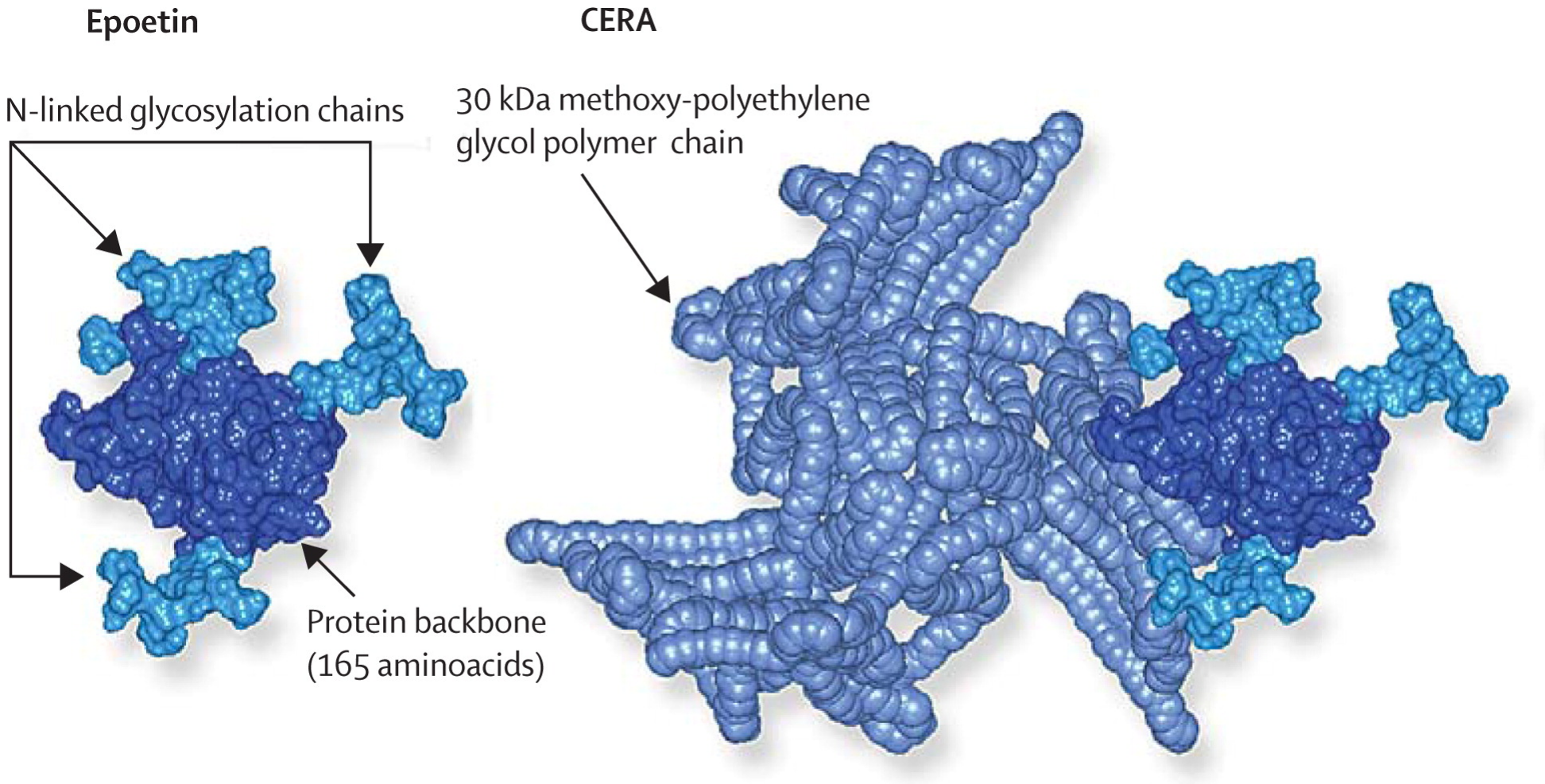
2001

Darbepoetin alfa approved by U.S. FDA

2007

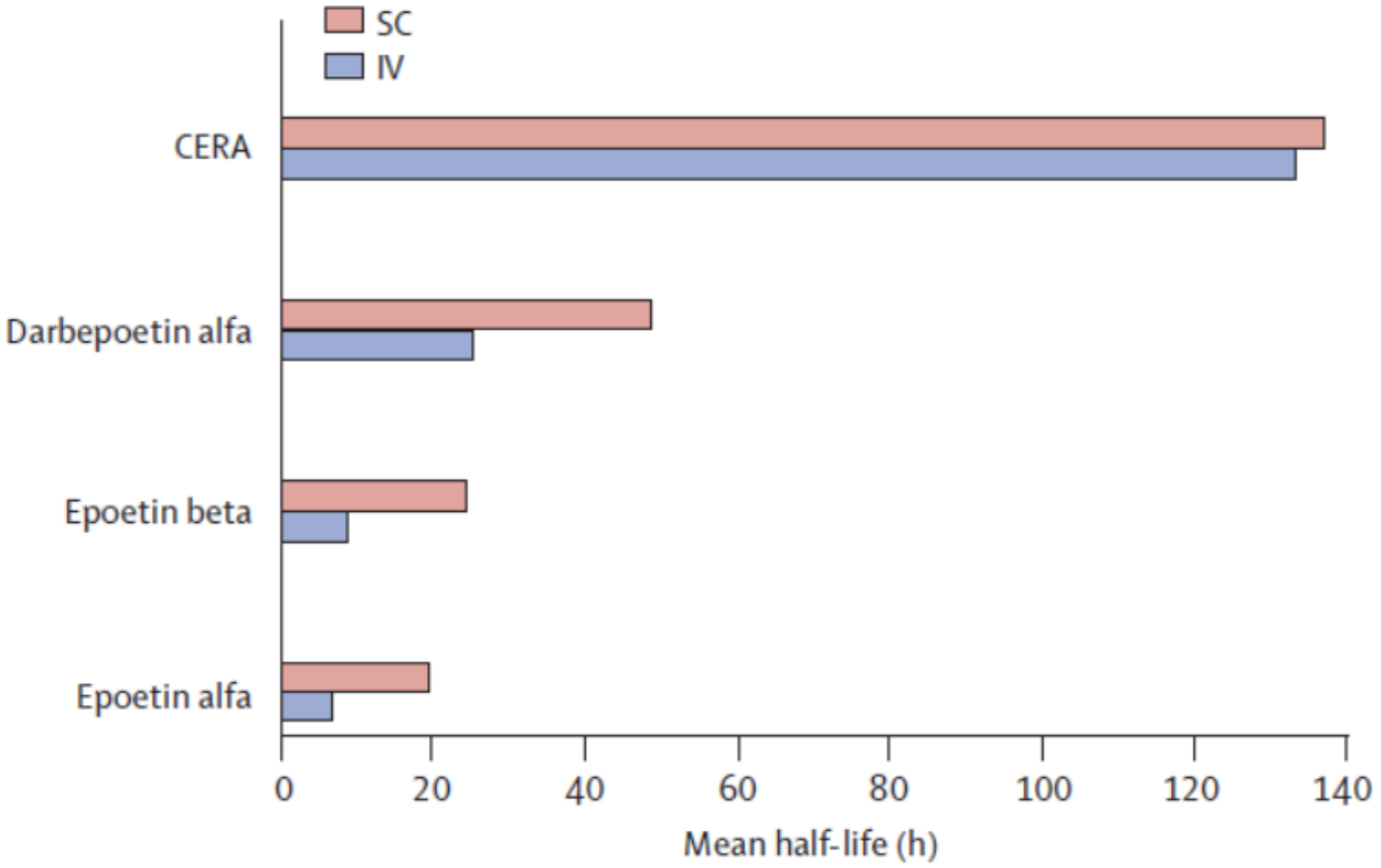
CERA approved by U.S. FDA

Methoxy polyethylene glycol-epoetin beta (Mircera® Hoffman-La Roche)
SC or IV administration



- Integration of a large methoxy polyethylene glycol polymer chain
- Extended half-life of up to 130 hours when given SC, 90 hours IV
- Allows for a monthly dosing regimen

Mean in vivo half-lives of available erythropoiesis stimulating agents



S. Elliott *et al.*, "Enhancement of therapeutic protein in vivo activities through glycoengineering," *Nat. Biotechnol.*, 2003.

ESA Initiation

3.4.5: For all pediatric CKD patients, we suggest that the selection of Hb concentration at which ESA therapy is initiated in the individual patient includes consideration of potential benefits (e.g., improvement in quality of life, school attendance/performance, and avoidance of transfusion) and potential harms. (*2D*)

- ESA initiation for hemoglobin 9-10 g/dl (90-100 g/l)
- *Children: maintain hemoglobin 11-12 g/dl (110-120 g/l)

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KDIGO Clinical Practice Guideline for Anemia in Chronic Kidney Disease. *Kidney International* 2(4): August 2012

- **Physicians and their patients with chronic kidney disease should weigh the possible benefits of using ESAs to decrease the need for red blood cell transfusions against the increased risks for serious adverse cardiovascular events. For each patient, individualize dosing and use the lowest dose of ESA sufficient to reduce the need for transfusion.**

- **For patients with the anemia of chronic kidney disease NOT on dialysis**
- Consider starting ESA treatment only when the hemoglobin level is less than 10 g/dL and when certain other considerations apply
- If the hemoglobin level exceeds 10 g/dL, reduce or interrupt the dose of ESA.

- **For patients with the anemia of chronic kidney disease on dialysis**
- Initiate ESA treatment when the hemoglobin level is less than 10 g/dL.
- If the hemoglobin level approaches or exceeds 11 g/dL, reduce or interrupt the dose of ESA.

"WARNING: ESAs INCREASE THE RISK OF DEATH, MYOCARDIAL INFARCTION, STROKE, VENOUS THROMBOEMBOLISM, THROMBOSIS OF VASCULAR ACCESS AND TUMOR PROGRESSION OR RECURRENCE"

See full prescribing information for complete boxed warning.

ESA Dosing

- Goal rate of hemoglobin increase:1-2 g/dL/month
- Epoetin alfa or beta
  - 20-50 IU/kg/dose three times weekly IV or SC
- Darbepoetin alfa
  - 0.45 µg/kg SC or IV weekly
  - 0.75 µg/kg SC or IV every 2 weeks

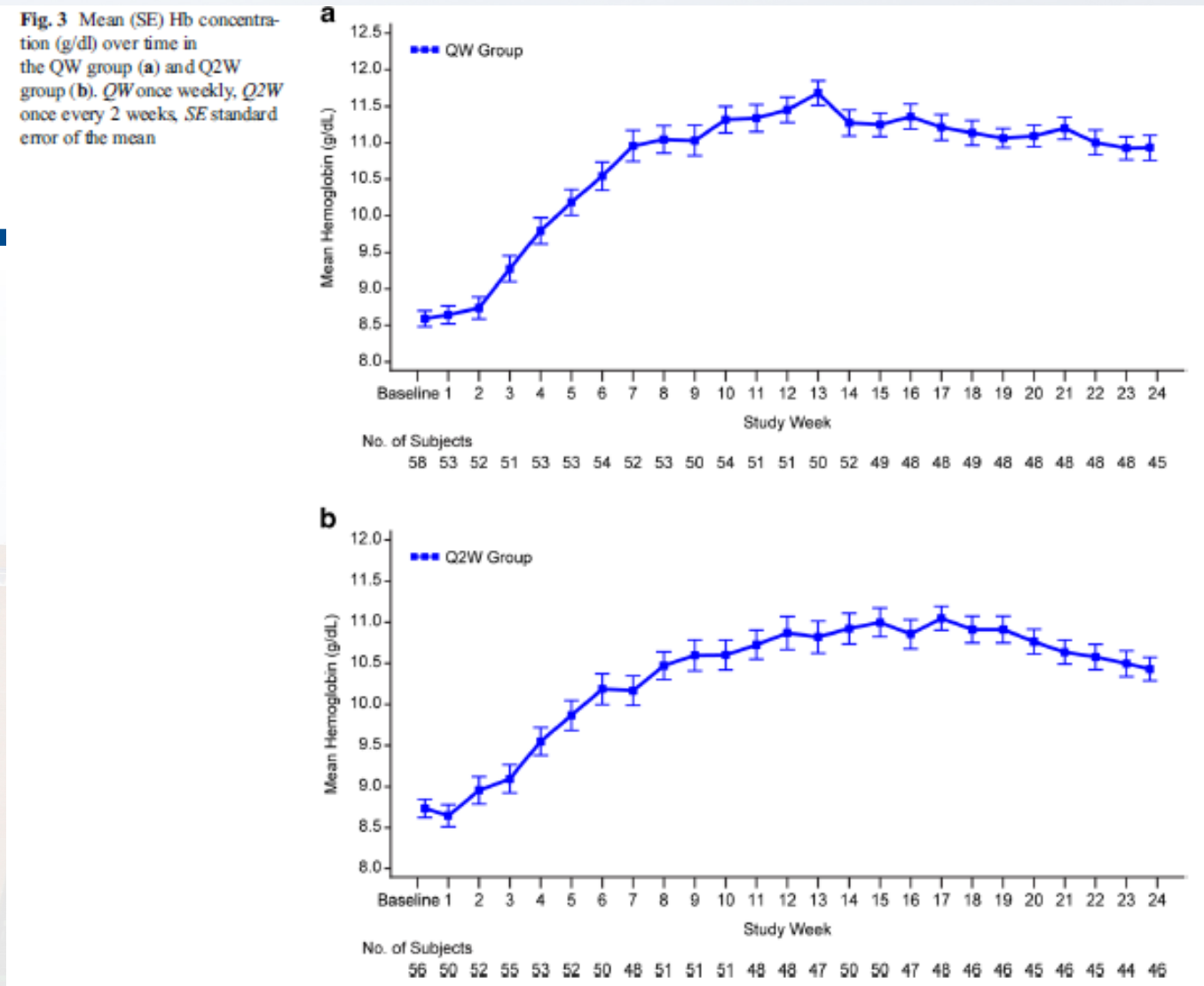

Fig. 3 Mean (SE) Hb concentration (g/dl) over time in the QW group (**a**) and Q2W group (**b**). *QW* once weekly, *Q2W* once every 2 weeks, *SE* standard error of the mean

- Darbepoetin alfa can be safely administered either weekly or q 2 weeks in ESA-naïve pediatric pts to achieve Hgb targets of 10-12.

Warady et al. De novo weekly and biweekly darbepoetin alfa dosing in pediatric patients with chronic kidney disease. *Pediatric Nephrology*. 2018;33:125-137.

ESA Dosing

- 275-350 units/kg/week in infants
  - Koshy et al. Anemia in children with CKD. *Ped Neph* 23: 2008
- 200-250 units/kg/week in older children
  - Koshy et al. Anemia in children with CKD. *Ped Neph* 23: 2008
- Children and adolescents on HD may require higher absolute doses than adults despite lower body weight
  - Bamgbola et al. Analyses of age, gender, and other risk factors for Epo resistance. Ped Neph 24:2009
- Increased drug clearance with growth?

