COURSE 04: NEURO-INTERVENTIONAL RADIOLOGIC MANAGEMENT OF VASCULAR ANOMALIES

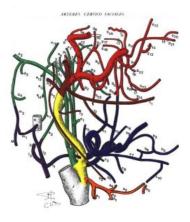
Dr. Alejandro Berenstein

Introduction: Neuro-interventional radiologic management of vascular anomalies

This module highlights the role of endovascular treatment in vascular lesions of the head and neck. The use of transarterial embolization, direct percutaneous embolization, and sclerotherapy will be illustrated.

The first and most important step in treatment is to establish the correct diagnosis.

In this module, we will review the angiographic appearance and imaging used for diagnosis and therapy planning.



Objectives

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

- $^{>}$ Identify the different vascular lesions affecting the head and neck
- > Describe the role of embolization and sclerotherapy
- > Define indications for management of vascular lesions
- > Classify vascular malformations
- > Explain when and how to intervene in vascular malformations
- > Recognize the various types of vascular malformations
- > Choose the best treatment, or combination of treatments, for vascular malformations

Clinical & angiographic classification of congenital vascular lesions

PROLIFERATIVE MALFORMATIONS

Hemangioendothelioma
KMS

ERRORS OF MORPHOGENESIS

Capillary	hypervascular
High flow (AVM)	with fistulization
Capillary	small veins & "cavernous " spaces
Venous	"cavernous" venous lakes
Hemangiolymphangioma (Veno-lymphatic)	mass & spontaneous bleeding
Lymphatic Malformations	cystic: uni or multicompartmental macrocystic / microcystic

Patient problems

- > Cosmetic deformity
- > Hemorrhage
- > Functional problems (airway, feeding, speech/vision/hearing)
- > Multiple surgical procedures

Vascular malformations

Goals of treatment

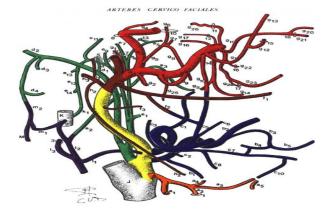
- > Cosmetic improvement
- > Management of hemorrhage
- > Management of ischemic pain
- > Functional restoration
- > Stabilization and/or regression

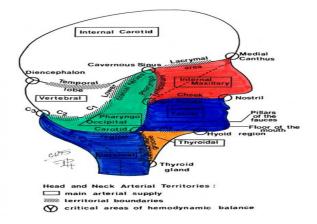
Treatment

- > Total resection
- > Total endovascular obliteration
- > Combined treatment
- > Palliation (bleeding, fistulization, ischemic ulcerations)

Functional vascular anatomy of the head and neck

A detailed knowledge of head and neck vascular anatomy, including potential collateral pathways, is essential to perform safe embolizations.





Endovascular interventions

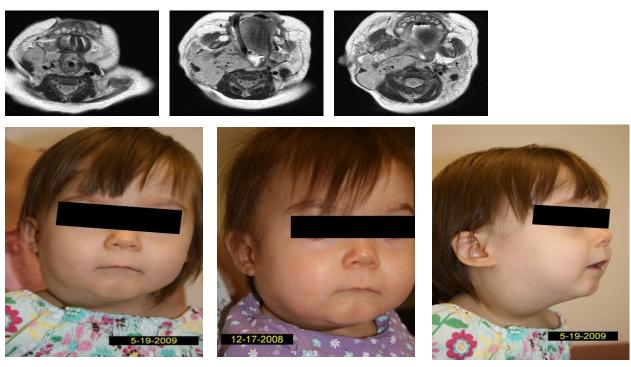
- > Transarterial embolization
- > Transvenous embolization
- > Direct percutaneous embolization
- > Sclerotherapy
- > Pre-operative
- > Curative
- > Palliative
- > Post-operative

Hemangioma

With the introduction of Propranolol, there is less need for interventional neuro-radiologic treatment of infants with Infantile Hemangioma. There are isolated incidences where more aggressive approaches have been used, as in the two cases that are being presented here.

Parotid and parapharyngeal hemangioma

2-month old girl with a parotid and parapharyngeal hemangioma



After six months, the parotid and parapharyngeal hemangioma continue to proliferate, despite systemic corticosteroid treatment.

Placed on Propanolol.

Six months later, the lesion continued to proliferate.





Bleomycin sclerotherapy for the hemangioma

21-month old girl with parotid hemangioma.

