Anomalous retinal vessels can lead be a diagnostic dilemma for many practitioners. Nonetheless, proper identification is important, as these vessels can be a sign of potentially life-threatening conditions. This course will review and provide current information on common anomalous vessels.

I. Collaterals - Microvessels that develop within the framework of existing retinal capillaries joining obstructed to non-obstructed adjacent vessels.
   - Pathogenesis: Increase flow within working capillaries or increased intravascular pressure within the capillary network adjacent to the obstructed vessel
   - Clinical Signs: Enlargement of the vessels that assume same caliber and cellular characteristic as obstructed vessel
     - Occur several weeks after the RVO
     - Single or multiple and frequently forms across the horizontal raphe
     - Fluorescein angiography- Collaterals DO NOT leak on FA
     - SDOCT/ OCTA
   - Misconceptions
     - Collateral vessels are benign
     - Truth- indicates underlying systemic diseases or prior ocular complication. Warrant medical work-up ~60% of patients developed collateral vascularization in BRVO
     - ONH collaterals are OPTOCILIARY SHUNT VESSELS
       - Shunt vessels- typically artery-vein connection without intervening capillaries
     - Correct term- Retinochoroidal Collaterals
       - CRVO/ Chronic glaucoma/ Chronic papilledema/ Compressive lesion- Glioma & Meningioma/ ONH Drusen
     - IRMA (intraretinal microvascular abnormalities) are NOT collateral vessels
       - Truth- Variation of collateral formation in Diabetic Retinopathy

II. Arterio-venous Malformation (AVM)- true shunt vessels (artery and vein connection). Also known as racemose hemangioma
   - Pathogenesis: Rare congenital high-flow vascular anomalies (Magnus 1874).
   - Clinical Sign
     - Unilateral involvement- isolated or widespread
     - Marked arterial and venous dilation
- Type 1- involves a capillary plexus connecting artery and vein.
- Type 2- Direct AVMs with no capillary plexus
- Type 3- is characterized by widespread retinal AVMs
  - The presence of stage 3 AVM with ipsilateral facial and intracranial vascular malformations is Wyburn–Mason Syndrome
  - Complications:
    - Intraretinal hemorrhages, exudation, RVO, aneurysm formation, vitreous hemorrhage, NVG
  - FA and OCT evaluation
  - Misconceptions- AVMs are benign
  - AVMs require neurologist consultation/ MRI

III. Retinal Arterial Macroaneursym (RAM)
- Idiopathic or acquired focal dilation of retinal artery
- Within first three bifurcations of the central retinal artery)/ often found on the temporal retina
- Elderly females
- Associated with hypertension or atherosclerosis
  - Up to 80% of patients presenting with RAM are shown to have hypertension
- Clinical finding: Multilayer hemorrhage, exudation, and macular edema)
- FA/ICG
- SD/OCT or OCTA
- Unpredictable natural history
  - Spontaneous regression or recurrent with vision loss
  - Anti-VEGF therapy
  - RAMs are misdiagnosed at a rate of 75%
  - Clinical pearl: look for leakage and/or hemorrhages surrounding an arterial area

IV. Retinal Telangiectasia
- Idiopathic Macular telangiectasia (IMT) / formerly known as idiopathic juxtafoveolar retinal telangiectasia (IJRT)
  - A retinal vascular malformation-irregular ectasia and dilation of retinal capillaries
  - Type I- Aneurysmal telangiectasia
  - Type II- Perifoveal telangiectasia (non-proliferative)
    - OCT- Internal limiting (ILM) drape
    - Müller cell loss
    - Observation
  - Type II- proliferative (CNV)
o Misconceptions - No medication work-up necessary for IMT
o Rule-out HTN and DM in patients with parafoveal hemorrhaging/microaneurysms. If these diseases are not present, then telangiectasia is the likely cause.

• **Leber’s Miliary Aneurysm**
  o Localized cluster of dilated capillaries and aneurysms and telangiectasia.
  o A variant of Coat’s disease - there is no exudation and minimal leakage
  o Affects one quadrants (superior temporal) in the mid-periphery
  o Male predilection with unilateral presentation presenting in 4th-5th decade of life
  o Typically asymptomatic, unless macula is involved

• **Coat’s Disease**
  o Mild to massive aneurysmal exudation, retinal hemorrhage, edema and exudates
  o Serous (exudative) retinal detachment can develop (common complication)
  o Predominately a unilateral presentation affecting males (85%) between the ages of 18 months and 18 years.
  o Affects the retinal periphery

V. **Familial retinal arterial tortuosity (Beyer 1958)**
  • Autosomal dominant (AD) disorder
  • Clinical finding - Tortuous small retinal arterioles with normal veins
    o Intra/preretinal hemorrhages
  • Usually 2nd-3rd order (after 1st bifurcation)
  • May be associated with coarctation of the aorta

**References:**