

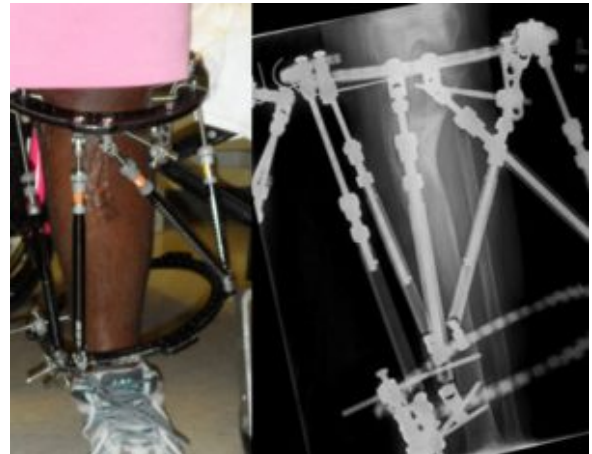
COURSE 09: ORTHOPEDIC MANAGEMENT OF VASCULAR ANOMALIES

Dr. Terry Amarol

Introduction: Orthopedic management of vascular anomalies

The intent of the module is to give an overview of the orthopedic clinical presentation of vascular malformations and currently available treatment options. Vascular lesions and malformations can have varying orthopedic manifestations based on the lesion type, musculoskeletal tissue type, location, and extent of involvement. Clinical impact can range from skin discoloration and soft tissue asymmetry to global limb or trunk involvement with significant multisystem morbidity.

Orthopedic pathology is the end result of a more complex systemic disorder which will require a multidisciplinary team approach to maximize ideal outcomes.



We have chosen to focus on topics most relevant to adult patients with vascular anomalies, to provide a background for the general practitioner and specialist, and to encourage an appreciation for circumstances under which patients should be referred to a dedicated vascular anomalies program. The module addresses the most common questions and clarifies misconceptions which arise in the management of these patients. Images of patients are used liberally in this course, since it is important to recognize the appearance and identify the patterns of these lesions. We also include links to relevant internet sites to reinforce knowledge and provide ongoing resources for learning.

Objectives

Upon successful completion of this activity, participants should be able to:

- > Identify and describe the location of the vascular lesion regarding the type of tissue and limb involved
- > Define the vascular lesion type, natural history, and potential associated medical issues
- > List and explain the potential orthopedic treatment options based on the type and location of the lesion
- > Classify lesions based on the current classification system from the ISSVA
- > Locate the source lesion leading to the downstream pathology
- > Recognize early manifestations of the lesion
- > Choose the appropriate timing regarding other medical specialists' input

Aggressive lesions

Tumors with varying degrees of activity

- › Benign
- › Locally aggressive or borderline
- › Malignant

Aggressive lesions: treatment

Principles for treatment of aggressive lesions are:

- › Complete removal to prevent recurrence
- › Limb preservation to maintain form and function
- › Survival of the patient

Static vascular malformations

Static vascular malformations are classified by

- › Predominant vessel type
 - › Arterial
 - › Venous
 - › Capillary
 - › Lymphatic
 - › Combined (more than 1 vessel type)
- › Impact on major vessels
- › Complexity with associated other anomalies (skeletal, CNS, major organ systems)

Management of vascular malformations with musculoskeletal involvement

Major clinical issues affecting treatment strategies:

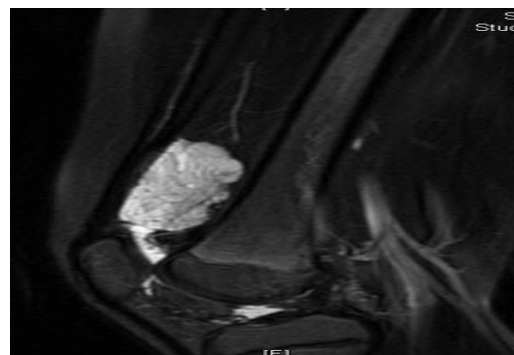
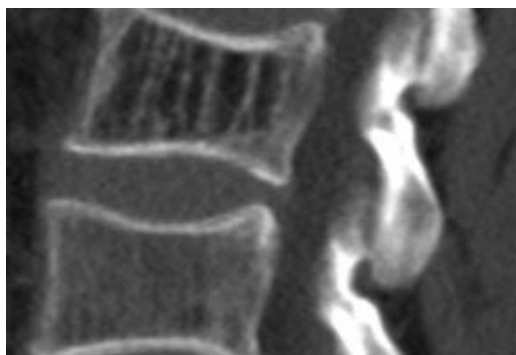
- > Deformity
 - > Bone destruction and maldevelopment
 - > Joint destruction and arthritis
- > Function
 - > Contractures
 - > Decrease muscle strength
 - > Limited mobility and use due to bulk and shape
- > Pain
 - > Caused by deformity and contractures
 - > Caused by sensory nerve involvement



