

# COURSE 07: SURGICAL MANAGEMENT OF VASCULAR MALFORMATIONS OF THE HEAD AND NECK

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## Introduction: Surgical management of vascular malformations of the head and neck

A 9 year old who presented with an untreated LM of her tongue and pre-mandibular Area. She also had an open bite. We performed a tongue reduction followed by a resection of her premandibular area and this was followed with an orthognathic correction of her bite. The treatment of vascular malformations is in a state of flux.



In some cases, medical management is rapidly replacing sclerotherapy and surgery as a first line modality for lymphatic malformations. With the discovery of more biochemical pathways in the pathophysiology of these disorders, newer drugs will become available. Surgery remains an important modality for these complex cases. However, surgery is rarely a single modality; most often, it is used in conjunction with interventional radiological techniques. The role of surgery is to completely remove disease. This is only possible in a minority of cases with localized disease. Another role of surgery is to improve the appearance of the patient. This may involve treating the oral cavity vesicle, correcting skeletal distortion, decanulation of a patient with a tracheotomy, and treating orbital disease.

Intraoperative hemorrhage has always been a major factor preventing the widespread use of surgery for the treatment of vascular malformations. A modification of surgical techniques, the advent of intraoperative local and systemic hemostatic agents, and the use of pre-operative sclerotherapy/embolization have changed this. In attempting surgical removal of a portion, or all, of the vascular malformation, pre-operative planning is essential. The aim of surgery must be clearly defined, whether palliative or curative. We intend to teach the subject through the liberal use of cases, each selected to illustrate a principle.

# Objectives

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

- › Distinguish the role of surgery in the management of patients with vascular anomalies
- › Identify some of the treatment options and techniques used to treat these patients
- › Assess common pitfalls and complications
- › Determine how to prevent and treat some of these complications
- › Ascertain the correct diagnosis of a vascular anomaly

# Lymphatic malformations

Treatment modalities include:

- > Direct stick embolization (sclerotherapy)
- > Pharmacotherapy (Rapamycin)
- > Surgery

Lymphatic malformations may be:

- > Superficial
- > Deep
- > Compound

Treatment depends on:

- > Depth
- > Anatomic location
- > Extent
- > Previous treatment

Treatment decisions should be made by a multidisciplinary team that includes:

- > Interventional radiology
- > Surgery
- > Hematology/oncology
- > Other subspecialties as needed

Most patients are treated with more than one modality.

- > Surgery is rarely a sole modality of treatment.
- > Most patients are treated with a combination of sclerotherapy and surgery.
- > For the most part, surgery is used to remove a large volume of disease and thereby reduce the amount of sclerotherapy necessary.
- > Rapamycin is a new treatment and its exact role has yet to be determined.
- > Many units will start treatment with a course of Sirolimus.

# Lymphatic malformations

Superficial disease is treated with intralesional Bleomycin (3mg/ml, injected with a 27G needle). One or two treatment sessions may be required.



A patient with mainly superficial LM of the tongue.

Pictured (left): before treatment

Pictured (right): 3 months after two intralesional Bleomycin injections (18 mg. total was used)

## Sclerotherapy

- › Extensive deep disease is usually treated with a combined approach of sclerotherapy and surgery.
- › The role of surgery is to remove bulk, leaving minimal residual disease which can be managed with sclerotherapy.



A patient with an extensive LM extending to the floor of mouth, treated surgically initially and then followed up with several rounds of sclerotherapy using Doxycycline and Bleomycin. She currently has no active disease.



## Lymphatic malformations (2)

Macro-cystic disease can be treated with sclerotherapy or surgery. Usually only one surgical procedure is necessary



A child with a macrocystic cervical LM, before and about 2 years after surgical excision.

## Lymphatic malformations (3)

In cases where it is not possible to remove all of the disease without significant disfigurement, enough disease is removed to achieve facial symmetry. The remaining disease can be treated with sclerotherapy periodically to maintain facial symmetry.

In some cases, a staged removal, as illustrated below, is appropriate.