Beta blocker and steroids were tried; no regression took place.

Direct injection of bleomycin was used which led to rapid improvement. This should be, at this time, considered an investigational treatment.

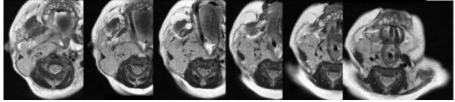






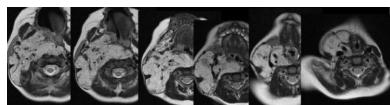
Additional images





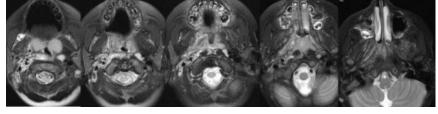
2-27-2008





5-20-2012





5-25-2012

Facial hemangioma

Facial hemangioma in a 3-year old. Surgical resection was aborted because of the risk of facial nerve injury.



Facial hemangioma

After 2 treatments and surgical resection



P.H.A.C.E.S.

PHACE Syndrome is an acronym for a distinctive syndrome that includes the following components:

- P osterior Fossa
- H emangiomas
- A rterial anomalies
- C oarctation/Cardiac abnormalities
- E ye abnormalities
- S ternal lesions

Right eye cataract

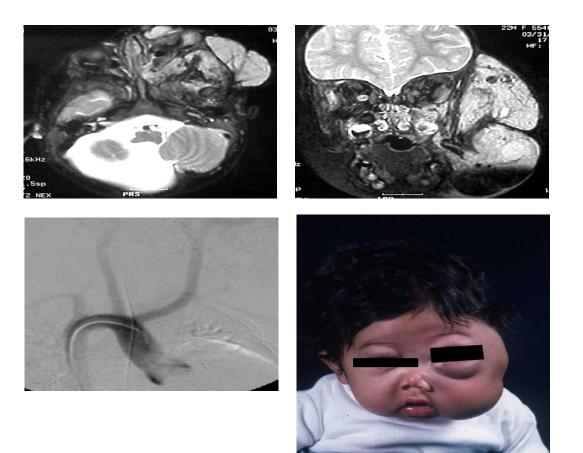
2-month old: right eye cataract, left eye glaucoma





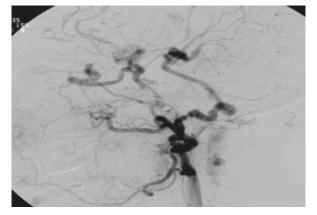
Right aortic arch

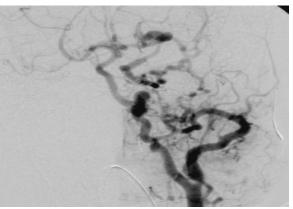
Right aortic arch, multiple hemangiomas, posterior fossa cyst



Arterial abnormalities

Arterial abnormalities, persistent hypoglossal artery, posterior cerebral (P2) aneurysm, stenosis of the petrous ICA





Transarterial embolization

After transarterial embolization, the venous hypertension and glaucoma resolved, and the hemangiomas started to involute.



Transarterial embolization

After transarterial embolization and surgical contour treatment



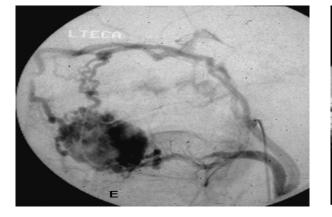


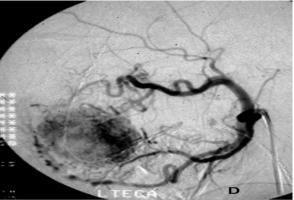
Non-involuting congenital hemangioma (NICH)



Hypervascular tumor blush

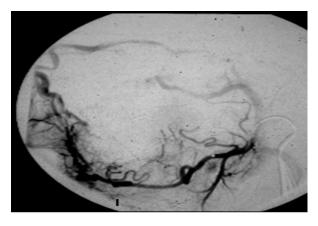
Arteriograms of non-ithout A-V shunt

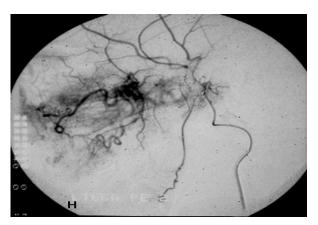




Hypervascular tumor blush (2)

After transarterial embolization





Non-involuting congenital hemangioma



After transarterial embolization



Hemangioendothelioma KMS after Rx platelets 8000



Hypervascular tumor

Hypervascular tumor with contrast material "pooling" without A-V shunting



After transarterial embolization; platelets raised to 95,000, permitting safe surgical removal



Vincristine

Most hemangioendotheliomas with KMS are treated with Vincristine.