ESA Dosing

- Make dose adjustments after 4 weeks of therapy
- No more often than q 2 weeks
- When a decrease in hemoglobin is necessary, decrease dose rather than hold therapy
- Long-acting ESAs – lower starting dose and less frequent adjustments

Efficacy and Long-Term Safety of C.E.R.A. Maintenance in Pediatric Hemodialysis Patients with Anemia of CKD

Michel Fischbach,[1] Elke Wühl,[2] Sylvie C. Meyer Reigner,[3] Zoe Morgan,[4] and Franz Schaefer[2]

Clin J Am Soc Nephrol 13: 81–90, January, 2018

- Phase II, open label, multicenter, multiple-dose study conducted at 28 sites in 10 countries
- 64 children aged 6-17 years on **chronic HD** received CERA (Mircera®) monthly
- Objective: identify a conversion factor for switching from previous ESAs (epoetin or darbepoetin) to CERA
  - Safety and efficacy

Efficacy and Long-Term Safety of C.E.R.A. Maintenance in Pediatric Hemodialysis Patients with Anemia of CKD

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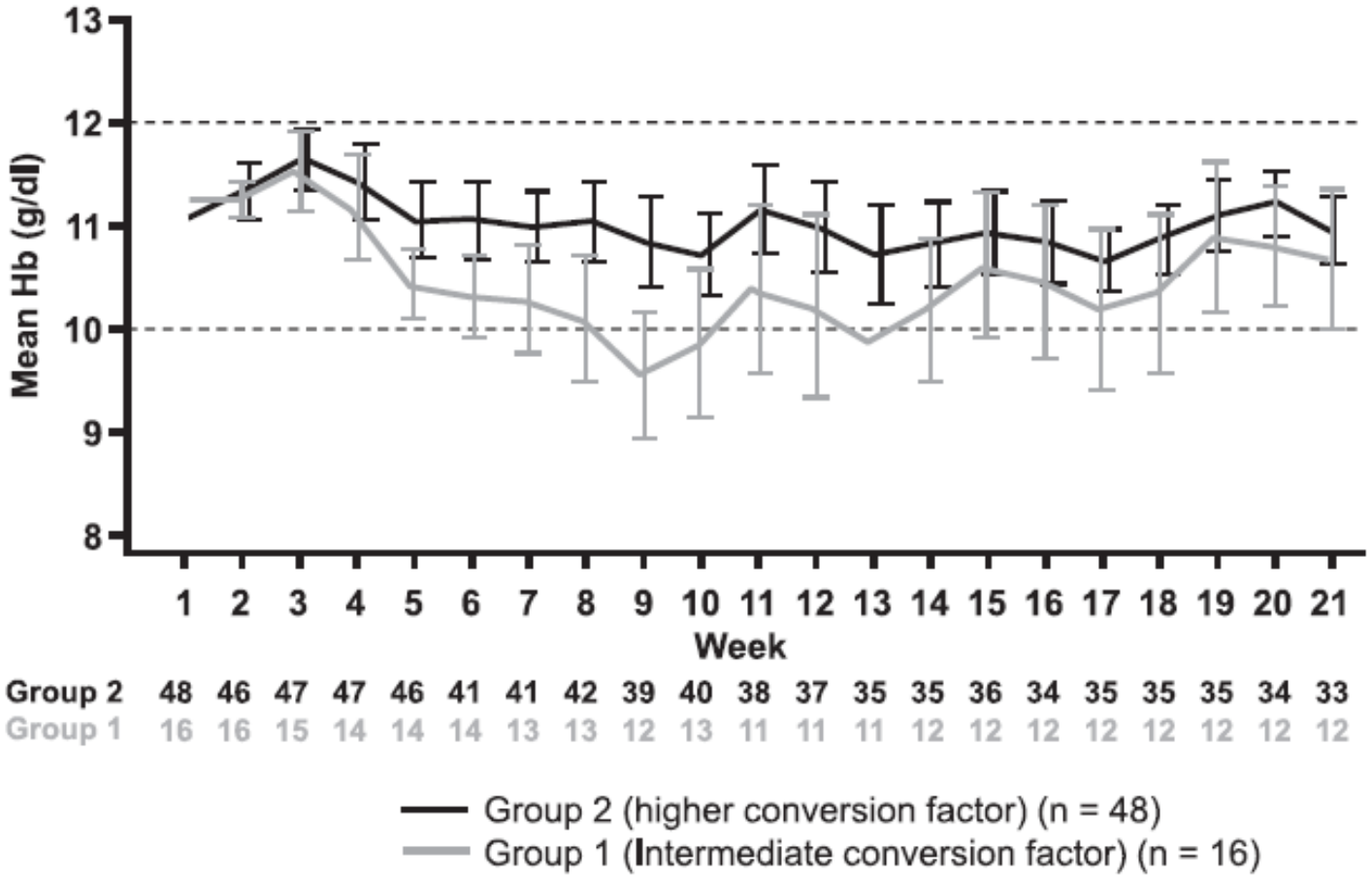

Figure 2. | Hemoglobin (Hb) was maintained within the target range in the higher conversion factor group during the core trial period. Mean Hb values with 95% confidence intervals are shown. Dashed lines indicate target range.

Efficacy and Long-Term Safety of C.E.R.A. Maintenance in Pediatric Hemodialysis Patients with Anemia of CKD

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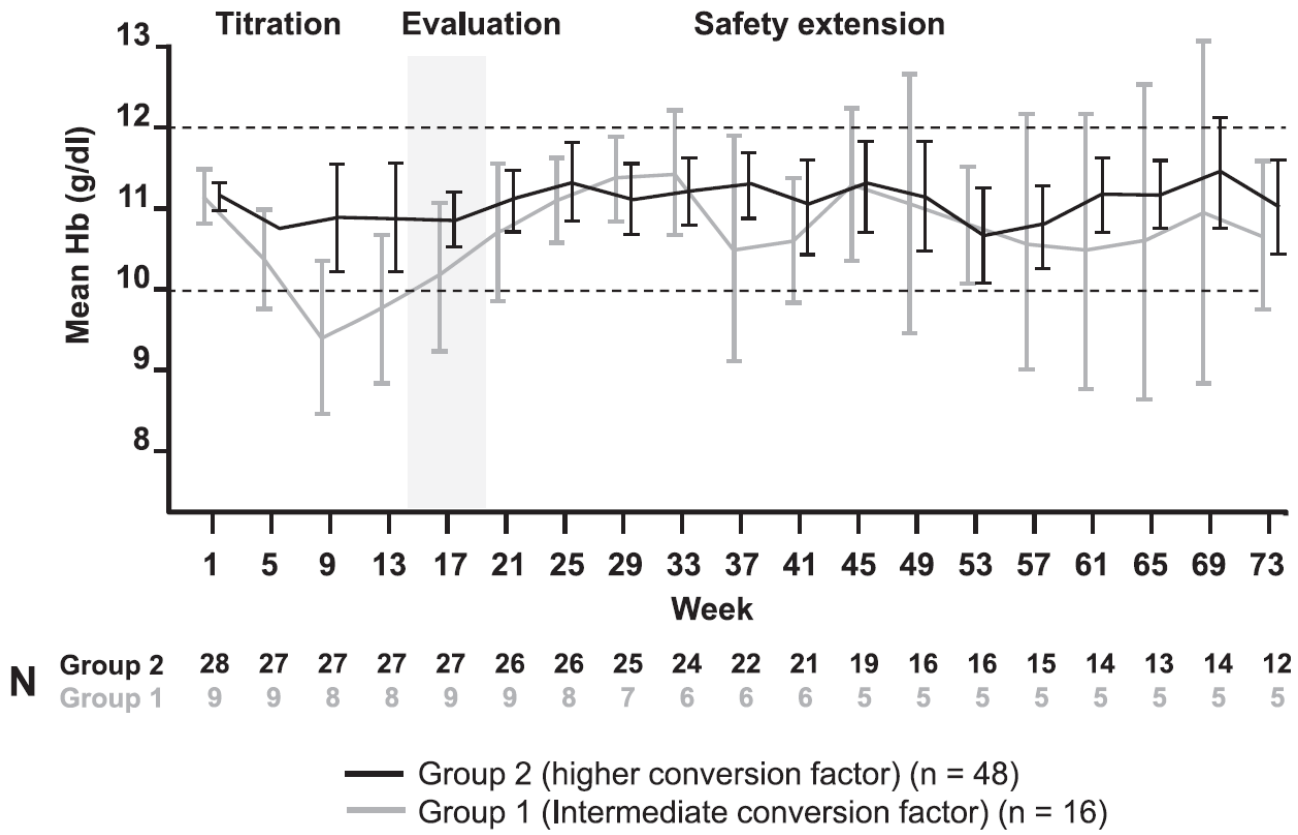


Figure 5. | Hemoglobin (Hb) concentrations were maintained in both groups in patients who entered the extension phase. Mean Hb values with 95% confidence intervals are shown. Dashed lines indicate the target range.

Efficacy and Long-Term Safety of C.E.R.A. Maintenance in Pediatric Hemodialysis Patients with Anemia of CKD

Michel Fischbach,[1] Elke Wühl,[2] Sylvie C. Meyer Reigner,[3] Zoe Morgan,[4] and Franz Schaefer[2]

Clin J Am Soc Nephrol 13: 81–90, January, 2018

On the basis of our results, patients aged 6–17 years with stable hemoglobin receiving darbepoetin alfa or epoetin alfa/beta can be switched to C.E.R.A. at a dose corresponding to 4 μg every 4 weeks for each 125 IU epoetin alfa/beta or 0.55 μg darbepoetin.

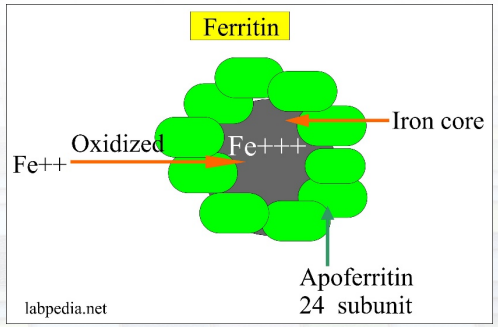
Iron Deficiency and Supplementation

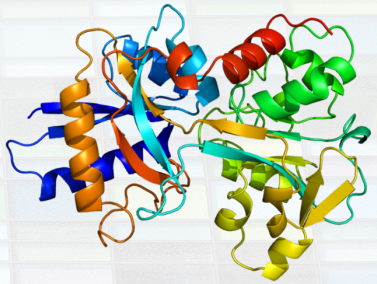
Iron Deficiency (ID)

- Correction of ID reduces severity of anemia of CKD
- Untreated ID is a frequent cause of ESA hypo-responsiveness
- Risk factors include:
  - Blood loss
  - Inflammation
  - Poor absorption of enteral iron

Biomarkers of Iron Availability

- Ferritin (serum)
  - Intracellular iron-storage protein
  - ⇧ by inflammation, iron overload
- Transferrin saturation (TSAT)
  - Transferrin binds to iron in plasma
  - Transports to bone marrow





JOHNS HOPKINS CHILDREN'S CENTER

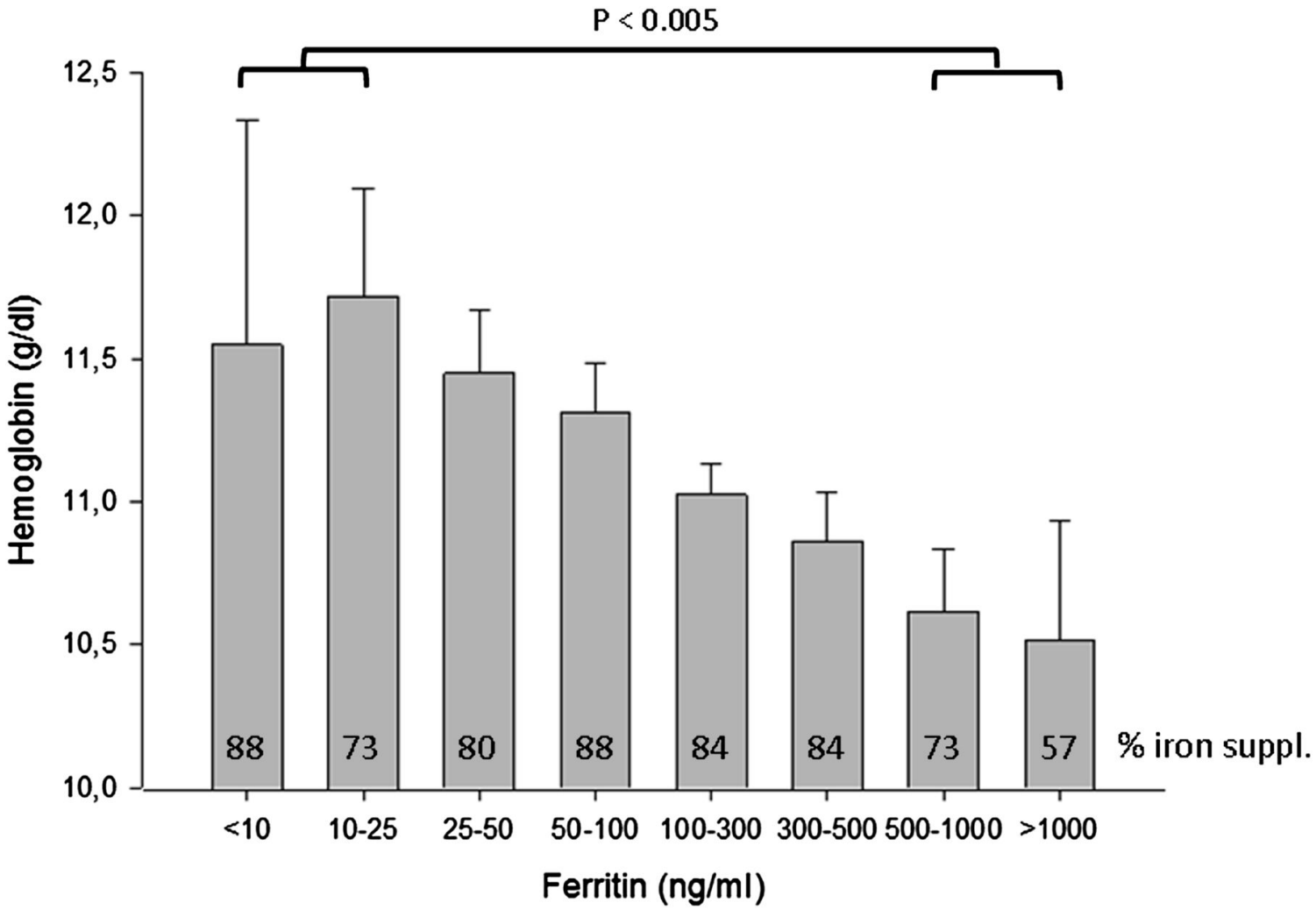
KDIGO Iron Targets

- In ESA-treated patients, iron supplementation to maintain
  - Ferritin ≥ 100 ng/mL
  - TSAT ≥ 20%
- Ferritin has limitations as a marker of *accessible* stored iron
  - Hepcidin-mediated iron blockade
  - Low ferritin = iron deficiency
  - High ferritin does not rule out iron blockade

KDIGO Iron Targets

- No routine iron supplementation for
  - Ferritin > 500
  - TSAT > 30%

Hb grouped by concomitant serum ferritin levels.



Dagmara Borzych-Duzalka et al. JASN 2013;24:665-676

Iron Supplementation: Route

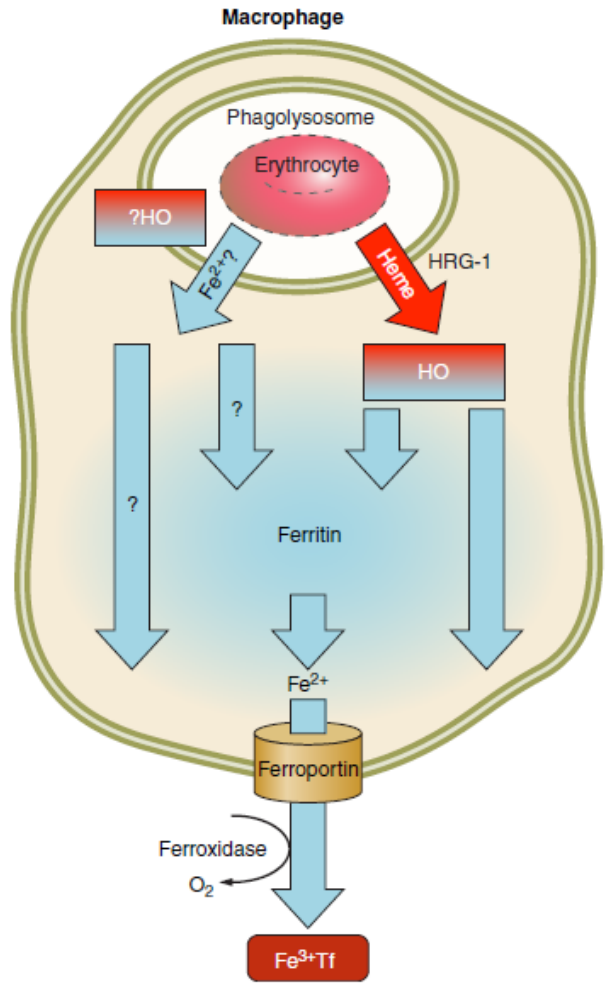
- Oral/Enteral
  - Pros: inexpensive, available, few adverse effects
  - Cons: poorly absorbed, adherence
  - Dosing: 2-6 mg/kg/day elemental iron
- Intravenous

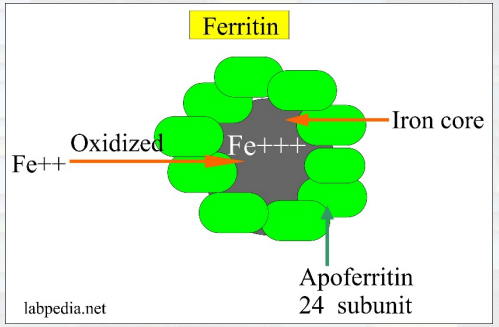
Iron Supplementation: Route

POTENTIAL BENEFITS OF IV IRON	POTENTIAL RISKS/ADVERSE EFFECTS OF IV IRON
• Decrease PRBC transfusion rates • Improved hemoglobin (and associated improved QOL?) • Decrease in required ESA dose • Adherence assured	• Anaphylaxis • Oxidative stress, endothelial dysfunction • Cellular iron deposition • Pro-oxidant cell-signaling • Infection • Cost, burden of visits, monitored administration

Goals of Iron Supplementation

- Avoid depletion of iron stores
- Prevent iron-restricted erythropoiesis





Ganz. Systemic Iron Homeostasis (2013) Physiol Rev 93:1721-1741.

