

Multidisciplinary Approach of Congenital Vascular Malformation -A New Role of a Vascular Center-

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B.B.Lee, MD, PhD, FACS

Professor of Surgery and Director

Center for Vascular Malformation and Lymphedema
George Washington University, Washington DC, USA

Adjunct Professor of Surgery

Uniformed Services University, Bethesda, Md, USA

Visiting Professor of Surgery

Johns Hopkins University, Baltimore, Md, USA

DISCLOSURE OF CONFLICTS OF INTEREST

BYUNG-BOONG LEE, MD, PhD, FACS

**Multidisciplinary Approach
of Congenital Vascular Malformation
-A New Role of a Vascular Center-**

I do not have any relevant financial relationships with any commercial interests.

Multidisciplinary Approach on the CVM

CVM is the vascular disorder of extreme variety with stigma of totally unpredictable behavior; “recurrence” is the trademark of CVMs.

- Enigma among the vascular disorders (E. Malan)
- Curse/condemned disease to the surgeon (E. Szylagyi)
- Ultimate challenge to the surgeon (S. Belov)
- **“Symbol of humiliation”**



Multidisciplinary Approach on the CVM

“Enigma in Modern Medicine”

CVM remains the most difficult and confusing diagnostic and therapeutic challenge through centuries due to;

- Wide range of the clinical presentation
- Unpredictable clinical course
- Erratic response to the treatment with high recurrence
- High morbidity of conventional treatment
- Confusing terminology without proper information on etiology, anatomy, and pathophysiology



Multidisciplinary Approach on the CVM

Wide range of the clinical presentation

- CVM is a group of birth defects often affecting more than one vascular system; capillary, arterial, venous, and/or lymphatic system with different characteristics and behaviors.
- CVM represents various birth defects developed in peripheral vascular system during various stages of embryogenesis as the result of a developmental arrest.
- *CVM is therefore, a unique vascular disorder known for the most difficult and confusing diagnostic and therapeutic challenge through centuries.*



CVM affecting various locations in various extents



VM



VM

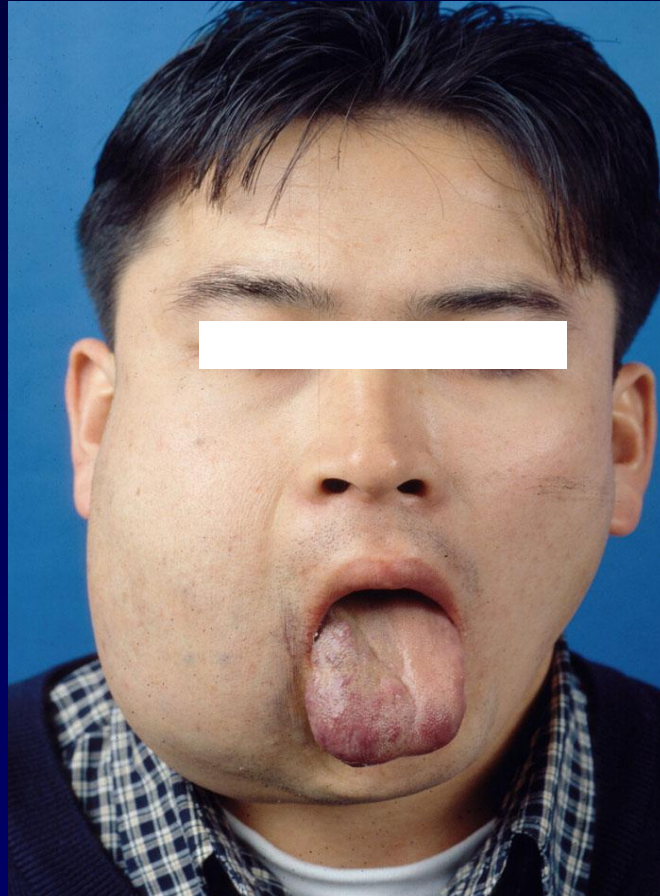


VM

CVM affecting various locations in various extents



VM



VM & LM



AVM



VM



VM



LM

CVM affecting various locations in various extents

Multidisciplinary Approach on the CVM

“Ultimate Challenge in Modern Medicine”

- Such unpredictable behavior with high recurrence is generally due to its pathognomonic/ embryological characteristics of the ‘extratruncular’ lesion as an embryonic tissue remnant.
- Its erratic response to the conventional treatment with high morbidity and high recurrence gave a notorious reputation as an enigma among many vascular disorders; “recurrence” became a trademark of CVM.