## Lymphatic malformations (4)

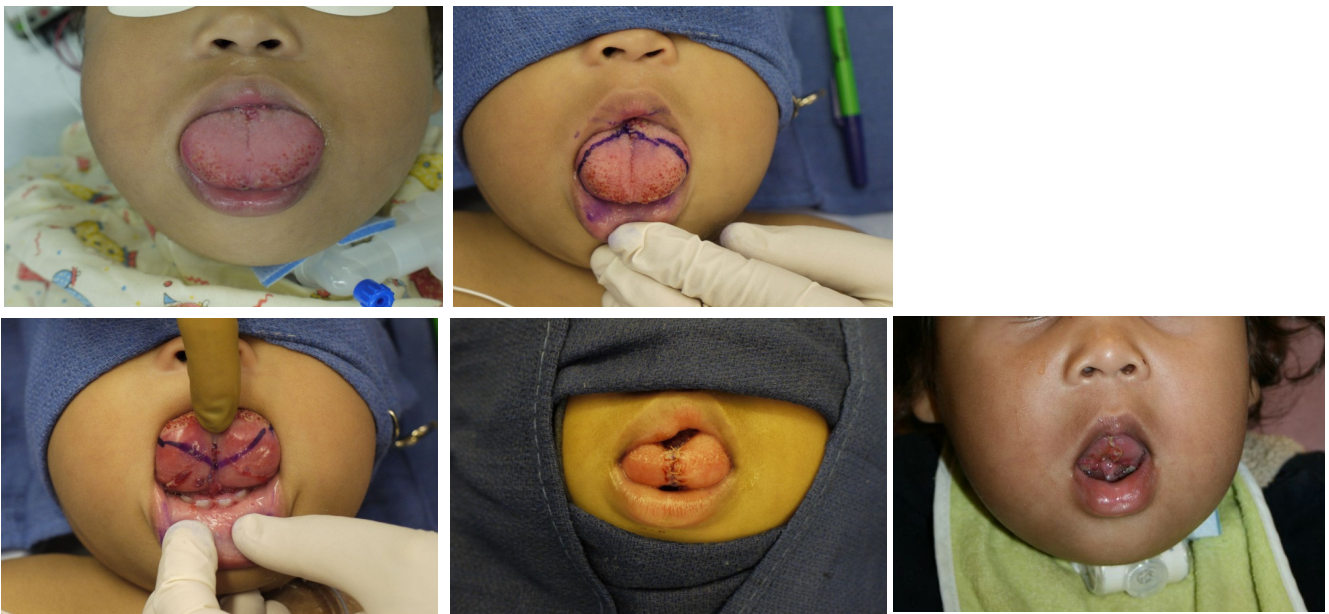


A 9-year old who presented with an untreated LM of her tongue and pre-mandibular area. She also had an open bite. A tongue reduction was performed followed by a resection of her premandibular area and, finally, an orthognathic correction of her bite.

## Lymphatic malformations (5)

Glossoptosis may be due to macroglossia, floor of mouth enlargement, or both.

Pictured: A case of glossoptosis due to macroglossia, corrected by the removal of a large wedge of tongue.



## Lymphatic malformations (6)

Correcting symmetry improves quality of life even in cases where the lesion does not involve the head and neck.



This patient presented early with a gluteal LM which was corrected surgically. The therapeutic objective was to remove enough LM to correct the contour. The patient is several years post-op, with only minimal recurrence

## Arteriovenous malformations

Arteriovenous malformations may be focal or diffuse.

Diffuse lesions may present with widespread involvement of an anatomical area or, more rarely, with several focal lesions in different anatomical locations.



Focal (one anatomical zone)



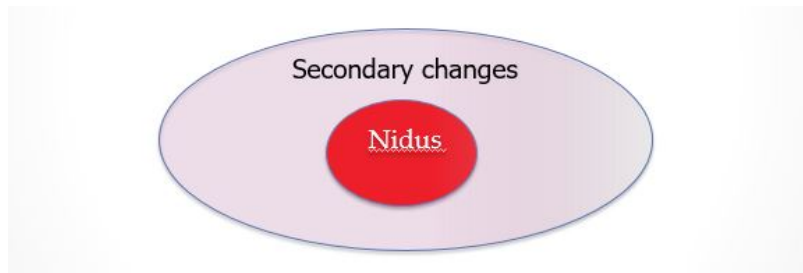
Diffuse (multiple anatomical zones) **Surgical objectives:**

- > Cure: only possible in focal lesions
- > Palliate: with diffuse lesions, improve quality of life

# Nidus

- › Pathophysiologically, an AVM is made up of a nidus and a surrounding secondary zone.
- › The nidus is where the arteriovenous shunting takes place. This is the “core” of the AVM. The surrounding vessels are secondarily dilated as a consequence of the shunting across the nidus and are referred to as the secondary zone.
- › During surgery, we aim to remove all of the nidus if this is possible. It is not necessary to remove tissue from the secondary zone.
- › It is often difficult to differentiate between the two zones. Intraoperatively, dissection within the nidus bleeds profusely and is difficult to control. This contrasts sharply with the secondary zone which bleeds far less and is easier to control.