Management strategies

Based on location and lesion type:

- > Joints
- > Bone
- > Spine
- > Muscle



Joint involvement

Intra-articular lesions

- › Vessel Type
 - › Venous
 - › Lymphatico-venous
- › Invasion of the joint space
- › Mass effect
- › Recurrent hemarthrosis
- › Synovitis and inflammation

Recurrent blood exposure to the joint surface leads to the cascade of joint destruction. Hemarthrosis leads to:

- › Synovitis and synovial hypertrophy
- › Hyaline cartilage destruction
- › Fibrosis of the synovial lining and joint capsule
- › Pathophysiology may be linked to an enzymatic cascade similar to rheumatoid arthritis

Clinical picture

- › Pain
- › Joint swelling (due to effusion of mass effect)
- › Contracture
- › Decreased use of the limb

Joint involvement (2)

X-ray findings:

- > Phleboliths
- > Joint effusion
- > Soft tissue shadows, suggesting a mass
- > Late finding would include arthritic changes

MRI:

- > Bloody effusion
- > Vascular lesion within the joint
- > Joint destruction (hallmark is hyaline cartilage erosion and destruction)
 - > Ligament breakdown due to end-stage arthritic changes
 - > Arthrofibrosis



Hemarthrosis

The main surgical modality is complete synovectomy.

- > **Open** (arthrotomy) may allow more extensive removal but with increased morbidity
- > **Arthroscopic** may result in incomplete synovectomy with potential for recurrence



Joint lesions: treatment team approach

Initial management is to prevent hemarthrosis and symptomatic treatment

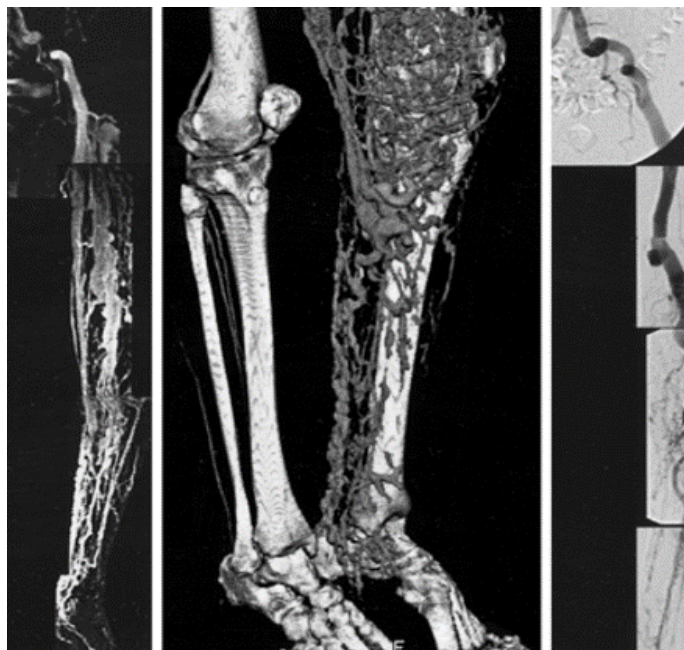
- > Physical therapy to improve ROM and strength
- > Compression garments to minimize swelling
- > Immobilization to prevent re-bleeding

Interventional radiological procedures

- > Manage the feeding vessels, decrease lesion size and impact
- > Often can provide definitive management with near complete eradication

Surgical management

- > Arthroscopic vs. arthrotomy
- > Resect intra-articular mass
- > Joint synovectomy
- > Contracture release
- > Cartilage repair
- > Distraction arthroplasty
- > Total joint replacement



Joint lesions: treatment (total joint replacement)

- > Last resort
- > Mainly for salvage since the patient population is relatively young
- > High complication rate
 - > Excessive blood loss (1-4 liters)
 - > Coagulopathic issues
 - > Wound complications and infections
- > Implant failure from malposition, poor bone quality, instability, and loosening due to poor incorporation of the bone cement



Muscle and soft tissue lesions

- > Involves lesions that mainly infiltrate muscle tissue
- > Main clinical picture is loss of strength and ROM
- > Pain is variable and main issue is loss of function
- > FAVA (fibro-adipose vascular anomaly)
- > Fibrofatty replacement of the muscle
- > Resection has been found to give more predictable results



Treatment

- > Lesion resection
 - > Debulking of the limb
- > Lengthening the motor unit to improve range of motion and function
 - > tendon lengthening
 - > Fascial lengthening
 - > Gradual stretching with external fixation

