

Aging in Marfan and Related Conditions

Juan Bowen, M.D.

Mayo Clinic

Rochester, MN

59 Year Old Woman With Marfan Syndrome

- Diagnosis in young adulthood
- Age 53
 - Family history aortic dissection
 - Aortic root 44 mm
 - Mitral valve prolapse with mild mitral regurgitation
 - Valve-sparing aortic root repair done



New Problem: Unsteady Gait and Imbalance

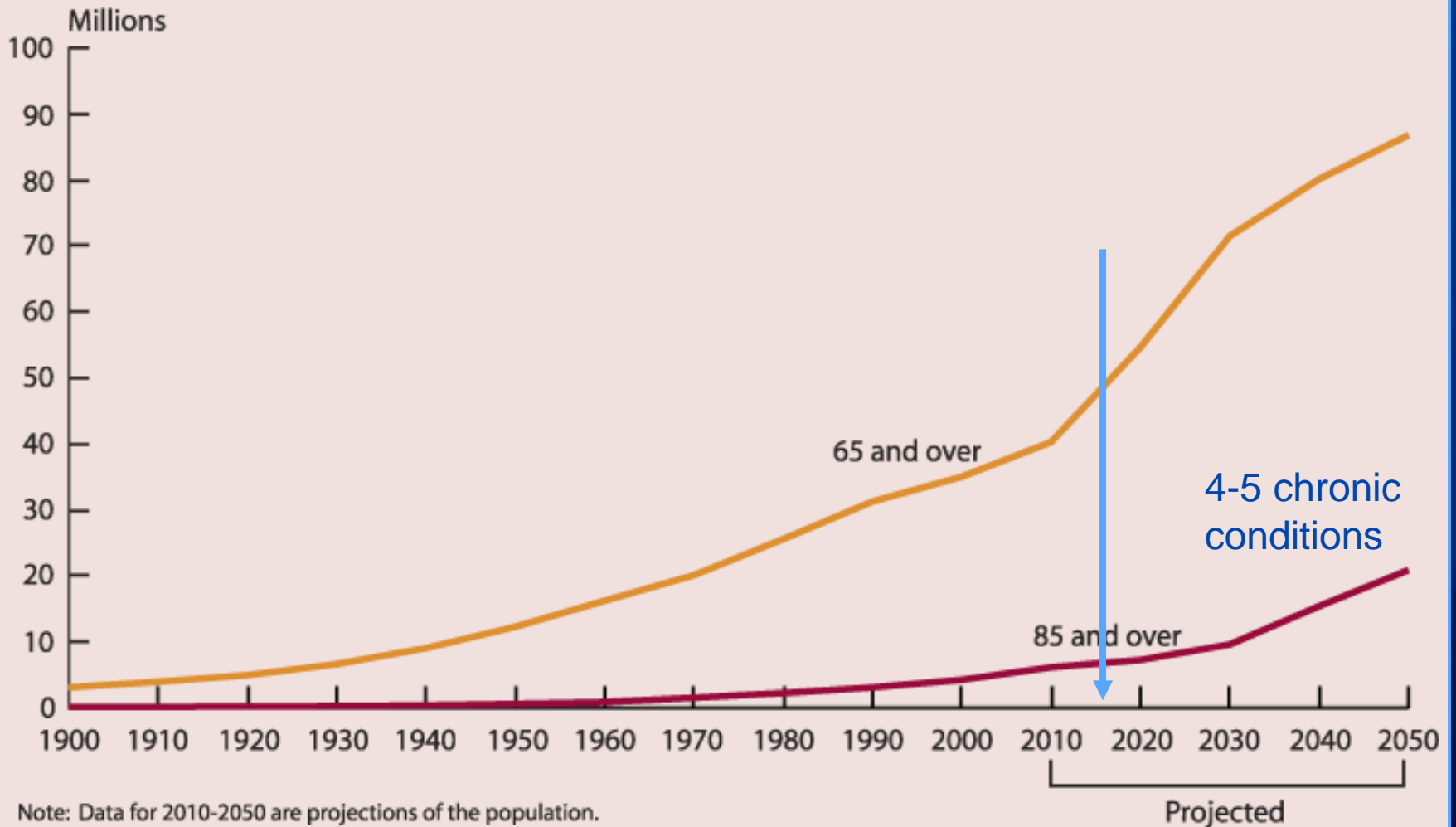
- Problems identified
 - Lumbar dural ectasia
 - Degenerative hip arthritis
 - Peripheral neuropathy



The Future for Ms. H.

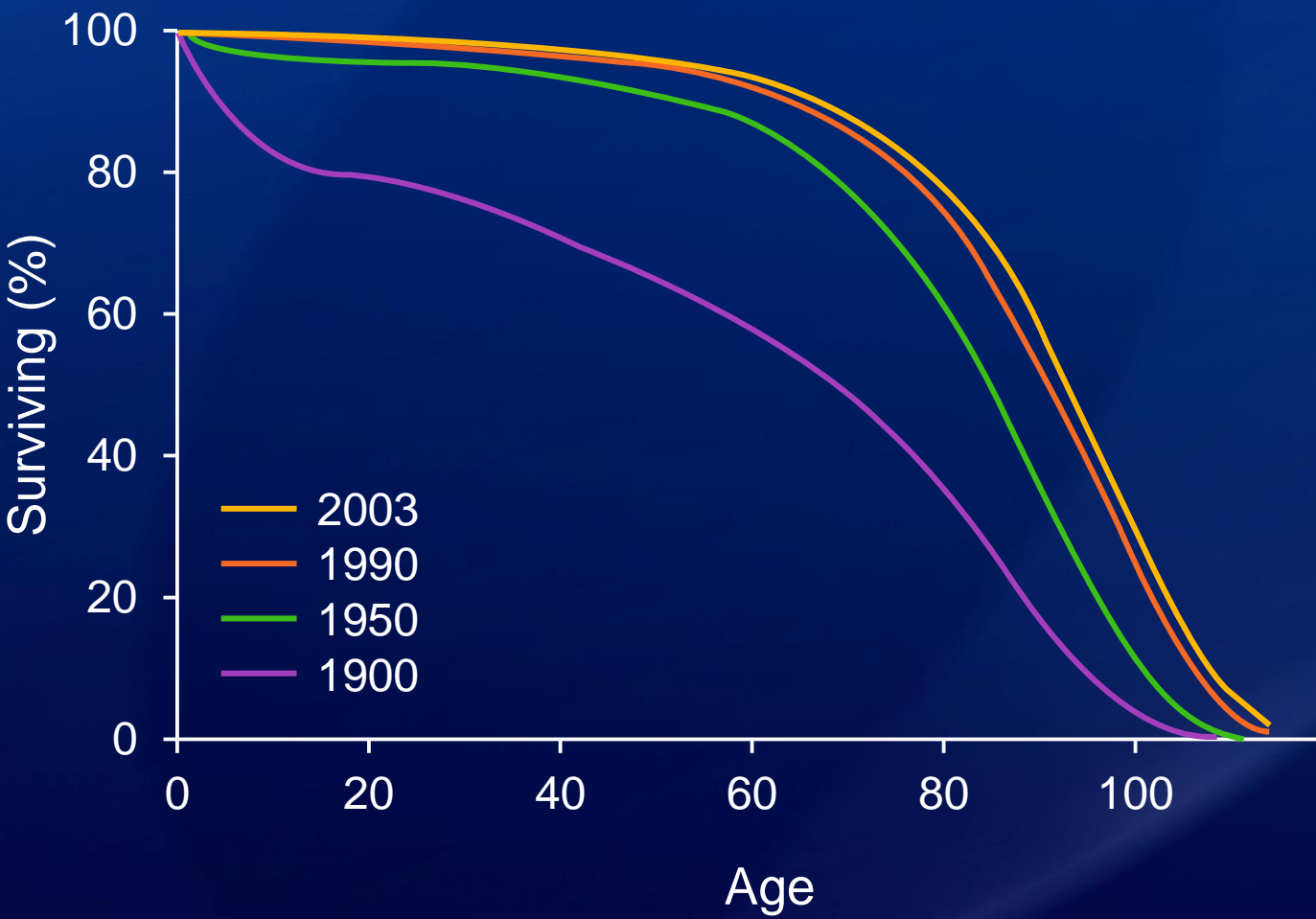
- Health issues related to aging.
- Health issues related to Marfan syndrome.

Number of people age 65 and over, by age group, selected years 1900-2000 and projected 2010-2050



Note: Data for 2010-2050 are projections of the population.
Reference population: These data refer to the resident population.
Source: U.S. Census Bureau, Decennial Census and Projections.

Life Expectancy in U.S.



Marfan Syndrome Life Expectancy

- 1972 – median survival 48 years
- 1993 – median survival 72 years

- Why?