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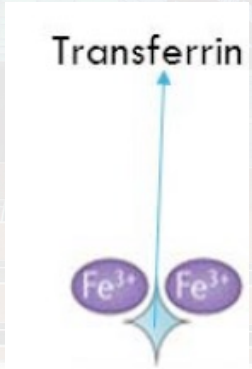

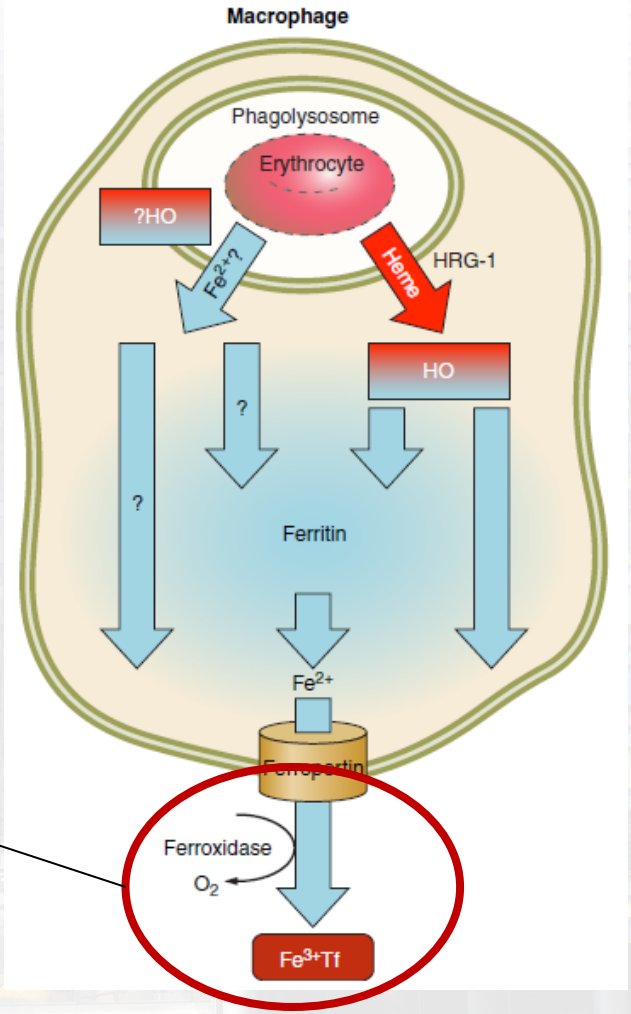

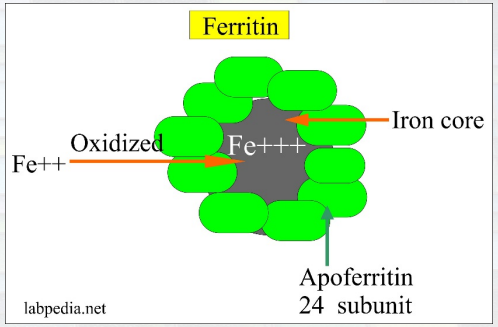

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Goals of Iron Supplementation

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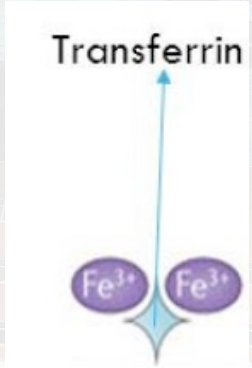

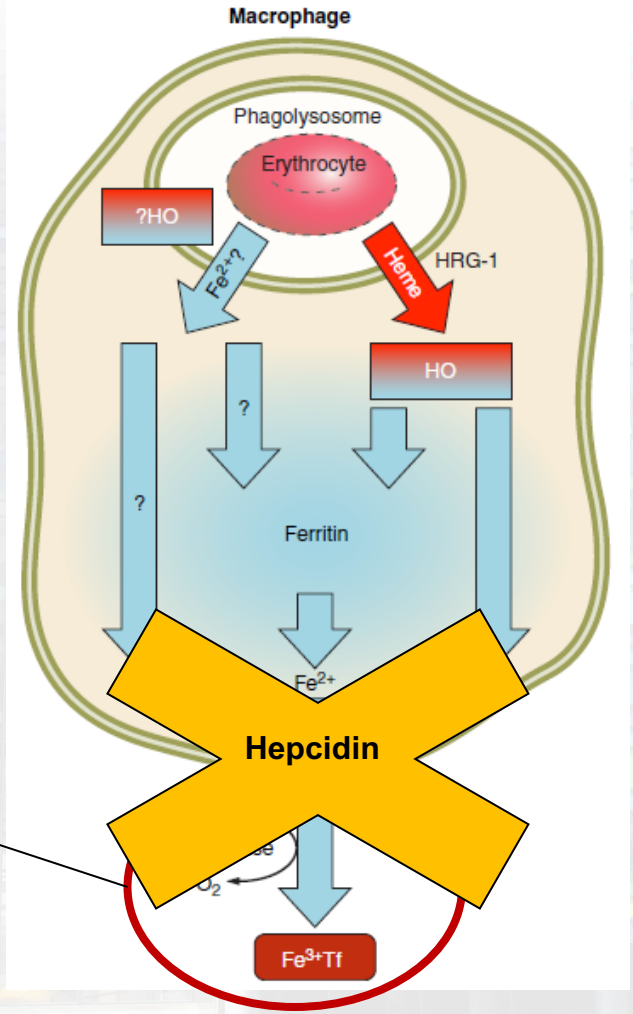

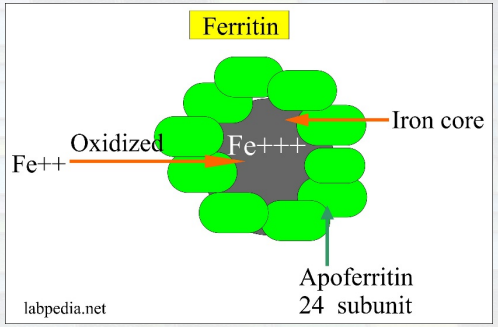

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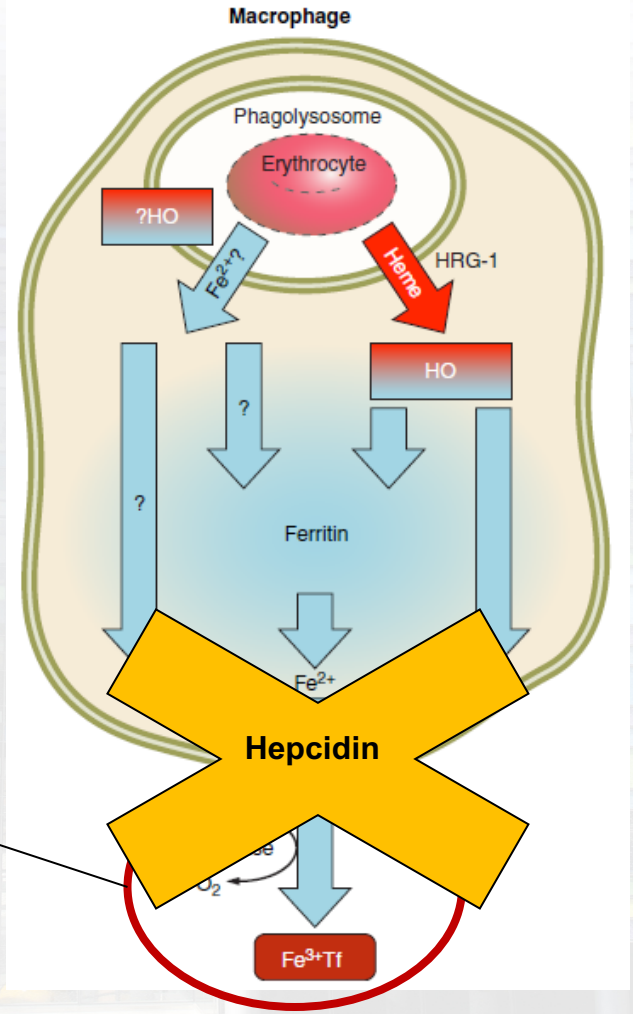
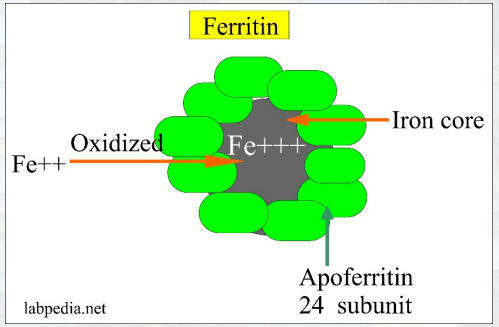
JOHNS HOPKINS CHILDREN'S CENTER

Goals of Iron Supplementation

- Avoid depletion of iron stores
- Prevent iron-restricted erythropoiesis

IV IRON
- Oxidative stress
- Increased inflammation
- Endothelial dysfunction
- Potentiating infection
- Tissue injury





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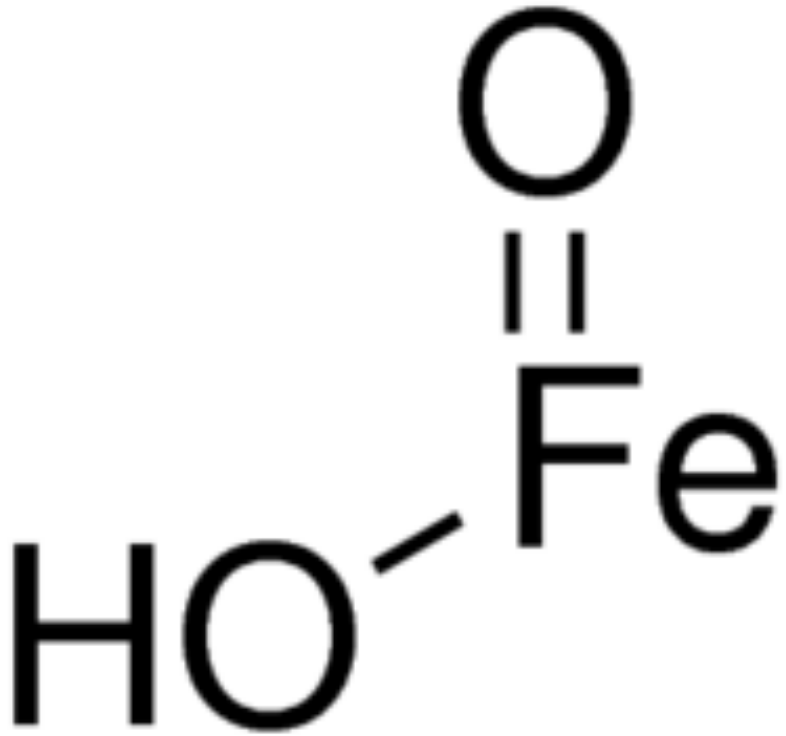
JOHNS HOPKINS CHILDREN'S CENTER

IV Iron: Safety

Table 1. Physiochemical char

Properties	Fer	n ose	Ferric Gluconate
Molecular mass (D)	7	000	200,000
Carbohydrate shell	Polyglu carbc		Gluconate, loosely associated sucrose
Median shell/particle diameter (nm)		3	8.6
Relative catalytic iron release		+	+++
Relative stability of elemental iron within the carbohydrate shell	High		Low
Relative osmolality	Isotonic		Hypertonic
Administration (iv push) rates	30 mg/	y 20 mg/min	12.5 mg/min
$t_{1/2}$ (h)	Approx		Approximately 1

D, daltons; nm, nanometer; iv, intraven



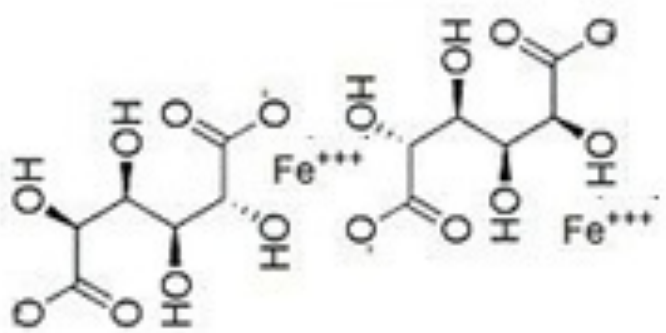
Charytan et al. Considerations and Challenges in Defining Optimal Iron Utilization in Hemodialysis. J Am Soc Nephrol 26:1238-1247:2015

IV Iron: Safety

Table 1. Physiochem

Properties	Ferric Gluconate
Molecular mass (D)	200,000
Carbohydrate shell	Gluconate, loosely associated sucrose
Median shell/particle diameter (nm)	8.6
Relative catalytic iron release	+++
Relative stability of elemental iron within the carbohydrate shell	Low
Relative osmolality	Hypertonic
Administration (iv push) rates	g/min 12.5 mg/min
$t_{1/2}$ (h)	Approximately 1

D, daltons; nm, nanometer; iv, intravenous.



IRON SUCROSE COMPLEX
CAS #8047-67-4

Charytan et al. Considerations and Challenges in Defining Optimal Iron Utilization in Hemodialysis. J Am Soc Nephrol 26:1238-1247:2015

JOHNS HOPKINS CHILDREN'S CENTER

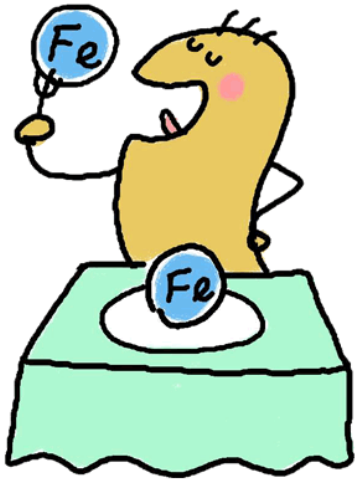
IV Iron: Safety

Iron during infection

2.4: Avoid administering IV iron to patients with active systemic infections. (*Not Graded*)

KDIGO Clinical Practice Guideline for Anemia in Chronic Kidney Disease. *Kidney International* 2(4): August 2012

- Iron is essential for bacterial growth
- May impair host immune response by decreasing PMN and T-cell function



AJKD

Special Report

Diagnosis and Management of Iron Deficiency in CKD: A Summary of the NICE Guideline Recommendations and Their Rationale

Laura E.K. Ratcliffe, MRCP,[1] Wayne Thomas, FRCPath,[2] Jessica Glen, MSc,[3] Smita Padhi, MBBS, MPH,[3] Ben A.J. Pordes, BSc,[3] David Wonderling, MSc,[3] Roy Connell, RN (Child), MSc,[4] Suzanne Stephens, MBBS, FRCPCH,[5] Ashraf I. Mikhail, MD, FRCP,[6] Damian G. Fogarty, MD,[7] Jan K. Cooper,[8] Belinda Dring, BSc, MPH,[4] Mark A.J. Devonald, FRCPE,[4] Chris Brown, MPharm,[6] and Mark E. Thomas, FRCP[9]

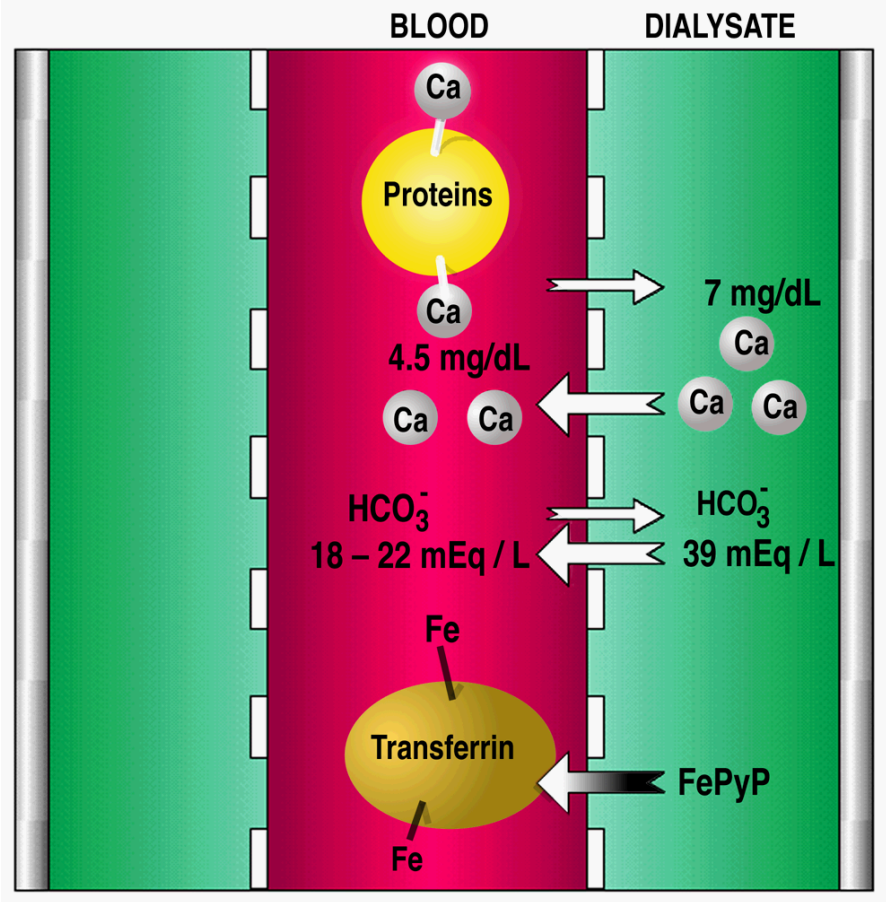
Am J Kidney Dis. 67(4):548-558.