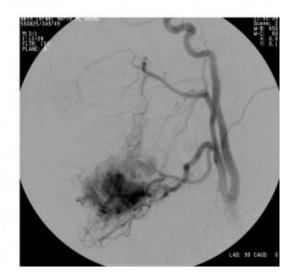
NBCA



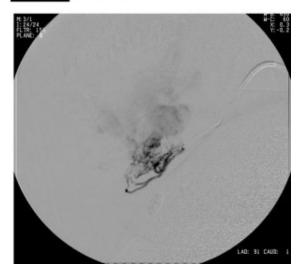
(Upper photo) A 15-year old female presented with severe bleeding after placement of braces. (Lower photo) Note the marked shrinkage of the AVM following embolization with liquid embolic (NBCA).



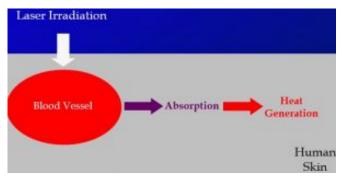
...>High-flow mandibular AVM

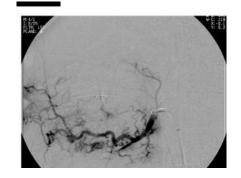


High-flow mandibular AVM presents with bleeding and loose teeth.



Super selective angiogram off the inferior mental artery, supplying the malformation through trans-bony anastomosis.

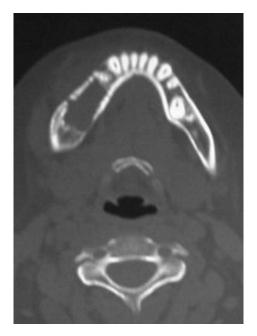




Last image hold following NBCA embolization;

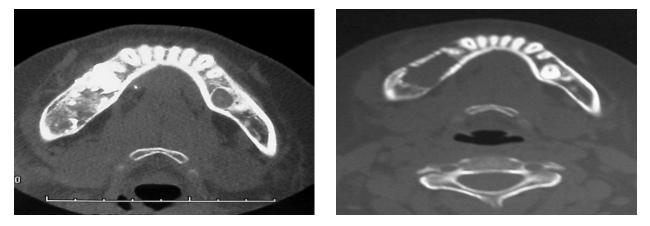
note that reaching the venous pouch is the best way to deal with a malformation.

Embolization treatment



Pre-emboliation:

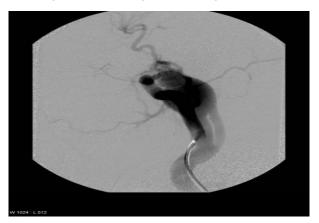
Note the destruction of bone prior to treatment.



Post-embolization treatment reveals bone remodeling.

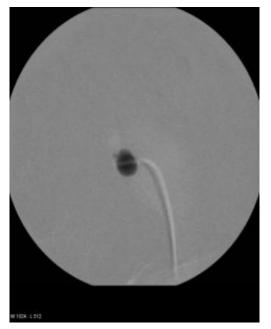
A-V fistula

2-year old asymptomatic child, noted by mother to have a pulsatile, electric-like sensation while kissing the child. High flow congenital A-V fistula is seen from the external carotid artery.

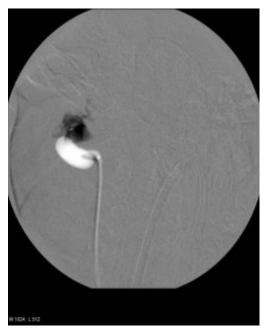




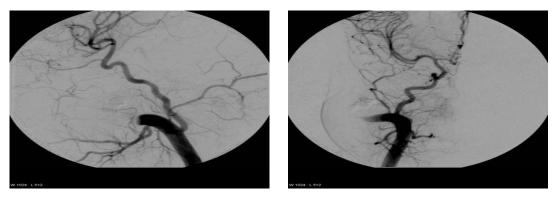
A-V fistula (2)



Double-lumen balloon catheter to control the flow; the balloon is inflated.