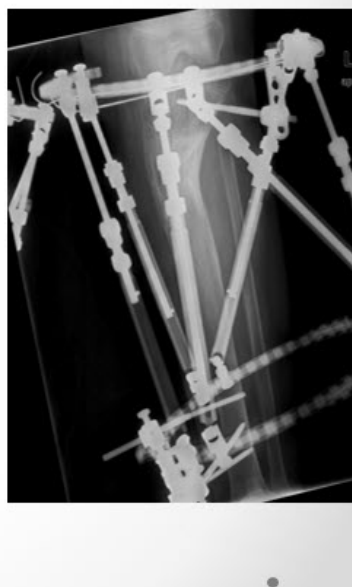
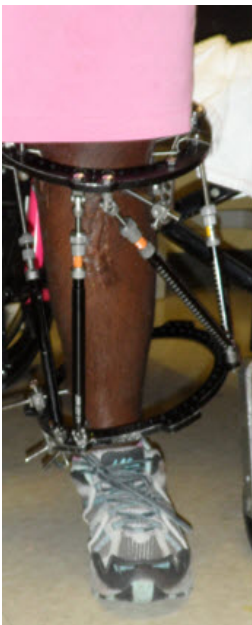


Bone and growth plate involvement

- › Global involvement of the limb with increased blood flow may cause a generalized overgrowth phenomenon
 - › Results in limb length discrepancy
- › Common causes:
 - › Klippel-Trenauney (capillary lymphatico-venous malformation)
 - › Parkes Weber

Treatment

- › Young patients with LLD >1cm
 - › Observation with close follow-up
 - › >1cm shoe lift
 - › 2cm with significant growth remaining with increasing difference; growth plate modulation
- › Older patients with little to no growth remaining with >3-4 cm
 - › Lengthening or shortening procedures
- › Severe overgrowth, or deformity
 - › Amputation
 - › Parkes Weber due to “steal” phenomenon of the high-flow lesion
 - › May be life-threatening
 - › Multidisciplinary team approach due to high potential for bleeding and complications



Spine deformity: scoliosis

Scoliosis can be caused by intrinsic or extrinsic factors.

- > Extrinsic
 - > LLD causing pelvic asymmetry resulting in compensatory scoliosis which later can become structural
 - > Extrinsic trunkal mass effect: mass pushes or shifts the spine
 - > Treatment is mainly focused on the extrinsic cause
- > Intrinsic
 - > Produced either from the intraosseous or intraspinal
 - > Intraosseous cause
 - > Intraosseous lesion
 - > Congenital syndromic lesion
 - > CLOVES (**C**ongenital, **L**ipomatous **O**vergrowth, **V**ascular malformations, **E**pidermal nevi and **S**coliosis/**S**keletal/**S**pinal anomalies)
 - > Intraspinal vascular malformations
 - > Intrathecal lesion

Orthopedic Treatment

- > Intraosseous
 - > Treatment is initially bracing for curves under 40 degrees
 - > Greater than 40 degrees *and* shows evidence of progression will require surgical stabilization
- > Intraspinal
 - > Lesion may have complex extradural/intradural involvement
 - > Possible local tether or mass effect with neurological symptoms may require neurosurgical intervention and possible orthopedic correction
 - > MRI evaluation is paramount

Mixed cases

Often, the patient presents a mixed picture involving multiple tissue types or limb sites.

In such cases, careful planning of treatment options is required, being mindful to minimize the complexity and number of surgical procedures.

Natural history

- › Based on the diagnosis and extent of involvement
- › Single tissue and single bone/joint involvement provide more predictable results
- › Aggressive lesions with global involvement require early diagnosis and careful planning
- › Some cases, regardless of the planning and aggressive approach, still require salvage options
 - › Total joint or joint fusion as last option to retain the limb
 - › Amputation to recover some quality of life and preservation of life

Progression

- › Initial presentation may be simple swelling with minimal limb inequality
- › Missed or delayed diagnosis with prolonged delay in management or mismanagement with lack of experience and knowledge can lead to late sequelae of the disease
- › Late manifestations:
 - › Severe deformity of long bones and/or severe limb inequality
 - › Worsening osteopenia and osteoporosis with high fracture risk
 - › Destructive arthritis of the joints, resulting in significant disability
 - › Extensive infiltration of the soft tissues, specifically muscles, leading to fibrosis, contracture, and nonfunctional motor units
 - › Severe enlargement of limb with soft tissue infiltration with tissue hypertrophy and/or edema,
 - › impairing function Skin and soft tissues compromise, leading to skin ulcerations, infection, and DVT

Summary

Orthopedic expertise with vascular malformations is useful to address:

