

FMD/Arterial Dissection Information and Support Group



**University Hospitals
Virtual Meeting
October 18, 2022
6:00-7:30 pm**

 University Hospitals
Harrington Heart & Vascular Institute
Cleveland | Ohio

Agenda

| | |
|------------------------------------|--|
| Welcome, Intros, Polling | Dr. Heather Gornik/All |
| Patient Stories | Multiple |
| Updates from the FMDSA | Pamela Mace, RN, Executive Director, FMDSA |
| Renal manifestations of FMD | Dr. Heather Gornik |
| Kidney Health/Ask the Nephrologist | Dr. Aparna Padiyar |
| Open Discussion/Q&A | All |
| Adjourn | |

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We are recording tonight's session to prepare a written transcript of tonight's content. No video will be used and no names will be included in the transcript.

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Introductions and Check-in, Polls

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Polls

1. Where are you joining us from tonight?
2. If you are joining us from outside the US/Canada, where are you?
3. Have you attended the University Hospitals FMD/dissection information/support group before?
4. What brings you to this group?
5. Are you a member of the FMD Society of America
6. Which of the following is your favorite type of Halloween candy?

Patient Stories

FMDSA Updates

Pamela Mace, RN

Save the Date: FMDSA 2023



FMDSA Annual Conference, May 19-20, 2023

Save the date! Next year's conference will be in-person and we'll be celebrating the FMDSA's 20th year!

We hope you will be able to join us in Cleveland for this special annual conference. Please check back for more information.

Watch the video presentations from our December 2021 Annual Conference

[SEE VIDEO PRESENTATIONS](#)

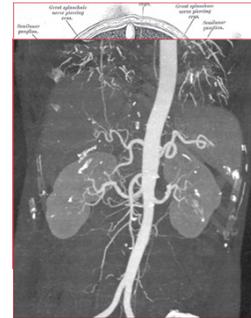


Renal Manifestations of FMD and Review of Terminology

Heather L. Gornik, MD

Renal FMD: Fundamental Anatomy

- Renal arteries arise from the abdominal aorta and supply oxygenated blood flow to the kidneys
- Patients have at least 1 renal artery to each kidney
 - ~1/3 of patients have more than 1 renal "accessory" artery



<https://en-academic.com/dic.nsf/enwiki/713110>

Unusual Anatomy! "horse shoe" kidney



Renal FMD: Imaging

- Imaging studies are used to assess the renal arteries for FMD
 - CT angiography (CTA)
 - MR angiography (MRA)
 - Doppler ultrasound (indirect)
 - Catheter-based angiography (the "gold standard")



Renal FMD

- Renal arteries the most common site of involvement of FMD in the body (or tied with the carotid/vertebral arteries)
- Type of FMD is now described based on how it looks on imaging (not under the microscope)
- Multifocal FMD more common than focal FMD
 - >95% of people on this call have multifocal FMD
 - Focal FMD is a disease of children and young adults
 - 1st described case of FMD was a young boy with the "focal" type of FMD

Multifocal FMD



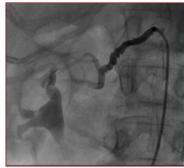
Focal FMD



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Potential Manifestations of Renal Artery FMD

- Incidental finding on imaging study
- Abdominal bruit
- High blood pressure (hypertension)
- Renal artery dissection
- Renal artery infarct
- Renal artery aneurysm
- Headaches, especially migraine type (common among patients with just renal FMD)




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Hypertension and FMD

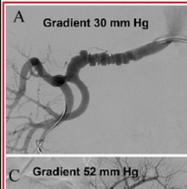
- Hypertension is **VERY** common among patients with FMD (67% in US Registry, 74% in European Registry*)
- Though a patient may have renal FMD and high blood pressure, the high blood pressure is not always caused by the FMD
 - Patients can have "essential" or "primary" hypertension due to genetic factors, weight, other related issues
 - Some patients with renal FMD do have high blood pressure caused by the renal artery FMD ("renovascular hypertension")
 - Important to identify whether the FMD (beads) in the renal artery is actually causing a significant narrowing/limitation of blood flow or is just a mild, innocent bystander
 - Tools available including pressure gradient assessment, intravascular ultrasound, OCT
 - Area of ongoing research

*Gornik HL, Persu AP, et al. *Vascular Medicine*. 2019;24:164.