Following the injection of NBCA into the fistula site.



Post-embolization control angiogram of the right common carotid artery demonstrates total obliteration of the fistula and preservation of all normal arteries.

Course 04: Lesson Quiz

- 1. Venous malformations:
- A. Involve only mucosa
- B. Involve mucosa and muscle
- C. Are usually focal
- D. Are frequently multifocal
- 2. With neuro-interventional therapy, diagnosis and treatment is based on:
- A. Angiographic appearance and imaging
- B. Imaging and clinical history only
- C. Clinical history of lesion
- D. Angiographic appearance and histology
- E. Hematologic findings
- 3. The first choice treatment for a hemangioma is:
- A. STS sclerotherapy
- B. Combination treatments
- C. NBCA embolization
- D. Propranolol

4. Which one of the following is NOT a problem identified by patients requiring neuro-interventional therapy?

- A. Cosmetic deformity
- B. Fibrofatty redundant tissue
- C. Fertility problems
- D. Functional problems
- E. Hemorrhage
- 5. Arteriovenous malformations:
- A. Are characterized by the presence of arteriovenous shunting
- B. Are the most frequent vascular malformation of the head and neck
- C. Can be treated with propanolol
- D. Are slow-flow lesions

6. Lymphatic malformations:

- A. May start as macrocystic, and have microcystic components
- B. Are all microcystic
- C. Are all mixed
- D. Are all macrocystic

- 7. Venous malformations:
- A. Present with a normal surface temperature
- B. Are easily compressible
- C. Will expand with Valsalva maneuvers
- D. Will have a bluish discoloration when involving skin or mucosa
- 8. Arteriovenous fistulas:
- A. Can be cured with sclerotherapy
- B. Are usually benign
- C. Are characterized by the presence of a nidus
- D. Are high-flow lesions best treated with fast-polymerizing tissue adhesives
- 9. The treatment of choice for venous malformation is:
- A. Surgical removal
- B. Radiosurgery
- C. Transarterial embolization and surgery
- D. Combination treatments, sclerotherapy, and surgery

AUTHOR PROFILES



Alejandro Berenstein, MD

Alejandro Berenstein, MD (Dr B.) was born in Mexico City in 1948, graduated from the UNAM, at present is Professor of Neurosurgery, Radiology and Pediatrics, Director of the Hyman-Newman Institute for Neurology and Neurosurgery at the Ichan School of Medicine at Mount Sinai Health System in NYC.

His contributions include the description of the functional vascular anatomy,

understanding and description of the diseases, and the development of multiple medical devices to treat them, now used throughout the world. He has authored over 150 publications in peer-reviewed journals, more than 25 chapters in the field, and 9 books, including the five-volume, groundbreaking textbook "Surgical Neuroangiography."

Among the more than 30 awards, in 1989 he received the Ramon y Cajal Award in Medicine for distinguished Latin American Physicians. In 1998 he received the first Cerebrovascular Luessenhop Award from the American Association of Neurological Surgeons (AANS) and the Congress of Neurological Surgeons (CNS) Section of Cerebrovascular Surgery. In 2001 he was awarded the Paolo Raimondi Award from the AANS/CNS on Pediatric Neurological Surgery, and is the only physician to have received both. In 2007 he received the Physician of the Year award from the Vascular Birthmarks Foundation for his work with children with vascular birth defects. In 2007, he received the Boston Scientific Neurovascular Lifetime Achievement Award for excellence in research and clinical applications; in 2008, he won the Heart of New York Stroke Spotlight Award from the American Heart and Stroke Association.

He is an Honorary Member of the Japanese Society of Neuroendovascular Therapy; in 2009 he received the first Serbinenko Golden Sign Award in Neurosurgery in Moscow. He has been visiting professor, both lecturer and faculty, on over 600 occasions. He became the first president of the World Federation of Interventional and Therapeutic Neuroradiology and third president of the American Society of Interventional and Therapeutic Neuroradiology. He has been the President of SILAN the Latin-American Society of Neuroradiology (2012-2013).

Alejandro Berenstein, MD, is Clinical Professor, Department of Neurosurgery, Radiology, and Pediatrics at the Ichan School of Medicine at Mount Sinai HCS in New York.

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Course 04: Lesson Quiz Answer Key

1. D 2. A 3. D 4. C 5. A 6. A

7. D

8. D

9. D