- Children not on an ESA and not on HD, treat with oral iron unless "intolerant" or target Hgb is not reached within 3 months
- On ESA and not on HD → trial of oral iron
- Offer IV iron to children on HD

Novel Routes of Iron Supplementation

Intradialysate Soluble Ferric Pyrophosphate Citrate (FPC) (Triferic®)

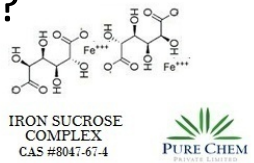
- Water soluble, no-carbohydrate shell, tightly complexed salt of Fe, electrostatically bonded to pyrophosphate
- Added to bicarbonate concentrate at each hemodialysis session
  - Dialysate with 110 µg/L iron
- Crosses the dialyzer membrane and donates iron directly to transferrin, bypassing hepcidin induced iron-sequestration
- Approved by U.S. FDA in 2015 for iron replacement and to maintain Hgb in adults on hemodialysis
- Also available in an IV formulation



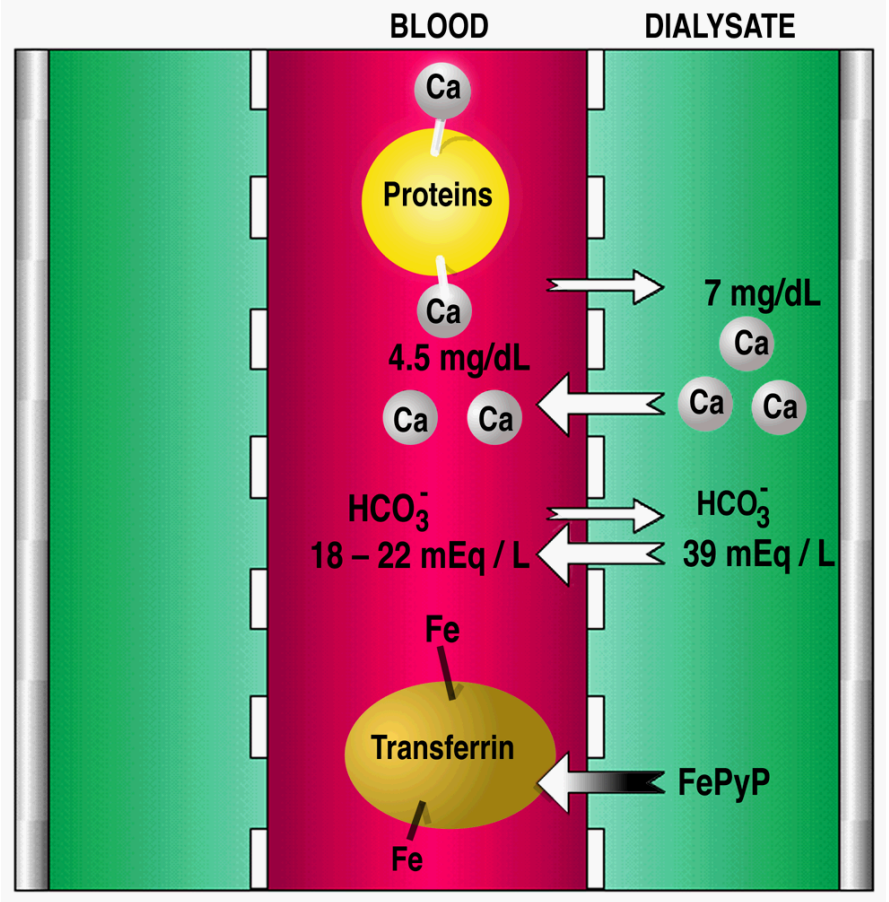
Courtesy of Dr. Ajay Gupta
via Dr. Brad Warady

Intradialysate Soluble Ferric Pyrophosphate Citrate (FPC) (Triferic®)

- FPC designed as a maintenance therapy, not repletion
  - Small doses of iron that are immediately bioavailable/bound to transferrin and rapidly delivered to iron-requiring tissue
- Different from parenteral iron products?
  - No carbohydrate shell



  - Iron more tightly bound to pyrophosphate compared to carbohydrate complexes
  - Rapidly bound to transferrin -> bone marrow, may avoid storage in reticuloendothelial system



Courtesy of Dr. Ajay Gupta
via Dr. Brad Warady

Pharmacokinetics of ferric pyrophosphate citrate administered via dialysate and intravenously to pediatric patients on chronic hemodialysis

Raymond D. Pratt[1] · Sarah Grimberg[1] · Joshua J. Zaritsky[2] · Bradley A. Warady[3]

Pediatric Nephrology (2018) 33:2151–2159

- Study Objectives
  - Evaluate pharmacokinetics and preliminary safety of FPC
  - Evaluate the dose of FPC delivered via dialysate in children on chronic HD
  - Examine the feasibility of IV administration of FPC in pediatric patients
    - Providing a dosing option for patients in HD systems which do not use liquid bicarbonate concentrate

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Pediatric Nephrology (2018) 33:2151–2159

- Study Design
  - Multicenter, open-label, two-period, single dose study
  - 2 week screening period followed by two sequential FPC treatment sessions
    - Single 0.07 mg/kg dose in D5W as a continuous IV infusion throughout the HD session
    - FPC added to dialysate to deliver a final dose of 110 µg/L throughout the HD session

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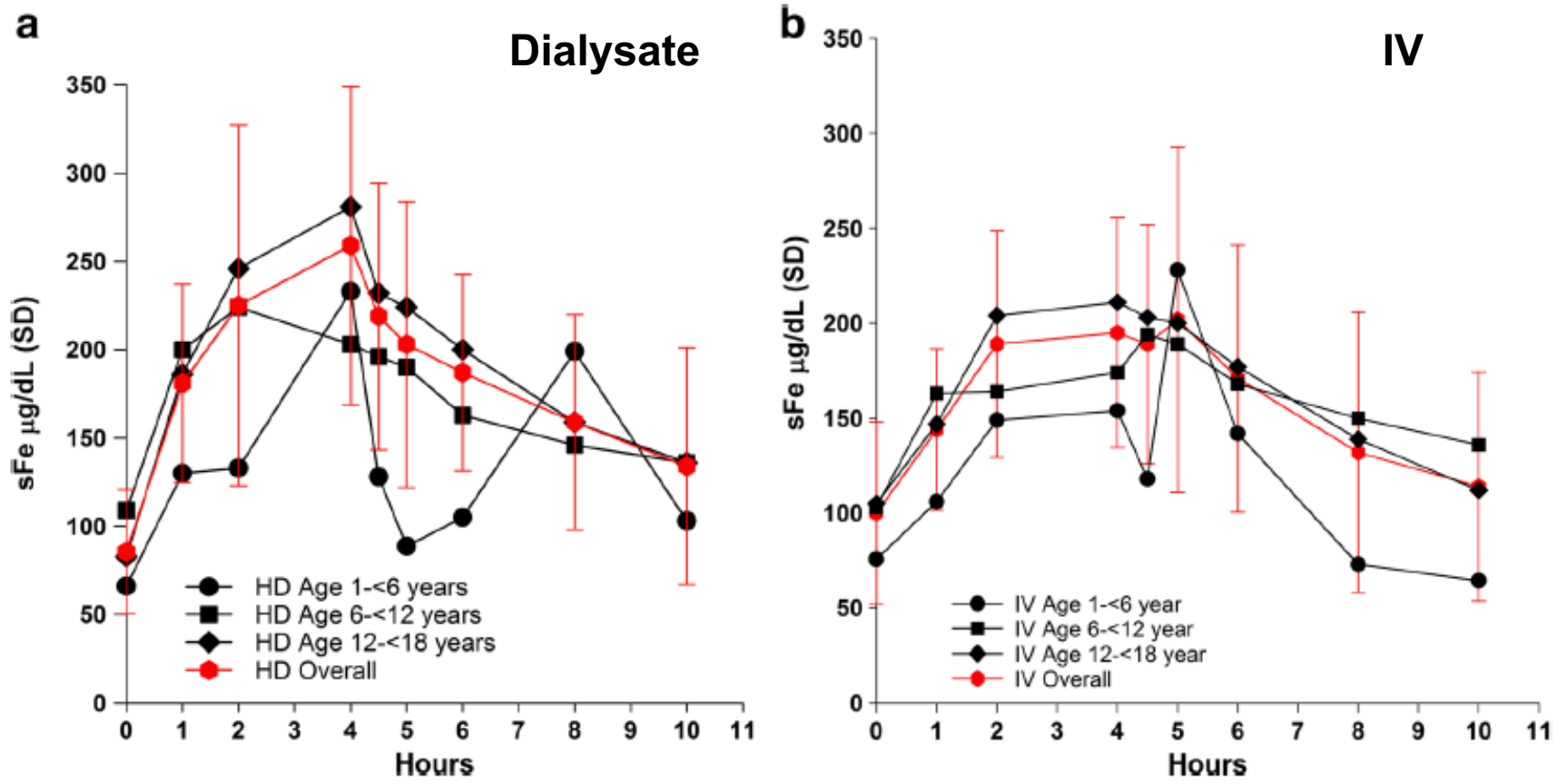

Fig. 1 Mean concentration-time plots for serum total iron (sFe) after administration of ferric pyrophosphate citrate (FPC) via hemodialysis (HD) at a concentration of 2 µM (110 µg/L) iron (**a**) and after intravenous (IV) administration of 0.07 mg Fe/kg of FPC by age group (years) and overall (**b**)

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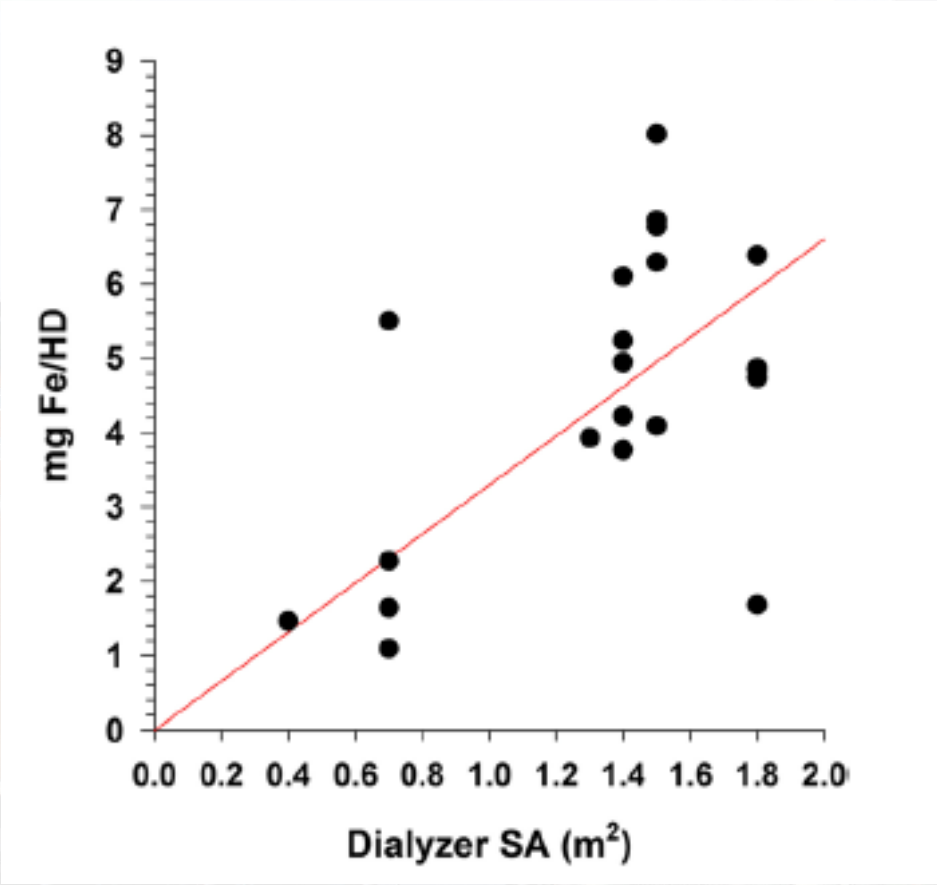