- Basic and clinical research leading to better diagnosis and management.

Major Causes of Death in U.S.

- Cardiovascular disease
 - Stroke
 - Heart attack
- Cancer
- Other

Health Behaviors Which Reduce Life Expectancy

- Tobacco use
- Alcohol use
- Obesity

Normal Aging

- Normal aging
 - Decreased reserve
 - Characteristic changes in body systems
- Marfan syndrome/related disorder
 - Specific issues

Quality of Life Issues

- Mobility
- Special senses
- Pain
- Fatigue
- Depression
- Anxiety

Body Systems

- Cardiovascular
- Respiratory
- Kidney
- Musculoskeletal
- Ocular
- Gastrointestinal
- Endocrine
- Neurologic

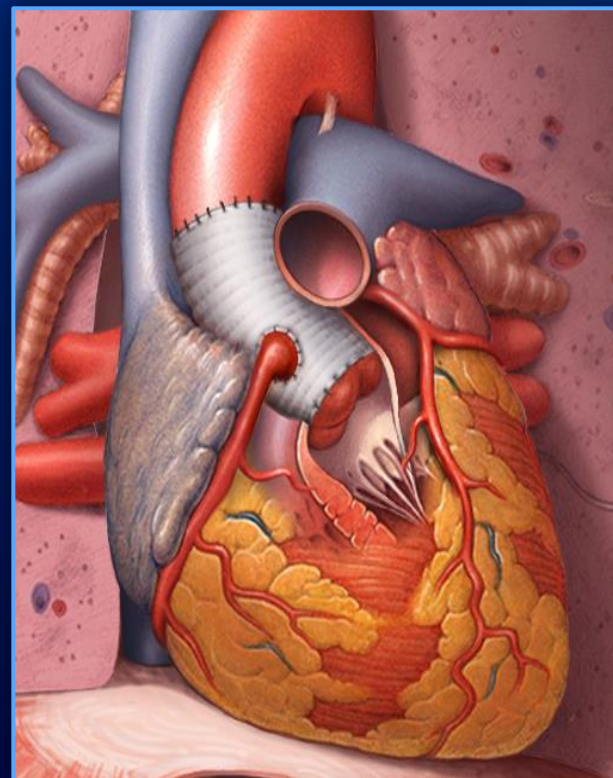
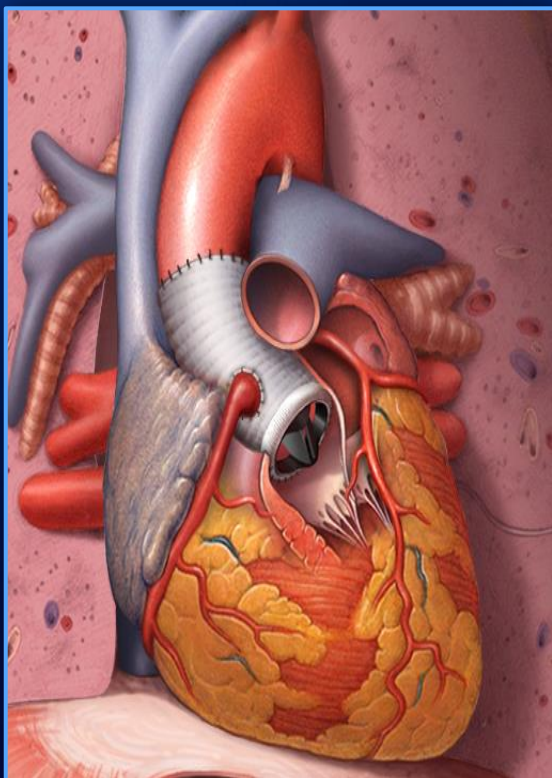
Aging of the Cardiovascular System

- Hypertension
- Heart valves
- Coronary arteries
- Conduction system
- Heart muscle

MFS/RD Cardiovascular System

- Aorta
- Heart valves
- Heart muscle

Preventive aortic repair

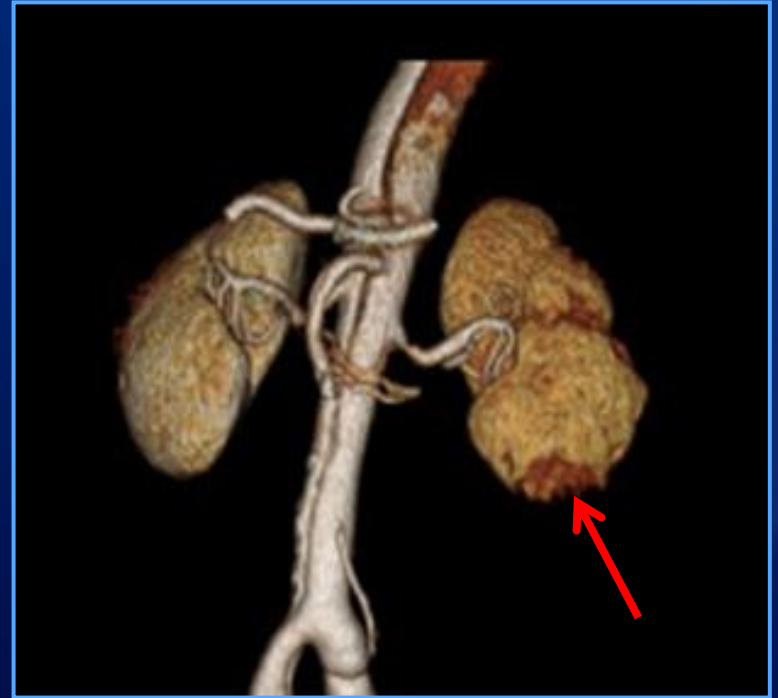
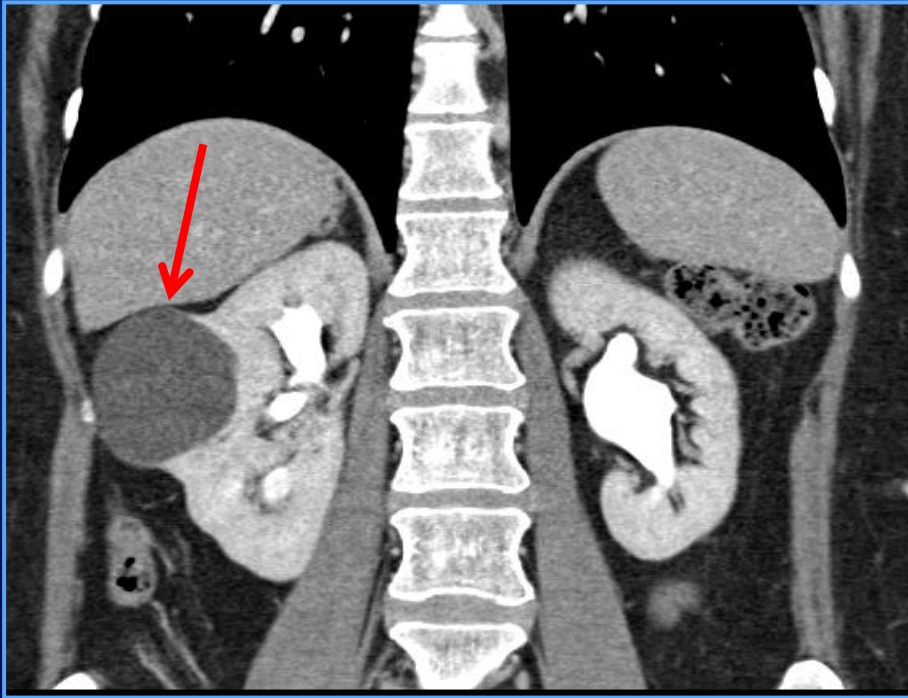


Aging of the Kidneys

- Expected decline in kidney function
- Hypertension
- Diabetes
- Toxicity from medications

MFS/RD Kidneys

- Benign kidney cysts
- Kidney damage
 - Hypertension
 - Diabetes
 - Vascular disease
 - Medications