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“Ultimate Challenge in Modern Medicine”

- Poor outcomes of the challenge to the CVM through earlier decades, mostly led by surgeons alone with cavalier approach based on limited knowledge/information, added much confusion on the CVM management with erroneous prejudice against the CVMs.



Multidisciplinary Approach on the CVM

Contemporary Concept

- Hamburg classification was established as a new classification fulfilling mandated condition with proper information on the etiology, anatomy, embryology, histopathophysiology of CVMs.
- This new classification provided right ground for contemporary concept on the CVMs; precise diagnosis of various CVMs became feasible based on new technology for the advanced management.
- Finally, a new concept of ‘**multidisciplinary team approach**’ emerged aiming at the prevention/control of ‘recurrence’ with minimally possible complications/morbidity as the ultimate goal of the management, as lonely surgeons dreamed for centuries.

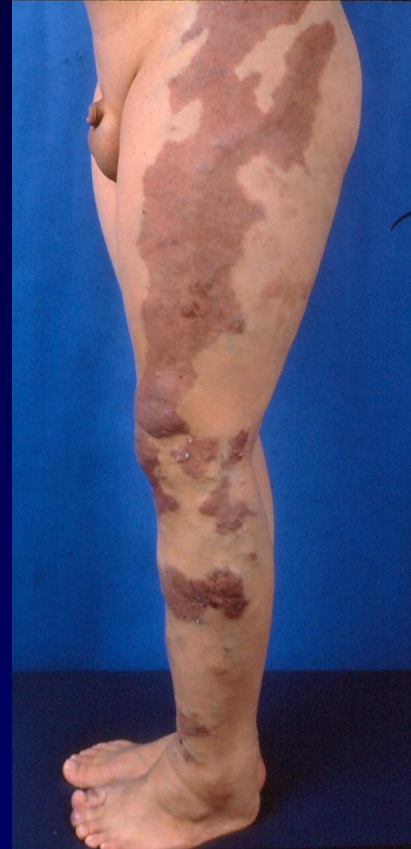
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Changing Concept on the CVM

Multidisciplinary Approach

- In order to improve its clinical outcome of surgical therapy alone, the endovascular therapy with various forms of embolo/sclerotherapy was adopted as multidisciplinary approach.
- New multidisciplinary approach has achieved full integration of various surgical and non-surgical/endovascular treatments.
- To accommodate this new approach effectively, a specialized care team has to be organized for ‘centralized’ management with new concept based on “Vascular Center” as the referral center.



Changing Concept on the CVM

Congenital Vascular Malformation (CVM) Clinic

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- Centralized management system by specialized care team as “Referral Center” with 17 related clinical subspecialties involved.
- Symbol of extensive multidisciplinary approach to the old problem with new concept for the advanced care.
- Fully experimented and proved for its efficacy and endorsed as an ideal system for the contemporary management of CVMs by Society for Vascular Surgery.



Changing Concept on the CVM

CVM Clinic - Multidisciplinary Team

Vascular Surgery, Pediatric Surgery, Orthopedic Surgery, Plastic & Reconstructive Surgery, Oral-Maxillar-Head & Neck Surgery, Physical Medicine & Rehabilitation, Anesthesiology, Pathology, Vascular Medicine/Angiology, Cardiology, Pediatrics, Genetics, Dermatology, Psychiatry, Interventional Radiology, Diagnostic Radiology, Nuclear Medicine.



