2019 MID-ATLANTIC
CONFERENCE

9th ANNUAL CURRENT CONCEPTS IN

VASCULAR THERAPIES

2019

Hilton Virginia Beach Oceanfront
Virginia Beach, Virginia

MAY 2-4

2019 MID-ATLANTIC
CONFERENCE

9th ANNUAL CURRENT CONCEPTS IN
VASCULAR THERAPIES

2019

C Scott McEnroe, MD, FACS

Medical Director
Vein Center of Virginia

Sentara Vascular Specialists
Sentara Medical Group

May 3, 2019

Klippel-Trenaunay, Park-Weber, Nutcracker Syndromes

Klippel-Trenaunay Syndrome

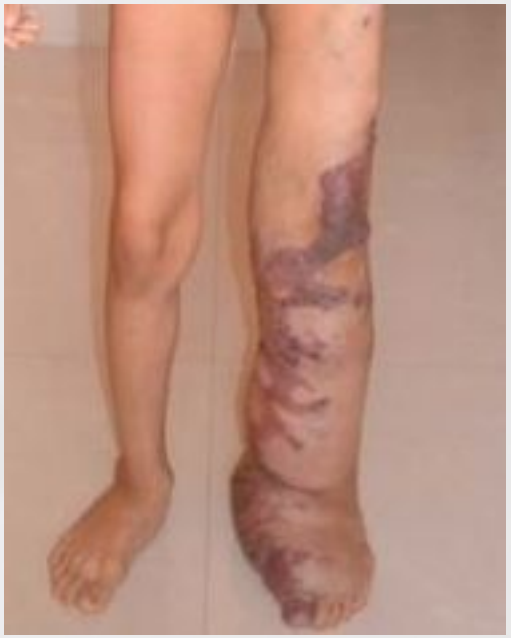
- **Overview**
  - Rare, congenital disorder, characterized by complex, low flow vascular malformations
  - 1:100,000, equal male/female
  - Not inherited
  - Genetic mutation in PIK3CA gene during early cell division
    - Responsible for blood vessel, soft tissue, bone development
      - Results in overgrowth

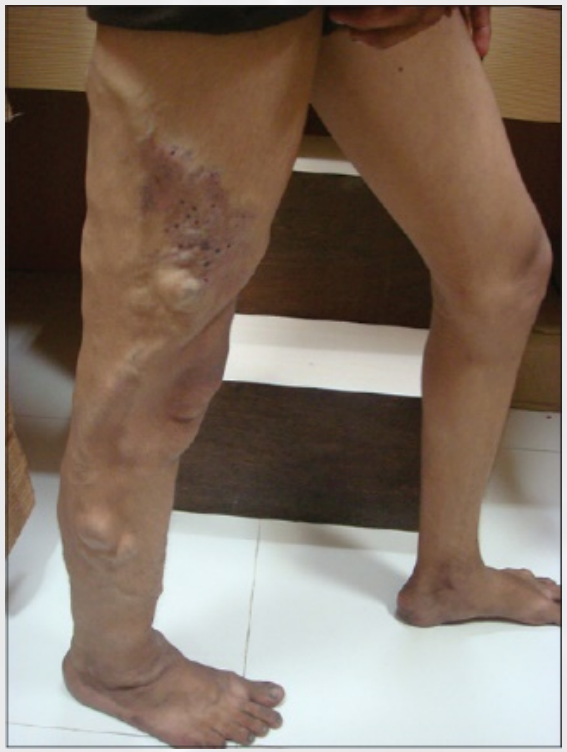
Klippel-Trenaunay Syndrome

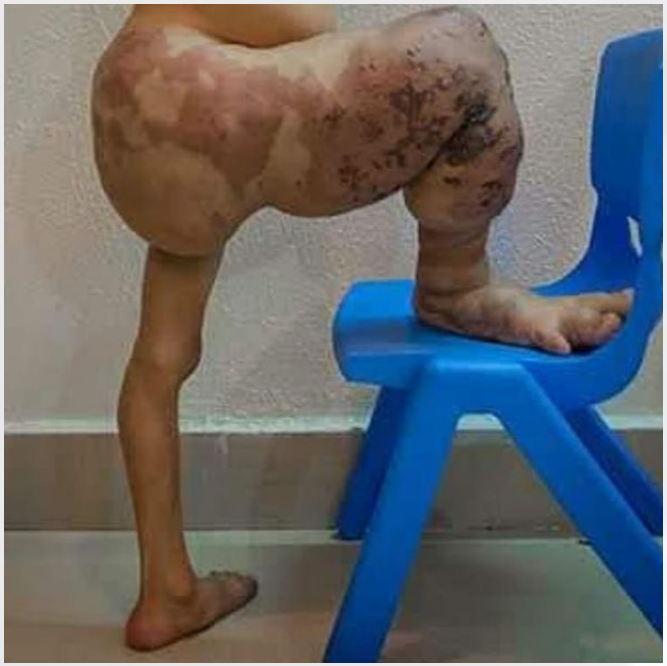
- **Characteristic Features**
  - Port-wine stains: flat, pale pink to deep maroon
  - Venous malformations:
    - 90 % one LE involved only
    - Can involve UE's, abdomen, pelvis, viscera
    - Large varicose veins
    - *May* have aplasia of deep venous system
      - Results in overgrowth
  - Hypertrophy of bone, soft tissue: overgrowth of affected limb
  - Lymphatic system abnormalities

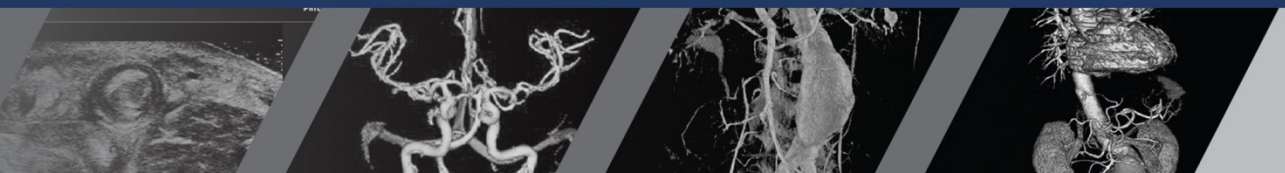
Klippel-Trenaunay Syndrome

- **Characteristic Features**









Klippel-Trenaunay Syndrome

- **Symptoms/Complications**
  - Port-wine stains: bleeding, portal for infection
  - Venous malformations
    - Pain, edema
    - Venous ulceration
    - Superficial phlebitis
    - DVT, PE
  - Overgrowth of affected limb
    - Pain, heaviness, difficulty ambulating, hip/spine disorders
  - Lymphatic system abnormalities
    - Edema, ulceration, cellulitis

Klippel-Trenaunay Syndrome

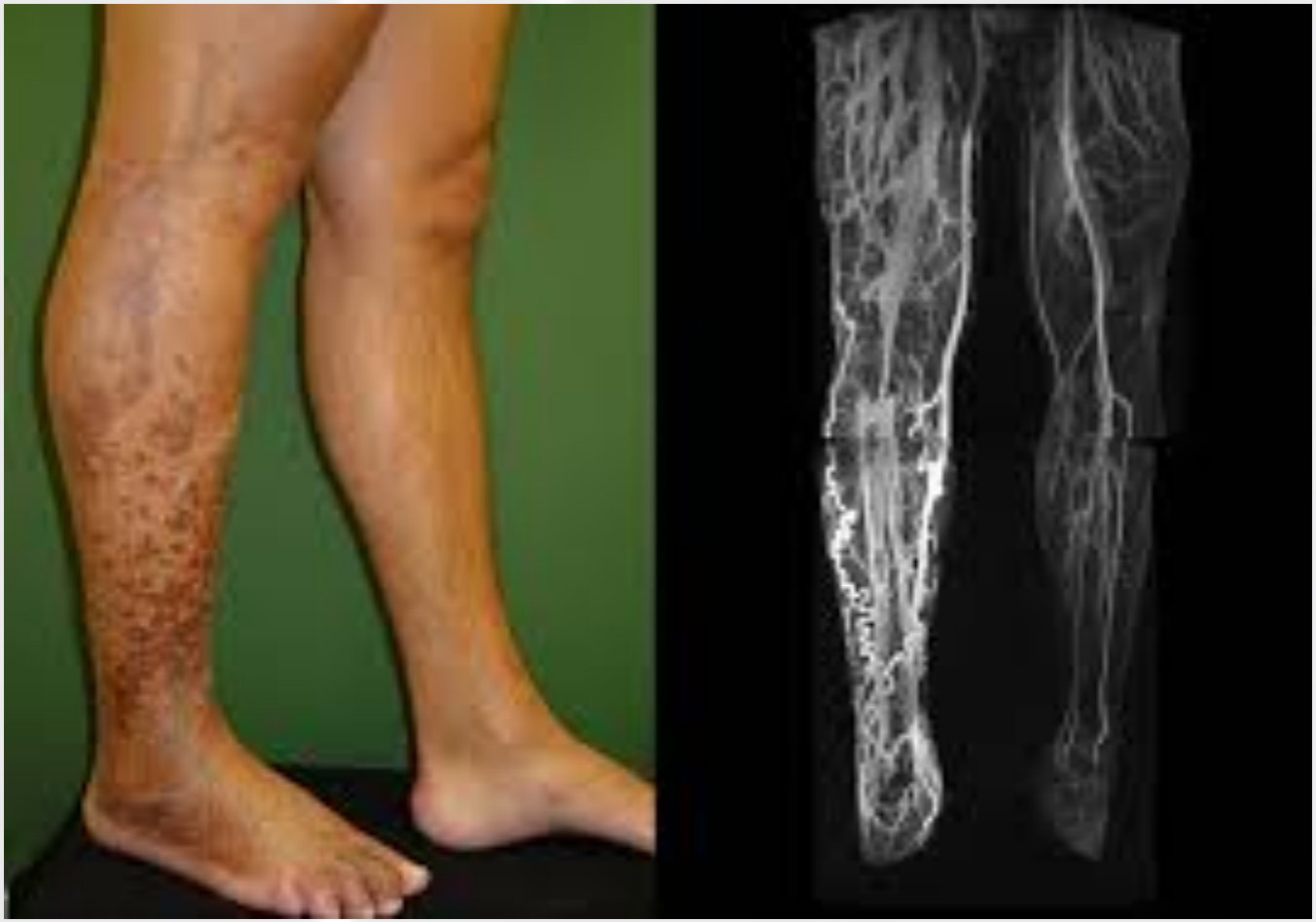
- **Presentation**
  - Venous insufficiency: 90 %
  - Limb hypertrophy: 90 %
  - Cutaneous hemangioma: 90 %
  - Pulmonary embolus: 30%
  - GI/vaginal bleeding, hematuria: 30 %
  - Coagulopathy: 7%
    - Hypercoagulable
    - DIC: stasis/platelet trapping