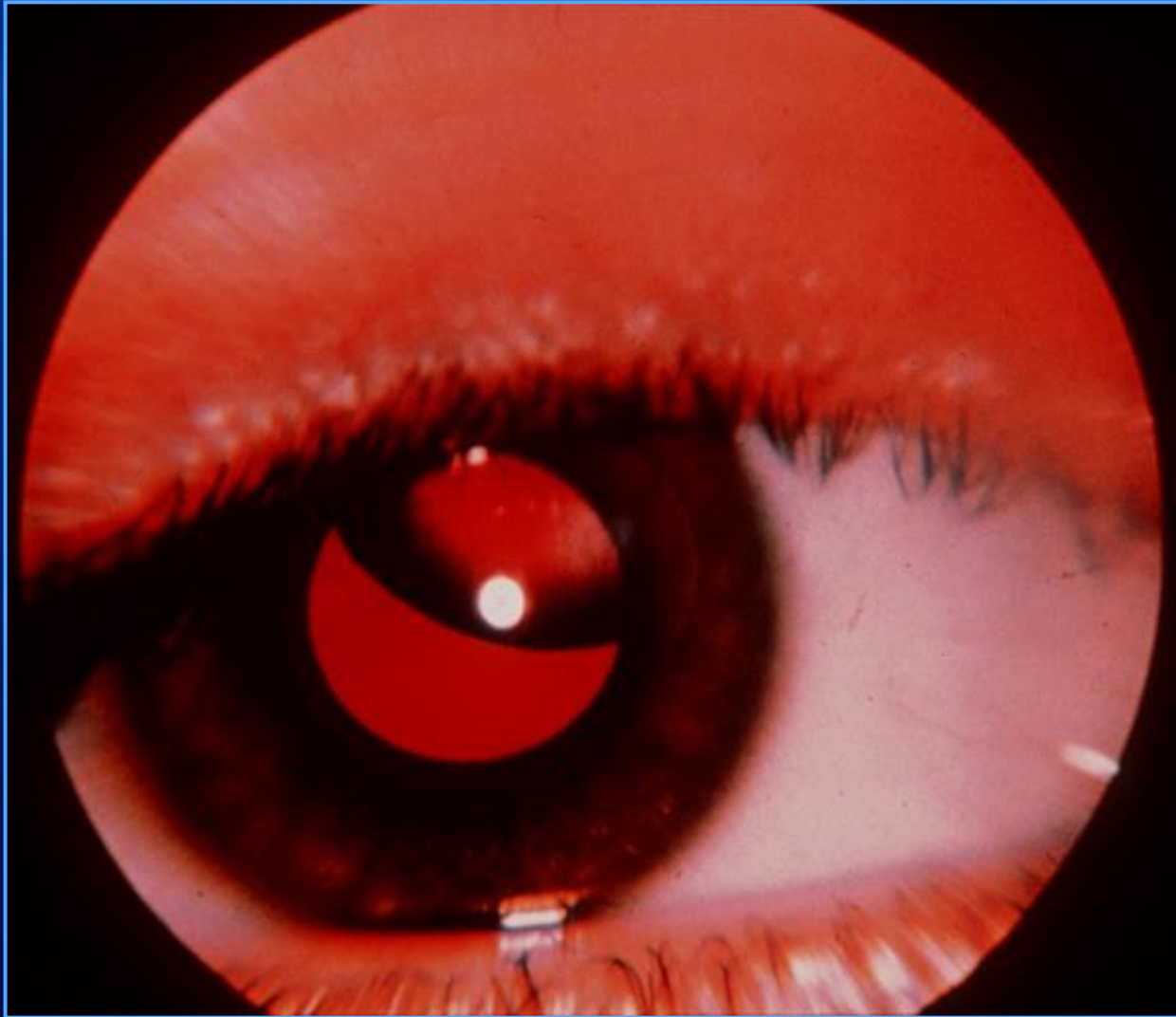


Aging of the Eye

- Cataract
- Glaucoma
- Macular degeneration

MFS/RD Eye

- Myopia
- Retinal tears
- Lens dislocation
- Glaucoma



Iridodonesis



Aging of the Musculoskeletal System

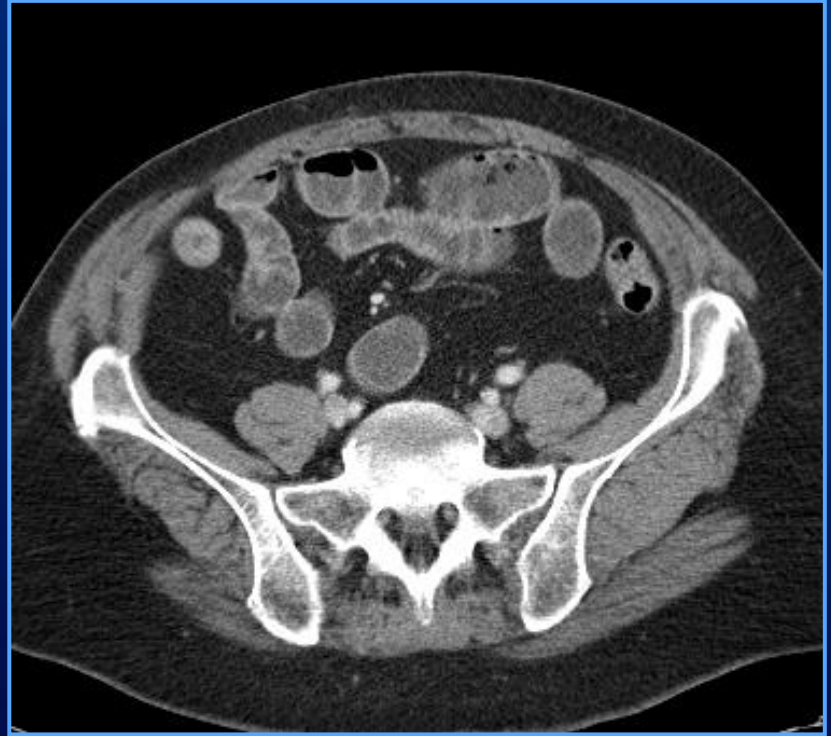
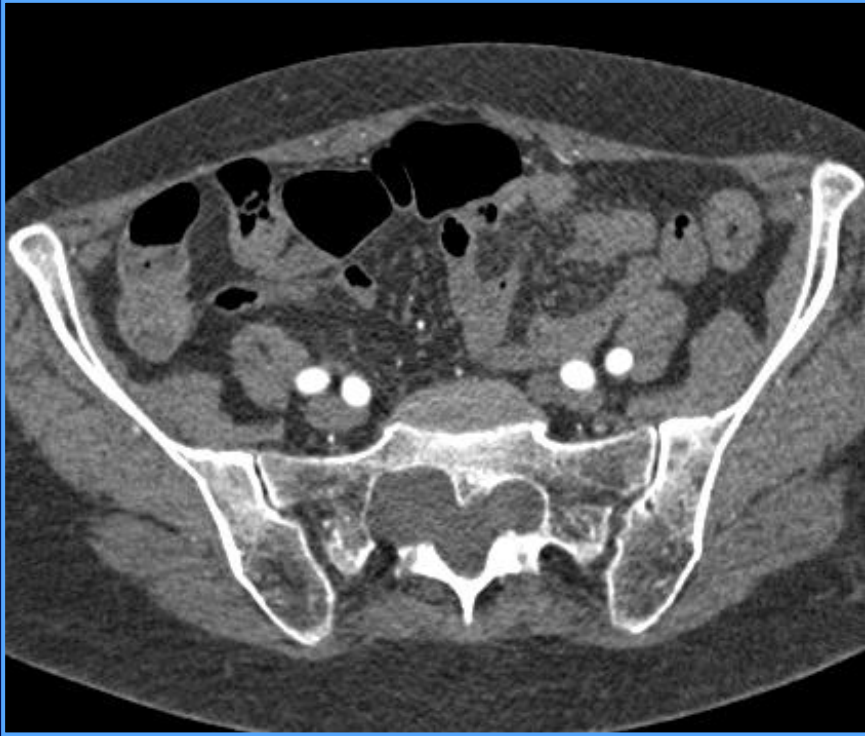
- Degenerative arthritis
 - Spine
 - Hips and knees
- Decreased muscle mass

MFS/RD Musculoskeletal

- Spine
 - Scoliosis
 - Chest wall
 - Dural ectasia
- Joints
 - Accelerated degenerative arthritis
- Muscles
 - Myopathy