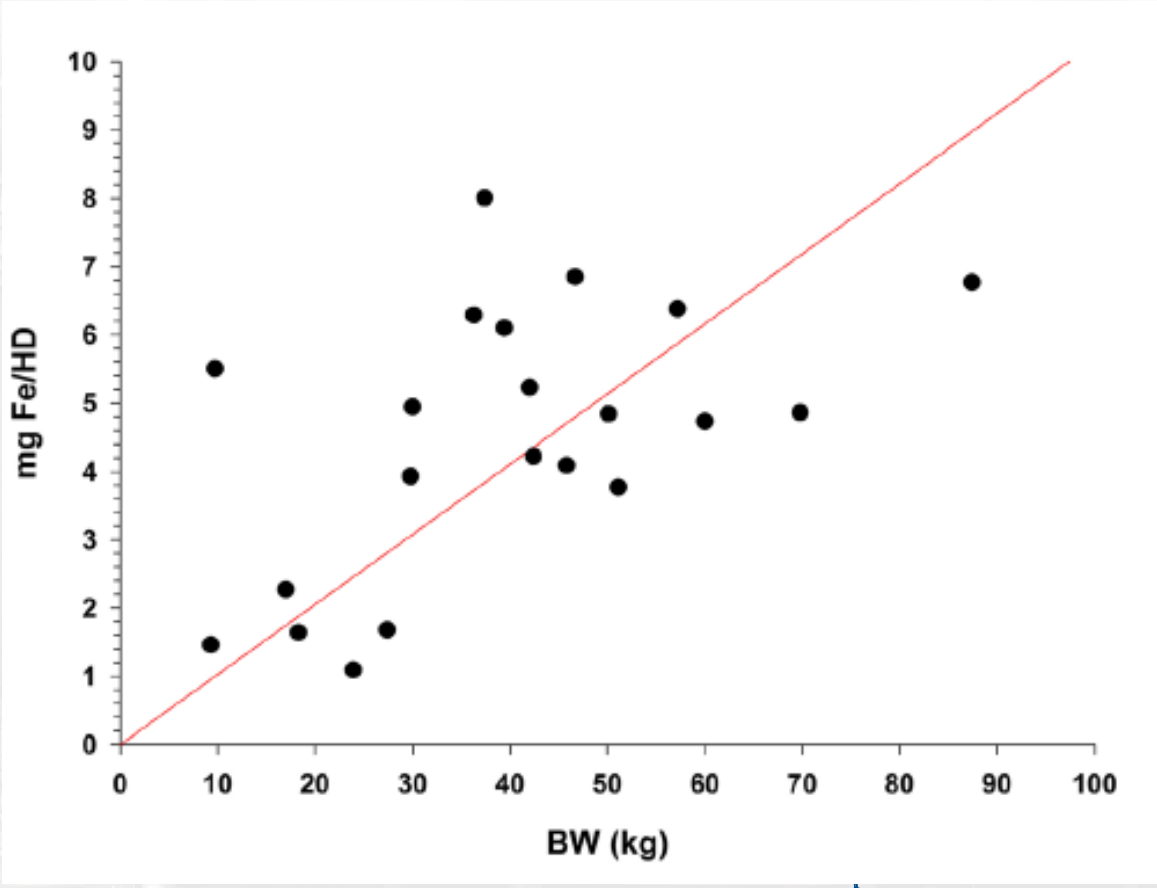

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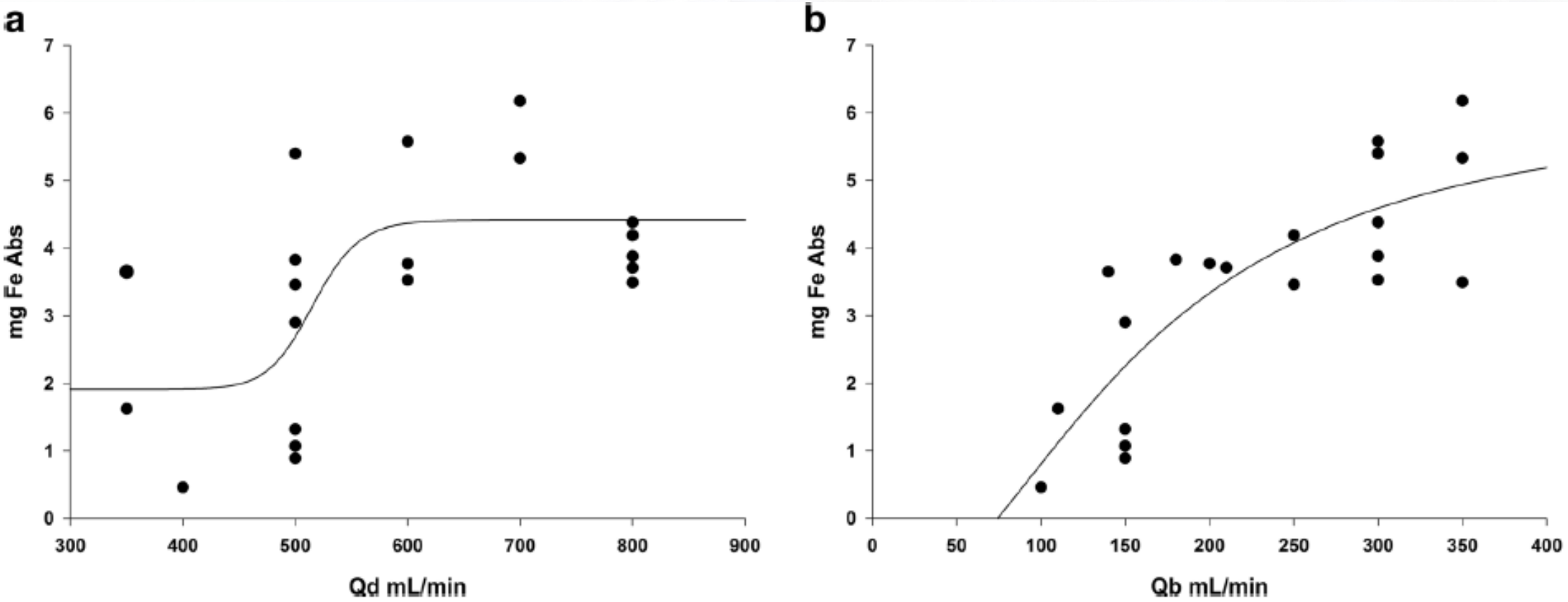

Fig. 3 Amount of iron (Fe) administered during hemodialysis (HD) by dialysate flow rate (Qd) (**a**) and blood flow rate (Qb) (**b**). Regression lines are fitted by nonlinear regression in SigmaPlot V14.0

Pharmacokinetics of ferric pyrophosphate citrate administered via dialysate and intravenously to pediatric patients on chronic hemodialysis

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Pediatric Nephrology (2018) 33:2151–2159

Table 2 Baseline-corrected noncompartmental pharmacokinetic parameters of serum total iron in pediatric patients after administration of ferric pyrophosphate citrate intravenously and via dialysate

Route of administration	Pharmacokinetic parameter[a]		
	C_{max} (µg/dL)	AUC_{last} (h µg/dL)	$t½$ (h)
Intravenous ($N=21$)	114 (53.7)	419 (101.6)	1.60 (190.1)[b]
Via dialysate ($N=20$)	166 (54.3)	682 (82.9)	1.98 (60.6)[c]

AUC_{last} area under the serum concentration-time curve from time zero to the time of the last quantified concentration, *C_{max}* maximum drug concentration in serum, *CV%* percent coefficient of variation, *$t½$* terminal phase half-life

[a] Values are reported as geometric mean (geometric CV%)

[b] $n=8$

[c] $n=10$

- Total iron exposure was greater after FPC administration via dialysate than after IV administration for all patients
- Weight-normalized amount of iron delivered via dialysate was ~ 0.06-0.10 mg/kg

Pharmacokinetics of ferric pyrophosphate citrate administered via dialysate and intravenously to pediatric patients on chronic hemodialysis

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Pediatric Nephrology (2018) 33:2151–2159

- Safety
  - No SAE's, no interruptions or discontinuations for AE's
  - All AE's reported at low incidence, most mild, and not attributed to FPC
  - 1 drug-related event reported
    - Moderate axillary pain

Pharmacokinetics of ferric pyrophosphate citrate administered via dialysate and intravenously to pediatric patients on chronic hemodialysis

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Pediatric Nephrology (2018) 33:2151–2159

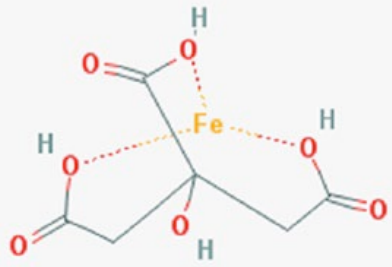
The recommended initial dose of FPC for future studies in pediatric patients with CKD-5HD is 2 μM (110 μg iron/L) in dialysate or 0.1 mg iron/kg IV during HD, using weight-based dosing for patients weighing < 50 kg and 6.75 mg IV for patients weighing > 50 kg.

NAPRTCS
North American Pediatric Renal Trials and Collaborative Studies

- Efficacy trial planned

Ferric Citrate

- Iron-based oral phosphate binder
- Approved by the U.S. FDA in 2014 for use as a phosphate binder in adults on dialysis
- Ferric ion dissociates in the GI tract and combines with dietary phosphorus and is excreted as ferric phosphate



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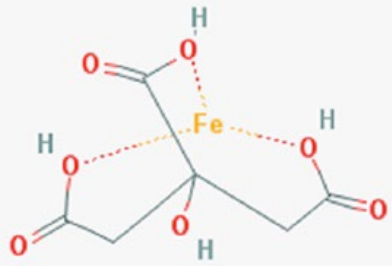
Ferric Citrate

- Some of the ferric ions dissociated from ferric citrate are reduced by the bowel mucosa to ferrous iron and absorbed through the duodenal brush border – ferroportin channels
- Data in adults that ferric citrate in dialysis patients is associated with increased transferrin saturation, decrease IV iron requirement, and decreased ESA dose

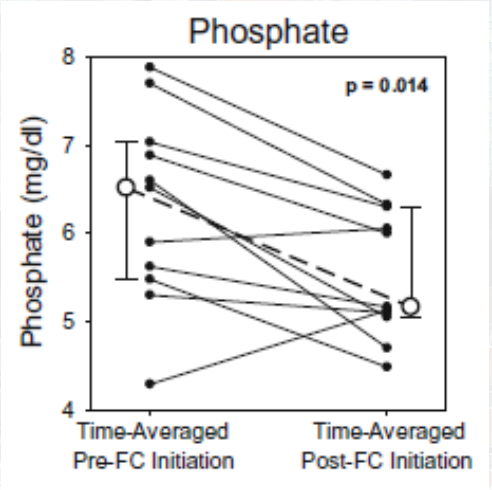


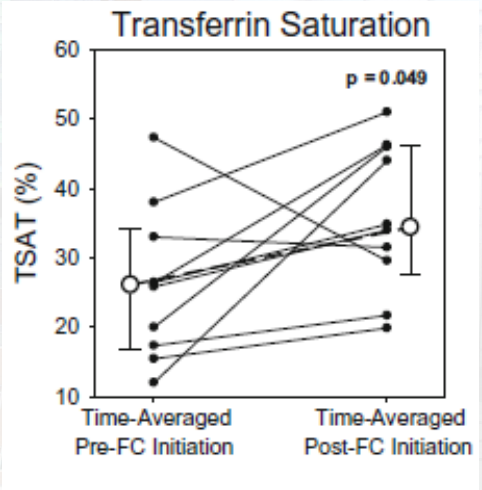
Clinical experience with the use of ferric citrate as a phosphate binder in pediatric dialysis patients

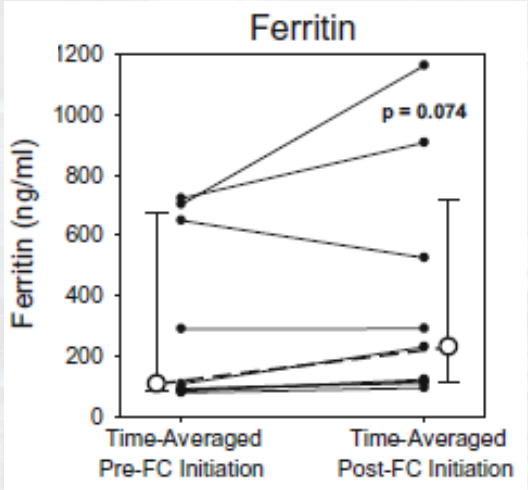
Mark R. Hanudel[1] · Marciana Laster[1] · Georgina Ramos[1] · Barbara Gales[1] · Isidro B. Salusky[1]

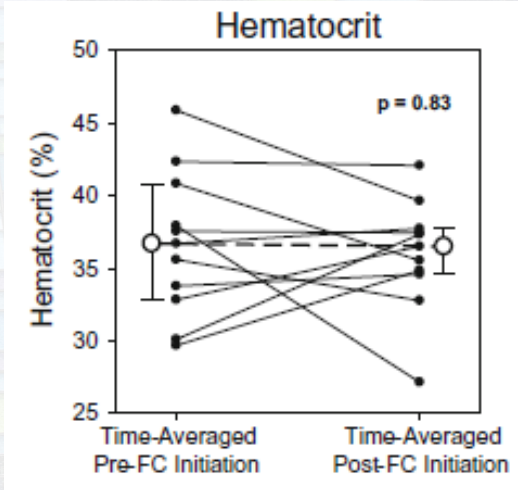
Pediatr Nephrol (2018) 33:2137–2142

- Retrospective analysis of 11 children 4-17 years of age on dialysis (HD and PD) who received ferric citrate as a phosphate binder 2015-2017 (off-label) for median treatment time 214 days



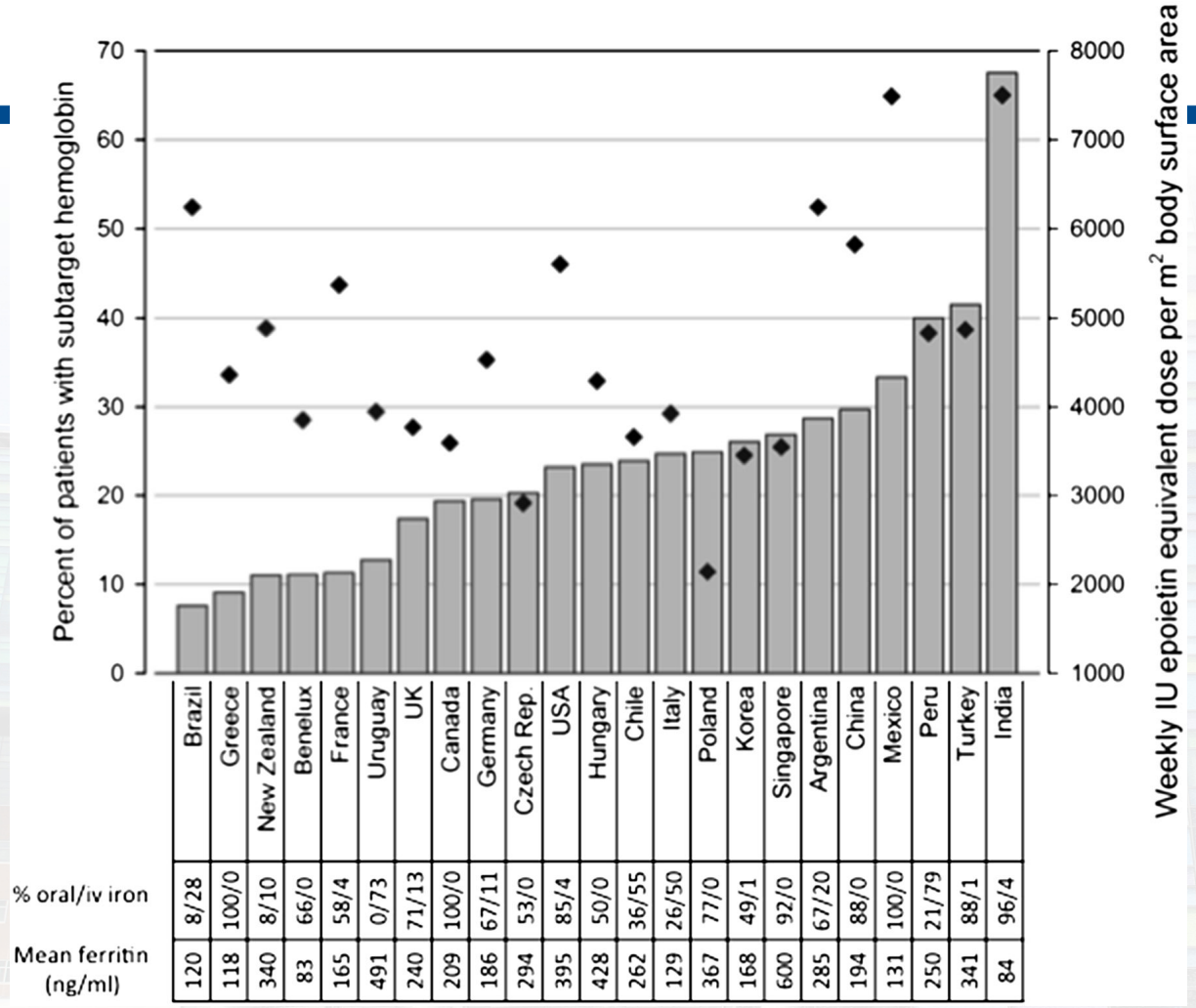






ESA Hyporesponsiveness

Regional variation of anemia control.



Dagmara Borzych-Duzalka et al. JASN 2013;24:665-676

ESA Hypo-Responsiveness

Initial ESA hyporesponsiveness

3.13.1: Classify patients as having ESA hyporesponsiveness if they have no increase in Hb concentration from baseline after the first month of ESA treatment on appropriate weight-base

3.13.2: In patients with ESA suggest avoiding repeat beyond double the initi

Subsequent ESA hyporesponsiveness

3.14.1: Classify patients as having acquired ESA hyporesponsiveness if after treatment with stable doses of ESA, they require 2 increases in ESA doses up to 50% beyond the dose at which they had been stable in an effort to maintain a stable Hb concentration. (*Not Graded*)

3.14.2: In patients with acquired ESA hyporesponsiveness, we suggest avoiding repeated escalations in ESA dose beyond double the dose at which they had been stable. (*2D*)

JOHNS HOPKINS CHILDREN'S CENTER

KDIGO Clinical Practice Guideline for Anemia in Chronic Kidney Disease. *Kidney International* 2(4): August 2012

ESA Hypo-Responsiveness

Some dialysis patients may have low Hgb levels due to dilution in red cell mass in volume overload rather than to an impaired response to ESA

Careful attention to volume status and “challenging” dry weight with increased ultrafiltration can clarify the contribution of volume overload to low Hgb concentration

KDIGO Clinical Practice Guideline for Anemia in Chronic Kidney Disease. *Kidney International* 2(4): August 2012

Pure Red Cell Aplasia

- Sudden onset of severe, transfusion-dependent anemia after at least 8 weeks of therapy
- Rare: 0.5 cases/10,000 pt years
- Neutralizing antibodies to ESA and endogenous EPO
- Rare with IV administration
- Treatment: stop ESA

Transfusion

- Balance risks and benefits to patient
- HLA sensitization

Acute clinical situations

- Acute severe hemorrhage
- Unstable coronary artery disease
- When rapid preoperative Hb correction is required

Chronic clinical situations

- Chronic anemia and ESAs are ineffective (hemoglobinopathies, bone marrow failure, ESA resistance)

