FMD Jeopardy

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Cleveland Clinic
Agenda

- FMD clinic at Cleveland Clinic: 2013 update
  - Local progress
  - Ongoing research activities

- An informal survey

- FMD Jeopardy
  - Prizes!
2013 Update

Cleveland Clinic FMD Program
Multi-specialty FMD Program

- 1st of its kind in the United States
- FMD clinic began August, 2008
  - Once a month → now twice a week
- Staffed by vascular medicine specialists as “FMD PCPs”
- Consultation with additional members of multidisciplinary team as needed
  - Consults driven by disease location, symptoms, and severity
  - Collaborative approach to challenging cases
  - Standardized approach to common FMD related problems
  - Development of experienced team of FMD providers
- Patients pre-screened prior to visit by FMD clinic nurse to optimize testing and consultation
- Regional, national, and international referrals
- Majority of patients are self-referred
Multi-specialty FMD Care Team

- Vascular medicine
- Interventional cardiology
- Vascular surgery
- Medical genetics
- Neurological institute
  - Stroke neurology
  - Neurosurgery
  - Headache program
- Psychiatry and psychology
- Nephrology
- High risk obstetrics
- Audiology (pulsatile tinnitus)
- Pathology
- Nursing
- Radiology
- Non-invasive vascular laboratory
- Administrative support
Activities of the FMD Program

- Increase awareness of FMD among Cleveland Clinic providers
- Educating physicians in training (residents, fellows) about FMD
- Educating nurses about FMD (CNEs program)
- Development of patient education materials
- Collaboration with FMDSA
  - FMD patient web chats (twice yearly)
  - Participation in annual FMDSA meeting
- FMD Research
  - U.S. Registry for FMD
  - FMD Biorepository
FMD Clinical Volume Growing

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FMD Weekly Warriors

Dr. Esther Kim
Kathy Petrarca
Ginger Davis
Kathy Murdakhaev
Neil Poria
Dr. Manju Raju (honorary member)

33 FMD Patients in 3 days!
The art tour stops by!
May 17, 2013
Research Roadmap for FMD
Spring, 2014

- 1 ½ day scientific symposium to be held in Cleveland, Ohio prior to FMDSA meeting in spring 2014
- Symposium chairs – Drs. Gornik and Olin
- State of the science lectures on FMD for clinicians and researchers
- 4 working groups composed of international experts in FMD will develop coordinated FMD research strategy
Research Roadmap for FMD Working Groups

1. Epidemiology (Dr. Esther Kim, chair)
   - What is true prevalence of FMD? Risk factors for development?

2. Genetics (Dr. Santhi Ganesh, chair)
   - Topics to include analysis plans for existing banked blood samples

3. Maximizing Registries to Advance FMD knowledge (Dr. Jim Froehlich, chair)
   - Future analysis plans of US Registry for FMD, collaboration with related vascular registries

4. Imaging and Clinical Management (Dr. Jeff Olin, chair)
   - What are top research priorities for specific clinical studies related to diagnosis and treatment?
FMD Jeopardy

- Fun!
- Prizes!
- Choose single best answer using audience response system
Part I: Warm-up

FMD Basics
Anatomy
Physiology
What do we see under the microscope in FMD?

1. Clusters of white blood cells infiltrating the blood vessel wall
2. Cholesterol filled cells known as “foam cells” and calcium deposits
3. Disruption of elastic fibers and muscle fibers in the middle wall with cystic areas
4. Thickening of the middle or inner layer of the vessel wall with extra collagen deposition
FMD Pathology - Medial Fibroplasia\textsuperscript{1,2} (elastic tissue stain)

\textsuperscript{1}Dr. Seymour Rosen in Kumar: Robbins and Cotran Pathologic Basis of Disease. 8\textsuperscript{th} Edition.

\textsuperscript{2}Casanegra AC, et al. SVM 2010.
What type of FMD is shown in this image?

1. Medial fibroplasia
2. Intimal fibroplasia
3. Aneurysmal FMD
4. Lateral FMD
Intimal and Medial Fibroplasia

Intimal fibroplasia or focal FMD

Medial fibroplasia or multifocal FMD
This Branch of Which Artery Shows Signs of FMD?

1. Internal carotid
2. External carotid
3. Vertebral
4. Basilar
To What Area of the Body Does the External Carotid Artery Supply Blood?