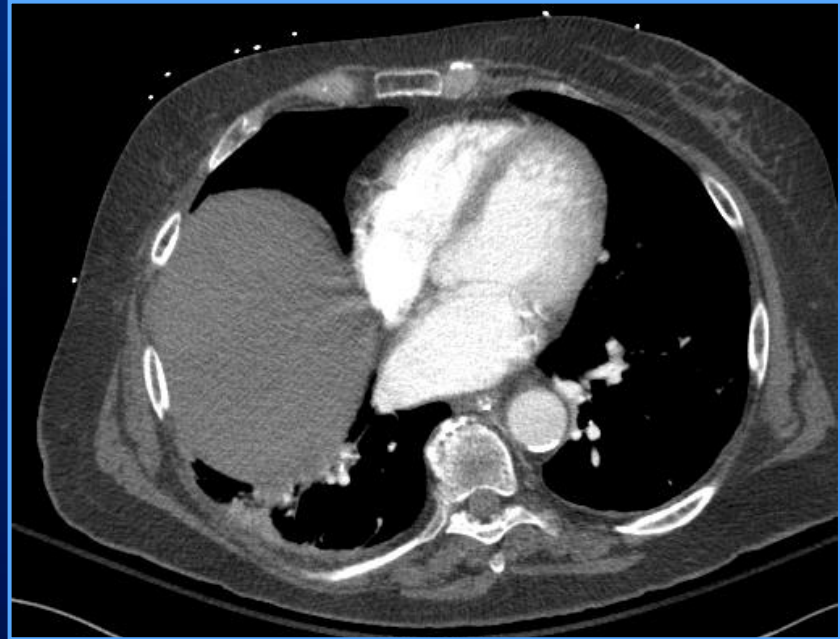


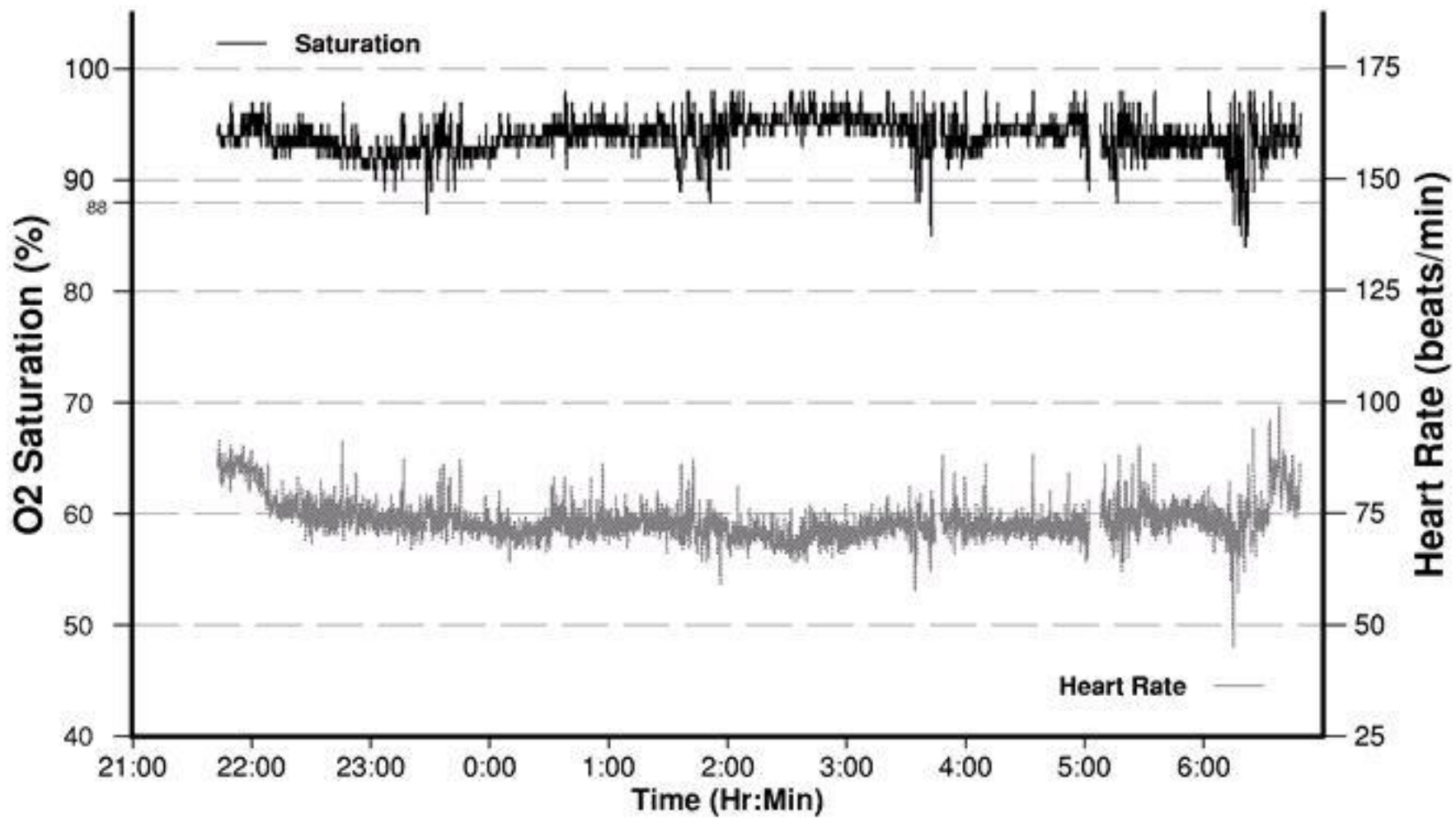
Aging of the Respiratory System

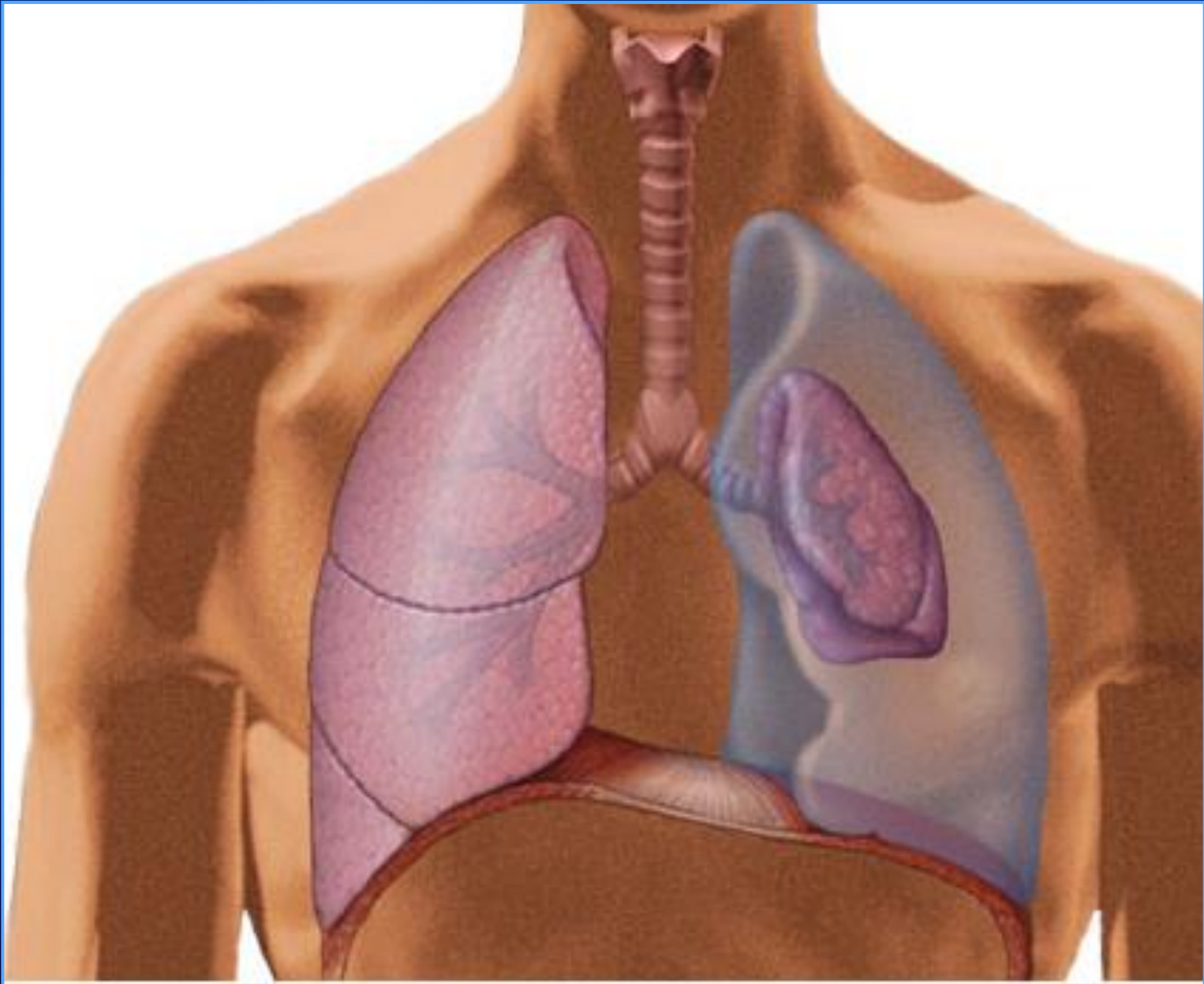
- Chest wall and lung function
- Asthma and emphysema