Transfuse

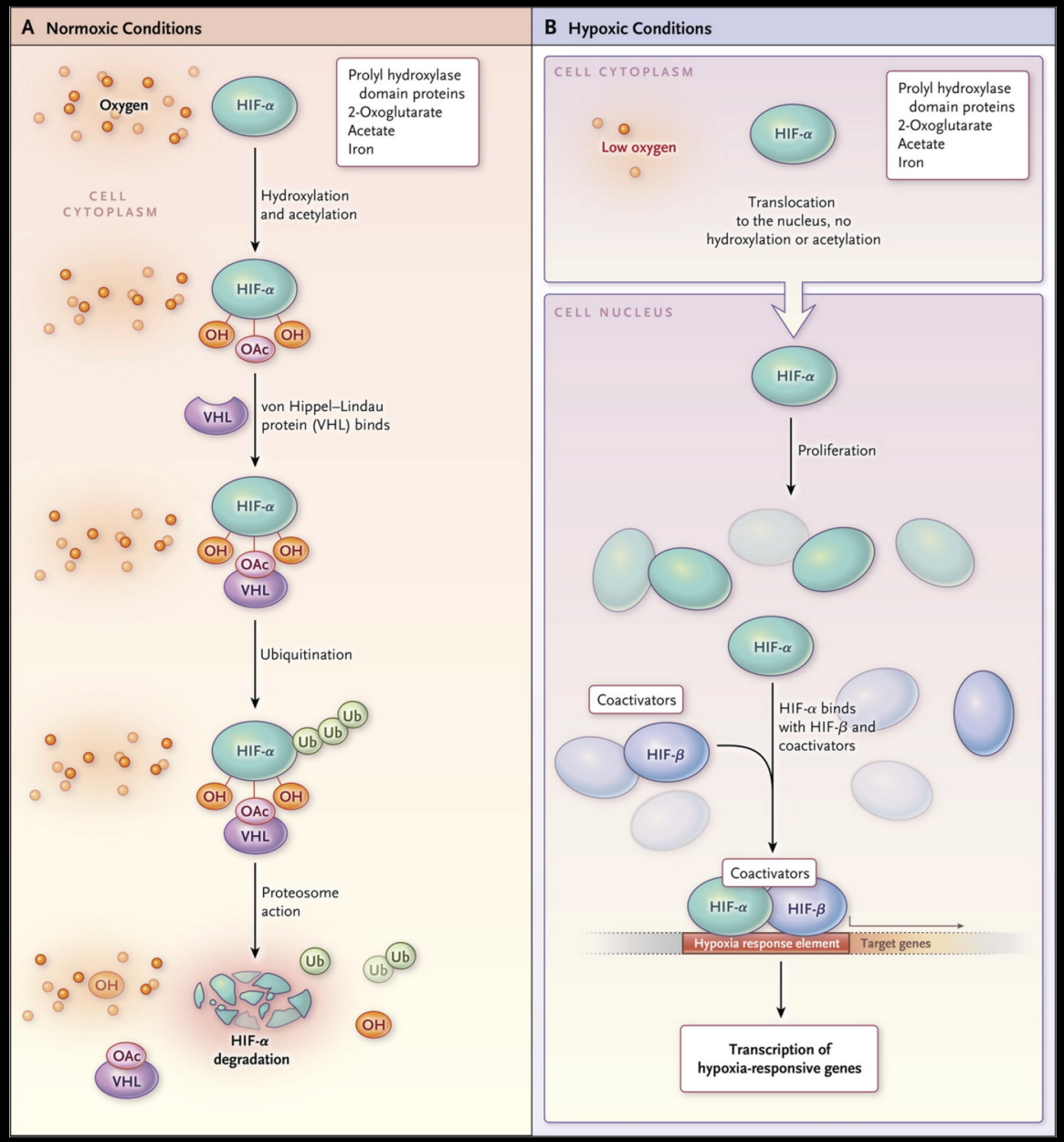
JOHNS HOPKINS CHILDREN'S CENTER

KDIGO Clinical Practice Guideline for Anemia in Chronic Kidney Disease. *Kidney International* 2(4): August 2012

rHuEPO- and Iron-Independent Anemia Therapy?

- Small-molecule hypoxia-inducible factor (HIF) stabilizers/prolyl hydroxylase inhibitors

Figure 2



Normoxia: HIF-α is rapidly (1/2-life 5 min) hydroxylated and degraded

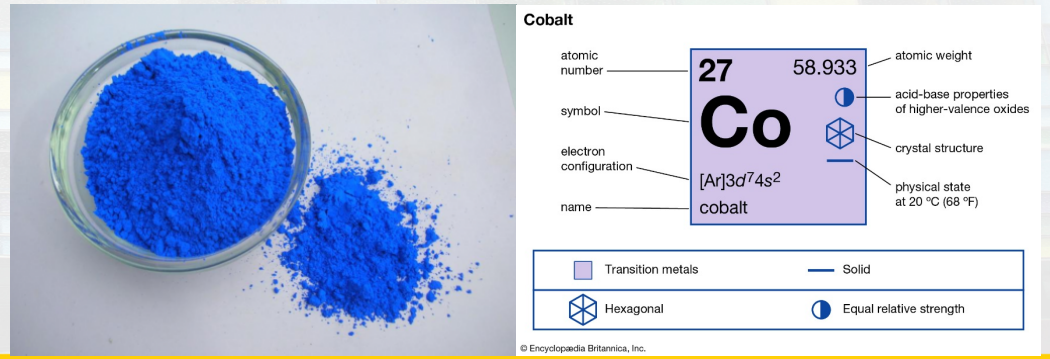
Hypoxia: HIF-α translocates to nucleus, binds HIF-β, activates hypoxia response element, EPO transcribed

American Journal of Kidney Diseases 2018 71, 423-435DOI: (10.1053/j.ajkd.2017.09.026)

rHuEPO- and Iron-Independent Anemia Therapy?

- Small-molecule hypoxia-inducible factor (HIF) stabilizers/prolyl hydroxylase inhibitors
- Administered orally in highly bioavailable preparations
  - Stabilize HIF and modulate HIF-controlled gene products
  - Stimulate endogenous EPO synthesis even in the setting of decreased renal oxygen consumption
  - Decrease hepcidin in adult trials

HIF stabilizer?



Seattle Children's
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Nutrition Management of Children on Dialysis

Kirsten Thompson, MPH, RDN
ADC Kansas City, February 7-11
Seattle Children's Hospital

No Disclosures

Learning Objectives

- Review factors that affect growth
- Review nutritional goals and recommendations for infants and children on dialysis
- Review aspects of diet requiring modification, and management strategies
- Describe approaches to achieve optimal nutritional status in children on dialysis

Focus of Nutrition Care

Overarching Goals:

- Achieve a normal pattern of growth and body composition through maintenance of optimal nutritional status
  - Avoidance/Correction of uremic toxicity, metabolic abnormalities, and malnutrition
- Reduce risk of chronic morbidities and mortality in adulthood

NKF, Am J Kidney Dis. 2009

Barriers to Optimizing Growth and Nutritional Status

- Malnutrition
- Metabolic acidosis
- Anemia
- Fluid and Electrolyte Imbalance
- Long-term use of corticosteroids
- Alterations in bone metabolism

Malnutrition

Inadequate Intake and Nutrient Deficiencies

- Anorexia and poor appetite
  - Uremia, acidosis and anemia can cause taste alterations
- Oral food aversions and dislike of solid foods
- GI Disturbances
  - GERD, nausea/emesis, constipation, diarrhea
  - Delayed gastric emptying
  - Increased IP pressure during PD
  - Food Allergies/Intolerances
- Fluid and dietary restrictions limiting food availability and variety
- Vitamin, mineral and protein losses through dialysis
- Cultural influences
- Psychosocial issues
  - Depression, financial instability, food insecurity, stressful living situation

Phases of Growth

Phase	Fetal	Infant	Child	Pubertal
From	Conception to Birth	Birth to 18 months	18 months to 12 years	Onset of Puberty
% of total growth	30	15	40	15
Dependent on	Nutrition Placenta	Nutrition Good health	Growth hormone Thyroid hormone Good health	Growth hormone Testosterone /Estrogen Good Health

Growth Pattern and Dietary Intake of Children with CKD

>80% DRI –
- **Normal** growth

<80% DRI –
- **Reduced** growth velocity

<40% DRI –
- **Cessation** of growth

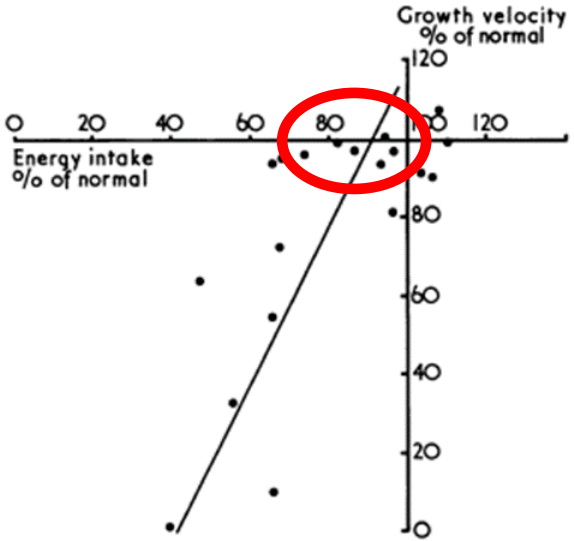


FIG. 3—Relation between growth velocity, expressed as percentage of expected 50th centile velocity, and energy intake, expressed as percentage of that recommended for same age. ($r=0.72$; $P<0.001$.)

Betts & Magrath, BMJ 1974

Growth Assessment

- Estimated dry weight and weight for age %ile or SDS (std deviation score)
  - Consider fluid status
- Recumbent Length (<2 years) or standing height-for-age (>2 years) %ile or SDS
  - Calculate mid-parental height to evaluate growth potential
- Head circumference-for-age %ile or SDS (up to 36 months)
- Weight-for-length (<2 years) or BMI (>2 years) for height age %ile (age at which height is at 50%ile)
  - <5%ile classified as underweight
  - BMI <5%ile and >95%ile for age associated with increased morbidity and mortality
- Length/height and weight velocity for age percentile

Growth Assessment Frequency

- Growth assessment should be performed twice as often as that of a healthy child of the same age

Table 1. Recommended Parameters and Frequency of Nutritional Assessment for Children with CKD Stages 2 to 5 and 5D

	Minimum Interval (mo)									
	Age 0 to <1 y			Age 1-3 y			Age >3 y			
Measure	CKD 2-3	CKD 4-5	CKD 5D	CKD 2-3	CKD 4-5	CKD 5D	CKD 2	CKD 3	CKD 4-5	CKD 5D
Dietary intake	0.5-3	0.5-3	0.5-2	1-3	1-3	1-3	6-12	6	3-4	3-4
Height or length-for-age percentile or SDS	0.5-1.5	0.5-1.5	0.5-1	1-3	1-2	1	3-6	3-6	1-3	1-3
Height or length velocity-for-age percentile or SDS	0.5-2	0.5-2	0.5-1	1-6	1-3	1-2	6	6	6	6
Estimated dry weight and weight-for-age percentile or SDS	0.5-1.5	0.5-1.5	0.25-1	1-3	1-2	0.5-1	3-6	3-6	1-3	1-3
BMI-for-height-age percentile or SDS	0.5-1.5	0.5-1.5	0.5-1	1-3	1-2	1	3-6	3-6	1-3	1-3
Head circumference-for-age percentile or SDS	0.5-1.5	0.5-1.5	0.5-1	1-3	1-2	1-2	N/A	N/A	N/A	N/A
nPCR	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	1*

Abbreviation: N/A, not applicable.
*Only applies to adolescents receiving HD.

American Journal of Kidney Diseases, Vol 53, No 3, Suppl 2 (March), 2009: pp S16-S26

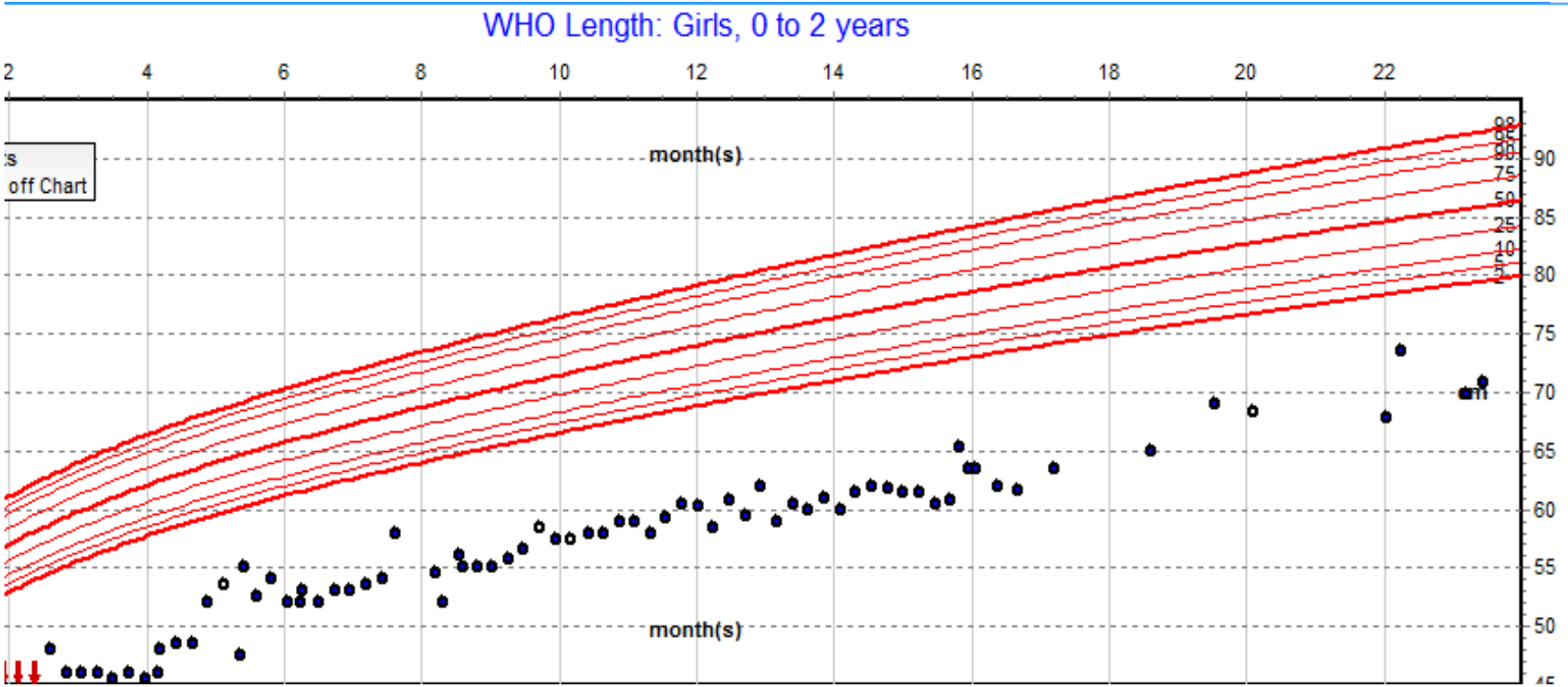
Plotting Growth

Infants (WHO growth charts 0-24 mo)	Pediatrics/Adolescents (CDC growth charts 2-18 years)
Weight-for-age	Weight-for-age
Length-for-age	Height-for-age
Weight-for-Length	BMI-for-age
Head Circumference-for-age	

****Prematurity**

- Use Fenton Premature growth charts up to 50 weeks
- Plot weight and length for corrected age up to 36 months

Example


WHO Length: Girls, 0 to 2 years
2
4
6
8
10
12
14
16
18
20
22
month(s)
off Chart
98
95
90
75
50
25
10
5
2
90
85
80
75
70
65
60
55
50
month(s)

Nutrition Assessment

Other Tools:

- Mid Upper Arm Circumference (MUAC) – validated as marker of nutrition status in general pediatric population
- Waist-to-Ht Ratio (>0.49)
- Subjective Global (Nutrition) Assessment (SGA/SGNA)
- Nutrition Focused Physical Exam (NFPE)
- Bio-electric Impedance Analysis (BIA)
- Bioimpedance Spectroscopy (BIS)

Mastrangelo, Ped Neph 2013; Eng, NDT 2017; Addo, AJCN 2016; Modi, J Nutr 2015;Secker, AJCN 2007; Steiber, JRN 2007; Steiber, JRN 2004; Steiber, JRN 2004; Secker, JAND 2012; Secker, JRN 2011; Corkins, NCP 2015; Corkins, NCP 2016; Esper, NCP 2015

Obesity

Obesity, Dyslipidemia

- Increasing worldwide, including children with CKD
- International Pediatric PD Network (IPPN)
  - 19.7% prevalence of overweight/obesity in children at initiation of PD

- CKiD Data
  - Median energy/protein consumption exceeded recommendations in all age groups
  - 13% met activity goal
  - 98% exceed recommended screen time

NKF, Am J Kid Dis. 2009; Hui, Ped Neph 2017;Weaver, Semin Neph 2018; Schaar, Ped Neph 2020, Clark, Ped Neph 2015

Treating Growth Failure

- Metabolic abnormalities should be corrected and nutrition optimized prior to starting growth hormone
- Should be initiated pre-transplantation and pre-puberty
- KDOQI- guidelines for initiating growth hormone
- Varies by institution

NKF, Am J Kidney Dis. 2009

Estimating energy requirements

- 100% of the EER for chronological age at healthy weight
- Adjusted for PAL (physical activity factor) and body size
- Further adjustment based on rate of weight gain or loss
- Malnourished children typically have higher energy requirements to support “catch-up” growth
- Children on PD-
  - Dextrose absorbed from dialysate may need to be considered for infants and children who are gaining weight more quickly than expected.
- Special equations for children who are overweight/obese

NKF, Am J Kidney Dis. 2009- KDOQI

Estimating energy requirements

Table 2. Equations to Estimate Energy Requirements for Children at Healthy Weights

Age	Estimated Energy Requirement (EER) (kcal/d) = Total Energy Expenditure + Energy Deposition
0-3 mo	EER = [89 × weight (kg) − 100] + 175
4-6 mo	EER = [89 × weight (kg) − 100] + 56
7-12 mo	EER = [89 × weight (kg) − 100] + 22
13-35 mo	EER = [89 × weight (kg) − 100] + 20
3-8 y	Boys: EER = 88.5 − 61.9 × age (y) + PA × [26.7 × weight (kg) + 903 × height (m)] + 20
	Girls: EER = 135.3 − 30.8 × age (y) + PA × [10 × weight (kg) + 934 × height (m)] + 20
9-18 y	Boys: EER = 88.5 − 61.9 × age (y) + PA × [26.7 × weight (kg) + 903 × height (m)] + 25
	Girls: EER = 135.3 − 30.8 × age (y) + PA × [10 × weight (kg) + 934 × height (m)] + 25

Source: ref 175.