Changing Concept on the CVM

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Treatment Strategy - Multidisciplinary Approach

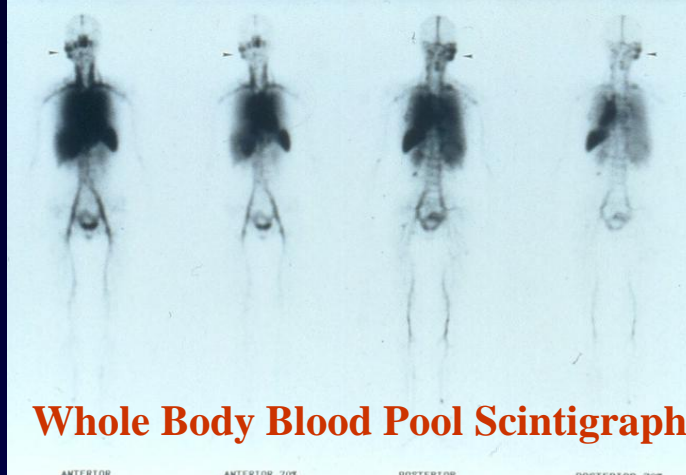
- Not every CVM lesion should be considered for the treatment; the only lesion with justified indications should be assessed by the multidisciplinary team as a treatment candidate.
- Only when the team consensus should expect the benefit by the treatment to exceed the risk of the complication and morbidity by the treatment, less risky therapy should be tried first.
- Controlled aggressiveness is warranted for the CVM in general regardless its condition even for AVM.



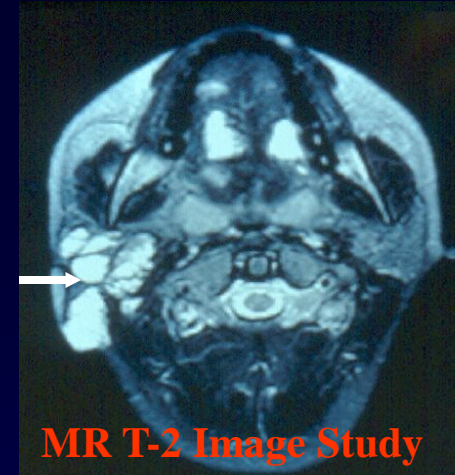
Changing Concept on the CVM

Treatment Strategy - Multidisciplinary Approach

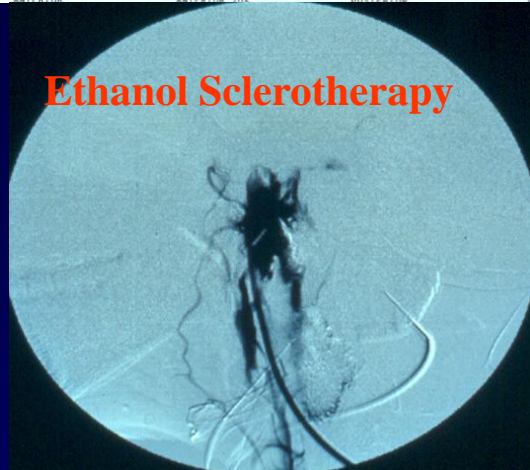
- Traditional open surgical/excisional therapy and endovascular therapy are now fully integrated as a total care management of the CVM as the most effective means. (e.g. embolo/sclerotherapy for extratruncular lesion: angioplasty and stent for truncular lesion)
- Endovascular therapy is now the treatment of the choice to non to poor surgical candidate with extensive lesions beyond deep fascia with involvement of muscle, tendon and bone as diffuse infiltrating extratruncular CVM lesions as an independent therapy.