MFS/RD Respiratory

- Chest wall effects on lung function
- Sleep apnea
- Pneumothorax





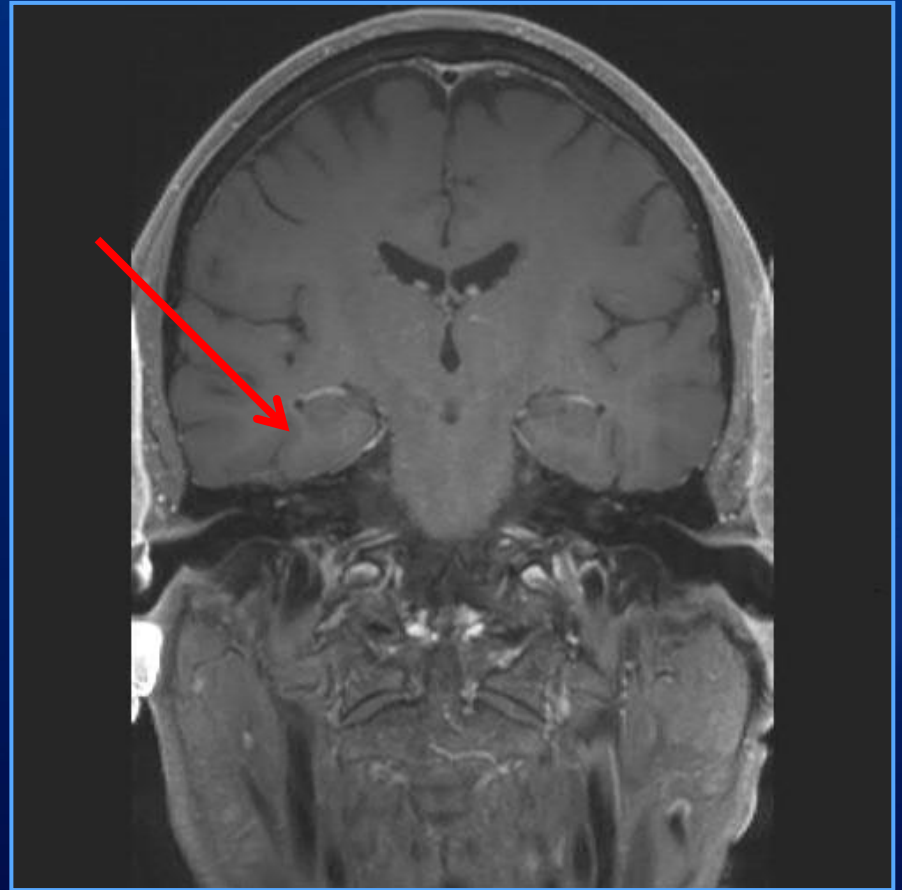
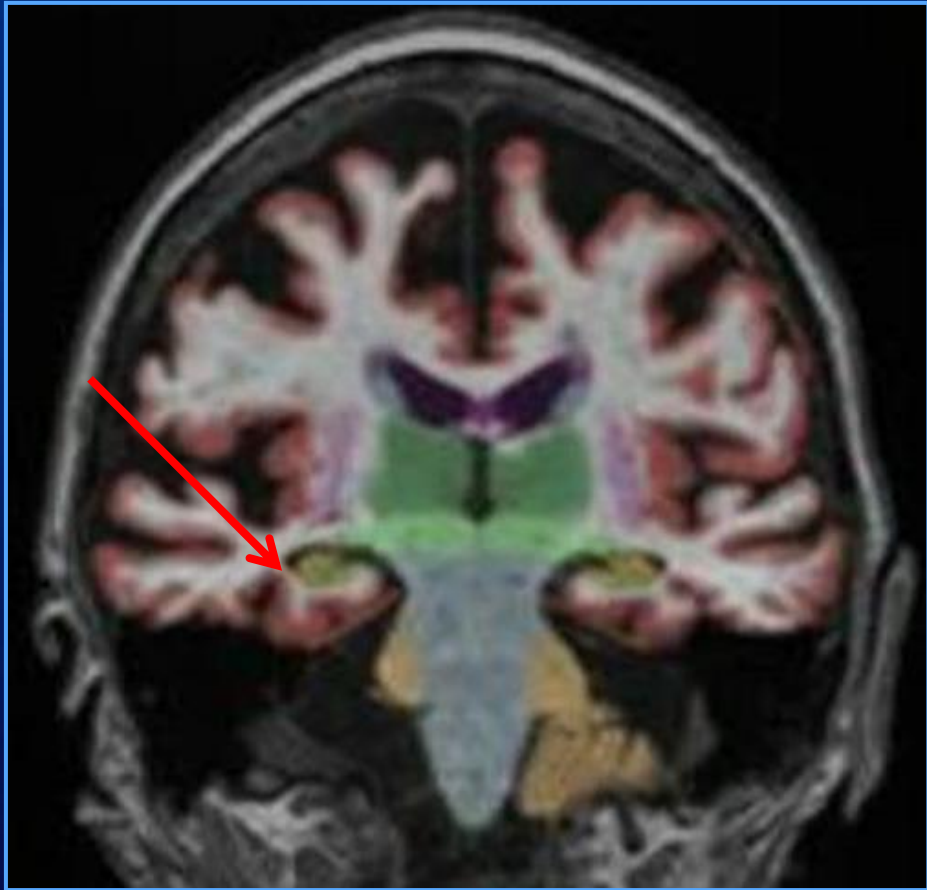


Aging of the Nervous System

- Imbalance
- Parkinson's disease
- Peripheral nerves
- Cognitive function

Cognitive Function

- Attention, processing speed, and episodic memory decline in normal aging
- An active and engaged lifestyle early in life is protective
- Continued physical activity is protective



MFS/RD Nervous System

- Central nervous system
 - No known changes
- Peripheral nervous system
 - Effects from dural ectasia