1. Shoulder
2. Face and scalp
3. Frontal lobe of the brain
4. Occipital lobe of the brain
What is the Circle of Willis?

1. Network of blood vessels supplying blood to the intestines
2. Part of the aorta where the coronary arteries come off
3. Where the carotid and vertebral arteries communicate to cross cover in the setting of severe blockages
4. Cluster of small arteries that filter blood to make urine
Communicating Blood flow to the Brain: The Circle of Willis

Vertebral arteries
In which of the following blood vessels is FMD least common?

1. Vertebral artery
2. Internal carotid artery
3. Left circumflex coronary artery
4. Renal artery
The arrow is pointing to which blood vessel?

1. Abdominal aorta
2. Renal artery
3. Common iliac artery
4. External iliac artery
CTA: 66 year-old woman with bilateral femoral bruits
Which of the following blood tests is the best marker of the severity of disease in a patient with FMD

1. Erythrocyte sedimentation rate
2. C-reactive protein
3. Creatinine
4. None of the above
Part II: In the clinic

Medical history
Symptoms
Physical examination
What is pulsatile tinnitus?

1. Rhythmic swooshing sound in the ear
2. Pain in the abdomen after eating
3. Pain in the arms or legs that comes on with exertion and is alleviated by rest
4. Shortness of breath that occurs while lying down
What is shown in this image?

1. Horner’s syndrome
2. Bell’s palsy
3. Trigeminal neuralgia
4. “Blown pupils”

Image courtesy: www.radiopaedia.org
In the FMD clinic, what does Horner’s syndrome suggest?

1. Severe carotid artery stenosis
2. Carotid dissection
3. A diagnosis other than FMD, such as multiple sclerosis
4. Poorly controlled hypertension
What is a dissection?

1. Tear of a blood vessel that can lead to narrowing or complete blockage
2. Enlargement of a blood vessel to more than 1.5 times its normal size
3. Blood clot (thrombosis) of a blood vessel
4. Rupture of an aneurysm
Which of the following is most likely to be a symptom directly related to FMD:

1. Fatigue and loss of energy
2. Sudden weakness or numbness in the right arm
3. Heart palpitations
4. Pain in the joints of the hands and feet
What is a normal blood pressure for a 13 year-old girl?

1. It depends on her height
2. It depends on her weight and whether or not she has started to menstruate
3. <140/90 mm Hg
4. <130/80 mm Hg
## Blood Pressure Levels for Girls by Age and Height Percentile (Continued)

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Source: NIH/NHLBI also see FMDSA website
Which Artery Can a Doctor Palpate (feel) on Physical Examination

1. Vertebral artery
2. Renal artery
3. Left main coronary artery
4. Brachial artery
Fibromuscular Dysplasia of the Brachial Artery

Which of the following blood tests is the best marker of kidney function?

1. Potassium
2. Creatinine
3. Blood urea nitrogen
4. Plasma renin level
Participant Scores

1000  Participant 1DD303
Part III: FMD Imaging
What is this?

1. Intravascular ultrasound
2. Carotid duplex ultrasound
3. Gadolinium enhanced MRA
4. CT angiogram during “run off” phase
What is Happening in this Picture

1. Patient receiving new treatment for swooshing in the ears
2. Ultrasound of arteries inside the brain
3. Activation of coils for treatment of brain aneurysm
4. Cutting edge portable MRA of the brain
Transcranial Doppler (TCD)

- Low frequency ultrasound used to listen to blood flow in the brain arteries and Circle of Willis through areas of the skull
- Does not directly visualize arteries, but can assess adequacy of flow and rule out narrowings or blockages
- Helpful to evaluate flow in the vertebral and basilar arteries in the back of the neck and brain
- Can assess for embolization or spasm of brain arteries
What Test Was Used to Diagnose Carotid Multifocal FMD?

1. CT Angiogram
2. Carotid duplex ultrasound
3. Gadolinium enhanced MRA
4. Catheter (digital subtraction) angiogram
What is this?

1. FMD under the microscope
2. Renal Doppler
3. IVUS ultrasound images taken from inside an artery
4. Simulation of the latest ride at Disneyland
What Symptom Might this Patient Be Complaining of?
What Symptom Might this Patient Be Complaining of?

1. Chest pain and shortness of breath
2. Severe neck pain and headache
3. Abdominal pain and weight loss
4. Pain in the flank (sides)
Coronary Artery Dissection with FMD (?)
What is Shown in This CT Scan?