## Focal lesions

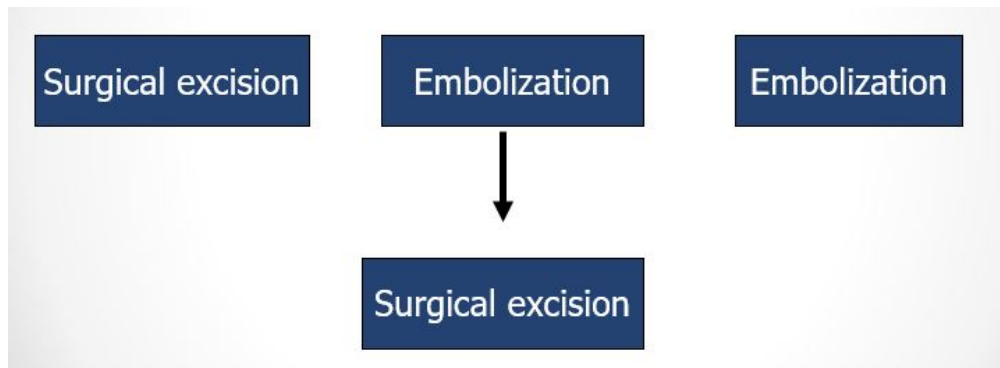


A diagram depicting a nidus surrounded by secondary changes. It is only necessary to remove the nidus. Sometimes it is difficult to distinguish the nidus from secondary changes.

- › Surgical intervention should remove the entire “nidus.”
- › It is not necessary to remove secondary vessels (those which have enlarged as a result of shunting across the nidus).



## Treatment of arteriovenous malformations



In all but the very smallest of lesions requiring surgery, pre-operative embolization is undertaken about 24 hours before surgery.



This case clearly demonstrates the principle of removing only the nidus. A patient with an extensive hemorrhagic AVM of her lower lip was seen. Multiple episodes of embolization had been administered preceding her referral. Clinical examination performed at the time of surgery determined that the nidus was centrally located in the patient's right hemilip. This was excised in a wedge resection. In the second photo, the patient is seen just prior to extubation. Despite some persistent swelling, the patient improved, and the last photo depicts her appearance about a year later.

## A 3-year old with a focal AVM



A 3-year old with a focal AVM of what appears to be her entire upper lip. On closer examination, the nidus was situated centrally in the lesion. It was thus not necessary to remove a large portion of her lip. She was seen again 14 years later with no recurrence. A small cosmetic procedure was done to improve her appearance (last image).

## Removal of the nidus is not possible

In some cases, complete removal of the nidus is not possible without causing severe disfigurement. In this patient our aim was to remove as much disease as possible to improve the child's appearance. Residual disease can be controlled with embolization. Further procedures are therefore indicated.



## Diffuse AVM

A more diffuse AVM involving the patient's upper lip and cheek. Multiple surgical resections were undertaken to remove all identifiable nidus. The patient is now 5 years post-treatment and has not yet manifested with recurrence.



(Above: A patient with a diffuse AVM. His lesion was removed in stages. Each area was embolized pre-operatively and then removed. After several procedures, the patient was controlled. He declined further treatment, stating that he was controlled (4th image). He was not seen again for five years. At this time, he had minimal progression (final image).

## Difficult cases

Although the cases presented thus far all have good outcomes and their procedures seem simple, this is not always the case. In some cases, it may be necessary to perform extensive, ablative procedures in order to bring the disease under control. This may include amputation in cases where there is extensive disease of a limb. This has only happened twice in my career.

Despite multiple rounds of embolization and surgery, this patient's AVM progressed. Her facial skin eventually ulcerated and she needed an extensive resection with a free flap for reconstruction. The photo on the left shows her just prior to ulceration.