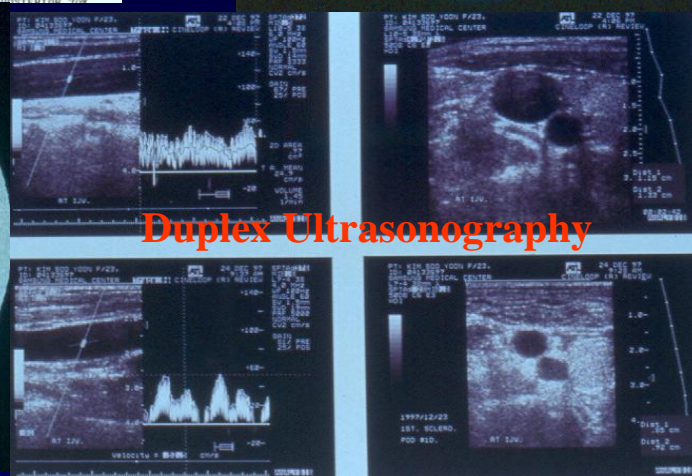
Whole Body Blood Pool Scintigraphy



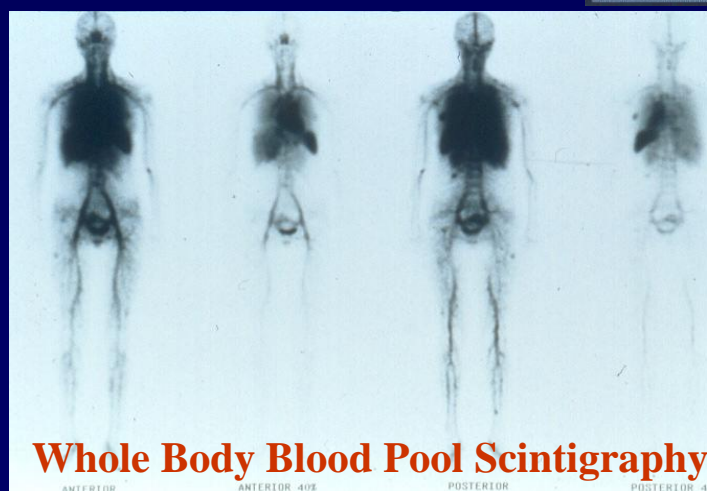
MR T-2 Image Study



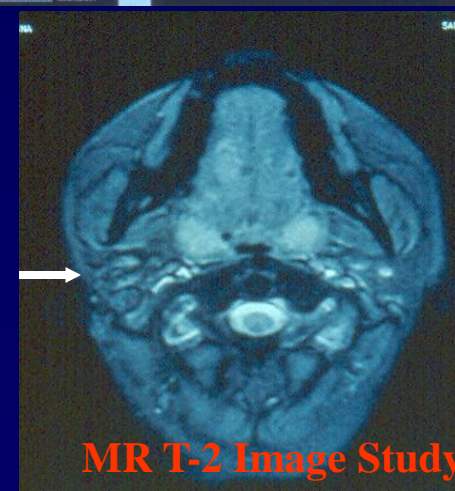
Ethanol Sclerotherapy



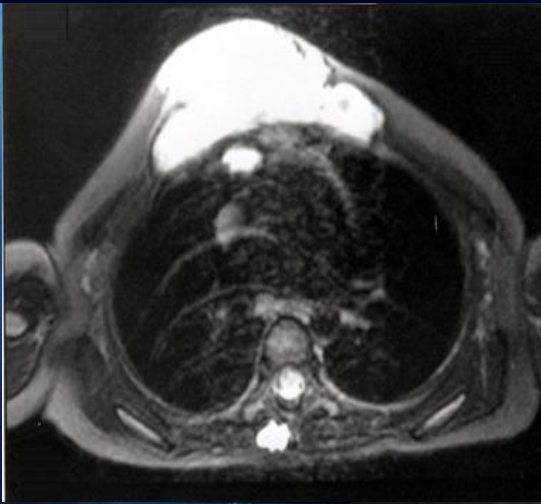