Klippel-Trenaunay Syndrome

- **Diagnosis**
  - Clinical exam: 2 or more signs
  - Duplex
  - MRI
  - CT
  - Venography
  - Arteriography
    - NORMAL
    - No arterial involvement

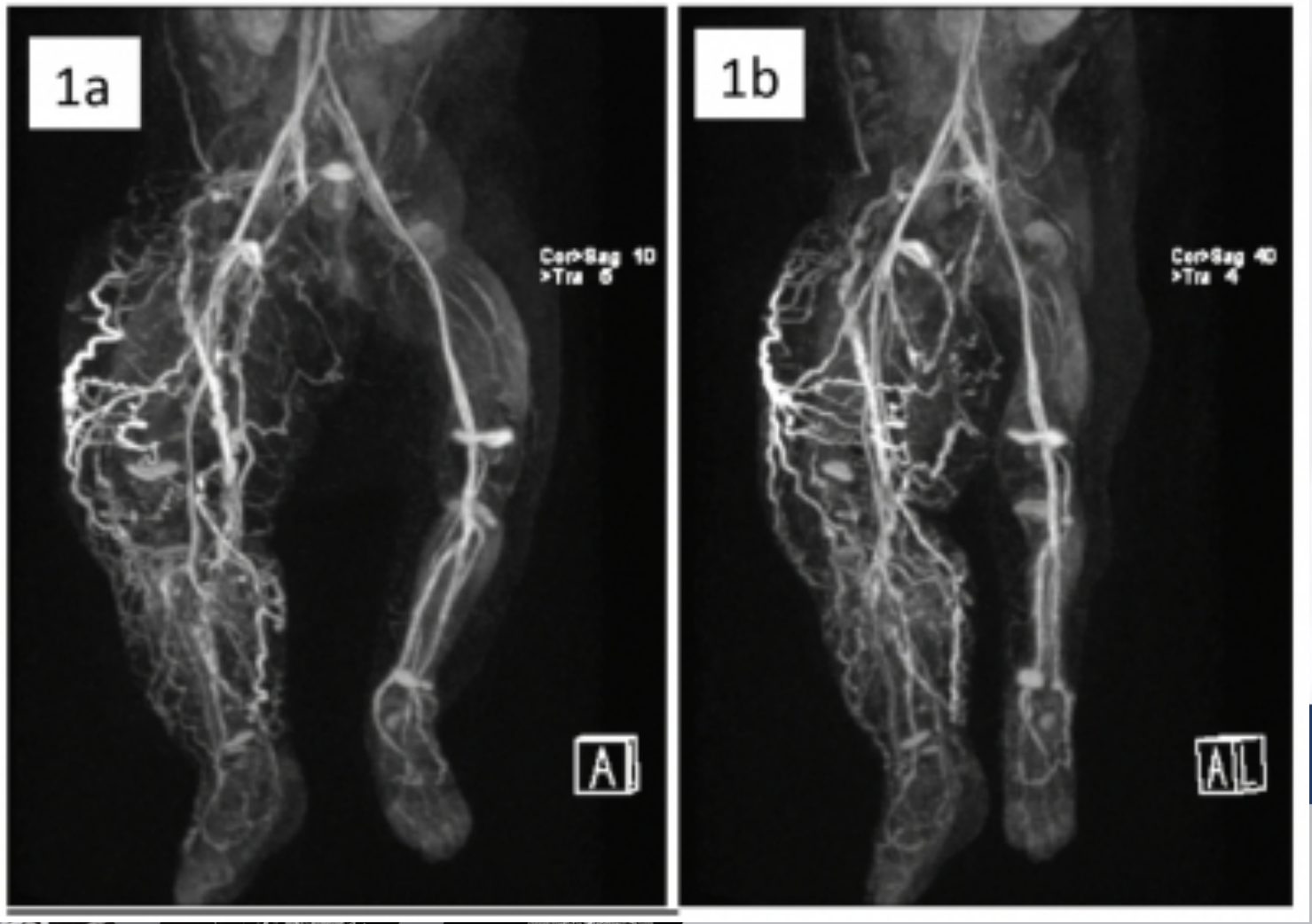
Klippel-Trenaunay Syndrome

- Diagnosis



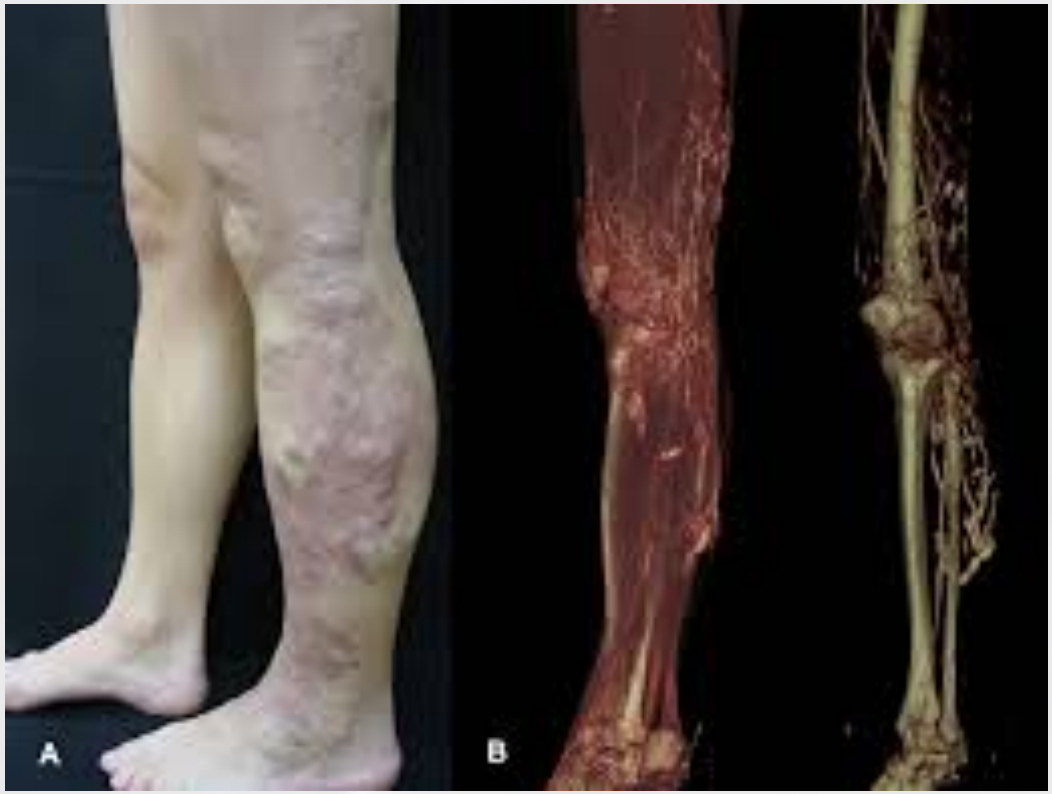
Klippel-Trenaunay Syndrome

- **Diagnosis**



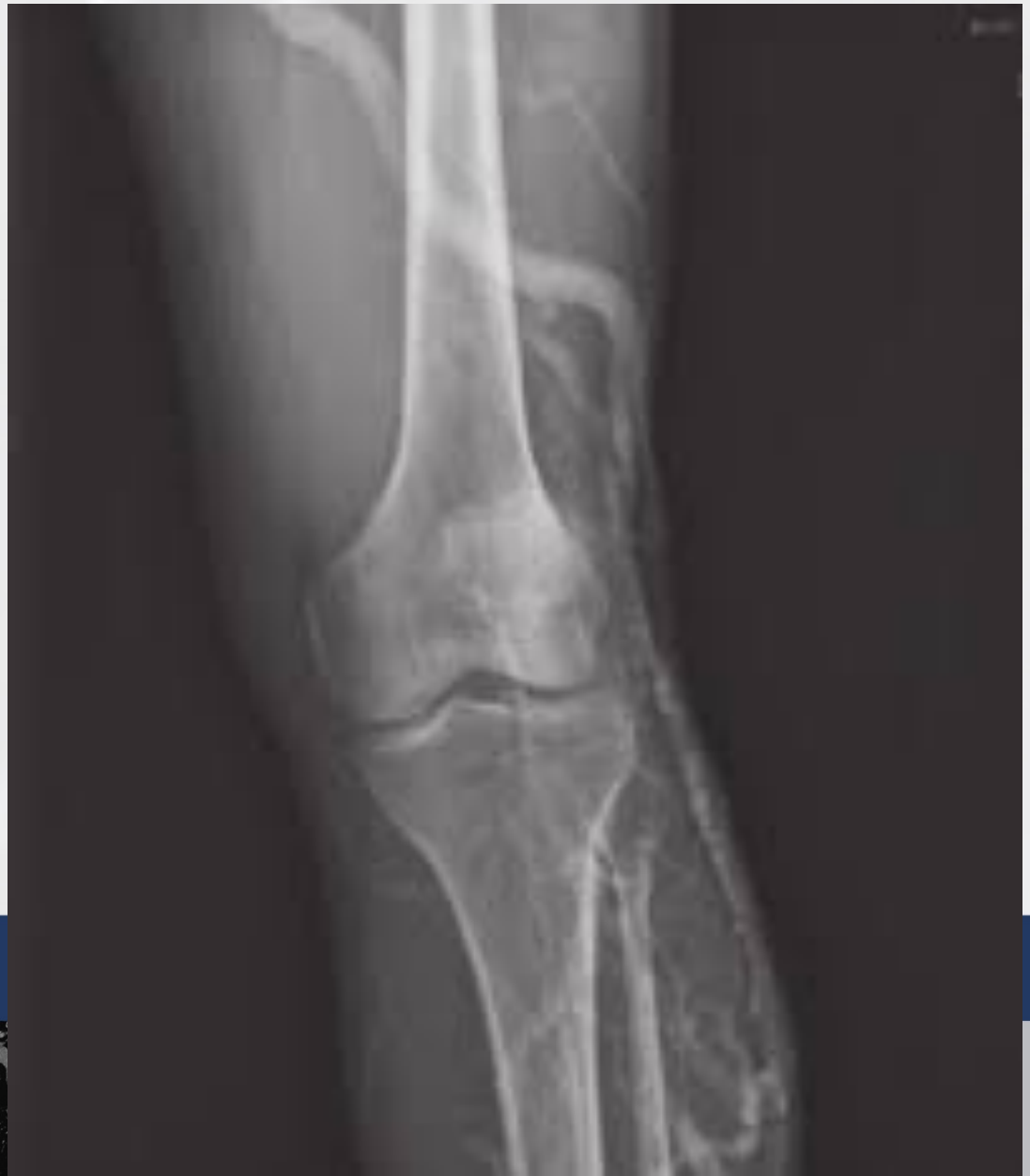
Klippel-Trenaunay Syndrome

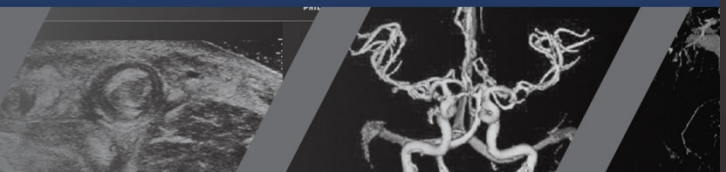
- **Diagnosis**



Klippel-Trenaunay Syndrome

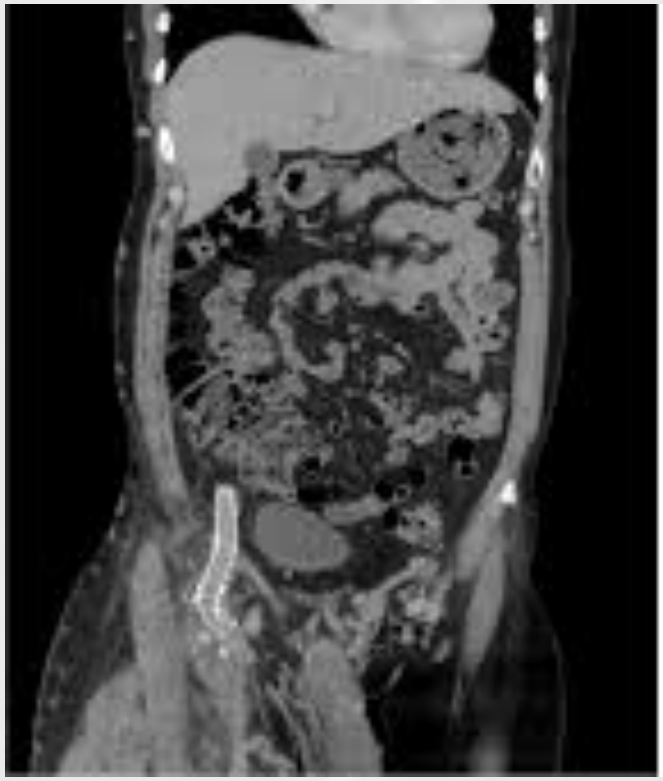
- Diagnosis

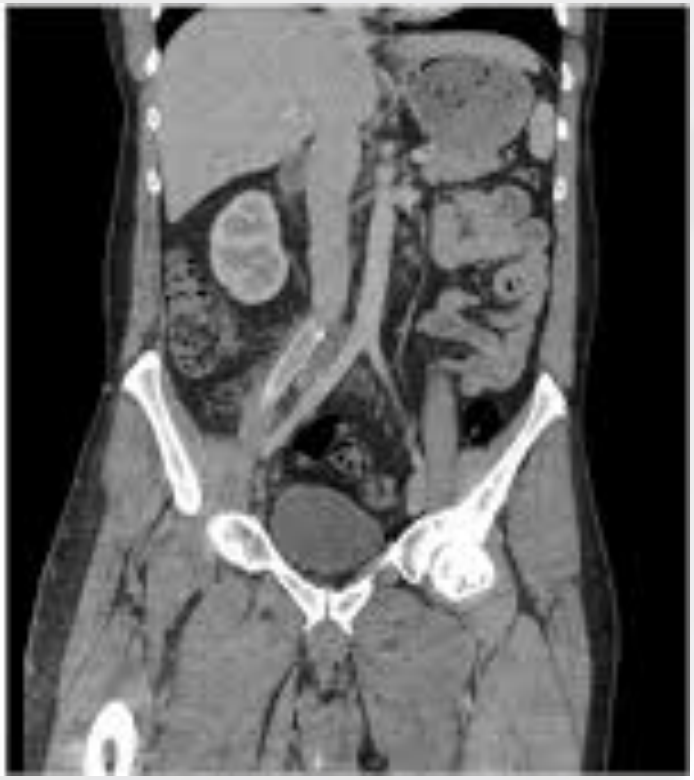




Klippel-Trenaunay Syndrome

- **Diagnosis**





Klippel-Trenaunay Syndrome

- **Treatment:** management of symptoms, complications
  - Compression: hose, Unna's Boot, pneumatic devices
  - Venous intervention: ablation, sclerotherapy, surgery
    - Confirm presence of deep venous system first
  - Manual Lymphatic Drainage
  - Orthotic shoes
  - Orthopedic intervention to disrupt growth plate
  - Laser, IPL
  - Immunosuppressive agents
  - NO supplemental hormones, BCP, close monitoring of pregnancy

Parkes-Weber Syndrome

- **Overview**
  - Rare, congenital disorder, characterized by complex, ***high flow*** capillary malformations and arteriovenous fistulas
  - Incidence unknown, but less common than KTS
  - Autosomal dominant pattern of inheritance
  - Genetic mutation in ***RASA1*** gene
    - Disrupts tightly regulated chemical signaling during development
    - Mechanism causing abnormalities not well understood