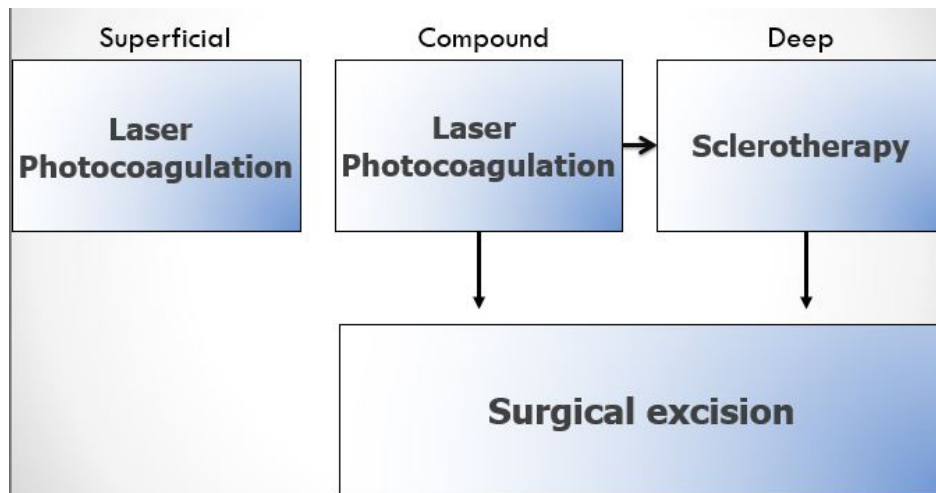


- › Embolization is also used in the management of AVM's.
- › This is most useful when used in conjunction with surgery although there are instances where embolization alone is effective.
- › The decision regarding treatment should always be made by a multidisciplinary team.



# Venous malformations

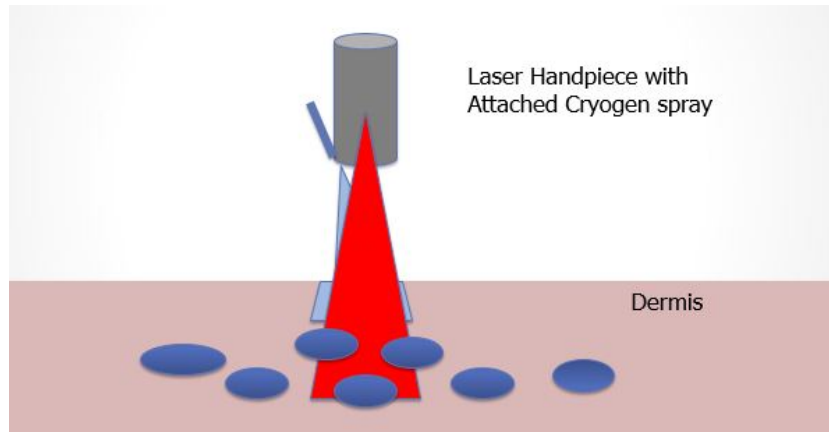
- > The term “venous malformation” applies to a “mixed bag” of lesions.
- > These may be focal or diffuse.
- > They may also be superficial, compound, or deep.
- > Venous malformations can be treated with a laser if they are superficial or if there is a superficial component.
- > Deeper lesions will require direct stick transcutaneous embolization as a primary treatment or, if surgery is indicated, as a pre-operative modality.
- > Pre-operative direct stick embolization is usually coordinated with the surgeon and performed 24 hours prior to surgery.



# Venous malformations

## Laser treatment

- > A Nd:YAG laser is used due to its wavelength (1064nm) and hence, its depth of penetration.
- > It will effectively treat up to a depth of about 1cm.
- > The surface will need to be cooled in order to prevent scarring. Either dynamic or contact cooling can be used.



This demonstrates the effect of dynamic cooling with a Nd:YAG laser. The cooling effect protects the dermis during irradiation with the laser.

## Before and after Nd:YAG laser treatment

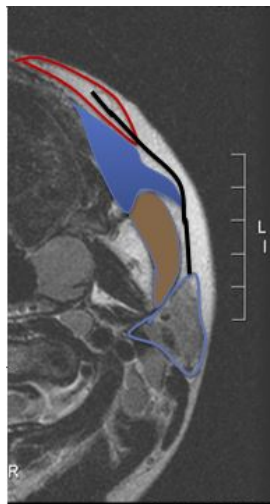


## Mid-facial mass



A child presents with a left mid-facial mass. The anatomical location of the lesion will determine the surgical approach.