Duplex Ultrasonography



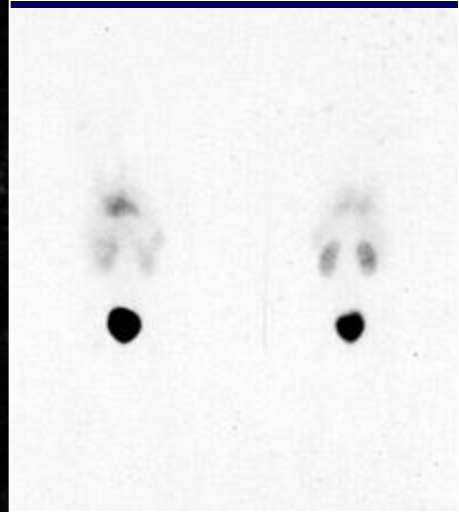
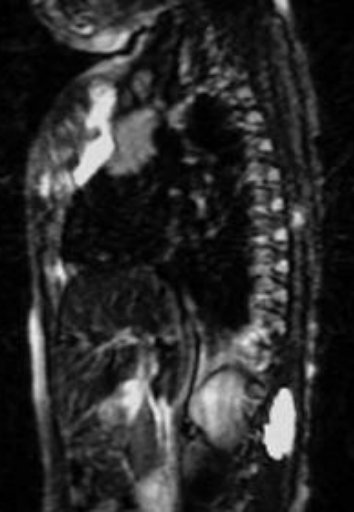
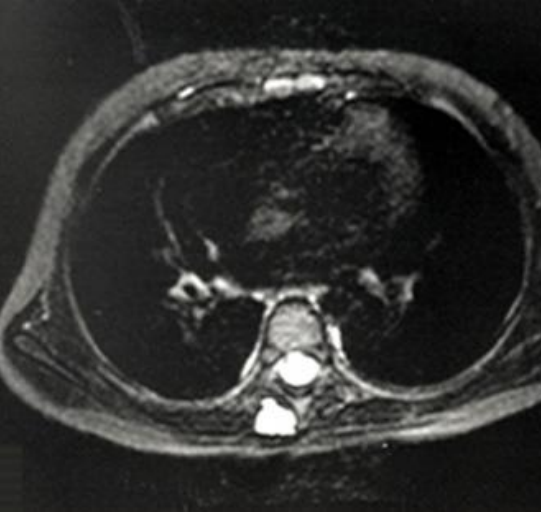
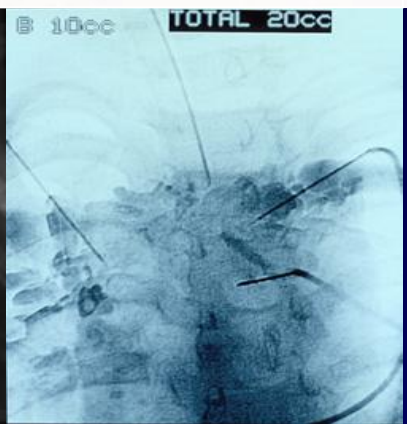
Whole Body Blood Pool Scintigraphy