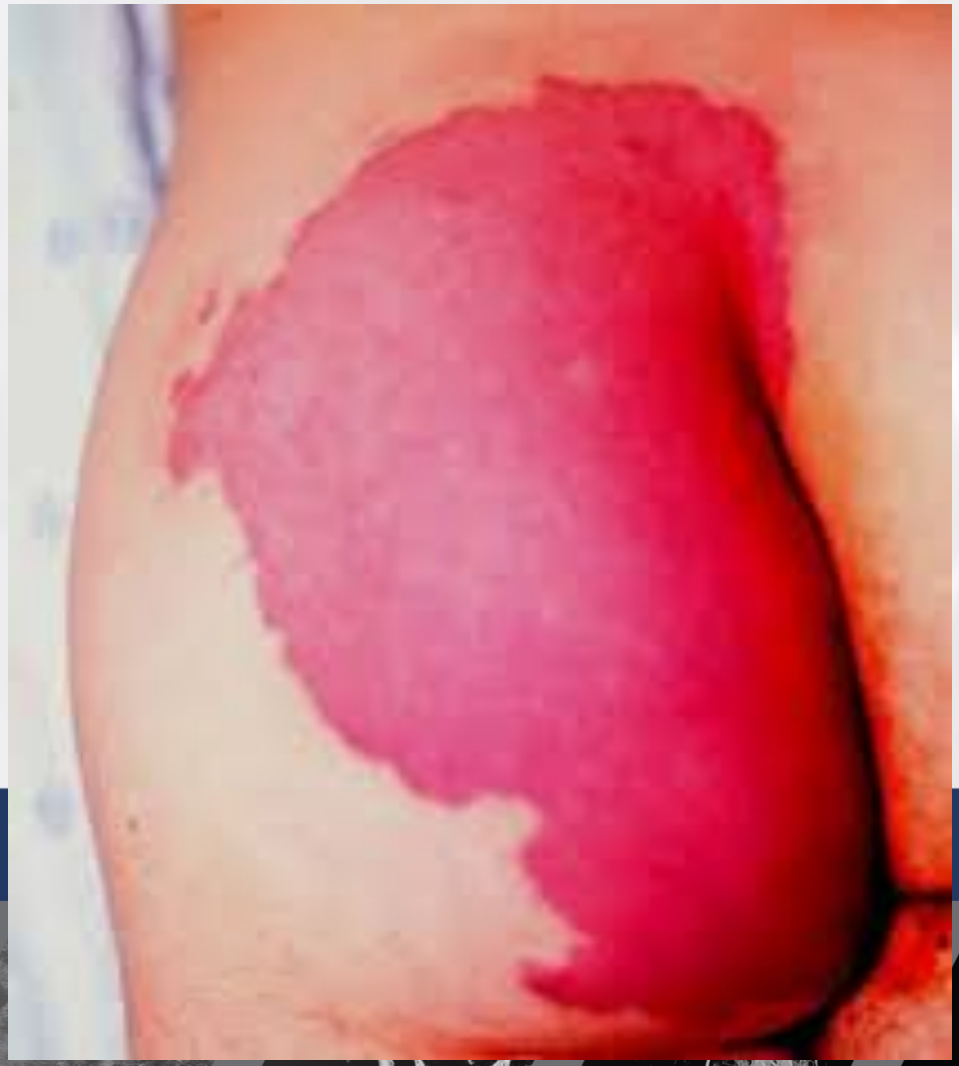
Parkes-Weber Syndrome

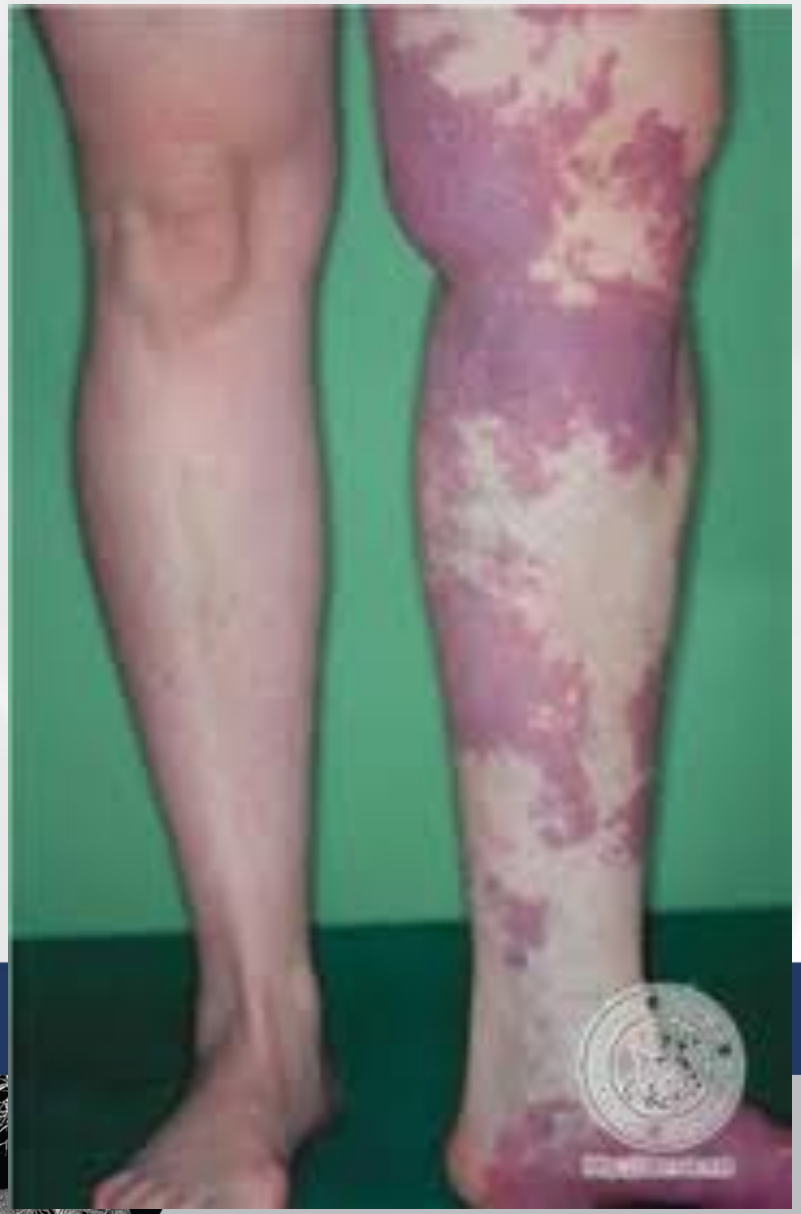
- **Characteristic Features**
  - Venous malformations
  - Lymphatic malformations
  - Hypertrophy of bone, soft tissue: overgrowth of affected limb
  - LE's most commonly involved, UE's more commonly than KTS
  - ***Multiple arteriovenous fistulas:*** occur anywhere
  - ***Capillary malformations: capillary arteriovenous fistula***