- › Limb lengthening inequalities
- › Limb deformity
- › Muscle infiltration resulting in mass effect, contractures, and pain
- › Joint involvement resulting hemarthrosis, synovitis, and arthritis
- › Back pain and spine deformity

Key points

Success is keyed to:

- › Early recognition
- › Early referral to an experienced team
- › Multidisciplinary surveillance and management to assemble a complete medical and surgical plan
- › Careful diagnosis and tailored approach
- › Aggressive medical management
- › Interventional radiology to provide focal, lesionally directed treatments
- › Surgical subspecialties to manage resectable lesions to provide symptomatic and, possibly, curative results

COURSE 09 QUIZ

1. A 27-year old female presents with occasional pain and swelling of the left knee with no history of trauma. The knee always feels full and the swelling never completely resolves. She has had a history of a joint aspiration in the Emergency Room in the past which was found to be mostly blood. What important issue has the largest impact regarding orthopedic prognosis for her knee?
 - A. She is at risk for severe arthrosis and joint destruction from the recurrent hemarthrosis.
 - B. There is ligamentous or tendon involvement which would lead to contractures.
 - C. She will develop a limb length discrepancy from the vascular infiltration.
 - D. She has an intra-articular fracture that has not healed leading to the recurrent swelling.
2. What is the first line of treatment once a vascular phenomenon has been determined to be the cause of the patient's orthopedic issues?
 - A. Total joint replacement for pain relief
 - B. Multidisciplinary involvement to assemble a complete medical and surgical plan
 - C. Arthroscopic evaluation to determine if hemarthrosis exists
 - D. Synovisc injections
3. Patients with AVMs undergoing total joint arthroplasty are at high risk for which complication?
 - A. Vascular compromise
 - B. Contracture
 - C. Implant failure
 - D. Poor satisfaction
4. How is limb length discrepancy best managed?
 - A. Ignoring it
 - B. Lengthening the short side
 - C. Shortening the long side
 - D. Based on the magnitude of the difference and the extent of the vascular lesions involvement of the limb
5. FAVA (fibro-adipose vascular anomaly) has been found have the most predictable results with what treatment modality?
 - A. Surgical resection
 - B. Radiation treatment
 - C. Embolization
 - D. Sclerotherapy
6. In preparation for scoliosis surgery for spine deformity caused by a vascular malformation, what would a work up include?
 - A. Genetic screening
 - B. MRI of the spine focusing on intraspinal issues such as tethering or mass effect
 - C. Renal ultra sound
 - D. Blood chemistries

7. Which lesion type will most often lead to a limb amputation as part of definitive treatment?
- A. Hemangioma
 - B. Klippel-Trenauney
 - C. Parkes Weber
 - D. CLOVES
8. What is the earliest orthopedic symptom of a vascular lesion?
- A. Unexplained recurrent fevers
 - B. Intractable vomiting
 - C. Joint pain and swelling with no previous history of trauma
 - D. Skin discoloration and staining
9. What strategies for managing patients with orthopedic manifestations of vascular malformations will be as successful as possible?
- A. Early amputation
 - B. Early detection and multidisciplinary approach
 - C. Multilevel osteotomies
 - D. Early total joint replacement
10. What is the most likely devastating orthopedic sequelae of intra-articular involvement of a vascular lesion?
- A. Hemarthrosis leading to end-stage arthritis
 - B. Tendon infiltration resulting in tendon rupture and disruption of the motor unit
 - C. Ligamentous degradation leading to joint instability
 - D. Skin lesions leading to infection

AUTHOR PROFILES



Terry Amaral, MD, Chief, Pediatric Orthopaedic Surgery Cohen Children's Medical Center

Dr. Amaral received his medical degree from the Albert Einstein College Medicine of Yeshiva University, and trained in Orthopaedic Surgery at Montefiore Medical Center. After completing a fellowship in Pediatric Orthopaedic Surgery at the Hospital for Special Surgery, Dr. Amaral joined the Montefiore Orthopedic faculty. He served as chief of pediatric orthopedic surgery at the Children's Hospital at Montefiore until 2014. He then joined Cohen Children's Medical Center of North Shore LIJ where he currently serves as Chief of Pediatric Orthopedic Surgery.

Dr. Amaral is an active member of many national orthopedic societies which include the Pediatric Orthopaedic Society of North America, Scoliosis Research Society, and the American Academy of Orthopaedic Surgeons, and has served as committee members for these as esteemed organizations. He has presented his work at national and international meetings and has been nominated and received best paper honors. His interests include complex limb deformity and limb reconstruction, complex spinal deformity which includes scoliosis and spondylololsthesis, and joint preservation surgery.

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COURSE 09 QUIZ ANSWER KEY

1. A
2. B
3. C
4. D
5. A
6. B
7. C
8. C
9. B
10. A