MR T-2 Image Study



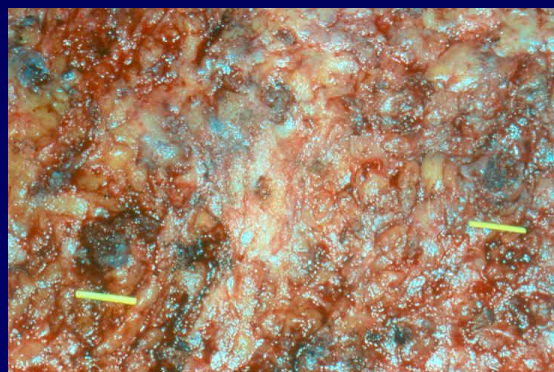
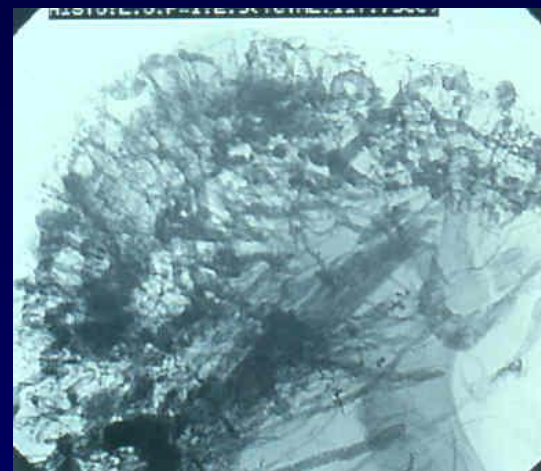
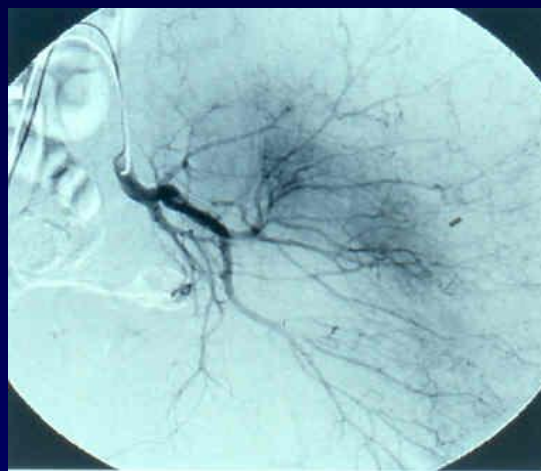
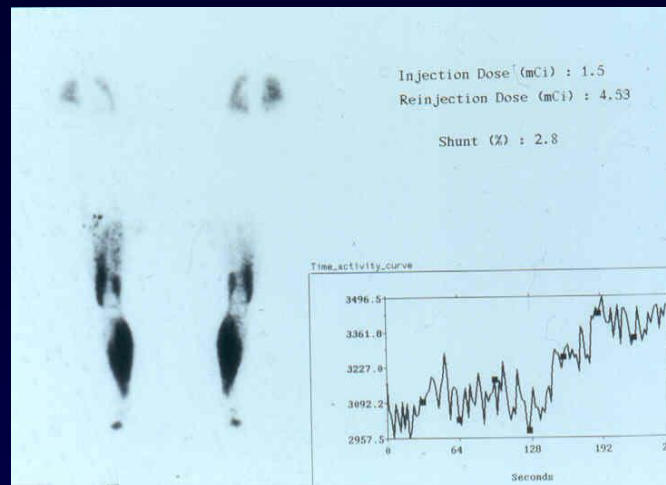
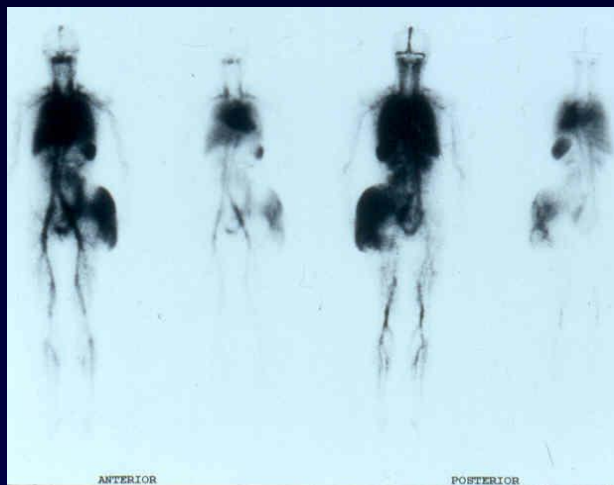
Extratruncular Venous Malformation