    -Port-wine stains: warm, bright red

Parkes-Weber Syndrome

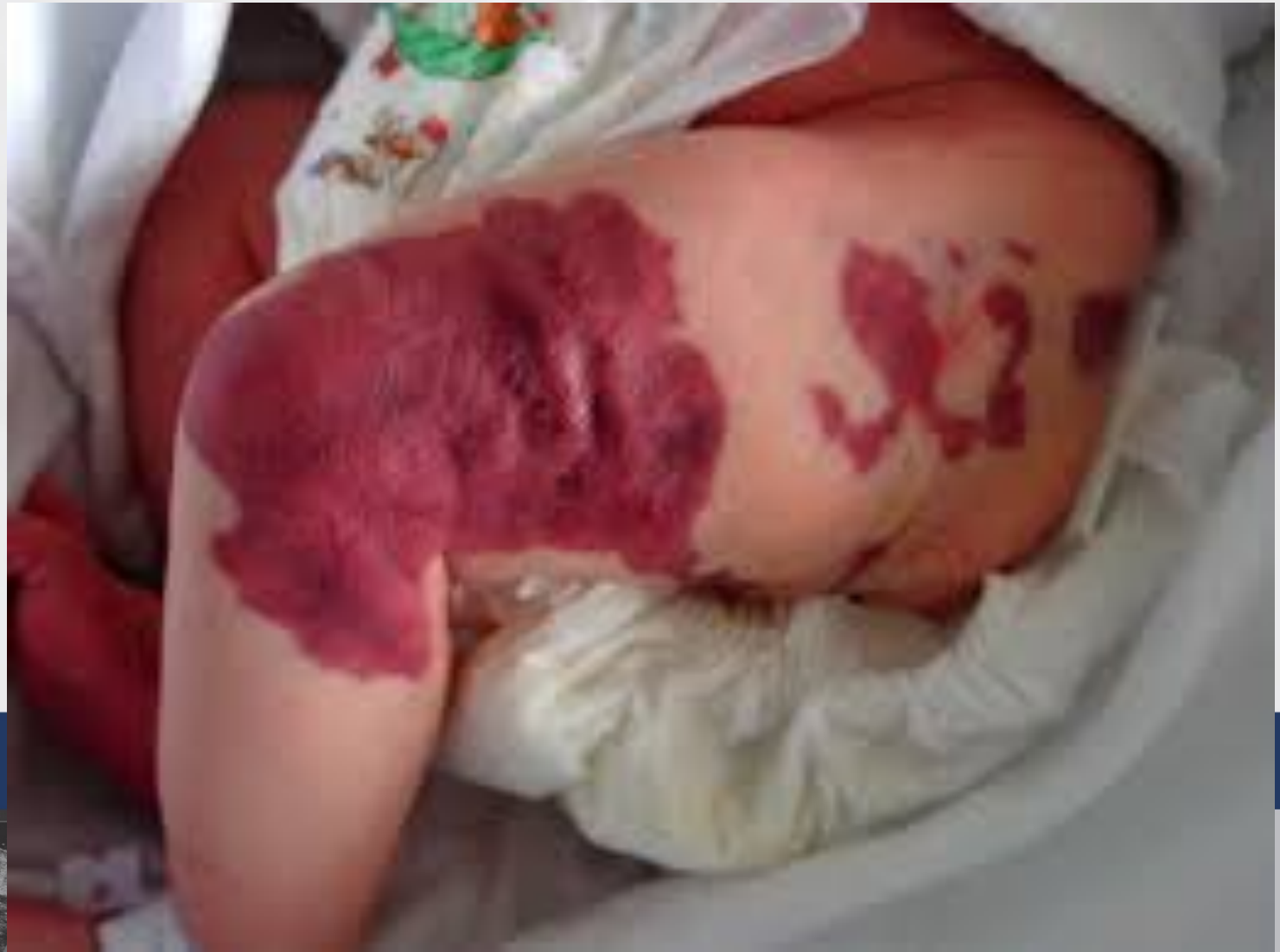
- **Characteristic Features**





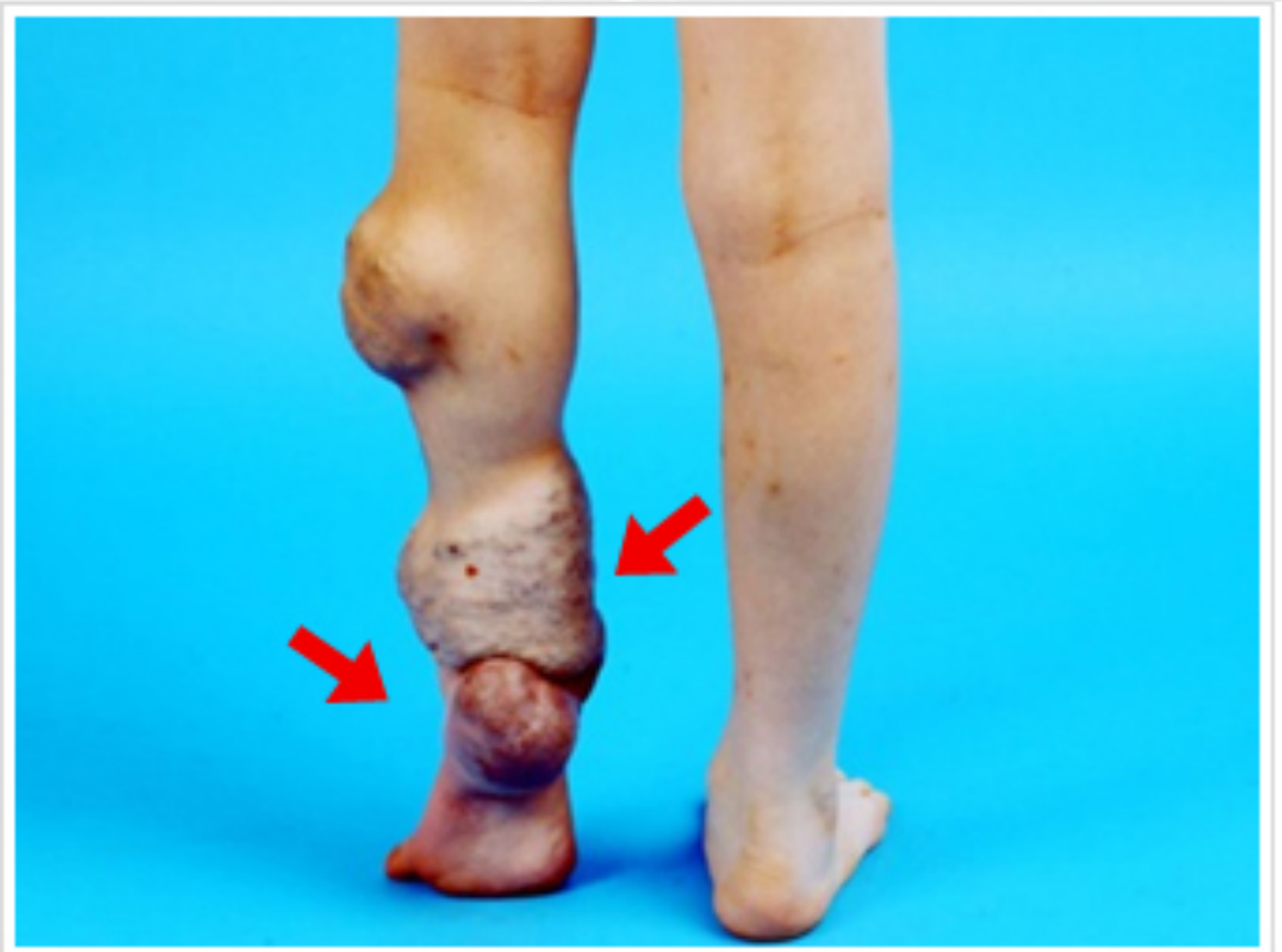
Parkes-Weber Syndrome

- **Characteristic Features**



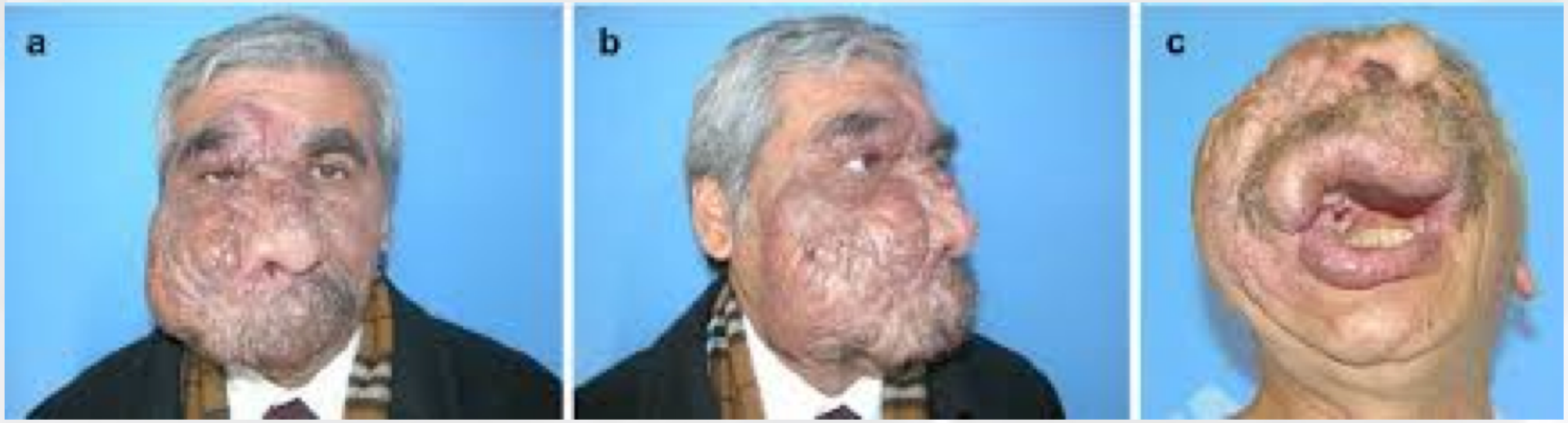
Parkes-Weber Syndrome

- **Characteristic Features**



Parkes-Weber Syndrome

- **Characteristic Features**



Parkes-Weber Syndrome

- **Symptoms/Complications: more serious than KTS**
  - Port-wine stains: more frequent cutaneous bleeding(AVF's)
  - Large arteriovenous fistulas
    - Pain, edema, ulceration
    - CHF
  - Overgrowth of affected limb
    - Pain, heaviness, difficulty ambulating, hip/spine disorders
  - Lymphatic system abnormalities
    - Edema, ulceration, cellulitis

Parkes-Weber Syndrome

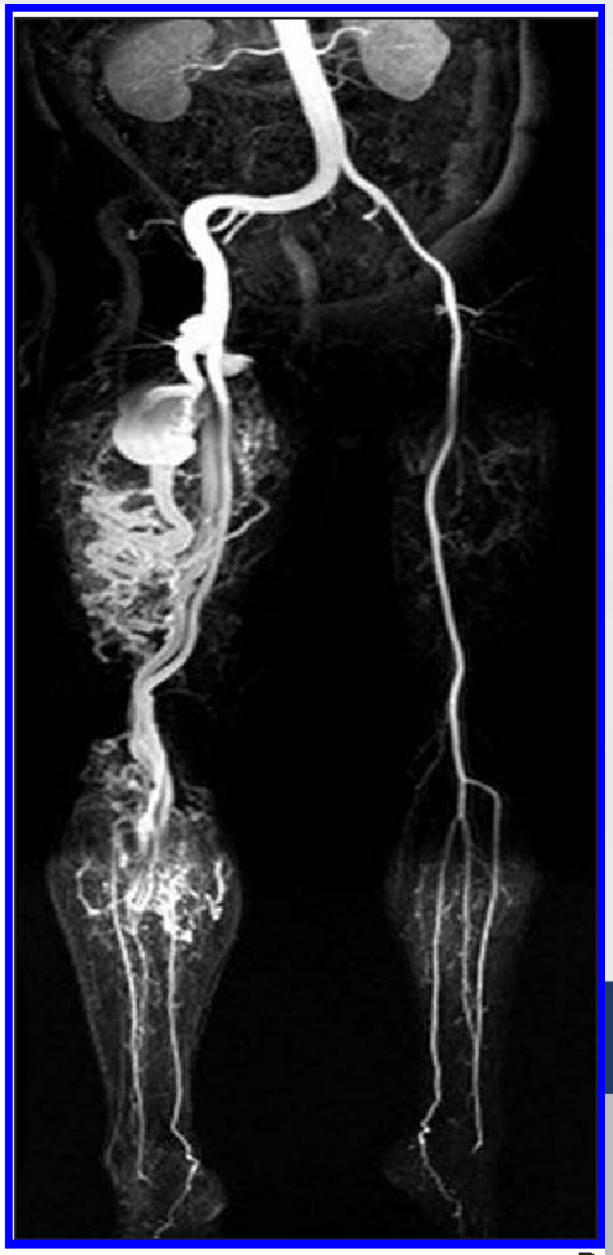
- **Presentation**
  - Frequent bleeding: 90 %
  - Limb hypertrophy: 90 %
  - Port-wine stains: 90 %
  - Arteriovenous fistulas: 90%
  - Varicose veins: 30 %
  - CHF: 7.5%

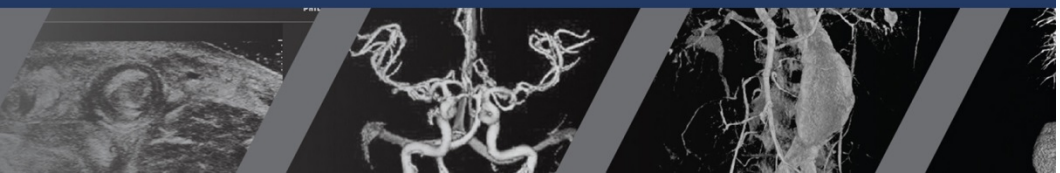
Parkes-Weber Syndrome

- **Diagnosis**
  - Genetic testing ***RASA1***
  - Clinical exam
  - Duplex
  - MRI
  - CT
  - ***Cardiac echo:*** ***high output failure***
  - Venography
  - ***Arteriography:*** ***abnormal, AVF's***