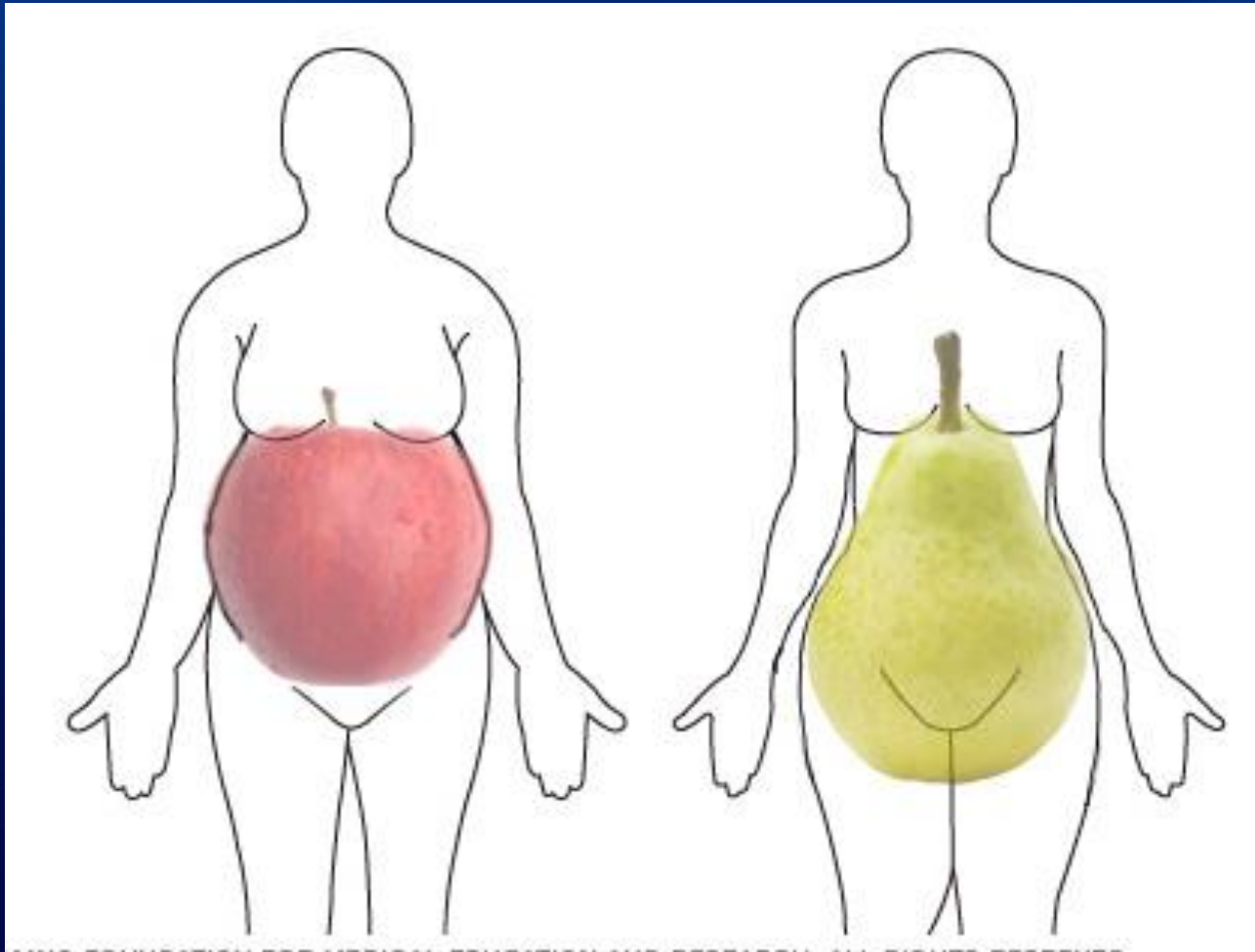
Aging of the Endocrine System

- Type 2 diabetes
- Thyroid
- Osteoporosis

MFS/RD Endocrine

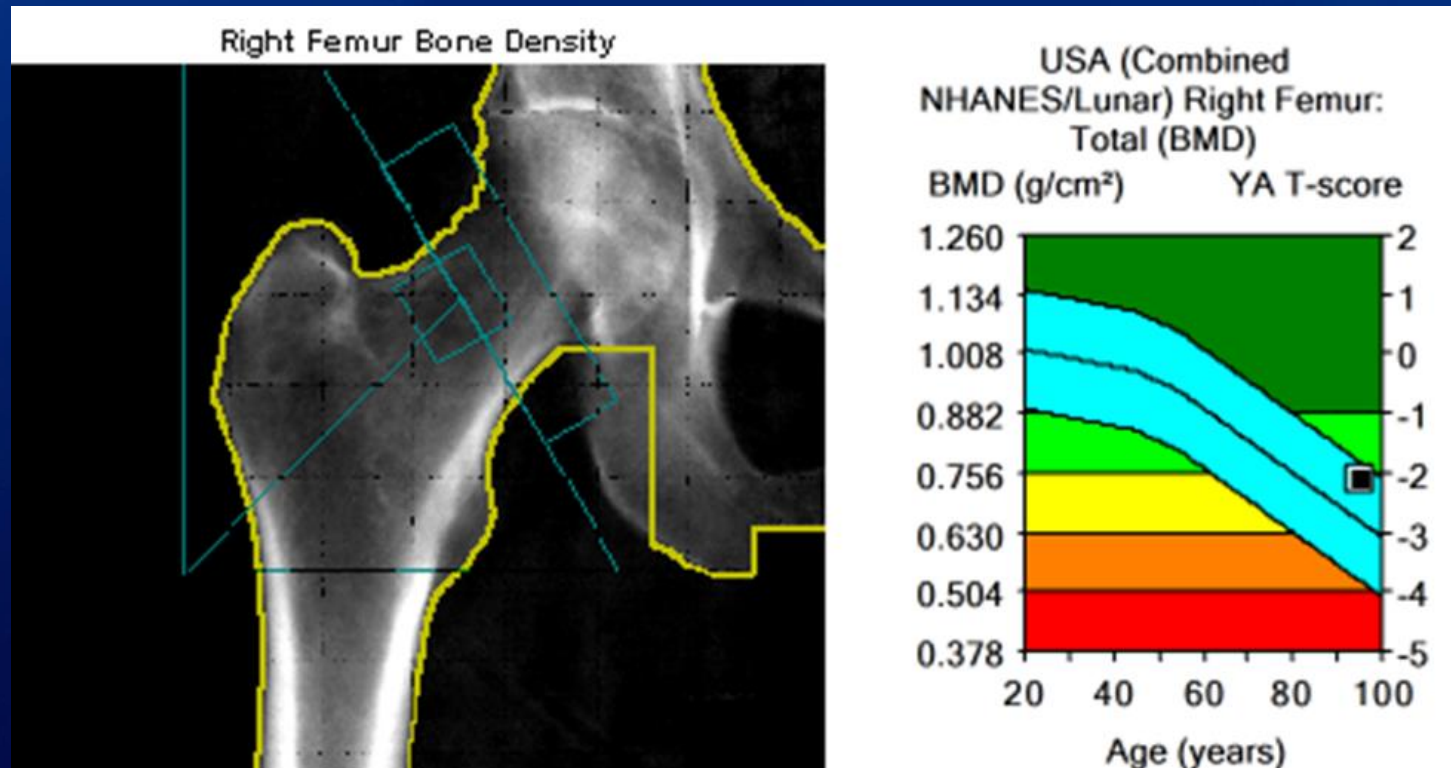
- Central obesity and risk of diabetes
- Osteoporosis

Variations in Fat Distribution



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Bone Density Measurement



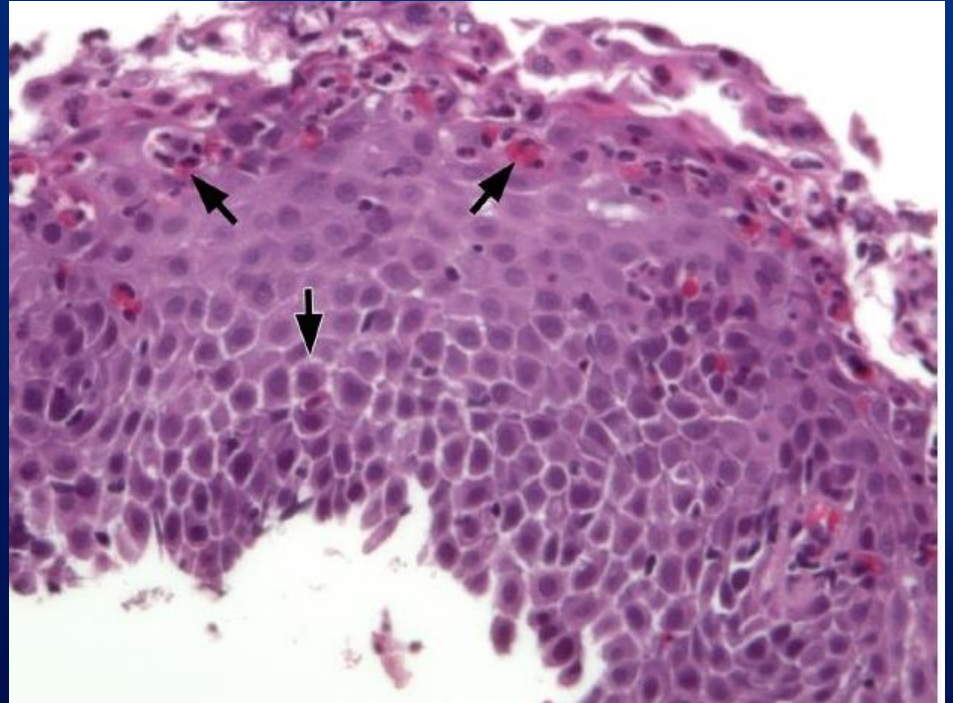
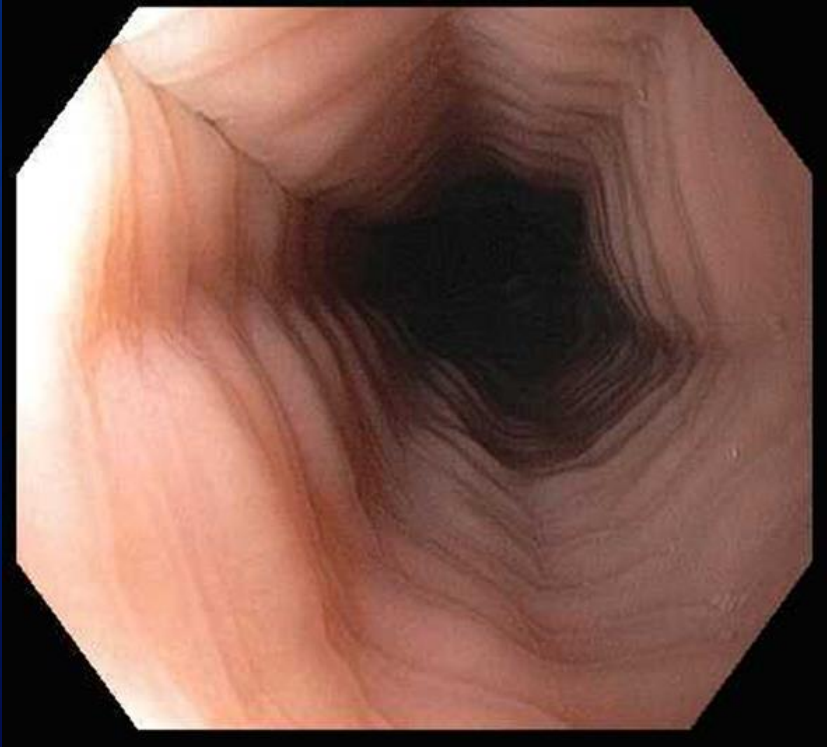
Aging of the Gastrointestinal System