NKF, Am J Kidney Dis. 2009

Recommended Dietary Protein Intake

Table 12. Recommended Dietary Protein Intake in Children with CKD Stages 3 to 5 and 5D

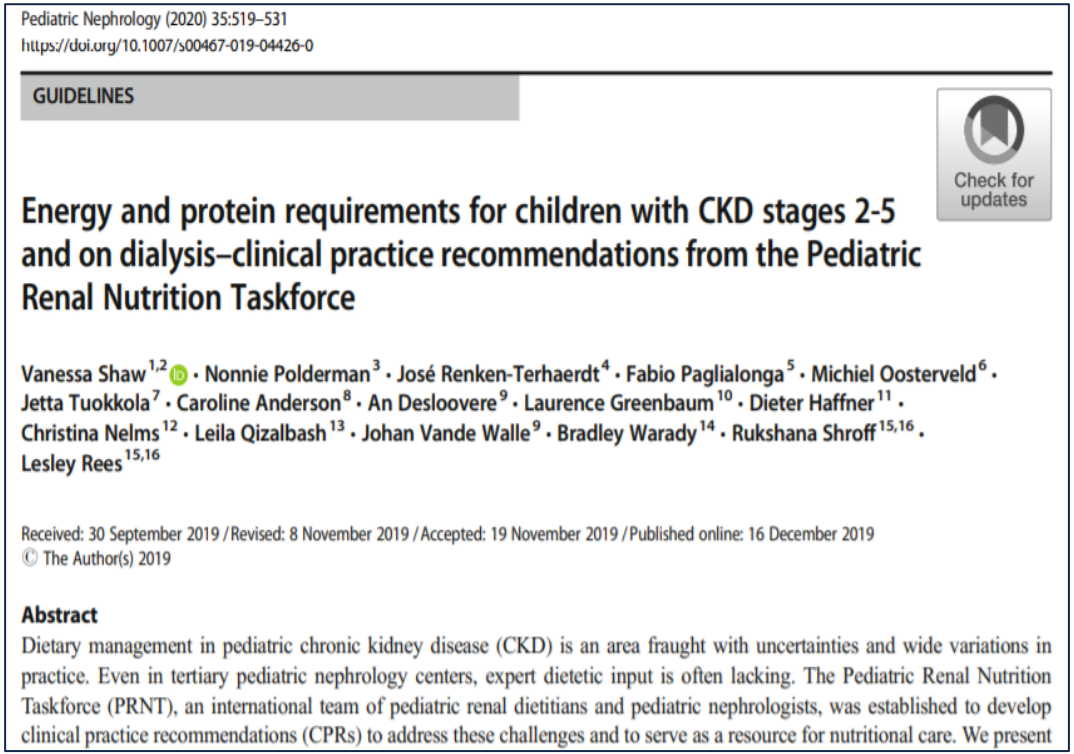
	DRI				
Age	DRI (g/kg/d)	Recommended for CKD Stage 3 (g/kg/d) (100%-140% DRI)	Recommended for CKD Stages 4-5 (g/kg/d) (100%-120% DRI)	Recommended for HD (g/kg/d)*	Recommended for PD (g/kg/d)†
0-6 mo	1.5	1.5-2.1	1.5-1.8	1.6	1.8
7-12 mo	1.2	1.2-1.7	1.2-1.5	1.3	1.5
1-3 y	1.05	1.05-1.5	1.05-1.25	1.15	1.3
4-13 y	0.95	0.95-1.35	0.95-1.15	1.05	1.1
14-18 y	0.85	0.85-1.2	0.85-1.05	0.95	1.0

*DRI + 0.1 g/kg/d to compensate for dialytic losses.
†DRI + 0.15-0.3 g/kg/d depending on patient age to compensate for peritoneal losses.

NKF, Am J Kidney Dis. 2009- KDOQI

Pediatric Renal Nutrition Taskforce (PRNT)

- International team of pediatric RDs and nephrologists
- Established to develop Clinical Practice Recommendations (CPRs) for energy and protein requirements for children with CKD 2-5 and 5D



Pediatric Nephrology (2020) 35:519–531
https://doi.org/10.1007/s00467-019-04426-0

GUIDELINES

Check for updates

Energy and protein requirements for children with CKD stages 2-5 and on dialysis–clinical practice recommendations from the Pediatric Renal Nutrition Taskforce

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Abstract
Dietary management in pediatric chronic kidney disease (CKD) is an area fraught with uncertainties and wide variations in practice. Even in tertiary pediatric nephrology centers, expert dietetic input is often lacking. The Pediatric Renal Nutrition Taskforce (PRNT), an international team of pediatric renal dietitians and pediatric nephrologists, was established to develop clinical practice recommendations (CPRs) to address these challenges and to serve as a resource for nutritional care. We present

CLINICAL PRACTICE RECOMMENDATIONS (CPRs)

Table 3 Summary of recommendations

	Category	Recommendation
1	Energy requirements	1.1 We suggest that the initial prescription for energy intake in children with CKD2–5D should approximate that of healthy children of the same chronological age. 1.2 To promote optimal growth in those with suboptimal weight gain and linear growth, we suggest that energy intake should be adjusted towards the higher end of the suggested dietary intake (SDI). 1.3 In overweight or obese children, adjust energy intake to achieve appropriate weight gain, without compromising nutrition.
2	Protein requirements	2.1 We suggest that the target protein intake in children with CKD2–5D is at the upper end of the SDI to promote optimal growth. The protein intake at the lowest end of the range is considered the minimum safe amount and protein intake should not be reduced below this level. 2.2 We suggest that the protein intake in children on dialysis may need to be higher than the SDI for non-dialysis patients to account for dialysate protein losses. 2.3 In children with persistently high blood urea levels, we suggest that protein intake may be adjusted towards the lower end of the SDI, after excluding other causes of high blood urea levels.

SDI for energy and protein: birth[a] to 18 years

Month	SDI[b] energy (kcal/kg/day)	SDI protein (g/kg/day)	SDI protein (g/day)	
0	93–107	1.52–2.5	8–12	
1	93–120	1.52–1.8	8–12	
2	93–120	1.4–1.52	8–12	
3	82–98	1.4–1.52	8–12	
4	82–98	1.3–1.52	9–13	
5	72–82	1.3–1.52	9–13	
6–9	72–82	1.1–1.3	9–14	
10–11	72–82	1.1–1.3	9–15	
12	72–120	0.9–1.14	11–14	
Year	SDI energy (kcal/kg/day)		SDI protein (g/kg/day)	SDI protein (g/day)
–	Male	Female		
2	81–95[c]	79–92[c]	0.9–1.05	11–15
3	80–82	76–77	0.9–1.05	13–15
4–6	67–93	64–90	0.85–0.95	16–22
7–8	60–77	56–75	0.9–0.95	19–28
9–10	55–69	49–63	0.9–0.95	26–40
11–12	48–63	43–57	0.9–0.95	34–42
13–14	44–63	39–50	0.8–0.9	34–50
15–17	40–55	36–46	0.8–0.9	Male: 52–65 Female: 45–49

SDI: *Suggested Dietary Intake* – based on the range from various international bodies research recommendations

Monitoring Protein Status

- Albumin and pre-albumin
  - Acute phase proteins
    - Suppressed in the setting of inflammation and edema
  - Low levels associated with increased mortality and morbidity
  - Not good markers of malnutrition and nutritional status
- Assess BUN: Creatinine
- Nitrogen balance studies
- nPCR = normalized protein catabolic rate (adolescents on dialysis)

Evaluating Dietary Intake

- Methods of assessing intake
  - 24-hour food recall
  - 3 day food record
  - Food frequency questionnaire
  - iPhone apps

- Early identification of food preferences, allergies and intolerances
  - Important to create an individualized meal plan

Feeding in Infants and Toddlers

- Breastfeeding/Expressed Breast Milk preferred method for feeding
- Whey dominant infant formulas recommended if EBM not available
  - Low electrolyte and mineral formulas if K/phos restrictions needed
  - Fortify with formula powder or modular products to meet nutrition goals if fluid restrictions indicated
- Healthy infants show readiness for solids at 4-6 months
  - Frequently show delayed progression through normal stages of eating
  - Encourage families to follow the same eating and development timeline as that of a healthy child
    - Age appropriate introduction of solids
    - Minimize dietary restrictions if feasible and identify favorite foods

Feeding in school-age children

- Typically eat independently
- Continue oral stimulation and involve feeding therapy
  - Even if oral intake limited
- Consider school experience
- Include the child in discussions between caregivers and medical staff related to diet, nutrition, growth and medications

Adolescents

- Irregular eating patterns and meal skipping compromise patient's ability to meet nutritional needs
- Should be directly involved in meal planning and diet education
- Nutrition education should focus on cafeteria food, processed foods, fast foods, snacks and alternative drinks, high in sodium and phosphorus additives

Nutrition Support

- Most children on dialysis (especially infants and toddlers) require supplemental to full nutrition support to meet requirements
  - Start with oral supplements if possible
  - Formula regimen guided by age, CKD stage, electrolyte and mineral imbalances, fluid allowance, food allergies/intolerance, GI symptoms
  - Blended Tube Feeding (BTF) may be better tolerated in patients with GI disturbances

Nutrition Support Study

IPPN (International Pediatric PD Network)

2007-2009

- Analyzed growth in 150 patients on PD <2 yrs of age
  - 32 % NG
  - 25% PEG
  - 22% oral supplements
  - 21% no supplemental feeding

Results:

- PEG /NG had significantly higher Ht and BMI SDS
- Ht velocity was greater in the enterally-fed infants

Conclusion:

- Early institution of enteral feeding improves longitudinal growth in infants receiving chronic PD

Rees et al, JASN 2011

Comparison of Common Formulas

100 kcal Source (std kcal/oz)	ml	Prot (g)	Na (mg)	K (mg)	Ca(mg)	Phos (mg)
Cow's Milk	159	5.1	79	248	200	148
Human Milk (20)	142	1.5	25	75	46	20
Similac PM 60/40 (20)	147	2.2	23	80	56	28
Nephea Kids (37)	82	1.3	41	4	31	5
Kindergen (30)	100	1.5	46	24	22	19
Renastart (30)	100	1.6	49	22	21	19
Suplena (54)	56	2.5	44	63	59	40
Nepro (54)	56	4.5	59	59	59	40
Renalcal (60)	50	1.7	3	4	3	5

Modular Energy and Protein

Product	Nutrient	Form
Duocal	Carbohydrate/Fat	Powder
Solcarb	Carbohydrate	Powder
Complete amino acids	Protein	Powder
Microlipid	Fat	Emulsified Oil
MCT Oil	MCT Oil	Liquid
Liquid Protein	Protein	Liquid
Beneprotein	Protein	Powder

Blended Tube Feedings

- Food and beverage combined and blended to a consistency that allows it to flow through a feeding tube

Benefits

- Improvement in GI symptoms and bowel function (emesis, reflux, diarrhea)
- Can tailor the recipe to each patient's needs
- Parents love being able to give their kids real food

Challenges

- More time consuming
- Diet and fluid restrictions can make it difficult to meet nutrient requirements

Can be more expensive

Increased complications with feeding delivery

Intradialytic Nutrition Therapies

Intradialytic Parenteral Nutrition (+PO)

- Amino acids, Dextrose, Lipids
- ↑ Wt, BMI, %IBW
- ↑ oral caloric intake
- Costly
- Adverse events
  - Hyperglycemia
  - Lipid intolerance
  - Hypophosphatemia

Intradialytic Lipid Infusion (+PO)

- Lipids only
- ↑ albumin, pre-HD BUN, nPCR, cholesterol
- ↑ Wt velocity and SDS,↑ BMI
- Less costly
- No adverse events reported

Krause, JRN 2002; Goldstein, Ped Neph 2002; Orellana, JRN 2005; Haskin, JRN 2017

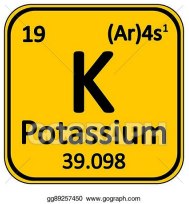
“Renal” Diet

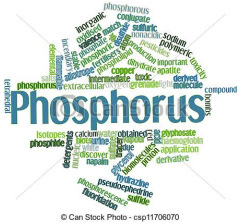
Modifications to:

- Phosphorus, sodium, potassium and fluid

There is no “one size fits all”

- Be as liberal as possible to start
- Implement restrictions as indicated
- Individualize for:
  - Age
  - Stage of development
  - Food preferences
  - Biochemistry
- More liberal if residual renal function, on PD or daily HD





“Renal” Diet

What **are** children with CKD actually eating?

CKiD Data

- Consuming more energy, protein, sodium and phosphorus than recommended
- Milk largest contributor to kcal, protein, phosphorus, and potassium
- Fast foods major contributors to fat, sodium, energy, and phosphorus

Important to educate patients and families about healthier food choices early to establish healthy eating habits later in life!