Parkes-Weber Syndrome

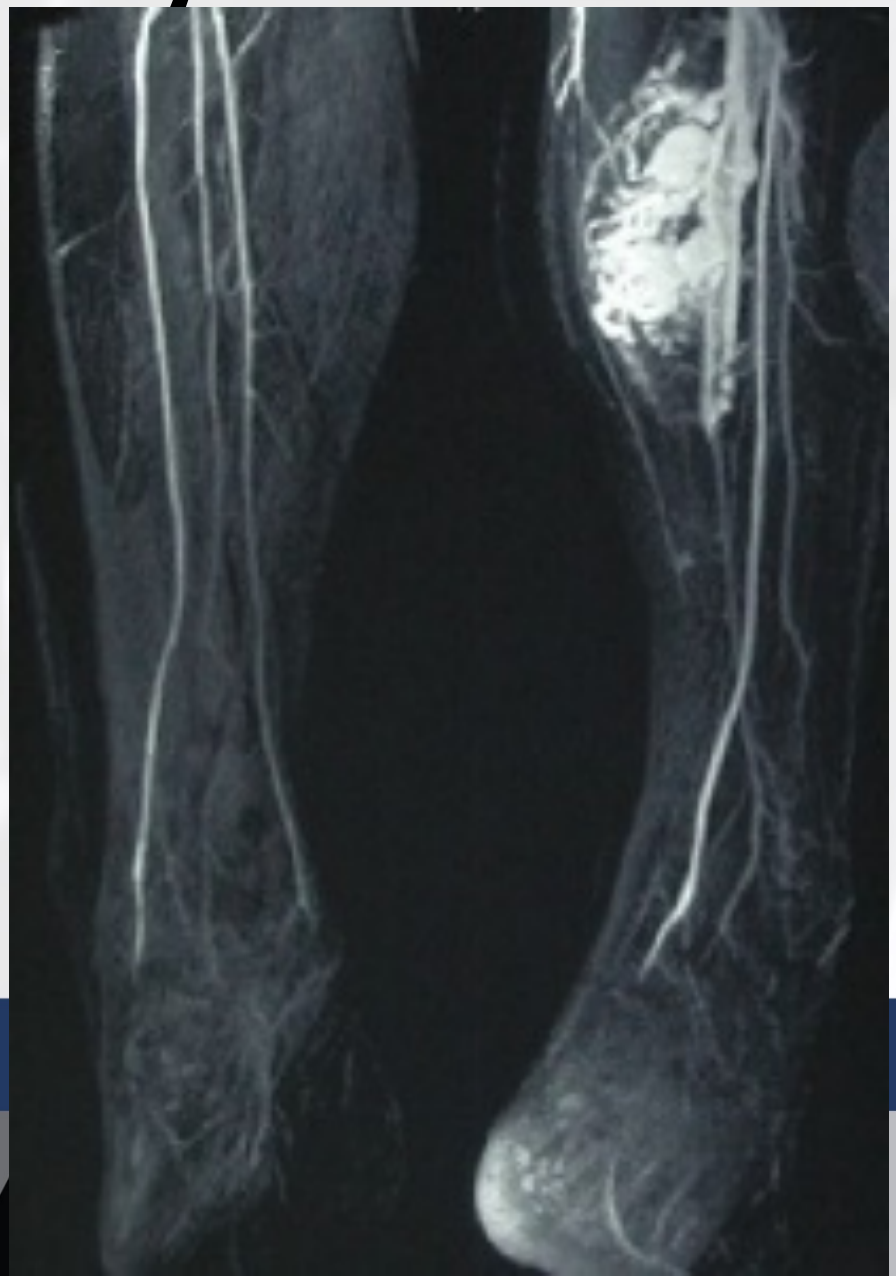
- **Characteristic Features**

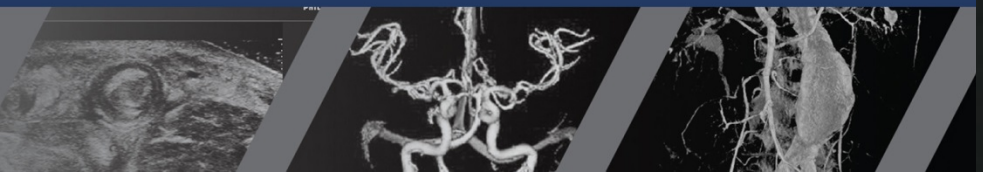




Parkes-Weber Syndrome

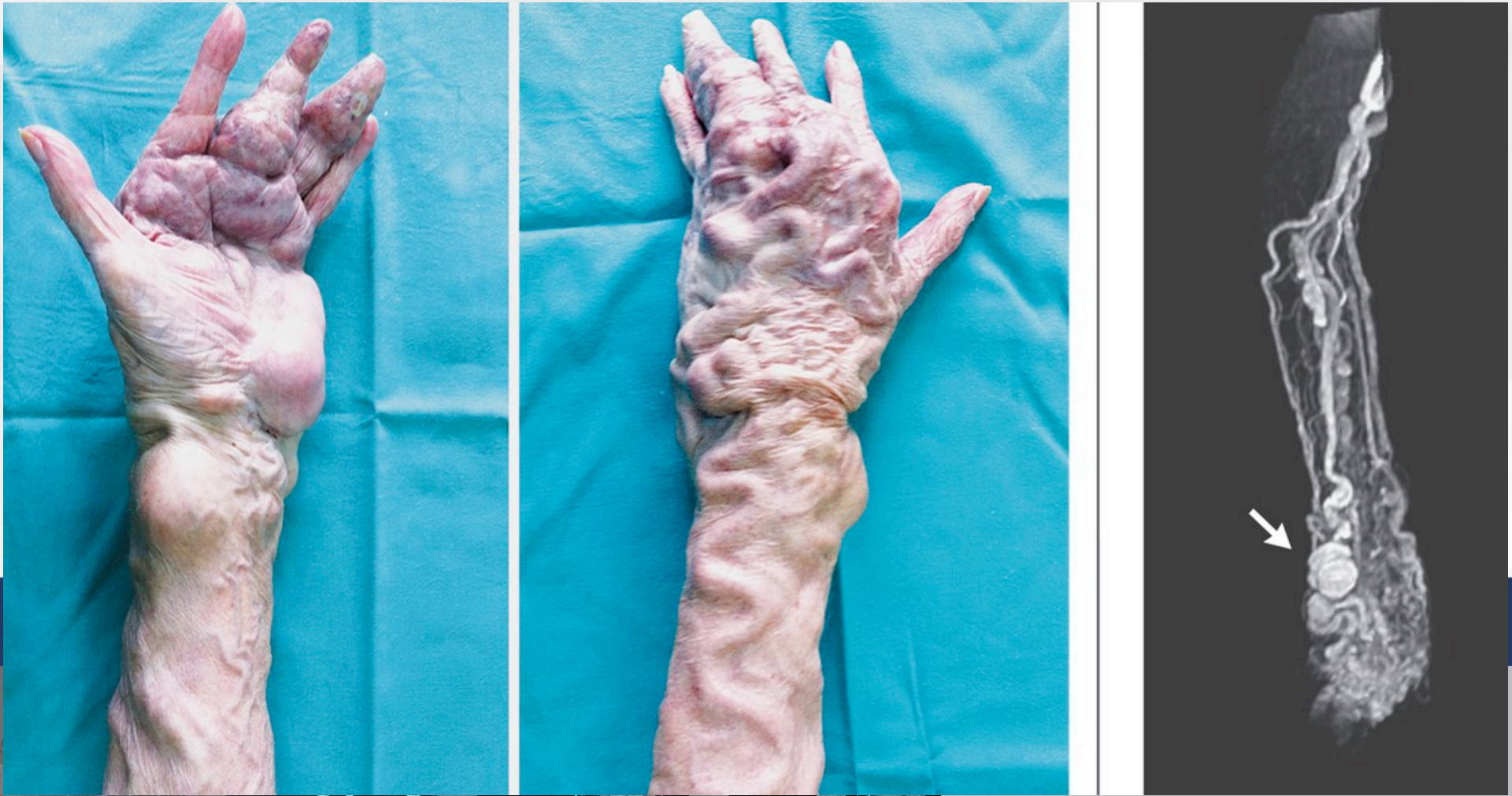
- **Characteristic Features**





Parkes-Weber Syndrome

- **Characteristic Features**



Parkes-Weber Syndrome

- **Treatment:** management of symptoms, complications
  - Compression: hose, Unna's Boot, pneumatic devices
  - Venous intervention: ablation, sclerotherapy, surgery
  - Arteriovenous fistulas: embolization, surgical interruption
  - Manual Lymphatic Drainage
  - Orthotic shoes
  - Orthopedic intervention to disrupt growth plate
  - Laser, IPL
  - Immunosuppressive agents
  - Cardiac evaluation: assess, manage high output CHF

Nutcracker Syndrome

- **Overview**
  - Rare disorder, characterized by compression of the left renal vein(LRV) between the aorta and superior mesenteric artery(SMA)
  - Incidence unknown
  - Associated with abnormal formation of LRV during 6-8th week of gestation and abnormal angulation of SMA
  - Symptomatic presentation peaks 2nd-3rd decade
  - Slightly more common in females

Nutcracker Syndrome

- **Characteristic Features**
  - Left renal vein hypertension
  - Left Gonadal vein dilatation, valvular incompetency
  - Pelvic varicosities

Nutcracker Syndrome

- **Symptoms/Complications**
  - Pelvic Congestion Syndrome
  - Painful intercourse
  - Left flank pain
  - Left lower extremity swelling
  - Labial varicosities
  - Left sided varicocele
  - Hematuria
  - Orthostatic proteinuria
  - Renal insufficiency
  - Infertility

Nutcracker Syndrome

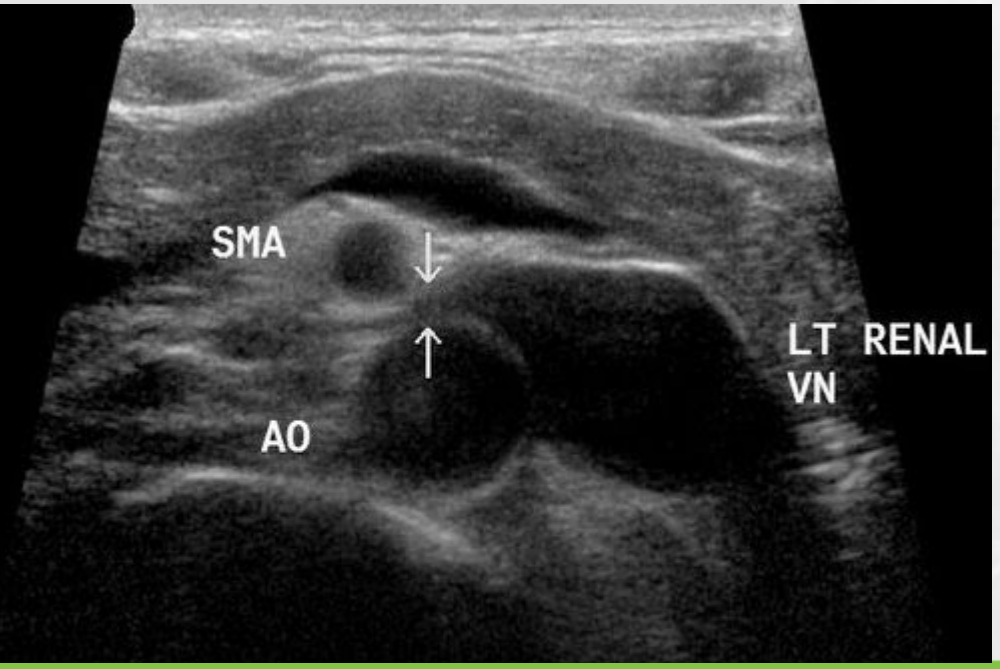
- **Presentation**
  - Incidental: “ Nutcracker Phenomenon “
  - Hematuria: micro, gross
  - Pelvic/left flank pain: sitting, standing, walking
  - Pelvic varicosities
  - Vulvar varicosities
  - Varicoceles

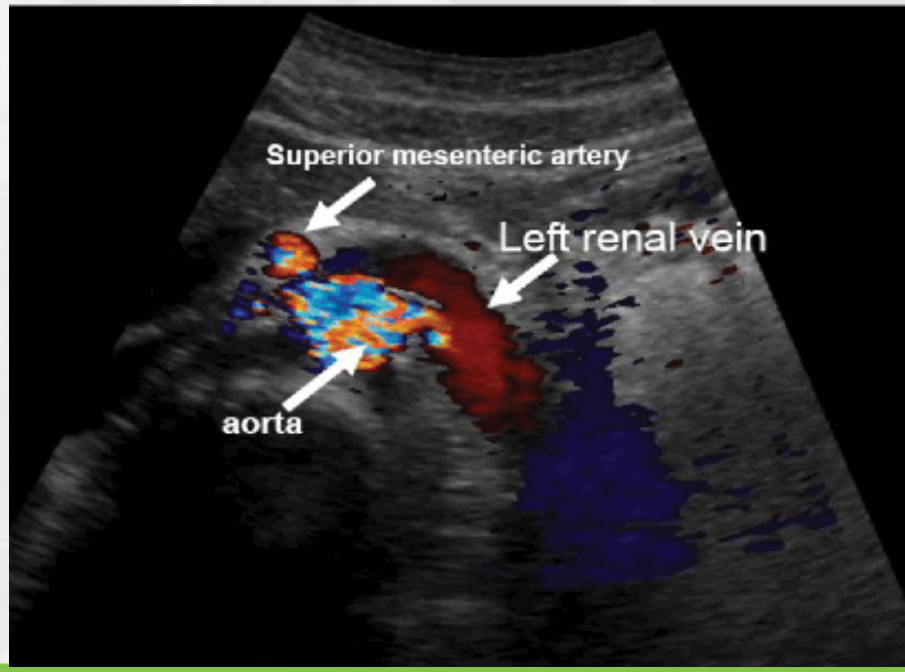
Nutcracker Syndrome

- **Diagnosis**
  - Exclude other etiologies for pelvic pain and hematuria
  - PVL: IVC duplex, LE venous insufficiency duplex
  - CTV/MRV
  - Venography with IVUS