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The Eyeball Cannot Accurately Assess Severity of Stenosis in Multifocal Renal FMD

A Gradient 30 mm Hg



B Gradient 50 mm Hg



C Gradient 52 mm Hg



D



Translesional pressure gradient measurements should be a routine component of the FMD renal angio protocol

Gradient = 19 mm Hg

Figure from: Gornik HL, Persu AP, et al. *Vascular Medicine*. 2019;24:164.

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Angiography/Angioplasty Protocol for Renal FMD

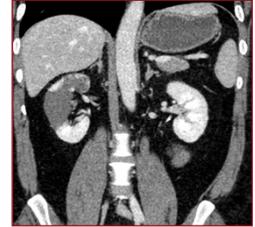
Table 6. Consensus protocol for catheter-based angiography and PTA in patients with renal artery FMD.

1. Flush aortogram (if prior cross-sectional imaging with CTA or MRA had not been previously performed) to look for all renal arteries and clearly profile the ostia of the renal arteries (with oblique views) prior to selective catheterization.
2. Selective renal arteriography, using multiple views, to visualize the entire renal vasculature, including for branch vessel involvement. **Kidney size, parenchymal perfusion, and assessment for renal artery aneurysm/dissection.**
3. A simultaneous, unstimulated, transluminal pressure gradient (between the distal renal artery and the aorta) should be measured, ideally with a pressure wire.⁴ If a pressure wire is not available, a small diameter end-hole catheter may be used for a pull-back pressure. In experienced centers, IVUS or OCT may also help to identify the severity of stenosis in patients with multifocal FMD.
4. A pressure gradient threshold of 10% of the mean (aortic) pressure can be used to decide whether to perform balloon angioplasty (i.e. PuffPa < 0.90).¹¹⁷ These parameters are extrapolated from the study of patients with atherosclerotic renal artery stenosis and have not been validated in patients with FMD.
5. For angioplasty, the initial balloon diameter used should be based upon the diameter of the distal normal renal artery using a calibrated catheter and quantitative vascular angiography software, IVUS, or OCT. The balloon diameter size should be incrementally increased by 0.5 mm until the transluminal gradient is resolved or until there is a < 10% mean transluminal gradient. **Angioplasty should be aborted if the patient experiences pain during balloon inflation or if a complication occurs.**
6. Renal artery stenting is generally not indicated in the setting of FMD and is limited for bail-out use to treat complications related to angioplasty (dissection, pseudoaneurysm, or rupture), in some cases of primary renal artery dissection, or for the treatment of a renal artery aneurysm.
7. At the end of the procedure, final angiograms are obtained using the same catheter and orthogonal views that were used for baseline angiography to assess for potential complications (renal artery dissection, pseudoaneurysm, rupture, renal emboli, or infarction).
8. This procedure can be performed on an outpatient basis most of the time. However, some patients may require monitoring overnight in the hospital.

University Hospitals Cleveland Clinic Gornik HL, Persu AP, et al. *Vascular Medicine*. 2019;24:164. Cleveland, Ohio

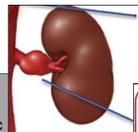
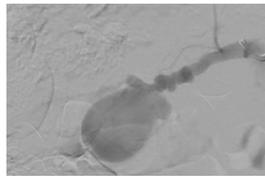
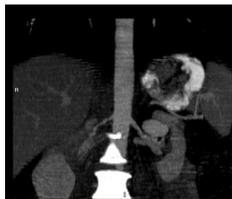
Renal Artery Infarction

- Lack of blood flow to a segment of the kidney causes death of kidney tissue or an "infarction" (analogous to a myocardial infarction/heart attack)
- Presents with flank pain, abdominal pain, nausea, vomiting, blood in urine
- Often due to a renal artery dissection (tear)
- Can also be due to clot formation in the renal artery or branches or an embolism from another part of the circulation (heart, aorta)
- Generally managed with medications, but intervention is needed in some cases



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Renal Artery Aneurysm



- Renal Artery**
- Symptomatic
 - Size > 3cm
 - All sizes
 - in women of childbearing age
 - in patients with refractory hypertension and renal artery stenosis

Recent guidelines have *increased* the threshold for repair in post-childbearing women. Most can be followed with periodic imaging, medications for blood pressure control, aspirin, and not smoking!

Repair can be open surgery or endovascular depending on anatomy

Chaer RA, et al. *J Vasc Surg*. 2020 72:35.