## Anatomical spaces of the mid-face



- > Parotid
- > Masseteric
- > Buccal fat space
- > Pre-maxillary

(The location of the facial nerve is shown in black.)

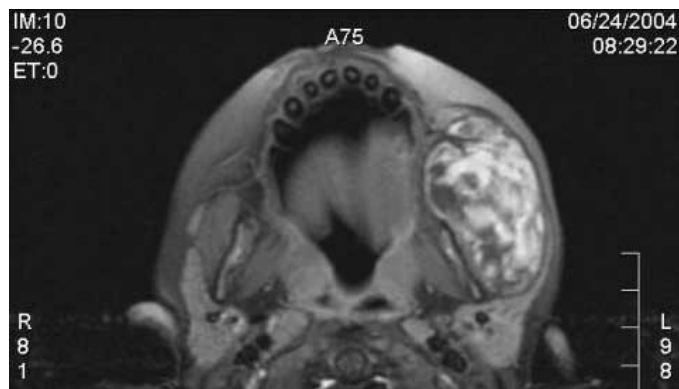
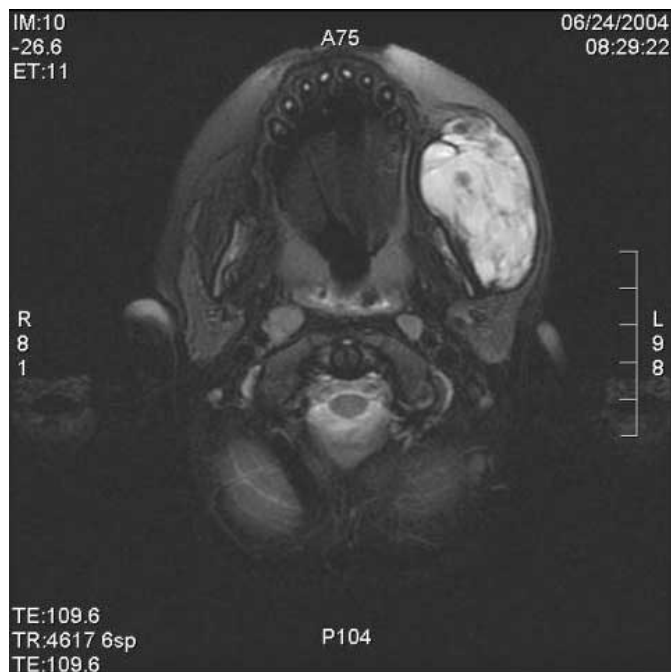
The correct identification of the anatomic space(s) involved will help determine the correct surgical approach and prevent complications.

# Surgical approach to mid-facial lesions

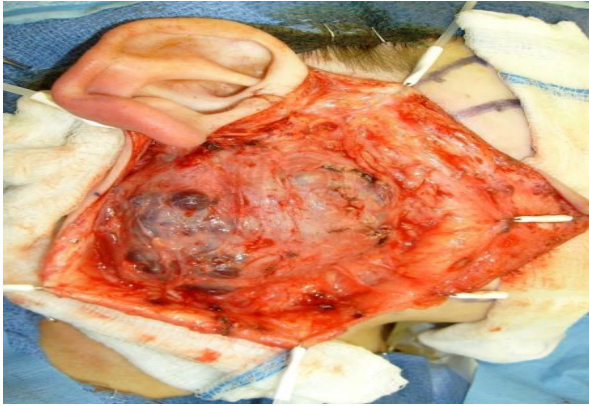
The location of the facial nerve branches with respect to the lesion is crucial.

Parotid lesions	Parotidectomy incision
Masseter	Extended Parotidectomy incision
Buccal fat space	Extended Parotidectomy incision
Premaxillary/mandibular space	Intraoral incision

## A masseteric venous malformation



## Venous malformations (2)



This child was sclerosed 24 hrs ago; we perform facial nerve monitoring, and lift a preauricular flap, starting from the periphery, remove the superficial lobe, and close up.



The same patient after surgical removal with VII preservation.

## Vascular malformations



A 24-year old with a masseteric venous malformation, before and after excision with VII preservation.