- Acid reflux
- Motility disorders

MFS/RD Gastrointestinal System

- Acid reflux
- Motility disorders

Gastrointestinal

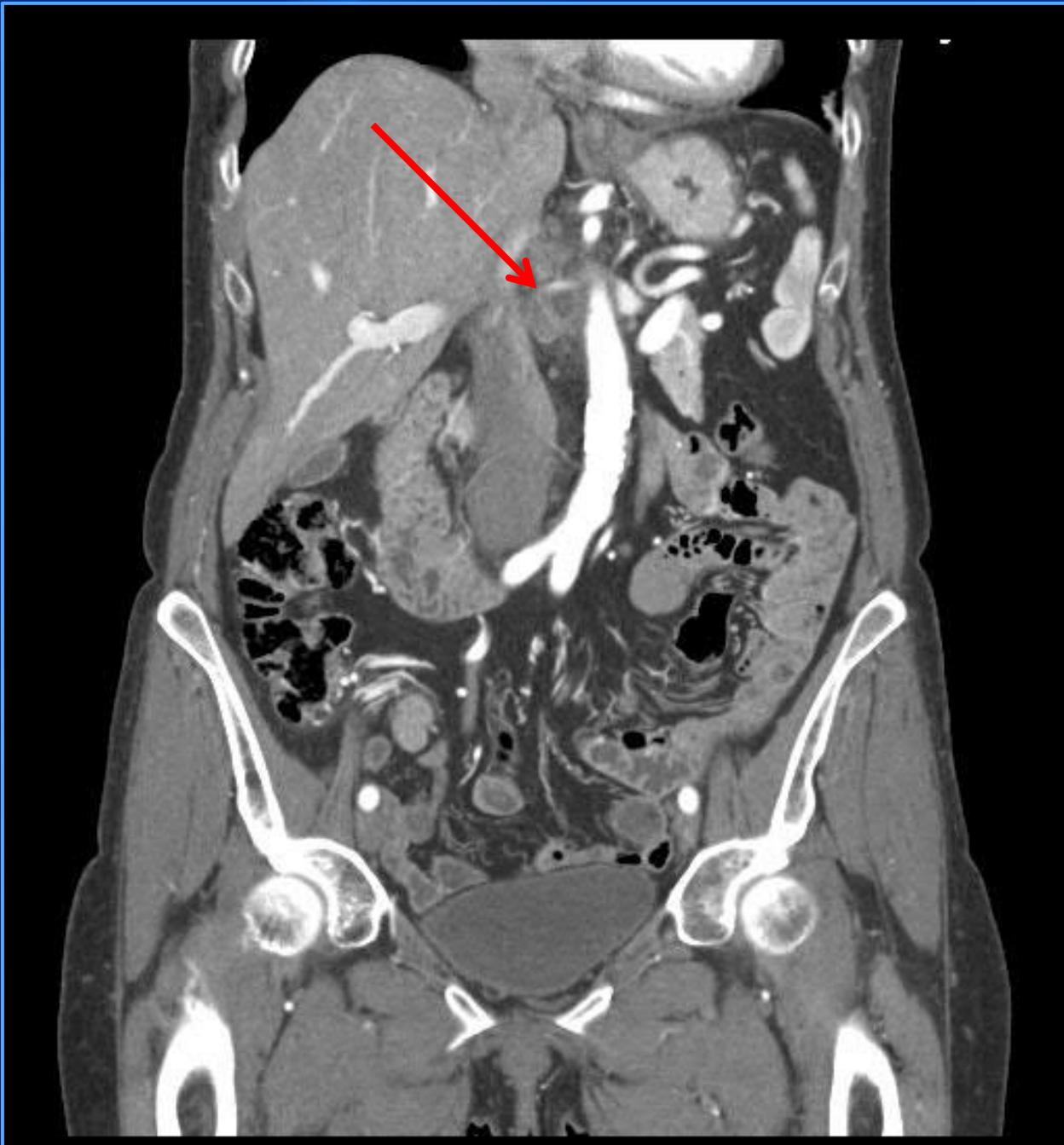


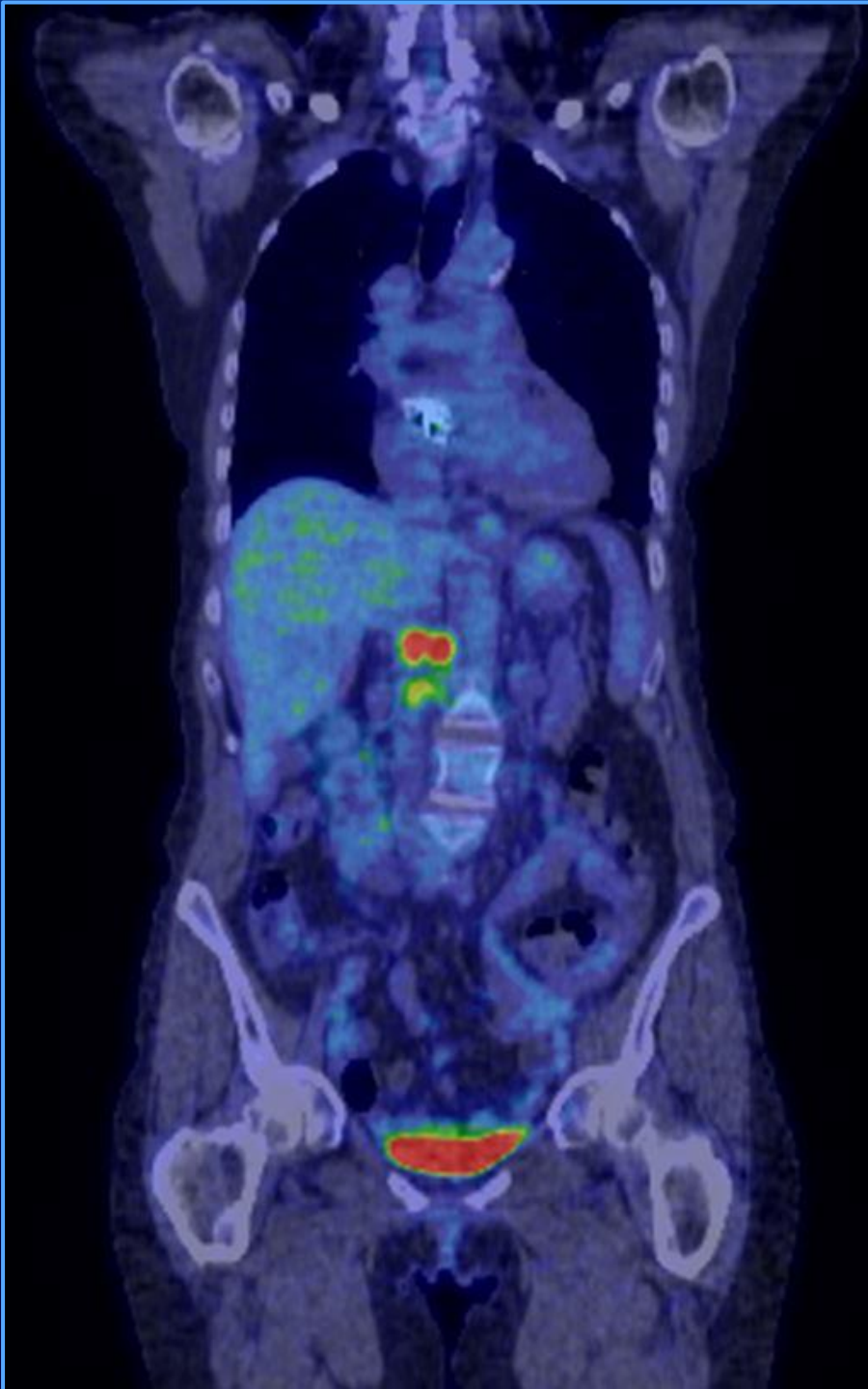
Aging of the Hematologic System

- Cell lines
 - Red blood cells
 - White blood cells
 - Platelets
- Most changes in blood counts are reactive and not primary bone marrow diseases
- Bone marrow malignancies