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Kidney Function and FMD

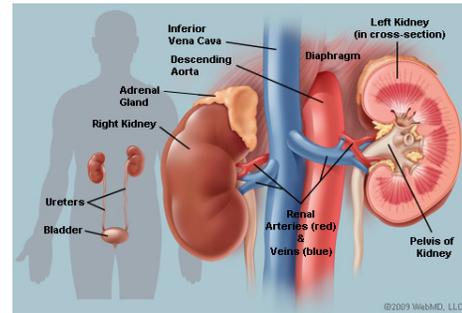
- Impaired renal function (aka chronic kidney disease or CKD) is actually *uncommon* among patients with FMD
 - 2012 US Registry: Only 1.6% of patients had "renal failure" at time of enrollment (Olin JW, et al. *Circulation*. 2012;125:3182).
 - 2019 US Registry: mean serum Creatinine 0.8 mg/dL and estimated GFR 83.6 ml/min/1.73 m² (Gornik HL, Persu AP, et al. *Vascular Medicine*. 2019;24:164)
- Nonetheless, we do monitor renal function as part of follow-up of renal FMD
- Maintaining kidney health is important for patients with FMD
 - Good control of blood pressure
 - Other "best practices" for healthy kidneys

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Maintaining Kidney Health and Preventing Chronic Kidney Disease

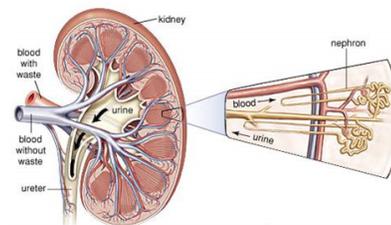
Aparna Padiyar MD
University Hospitals Division of Nephrology
October 2022



What is Chronic Kidney Disease?

- Progressive loss in kidney function over a period of months or years
- Kidney failure affects your whole body, and can make you very ill in its late stages
- Untreated kidney failure can be life-threatening

Each of Your Kidneys have MILLIONS of Tiny Filters called Nephrons



Kidney Diseases are Common, Harmful and often Treatable

Common: Between 8 and 10% of the adult population

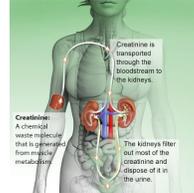
Harmful:

- Risk of progressive loss of kidney function that can lead to kidney failure which means regular dialysis treatment or a kidney transplant is needed to survive
- Increases the risk of premature death from associated cardiovascular disease (i.e. heart attacks and strokes)

Treatable: If CKD is detected early and managed appropriately, the deterioration in kidney function can be slowed or even stopped.

How is kidney function measured?

- Blood Creatinine level



- Glomerular Filtration Rate (GFR) can easily be estimated from measurement of the blood creatinine level

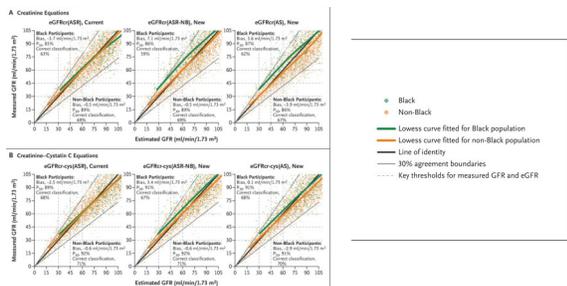
Stages of Chronic Kidney Disease

| Stage | Description | GFR Level |
|------------------------|--|-----------------------------------|
| Normal Kidney Function | Healthy Kidneys | 90ml/min or more |
| Stage 1 | Kidney damage with normal or high GFR | 90ml/min or more |
| Stage 2 | Kidney damage and mild decrease in GFR | 60 to 89ml/min |
| Stage 3 | Moderate decrease in GFR | 30 to 59ml/min |
| Stage 4 | Severe decrease in GFR | 15 to 29 ml/min |
| Stage 5 (ESKD) | Established kidney failure | Less than 15ml/min or on dialysis |