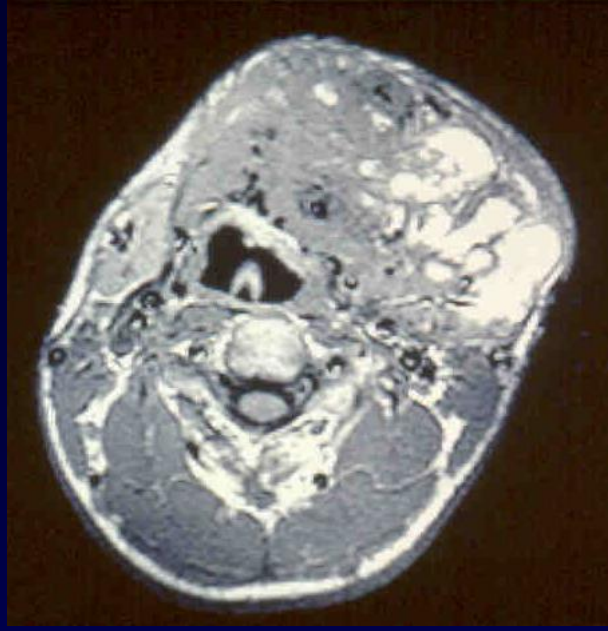


Changing Concept on the CVM

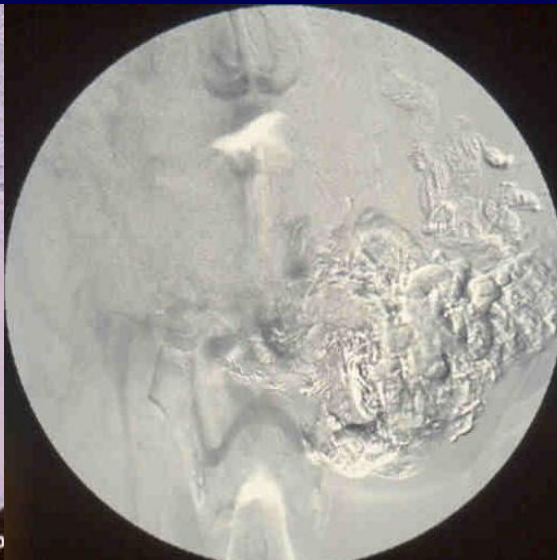
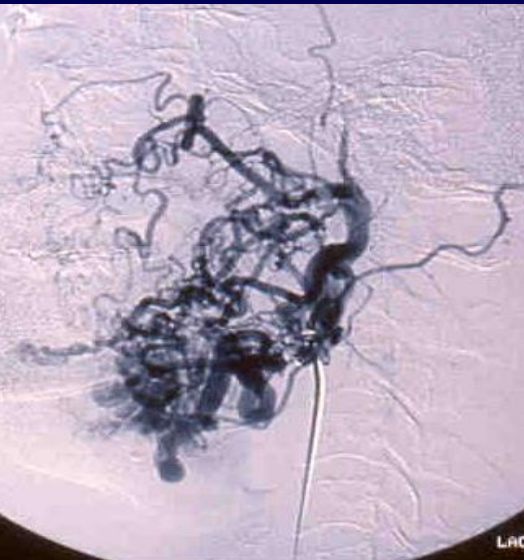
Multidisciplinary Approach

- The role of endovascular therapy is further expanded as adjunctive therapy to surgical candidate to improve the safety and effectiveness of surgical control with reduced morbidity.
- Active incorporation of the embolo/sclerotherapy pre- and/or post- operatively allows substantial expansion of the traditional role of surgical therapy alone especially for the infiltrating extratruncular form of CVM while maintaining acceptable range of surgical risk.





Infiltrating extratruncular lesions of AVM in left face



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Multidisciplinary Approach

- Treatment of CVM in general aims at the counter-action to the underlying primary pathology FIRST, especially for the correction of hemodynamic disturbances by the lesion.
- Treatment strategy has to be set up separately for the “primary” malformation itself first and then for their “secondary” disorders along the vascular and/or musculoskeletal and soft tissue system to follow.