MFS/RD Hematologic System

- Eosinophilic disorders in LDS

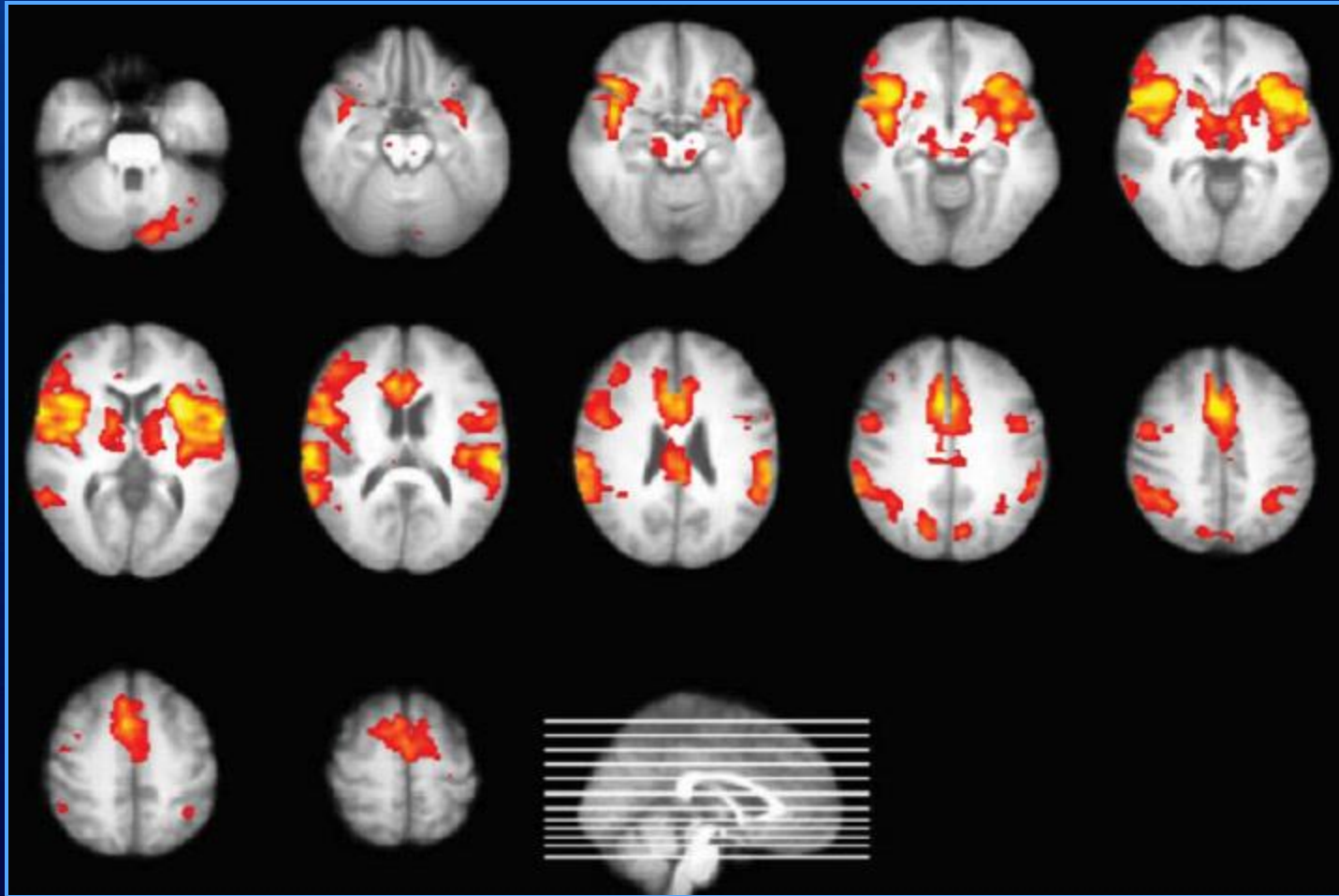




Aging and Mental Health

- Depression
- Anxiety
- Chronic pain
- Loneliness

Chronic Pain



MFS/RD Mental Health

- Depression
- Anxiety
- Chronic pain
- Disability

Medical Care

- Self care
- Primary care
- Specialty care

Exercise and Physical Activity in Marfan Syndrome and Related Disorders

Alan C. Braverman, MD

www.marfan.org

Medications

- Older people take 2 – 9 medications
- Issues
 - Altered drug processing
 - Drug-drug interactions
 - Drug-disease interactions
 - Supplements

The prevalence of obstructive sleep apnoea and its association with aortic dilatation in Marfan's syndrome

P Wordsworth,³ C Forfar,⁴ J R Stradling¹

CLINICAL STUDIES

and clinical impact of obesity in Marfan syndrome

nji T Yetman MD¹, Brian W McCrindle MD MPH²

Original article

Bone mineral density in Marfan syndrome. A large

Bertrand Moura^a, Florence Tubach^a, Moana Sulpice^a, Catherine Mouton-Rieu², Christine Muti^b, Bertrand Chevallier^d, Yasmine Oudiz^c, and the Multidisciplinary Marfan Syndrome Study Group

Psychology, Health & Medicine, May 2007; 12(3): 305-313

and visceral findings in patients with Marfan syndrome: a cross-sectional study

PhD^{2,3}, and Harold I. Litt, MD, PhD¹

Guillaume Jondeau, Jean-Marie Le Parc^{a,*}

Fatigue, cognitive functioning and quality of life in Marfan syndrome, a pilot study

SVEND RAND-HENTZ, HENRIK HOLM, and ARNSTEIN

Determinants of Quality of Life in Marfan Syndrome

PAOLO FUSAR-POLI, M.D., CATHERINE KLERSY, M.D., FRANCESCA STRAMESI, M.D., ALIRIA CALLEGARI, ELOISA ARBUSTINI, M.D., PH.D., PIERLUIGI POLITI, M.D., PH.D.

Original Article

Living with Marfan syndrome III. Quality of life and reproductive planning

Clin Genet 2002; 62: 110-120
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CLINICAL GENETICS
0009-9163

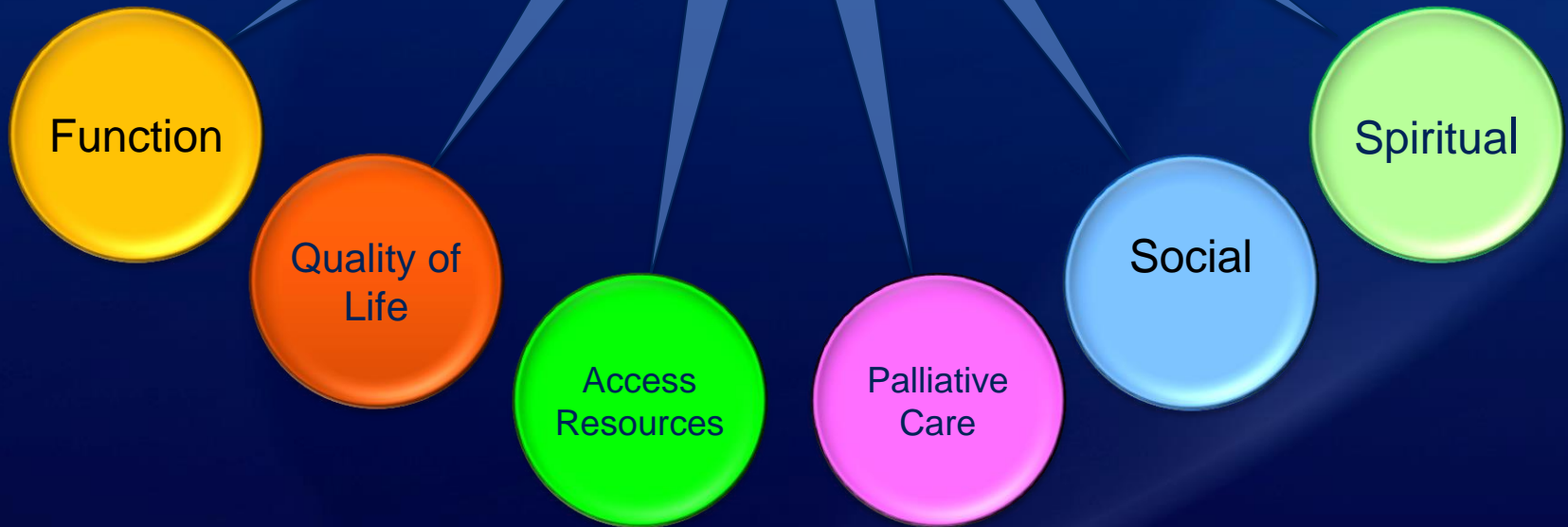
Decisions in Medical Care

- Evidence based
- Practice guidelines
- Clinical judgment
- Collaborative

Team Care



Geriatric Care





Canadian Marfan Association

Supporting Marfan Syndrome and Related Connective Tissue & Cardiovascular Disorders



EDUCATION • SUPPORT • RESEARCH

Helping Marfan patients since 1986



THE **MARFAN** FOUNDATION

Know the signs. Fight for victory.

Take Home Points

- Research is providing insights into aging
- Leading causes of death → CV disease and cancer
- Life expectancy in Marfan syndrome is now near normal.
- Everyone needs team-based primary care appropriate for the stage of life
- There are special concerns with Marfan syndrome and aging

Questions or Comments?

bowen.juan@mayo.edu