Multiple GFR Estimating Equations

| Equation | Creatinine Clearance (CrCl) | Estimated Glomerular Filtration Rate (eGFR) | | |
|----------------|--|--|--|--|
| | Cockcroft-Gault | Modification of Diet in Renal Disease (MDRD) 6 variable | Modification of Diet in Renal Disease (MDRD) 4 variable | Chronic Kidney Disease Epidemiology (CKD-EPI Creatinine) 11 th |
| | $CrCl = \frac{[(140 - Age) \times weight] \times 0.85 \text{ (if female)}}{SCr} \times 72$ | $eGFR = 175 \times (SCr)^{-1.154} \times (Age)^{1.176} \times 0.762 \text{ (if female)} \times BUN \times (Albumin)^{2.18} \times 1.18 \text{ (if Black)}$ | $eGFR = 175 \times (SCr)^{-1.154} \times (Age)^{1.176} \times 0.742 \text{ (if female)} \times 1.212 \text{ (if Black)}$ | $eGFR = 141 \times \min(SCr, 1)^{1.1} \times \max(SCr, 1)^{-1.154} \times 0.993^{(Age - 20)} \times 1.018 \text{ (if female)} \times 1.156 \text{ (if Black)}$ |
| Year Developed | 1976 | 1999 | 2006 | 2009 |
| Notes | -Studied in hospitalized men -Not adjusted for body surface area | -Studied in patients with CKD -Non-standardized SCR assay | -Studied in patients with CKD -Re-modeled to standardized SCR assay | -More accurate towards normal kidney function -As accurate as MDRD for those with <90ml/min/1.73m ² |
| | Weight in kilograms; Age in years SCr = serum creatinine; BUN = blood urea nitrogen; CKD = chronic kidney disease | | | |

Bias in All Equations



2021 NKF-ASN CKD-EPI

CKD-EPI Equation for Estimating GFR on the Natural Scale Expressed for Specified Sex, Standardized Serum Creatinine and Standardized Serum Cystatin C (From New Eng J Med 2021)

| Sex | Serum Creatinine (mg/dL) | Equation |
|--------|--------------------------|--|
| Female | ≤0.7 | $GFR = 142 \times (Scr/0.7)^{-2.45} \times 0.9938^{Age} \times 1.012$ |
| Female | >0.7 | $GFR = 142 \times (Scr/0.7)^{-1.208} \times 0.9938^{Age} \times 1.012$ |
| Male | ≤0.9 | $GFR = 142 \times (Scr/0.9)^{-1.208} \times 0.9938^{Age}$ |
| Male | >0.9 | $GFR = 142 \times (Scr/0.9)^{-1.226} \times 0.9938^{Age}$ |

THE IMPACT OF THE REMOVAL OF RACE IN EGFR EQUATIONS

Reduced eligibility for:

- Living donors
- Anti-cancer therapy
- Medications based on renal dosing

Expand eligibility for:

- Referral to a nephrologist
- Kidney transplant waitlist
- Coverage for kidney disease education

Fig. J. H. (2022). The Potential Impact of the New eGFR Equation in the United States and around the World. *Kidney News*, 14(1), 22-22.

Causes of CKD

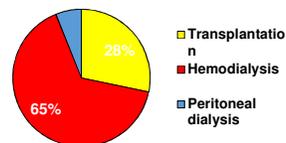
- High blood pressure (hypertension) 25%
- Diabetes 33%
- Other less common conditions:
 - Inflammation (glomerulonephritis)
 - Infections (pyelonephritis)
 - Inherited (such as polycystic disease)
 - Longstanding blockage to the urinary system (such as enlarged prostate or kidney stones).
- Some drugs can cause CKD, especially some pain-killing drugs (analgesics) if taken over a long time.
- Often doctors cannot determine what caused the problem.

Symptoms of CKD

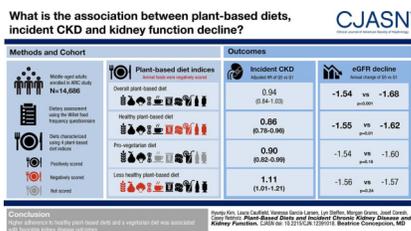
- A person can lose up to 90% of their kidney function before experiencing any symptoms.
- Most people have no symptoms until CKD is advanced.
 - Swollen ankles
 - Fatigue
 - Difficulty concentrating
 - Decreased appetite
 - Blood in the urine and foamy urine.

Treating CKD

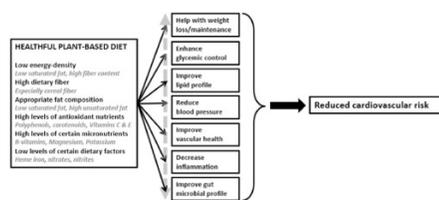
- There is no cure for chronic kidney disease.
- The main treatments are a proper diet, treating diabetes and hypertension, and medications.
- For those who reach ESKD, long term dialysis treatment or kidney transplantation are kidney replacement.