Nutcracker Syndrome

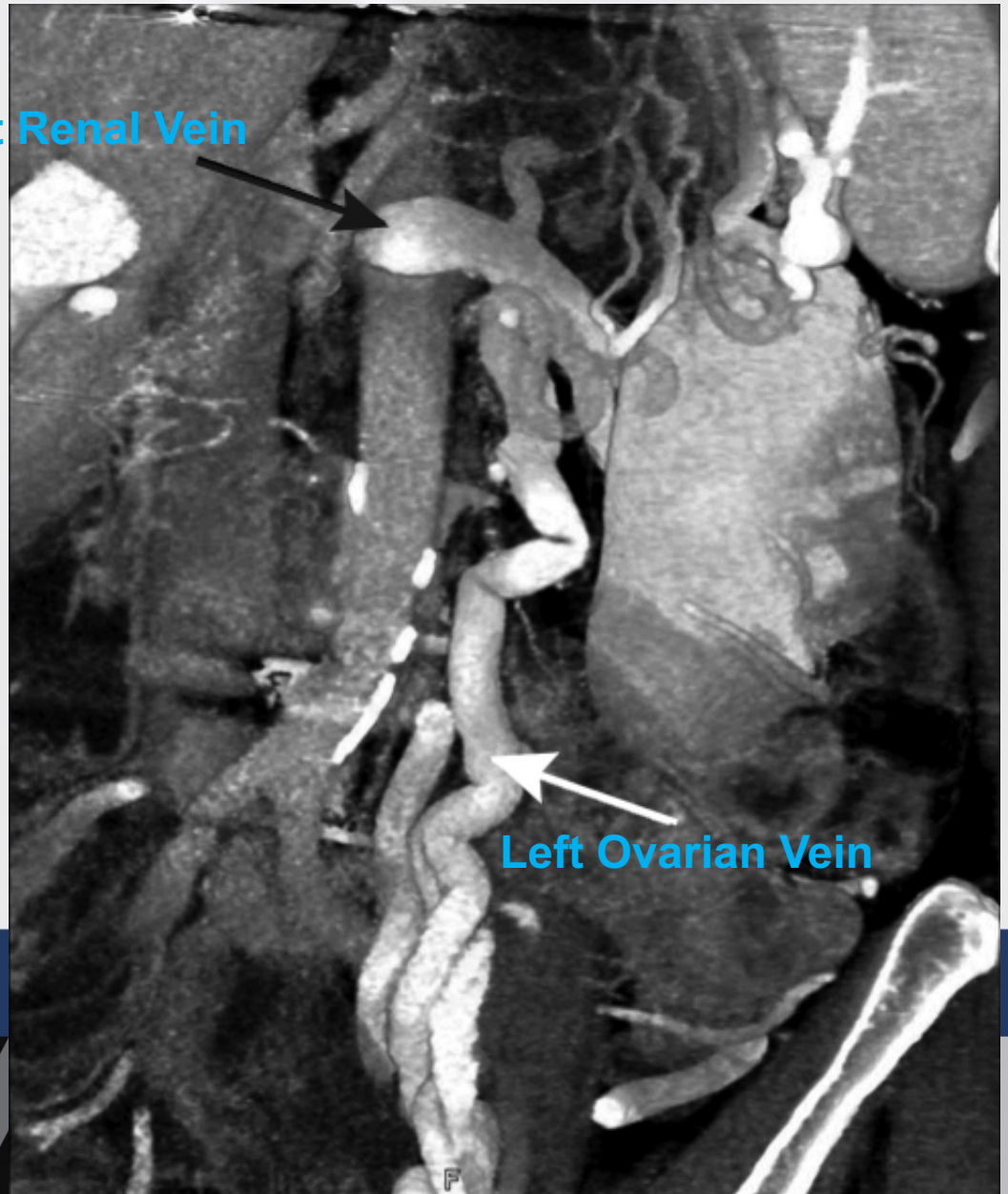
Diagnosis: IVC Duplex Scan

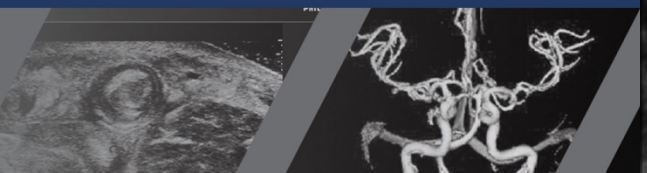




Nutcracker Syndrome

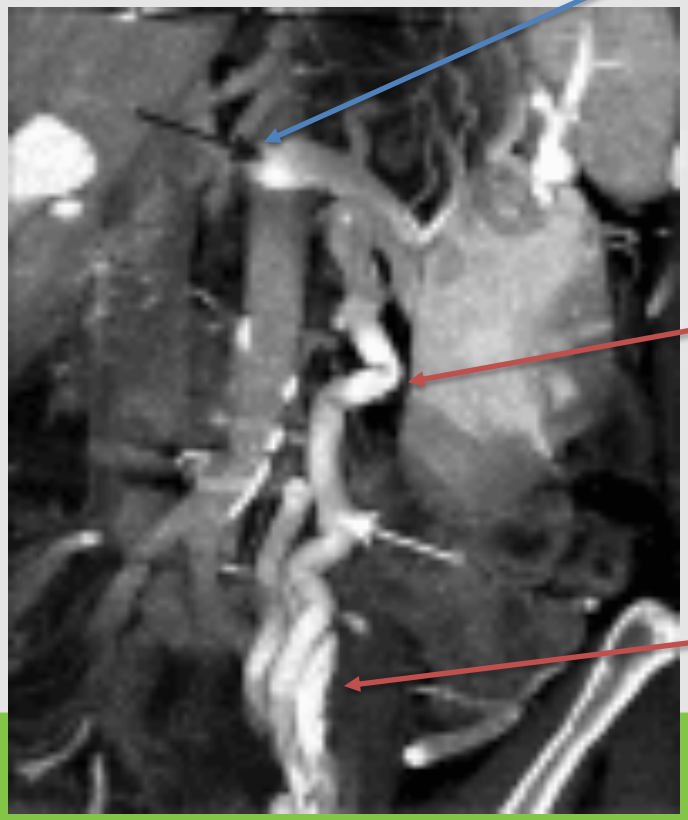
Diagnosis: MRV

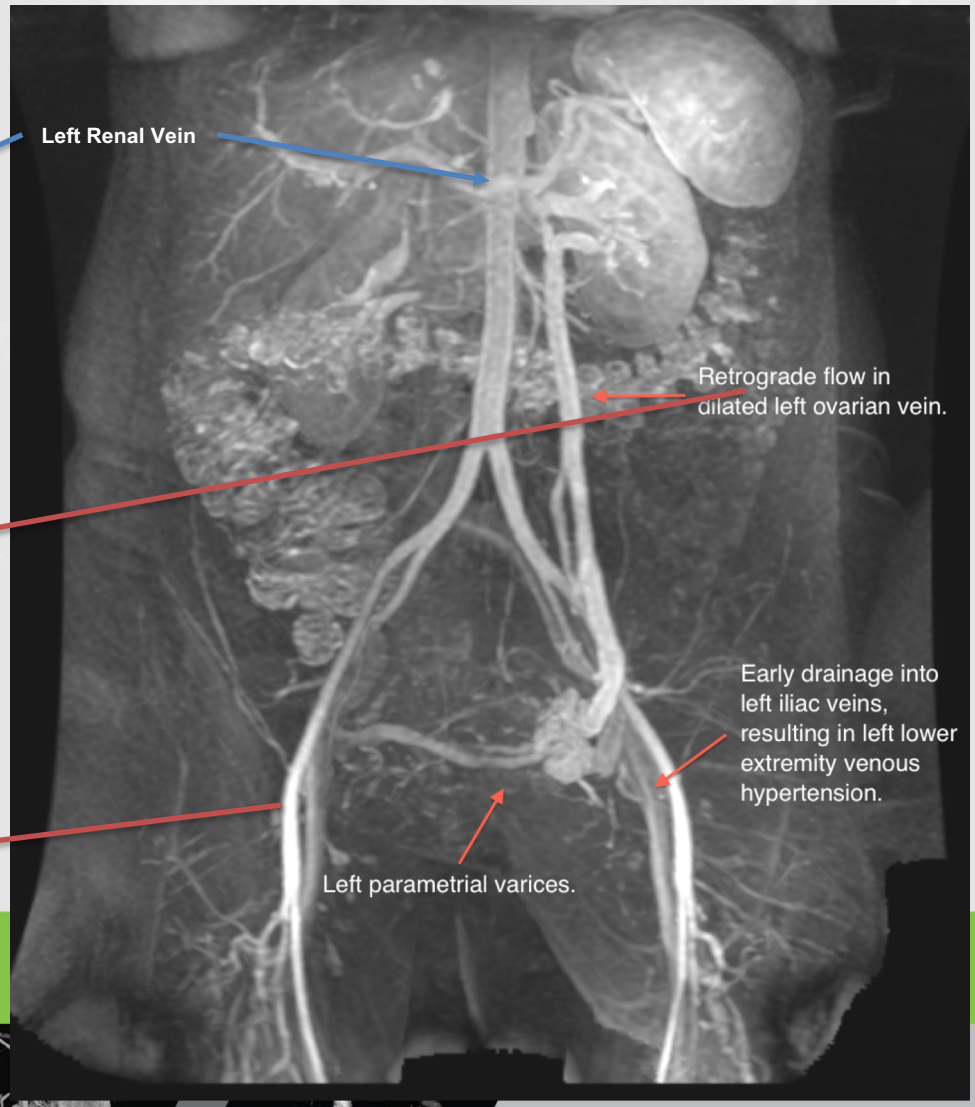




Nutcracker Syndrome

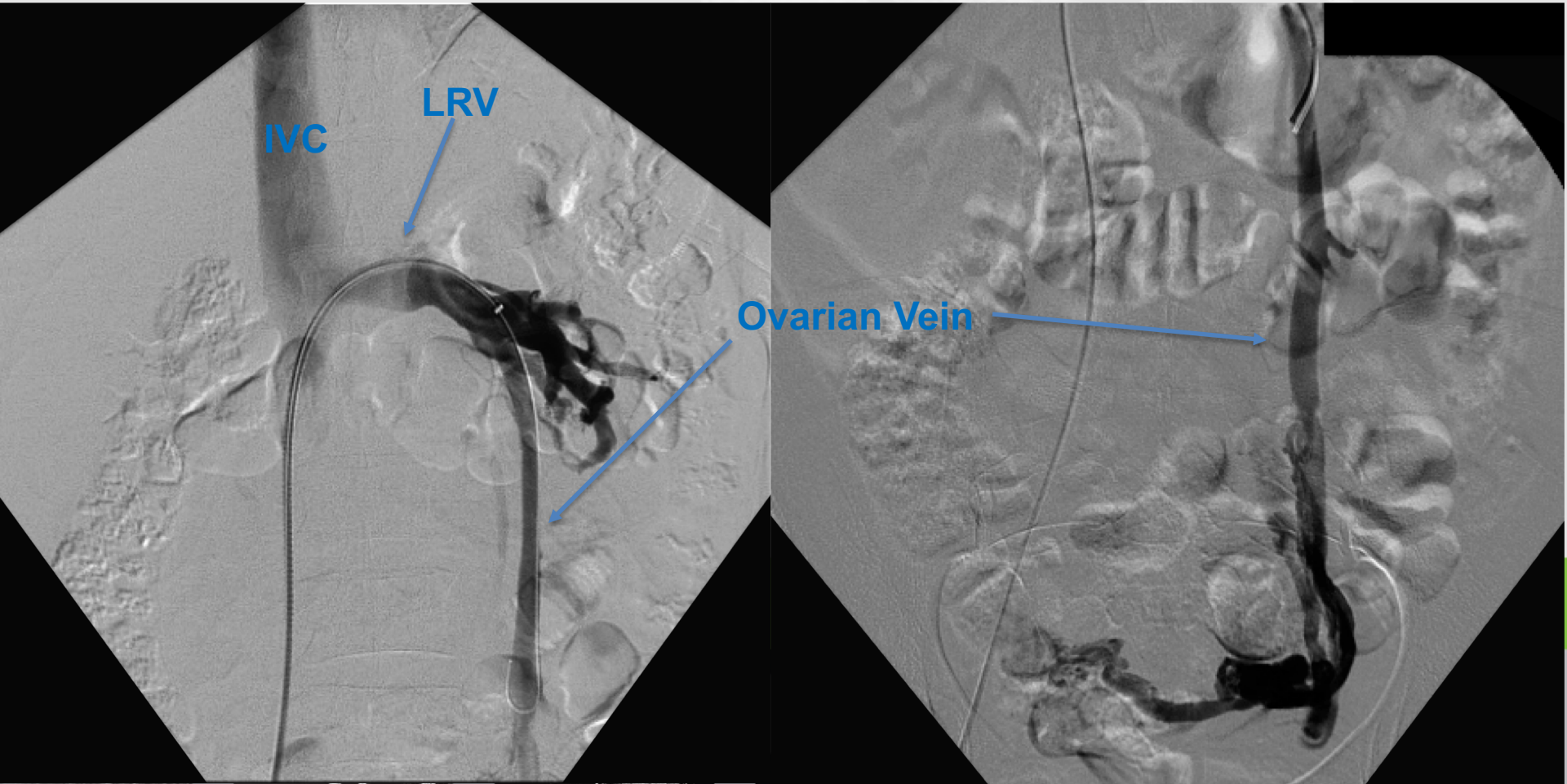
Diagnosis: MRV





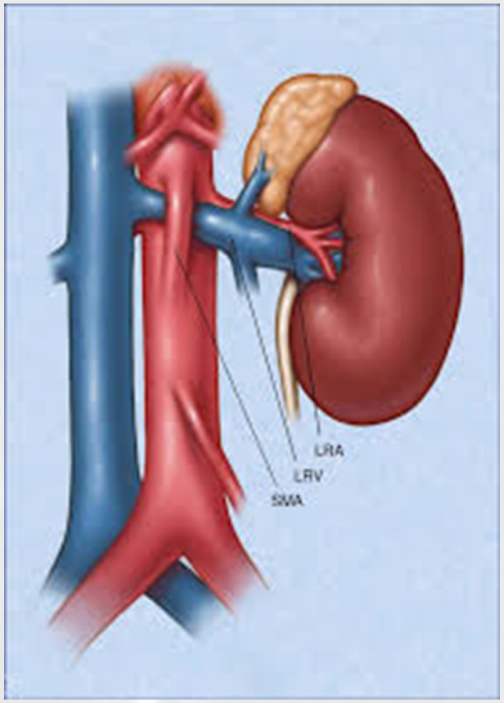
Nutcracker Syndrome

Diagnosis: Venography



Nutcracker Syndrome

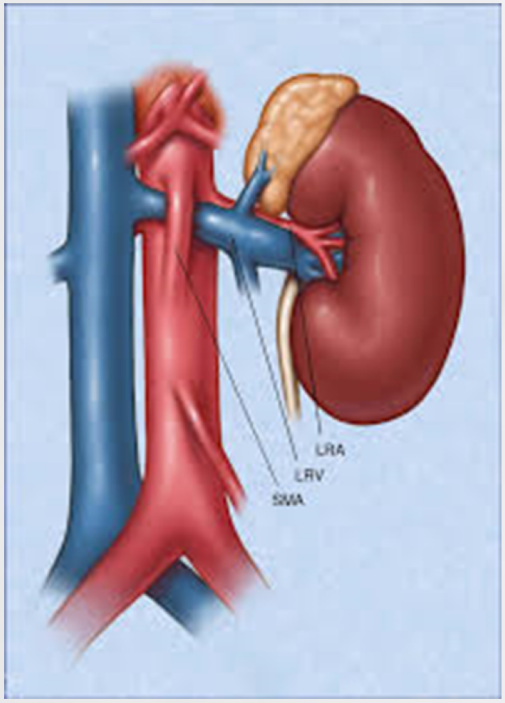
- **Treatment:** management of symptoms, complications

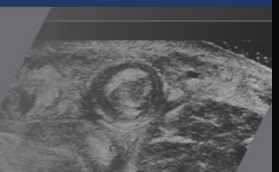


- Left renal vein bypass
- Renal autotransplantation