Hui, Ped Neph 2017; Chen, Ped Neph 2017

Sodium and Fluids

Polyuria & Na+ Wasting

- Obstructive uropathies
- Renal dysplasia
- Na+ depletion in infants on PD

Goals:

- Adequate hydration
- Na+ supplementation

✓ Promote muscle development, bone mineralization

✓ Prevent growth retardation

Na+ & Fluid Retention

- Primary glomerular disease
- Oliguric or anuric

Goals:

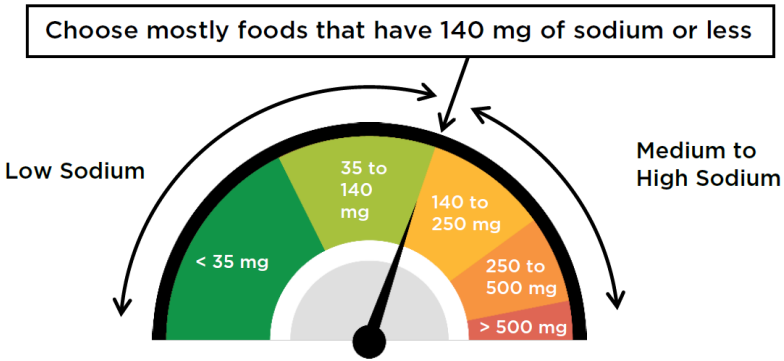
- Fluid restriction
- Na+ restriction

✓ Prevent volume overload, HTN

✓ Decrease risk of CVD and LVH

Managing Sodium

- Processed → Fresh
- Salt → Herbs and no Na+ spices
- Restaurants → Home prepared
- Limit Na+ to 1500-2000 mg daily (based on age)
- Read food labels



Managing Fluid

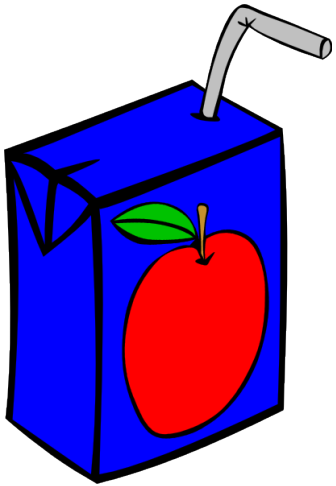
- Limit Na+ intake
- Small amounts divided through day
- Count obvious and hidden fluids
  - Fruits and vegetables
  - Popsicles
  - Soups
- Freshen mouth without drinking fluids

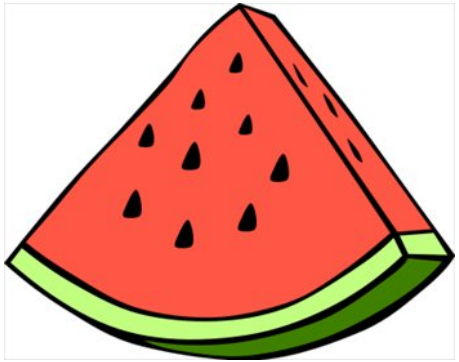
Potassium

- Reduce potassium intake from food
  - Consider potassium additives (potassium citrate)
- Use of adult renal formulas or modulars in combination with infant formulas
- Pre-treat and decant formula or EBM with sodium polystyrene sulfonate to partially remove potassium
  - Safer alternative to oral/rectal administration
  - Electrolyte derangements
  - Caution when using in patients needing sodium restriction
- Increased losses with PD

Low K+ (5-150mg/serving)	Serving Size	Medium K+ (150-250mg/serving)	Serving Size	High K+ (250-500+mg/serving)	Serving Size
2-3 servings/day		**1-2 servings/day**		**1 serving/day**	
FRUITS AND VEGETABLES					
Berries (Blue, black, raspberries, strawberries)	½ cup	Apple, Pear	1 medium	Banana , orange, nectarine, kiwi	1 medium
Pears canned, Pineapple, Applesauce	½ cup	Cherries	8-10	Tomato	1 medium
		Grapes	10-15	Tomato Paste	1/8 cup
Beans (green/wax), Corn	½ cup	Mango	½ medium	Potato	½ cup or 1 small
Cauliflower, Peas, Cucumber	½ cup	Watermelon	1 cup	Squash, yams, sweet potatoes	½ cup
Lettuce	1 cup	Broccoli, Brussel Sprouts, Carrots	½ cup	Avocado	¼ medium
				Salt substitute 1/4 tsp	¼ tsp
BEVERAGES					
Apple juice	½ cup	Grapefruit Juice, Grape juice (canned)	½ cup	Milk	1 cup
Grape juice (frozen), Cranberry juice	1 cup	Pineapple juice Apricot nectar	½ cup	OJ, Prune juice, Tomato Juice, V-8	½ cup
Crystal Light, Capri Sun, Kool Aid, Lemonade, Iced Tea	1 cup	Soy milk	½ cup	Instant Breakfast type drinks	1 cup

Potassium





Think portion size!

Think cumulative!

Phosphorus

- Early nutrition intervention key to addressing CKD-MBD and consequences
  - Cardiovascular disease
  - Poor transplant outcomes
  - Bone damage post transplant

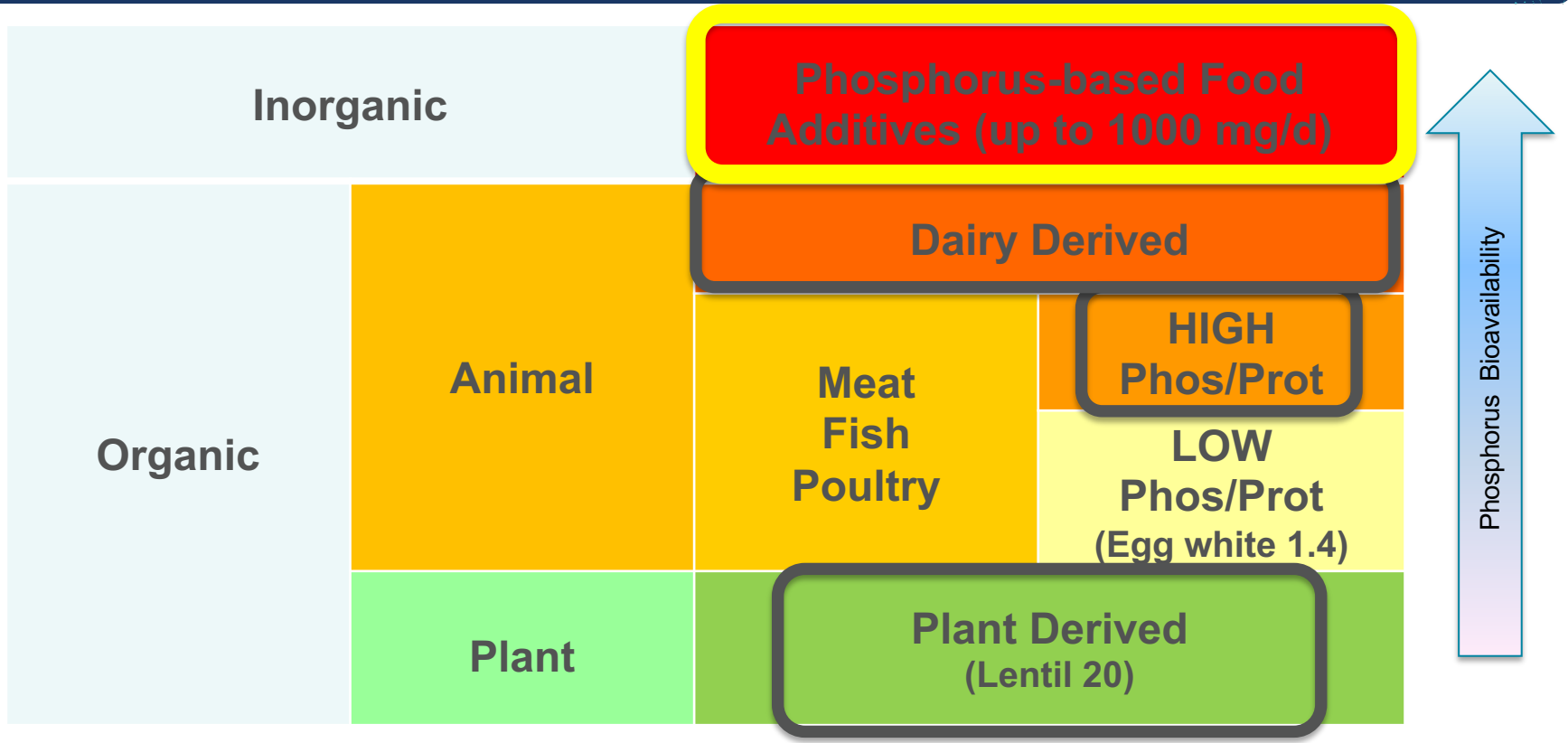
KDIGO CKD-MBD Update Work Group, Kidney Int 2017; Wesseling-Perry CJASN 2012; Wesseling-Perry NDT 2011

Phosphorus Management

Recommended Phosphorus Intake mg/d			
Age (y)	DRI (mg/d)	High PTH Normal Phos	High PTH High Phos
0-6 mo	100	<100	<80
7-12 mo	275	<275	<225
1-3	460	<460	<370
4-8	500	<500	<400
9-18	1250	<1250	<1000

NKF, Am J Kidney Dis. 2009

Sources of Phosphorus



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Adapted from Adema et al, JRN 2014; Carrigan JRN 2014; Uribarri, Semin Dial 2003

Phosphorus Management

Decrease Phosphorus Intake

Offer low phosphorus formulas. Delay introduction of cow's milk.	Limit intake of dairy Offer low phosphorus proteins	Limit or avoid sources of inorganic phosphorus (food additives)

Adjust phosphorus binder dose and timing to meals, snacks, tube feeds

Phosphorus Management

- Pre-treat and decant formula with Sevelamer (Similar to Kayexalate)
  - Can be useful in patients on continuous feeds or volume restrictions
  - Time intensive
  - Requires a fairly large dose of sevelamer for efficacy
  - Alters nutrient profile

Calcium

- Important role in bone health in children
- Adequate is necessary, excess should be avoided
  - KDOQI: Goal intake 100% DRI for age – max 200%
  - Consider Calcium burden from diet, formulas, medications

Age	DRI (mg/d)	Upper Limit (Diet + Binders)
0-6 months	200	<420
7-12 months	260	<540
1-3 years	700	<1000
4-8 years	1000	<1600
9-18 years	1300	<2500

NKF, Am J Kidney Dis. 2009; KDIGO CKD-MBD Update Work Group, Kidney Int 2017; Goodman, NEJM 2000

Calcium

Phosphorus Binder	Elemental Ca (% of total)	Elemental Ca (mg/dose)	Phosphorus Bound mg (mg per 100 mg Ca2+ delivered)
Calcium Acetate (667 mg)	25	167	45 (27 mg P/100 mg Ca2+)
Calcium Carbonate (1250 mg)	40	500	39 (8 mg P/100 mg Ca2+)

Vitamins

- Requirements = 100% DRI
- Adult renal formulas provide 100% of requirements without supplement
- Diet + Supplement < tolerable upper intake level (UL)
- Increased risk of deficiency
  - Intake limited by anorexia
  - Diet restrictions
  - Losses via dialysis
  - Interference with absorption, excretion, metabolism
- Children with CKD stage 5D should receive a water-soluble vitamin supplement

NKF, Am J Kidney Dis. 2009;

Vitamin D

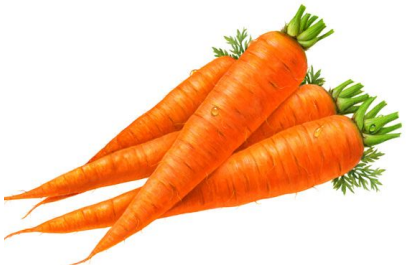
- High prevalence of Vit D insufficiency in children with CKD

- Current KDOQI Guidelines:
  - Measure 25-hydroxy vitamin D at least annually
  - If <30 ng/ml (75 mmol/L) supplement with D2 or D3
  - In the repletion phase, check PO4 and Ca levels after 1 month
  - When replete, supplement vitamin D continuously and monitor yearly

NKF, Am J Kidney Dis. 2009

Vitamin A

- Not removed by dialysis
- Rapidly absorbed and slowly cleared
- RBP catabolized in the renal tubules
- Vitamin A/RBP accumulation common and increases with stage of CKD
  - Elevated in majority of CKD and dialysis patients
- Sources
  - Preformed vitamin A (retinol) absorption rates =70-90%
    - Supplements/fortification, fish liver oil, liver, egg
  - Provitamin A (beta-carotene) absorption rates = 20-50%
    - Carotenoids from plant sources
    - Cannot cause toxicity
  - Vitamin A supplements contraindicated



Vitamin A

Signs/symptoms and complications of toxicity:

- Headaches, dry itchy skin, anorexia, bone pain, nausea/emesis
- Hypercalcemia
- Effects on bone
  - Increased hip fractures
- Intracranial hypertension
- Bulging fontanelle
- Pseudotumor cerebri
- Hepatomegaly

Vitamin A

- Intervention
  - Limit intake to DRI for age
  - Use modulars in formula to lower vitamin A administration
  - Promote intake of “real” food when appropriate
    - Formula and supplements contain high amounts of retinol
  - Use unfortified milks and foods

Other Vitamins

Vitamin E

- Commonly elevated in children on dialysis
- Insufficient evidence to recommend supplementation

Vitamin K

- Depleted with Antibiotic use
- Monitor for signs of deficiency

Water soluble vitamins

- No concrete guidelines/recommendations for specific monitoring
- Some institutions test periodically
- Recommend testing if signs/symptoms of deficiency or poor oral intake

NKF, Am J Kidney Dis. 2009; Fassinger, JRN 2010; Manichkavasagar, Ped Neph 2015; Joyce, Ped Neph 2018;

Minerals

MAGNESIUM

- Elevated levels found in dialysis patients

ZINC

- Commonly low in children on dialysis
- Varying response to supplementation
  - KDOQI- regular monitoring in patients on low protein diet or with poor intake
- Monitor signs/symptoms of deficiency/toxicity

COPPER

- High and low levels found
- No clear recommendations for supplementation
  - Measure levels if deficiency or toxicity is suspected

SELENIUM

- Commonly low in children on dialysis
- Monitor signs/symptoms of deficiency

Ponton-Vazquez, JRN 2017; Zwolinska, Ped Neph 2006; Filler, Ped Neph 2013; Tonelli, BMC Med 2009; Esmaeili, JRN 2019; Mekahli, CJASN 2009;

Education and Counseling

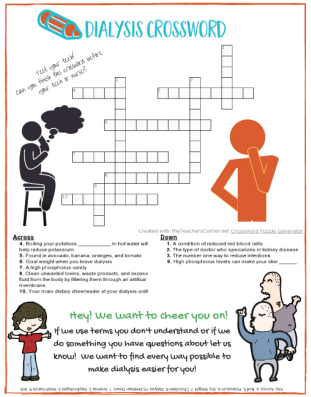
- Education with first intervention
- Frequent re-education
  - Be positive – focus on allowances
  - Incorporate personal preferences
  - Provide pleasure with food
- Role of cognitive function and developmental stage
- Health Literacy – child and caregivers

NKF, Am J Kidney Dis. 2009; Lum, Child, Care Health Dev 2017; Ellyn Satter Institute; Beto, Int J Neph and Renovasc Dis, 2016; Morris, J Ren Care 2015; Chen, CJASN 2018;

Education and Counseling

Learning styles

- Motivational Interviewing
  - Patient-centered goals
- Teach Back Method
- Creative Strategies
  - Technology, game-based learning
    - Apps, videos, games
  - Incentive programs
  - Multidisciplinary approach



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Swap This, for That: *Starbucks*

Swap it out

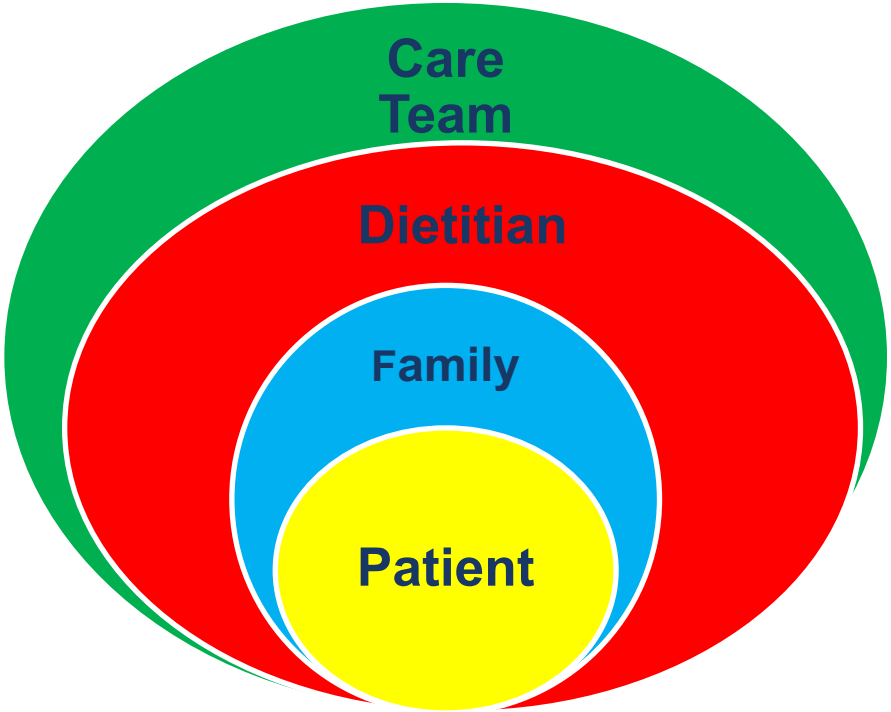
- Cold cream brew
- Nitro flat white
- Iced pumpkin spice latte
- Mocha
- White choc mocha
- Mocha or mocha cookie frap-

- Herbal tea
- Cold brew coffee
- Caffé vanilla or espresso Frap-puccino *with soymilk and no whip*
- Mango dragonfruit or strawberry acai lemonade or very berry hibiscus refresher
- Iced Americano
- Iced pumpkin spice latte *with soymilk and no whip*
- Iced caffé latte *with soymilk*
- Iced cinnamon dolce latte *with soymilk and no whip*
- Iced caramel cloud macchiato *with soymilk (caramel sauce has some dairy)*
- Matcha green tea latter *with soymilk*
- London fog tea latte *with soymilk*

Tips when choosing a drink

- Best choice: drinks without added milk or creamers
- If consume milk-containing coffee drinks, limit milk to 4 ounces
- Swap out cow's milk or cream for soy milk, almond milk or rice milk to help lower potas-sium and phosphorus content
- Some milk substitutes have added phospho-rus or calcium. Check the label/brand
  - Starbucks almond and coconut milk contain some phosphate additives

Nutrition Management of Children on Dialysis



Thank you

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