Dietary Prevention of CKD



How Does a Plant Based Diet Reduce CKD



Satija A, Hsu F. (2018) Plant Based Diets and Cardiovascular Health. Trends in Cardiovascular Medicine.

How to Eat a Plant-Based Diet

Eat These Foods

- Whole grains
- Legumes
- Vegetables
- Fruit
- Nuts
- Seeds

Avoid These Foods

- Meat
- Dairy
- Eggs
- Refined sugar
- Refined grains
- Saturated fat

MOVE YOUR WAY Adults need a mix of physical activity to stay healthy.

Moderate-intensity aerobic activity*

Anything that gets your heart beating faster counts.

at least **150 minutes a week**

Muscle-strengthening activity

Do activities that make your muscles work harder than usual.

at least **2 days a week**

AND

* If you prefer vigorous-intensity aerobic activity (like running), aim for at least 75 minutes a week.

If that's more than you can do right now, **do what you can**. Even 5 minutes of physical activity has real health benefits.

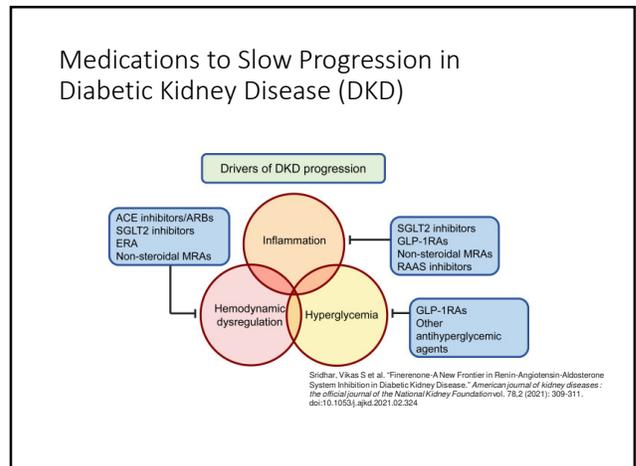
Walk. Run. Dance. Play. What's your move?

Physical Activity Guidelines For Americans. Department Of Health and Human Services. 2018.

Controlling Hypertension

Antihypertensive Drug Names

| Drug Class | Drug Names | Examples |
|--------------------------|------------|-----------------------------------|
| ACE inhibitors | "pril" | Lisinopril Enalapril |
| ARBs | "sartan" | Losartan Valsartan |
| Alpha Blockers | "osin" | Doxazosin Terazosin |
| Beta Blockers | "lol" | Metoprolol Labetalol |
| Calcium Channel Blockers | "dipine" | Amlodipine Nicardipine |
| Diuretics | "ide" | Furosemide Hydrochlorothiazide |



Avoiding Kidney Toxic Medications

| | | | | |
|---------------|----------------|----------------------------|-----------------------------|------------------------|
| | Acyclovir | Enalaprilat | Mesalamine | |
| | Ambisome® | Foscarnet | Methotrexate | herbal |
| | Amikacin | Gadopentetate dimeglumine® | Nafcillin | |
| | Amphotericin B | Gadoxetate disodium® | Piperacillin/tazobactam | |
| antibiotics | Captopril | Ganciclovir | Piperacillin | |
| | Carboplatin | Gentamicin | Siroliimus | iodinated contrast dye |
| | Cefotaxime | Ibuprofen | Sulfasalazine | |
| | Ceftazidime | Ifosfamide | Tacrolimus | |
| | Cefuroxime | Iodixanol® | Ticarcillin/clavulanic acid | |
| | Clodofovi® | Iohexol® | Tobramycin | |
| | Cisplatin | Iopamidol® | Tapiramate | |
| | Colistimethate | Ioversol® | Valacyclovir | |
| illicit drugs | Cyclosporine | Ketorolac | Valganciclovir | |
| | Dapsone | Lisinopril | Vancocycin | omeprazole |
| | Enalapril | Lithium | Zonisamide | |

In Summary...

- Early chronic kidney disease has no signs or symptoms.
- Chronic kidney disease usually does not go away.
- Kidney disease can be treated. The earlier you know you have it, the better your chances of receiving effective treatment.
- Blood and urine tests are used to check for kidney disease.
- Diet, control of hypertension and diabetes, and medications are important to slow the progression of kidney disease.
- Kidney disease can progress to kidney failure.

Open Q & A

Please Enter Your Questions into the "Chat" Box

Save the Date and Register Now!

Next Meeting Tuesday, January 17, 2023

Topic: SCAD, Cardiac rehabilitation