- SMA transposition
- Gonadal vein transposition
- Nephrectomy
- Left renal vein transposition
- Endovascular left renal vein stenting with gonadal vein embolization

Nutcracker Syndrome

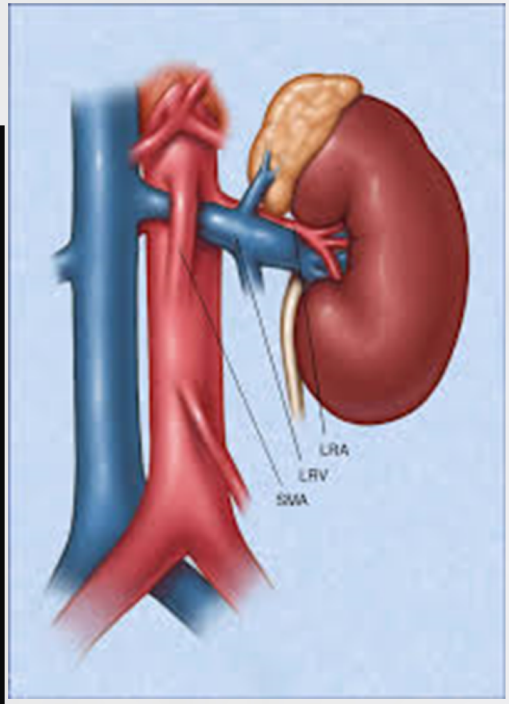
- **Treatment:**





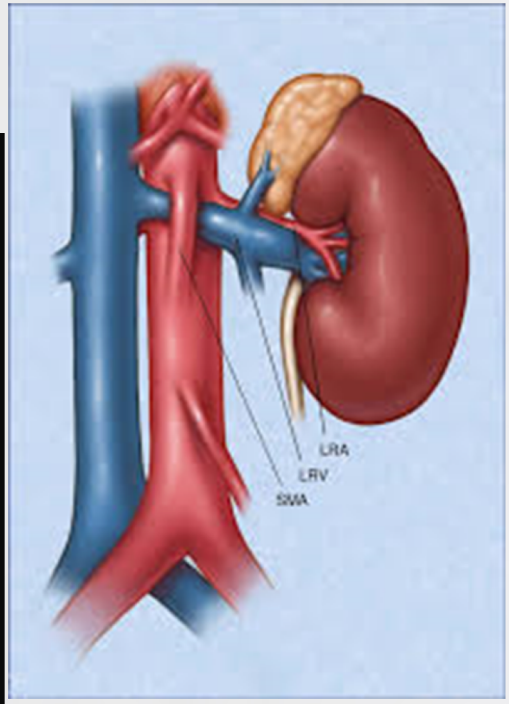
Nutcracker Syndrome

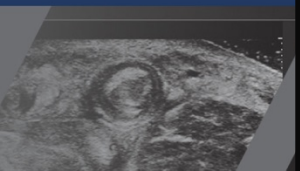
- **Treatment:**



Nutcracker Syndrome

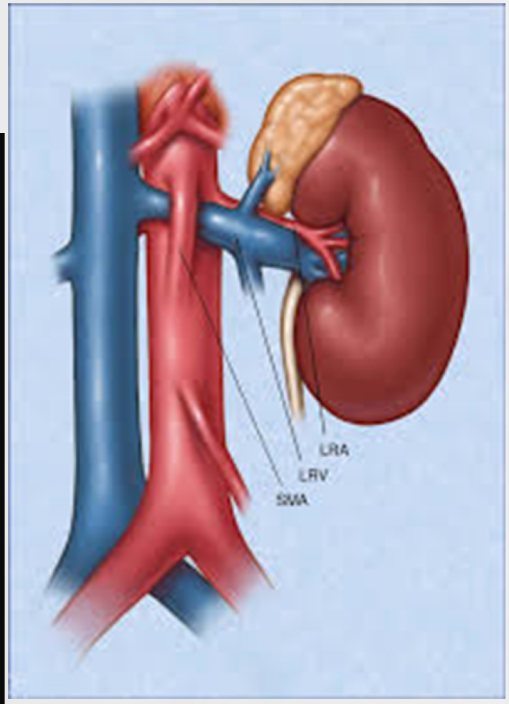
- **Treatment:**

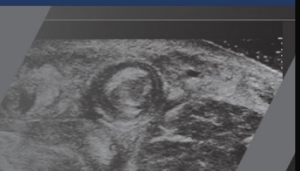




Nutcracker Syndrome

- **Treatment:**





Summary

Klippel-Treynaunay, Parkes-Weber, Nutcracker

Recognition

Evaluation

Management