1. Brain aneurysm
2. Carotid artery aneurysm
3. Carotid artery pseudoaneurysm
4. Carotid artery beading with some narrowing
Pseudoaneurysm = focal outpouching
Usually due to a prior dissection
Part IV: Approach to the Treatment of the FMD Patient
Each of these medications thins the blood by inhibiting the platelets, except…

1. Warfarin
2. Aspirin
3. Prasugrel
4. Clopidogrel
When might a stent be needed for treatment of renal FMD

1. Single kidney
2. Very long string of beads
3. Mobile right kidney
4. Renal artery dissection
5. Never
Goal for FMD is improvement of flow hemodynamics

*Do not try to straighten the beads*

*Only stent when necessary*
Case scenario

- A 51 year-old woman meets a new internist for the first time.
- She is healthy and takes no medications and has no neurological symptoms.
- The doctor hears a bruit in the right side of the neck.
- A carotid ultrasound is done in an experienced lab which shows evidence of FMD in both internal carotid arteries.

- Which of the following tests is most appropriate next step?
Please make your selection...

1. Catheter angiogram of the carotid arteries
2. Brain MRA to look for aneurysms
3. Transcranial Doppler
4. Coronary angiogram
Above what % stenosis should a patient with carotid FMD who has no symptoms except be sent for a carotid angioplasty

1. >50%
2. >70%
3. >80%
4. For 100% blockage
5. Almost never
Carotid Blockages Due to FMD vs. Atherosclerosis are Managed Differently

- Athero blockages more likely to embolize causing TIA or stroke
- Surgery (endarterectomy) or carotid artery stenting recommended for athero blockages:
  - Patient with > 50% stenosis with TIA or stroke
  - Asymptomatic patient with severe ICA stenosis (>70 or 80%)
- For FMD, we intervene for blockages only for patients with severe symptoms
Which Blood Pressure Medication has the Most Direct Effect on the Kidneys?

1. Beta Blocker
2. Calcium channel blocker
3. Angiotensin receptor blocker
4. Hydralazine
Which of the Following is the Least Important Factor in Determining When to Treat a Brain Aneurysm?

1. Aneurysm size
2. Aneurysm location
3. Prior history of brain aneurysm rupture
4. Presence of FMD/beading in the carotid artery below the brain aneurysm
Part IV: Lessons Learned from the Patient Registry
What is the Typical Delay Between Onset of Symptoms and Diagnosis of FMD?

1. 6 months
2. 1 year
3. 2 years
4. 4 years
5. 8 years
Men with FMD are More Likely than Women with FMD to:

1. Have an arterial dissection
2. Have vertebral FMD
3. Have a stroke
4. Have pulsatile tinnitus
If you check the renal arteries of a patient with carotid FMD, about ____ will have renal FMD too?

1. 1/4
2. 1/2
3. ~ 2/3
4. 3/4
Participant Scores

1300 Participant 1DD303

Congratulations
The FMDSDA Annual Meeting Gornik Poll
How are you connected to FMD?

1. I am an FMD patient
2. I am a loved one of an FMD patient
3. I am a health care provider and take care of patients with FMD
4. I have no direct connection with FMD, but I came to learn more about it
How Many FMDSA meetings have you attended?

1. My first
2. Two or three
3. Four or five
4. All six of them
5. I was there when the FMDSA founders met over a kitchen table
How old are you?

1. < 18 years
2. 18-40 years
3. 40-55 years
4. 55-70 years
5. > 70 years
What is your gender?

1. Female
2. Male
What is your occupation?

1. Health care provider
2. Teacher/education
3. Homemaker/full time mom or dad
4. Business or administration
5. Communications
6. Lawyer or law enforcement
7. Creative arts
8. Other
If you are a health care provider, are you a…

1. Nurse
2. Physician
3. Technologist
4. Medical assistant
5. Mental health provider
6. Other
How far did you travel to come to the FMDSA meeting?

1. < 50 miles
2. 50-200 miles
3. 200-500 miles
4. 500-1000 miles
5. > 1000 miles
If you are an FMD patient, who is your primary FMD specialist?

1. Family doctor, internist, or 1° care physician
2. Vascular surgeon
3. Medical vascular specialist or cardiologist
4. Nephrologist (kidney doc)
5. Neurologist (brain doc)
6. Radiologist
7. OB/GYN
8. Other
Does your primary FMD health care provider take care of other patients with FMD?

1. Yes
2. No
3. I don’t know
For FMD patients, in terms of the FMD patient registry…

1. I have enrolled
2. I have definite plans to enroll within the next year at a specific registry site
3. I would like to participate, but I need to learn more about it
4. I do not plan to participate
5. Other
What would you like to see more off at the next FMDSA meeting?

1. Lectures by health care providers on specific aspects of FMD
2. Research updates
3. Interactive Q and A with panels
4. Small group workshops or breakout sessions
5. More social events
Thank you FMDSA!