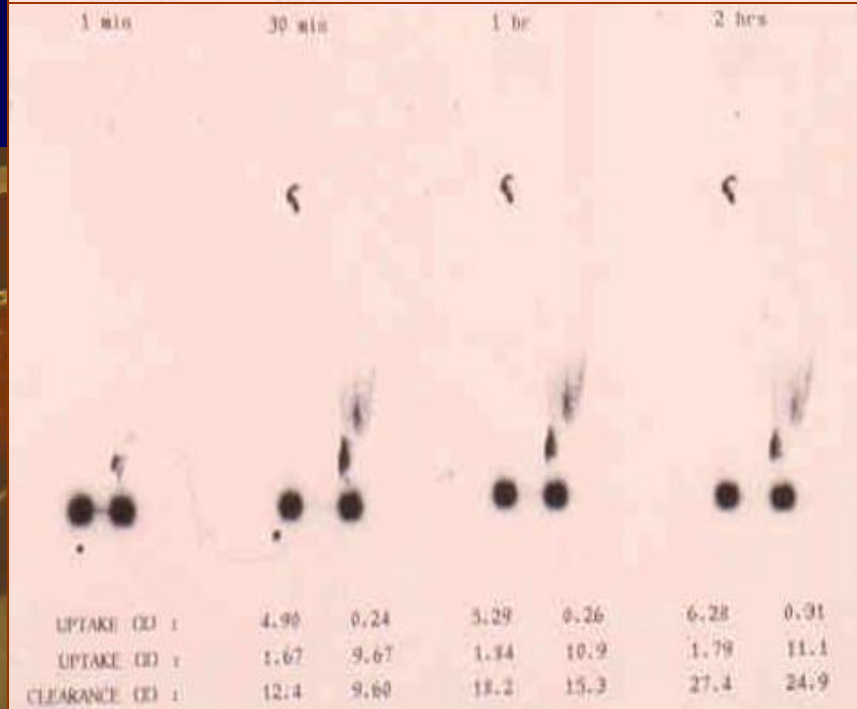
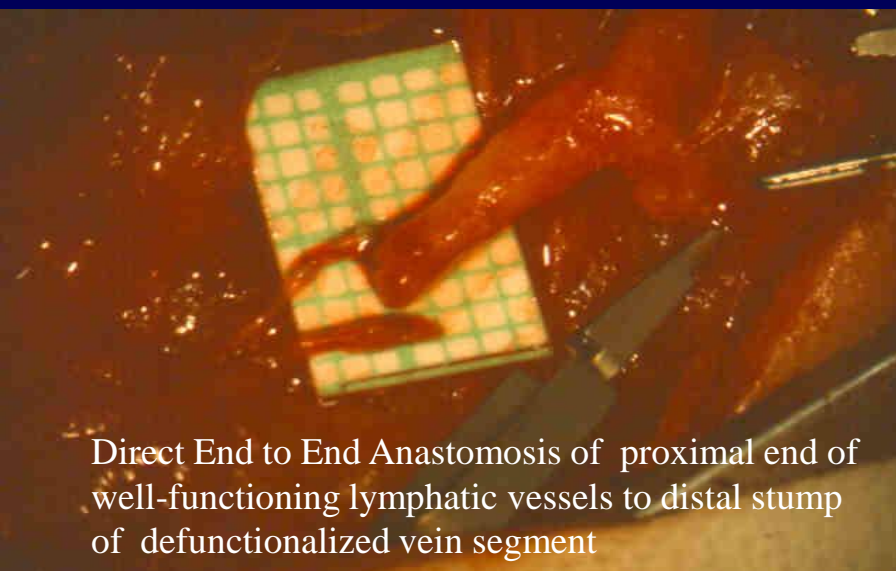
Changing Concept on the CVM

Multidisciplinary Approach - Surgical Therapy

- Vascular (hemodynamic) operation to correct hemodynamic derangement due to vascular defect should have a priority.
 - Reconstructive surgery
(e.g.) lympho-venous/lympho-lymphatic anastomotic surgery
 - Ablative surgery
(e.g.) removal of marginal vein; removal of vascular defects
- Non-vascular (non-hemodynamic) operation to correct the consequence of secondary impact by the CVM should follow.
 - (e.g.) orthopedic surgery - Achilles tendon lengthening
 - (e.g.) plastic & reconstructive surgery to correct cosmetic deformity



Lympho-venous Reconstructive Surgery





Marginal Vein - Truncular VM lesion



Changing Concept on the CVM

Conclusions

- Multidisciplinary approach with full integration of open surgical and endovascular therapy will be the main strategy for the contemporary CVM management.
- Team approach with new treatment strategy can achieve improved treatment results even to once tabooed lesion due to prohibitively high morbidity accompanied and deliver improved long term treatment results with a reduced morbidity and recurrence over the conventional approaches.

Thank you for your attention!