A young child with a venous malformation of the pre-maxilla and upper lip. The patient was treated with Nd:YAG laser treatment, followed 6 weeks later with preoperative sclerotherapy and surgical excision 24 hours later.

In cases where it is not possible to remove the entire lesion, our aim should be to improve quality of life.

Any residual disease can be treated with sclerotherapy or surgery.



## Course 07: Lesson Quiz

1. Concerning arterio-venous malformations, which of the following statements is true?
  - A. Sirolimus is usually helpful in treating AVM's and should be used as a firstline treatment.
  - B. Pre-operative embolization is not helpful because intraoperative bleeding is rarely problematic.
  - C. Embolization is often curative.
  - D. Focal AVM's can sometimes be completely removed.
2. A 3-year old presents with an extensive cervicofacial mixed macrocystic/microcystic lymphatic malformation. This patient should be treated in the following way:
  - A. A staged surgical approach should be undertaken.
  - B. An aggressive program of transcuteaneous direct stick embolization should be started.
  - C. The child should be considered for a course of Rapamycin.
  - D. No treatment should be recommended.
3. A 12-year old presents with a macrocystic cervical lymphatic malformation. He has had multiple rounds of direct stick embolization but has seen little or no response. A reasonable next step would be:
  - A. Consider starting Sirolimus.
  - B. Consider surgical resection of the lesion.
  - C. Repeat sclerotherapy, but with a different agent.
  - D. Repeat the MRI to check the accuracy of the diagnosis.
4. A 6-year old presents with an extensive craniofacial lymphatic malformation and glossoptosis that has been present since birth. The correct approach should be:
  - A. Schedule the child for a surgical tongue reduction procedure.
  - B. Start the child on Rapamycin immediately.
  - C. Determine whether or not the glossoptosis is due to macroglossia, floor of mouth disease, or both.
  - D. Start the child on a course of Sildenafil.
5. Which of the following is NOT part of differential diagnosis of a mid-face vascular mass?
  - A. Venous malformation
  - B. Hemangioma
  - C. Warthin's tumor
  - D. Arteriovenous malformation
6. Which of the following are not anatomic spaces of the mid-face?
  - A. Submandibular space
  - B. Buccal fat space
  - C. Masseteric space
7. Concerning venous malformations, which of the following statements is incorrect?
  - A. Surgery can be used as a single modality of treatment on some occasions.
  - B. Direct stick embolization is a useful treatment modality.
  - C. A multidisciplinary approach is always preferable to management planning.
  - D. Compound lesions should only be treated with surgery.

8. Which of the following statements is correct?
- A. A Nd:YAG laser with a surface-cooling device should be used to treat the deep component of a venous malformation pre-operatively.
  - B. A Nd:YAG laser is useful in treating the superficial component of a venous malformation.
  - C. A pulsed dye laser is useful in treatment the deep component of an AVM.
  - D. A Nd:YAG laser is useful for treating lymphatic malformations because the lymph component of an LM preferentially absorbs near infrared light.
9. Which of the following statements is incorrect?
- A. It is almost always necessary to embolize and AVM preoperatively to minimize operative blood loss.
  - B. In order to cure an AVM, it is necessary to remove both the nidus and the secondary changes.
  - C. An arteriovenous malformation is made up of a nidus surrounded by secondary changes.
  - D. The primary object of treatment is to improve quality of life.
10. The lifetime dose of Bleomycin should be less than:
- A. 400 mg
  - B. 200 mg
  - C. 250 mg
  - D. There is no limit in the amount of Bleomycin that a patient can receive.



## AUTHOR PROFILES

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**Milton Waner, M.D., B.Ch.(Wits, MD, FCS(SA))**

Dr. Milton Waner is an authority in the surgical management of hemangiomas and vascular malformations of the head and neck. He has over 100 peer reviewed publications, 30 textbook chapters, and countless presentations at national and international meetings. He has written and edited 4 textbooks, 3 of which are on hemangiomas and vascular malformations. Dr. Waner has been involved in this field for 30 years and pioneered the early management of hemangiomas. He is currently the Surgical Director of the AVM center at Lenox Hill Hospital where he is an attending surgeon in the Head and Neck Institute. He was formerly an endowed professor of Otolaryngology at the University of Arkansas for Medical Sciences.

Dr. Waner has won numerous awards for his work in the field of vascular malformations.

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## Course 07: Lesson Quiz Answer Key

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