

XV RÉUNION DE L'ASSOCIATION EUROPÉENNE CHIVA

ANGIODYSPLASIAS or VASCULAR MALFORMATIONS

Congenital usually non hereditary rarely familial
but more frequently somatic mutations
(Dysembryogenesis)

*C. Franceschi, C. Laurian, A. Bahnini, R. Delfrate,
M. Bricchi, C. Massoni, A. Bisdorff*

?

Confuse and
complex
nosography



Dysplasias

*Angiomatous
malformation
Lymphangioma*

Angioma

Hamartoma

Phacomatosis

Hemangioma

Neurecto-mesodermosis

?

Confuse and
complex
nosography



*Kasabach-Merritt
syndrome*

*Klippel-Trenaunay-Weber
syndrome*

Proteus syndrome

*Parkes-Weber
syndrome*

Vascular tumors

- Infantile hemangiomas
- Congenital hemangiomas (RICH and NICH)
- Tufted angioma (with or without Kasabach–Merritt syndrome)
- Kaposiform hemangioendothelioma (with or without Kasabach–Merritt syndrome)
- Spindle cell hemangioendothelioma
- Other, rare hemangioendotheliomas (epithelioid, composite, retiform, polymorphous, Dabska tumor, lymphangioendotheliomatosis, etc.)
- Dermatologic acquired vascular tumors (pyogenic granuloma, targetoid hemangioma, glomeruloid hemangioma, microvenular hemangioma, etc.)

Cambridge University Press
978-0-521-84851-0 - Color Atlas of
Vascular Tumors and Vascular
Malformations
Odile Enjolras, Michel Wassef and Rene
Chapot

Vascular malformations

Slow-flow vascular malformations:

- Capillary malformation (CM)
Port-wine stain
Telangiectasia
Angiokeratoma
- Venous malformation (VM)
Common sporadic VM
Bean syndrome
Familial cutaneous and mucosal venous malformation (VMCM)
Glomuvenous malformation (GVM) (glomangioma)
Maffucci syndrome
- Lymphatic malformation (LM)



Fast-flow vascular malformations:

- Arterial malformation (AM)
- Arteriovenous fistula (AVF)
- Arteriovenous malformation (AVM)

Complex-combined vascular malformations:

- CVM, CLM, LVM, CLVM, AVM-LM, CM-AVM

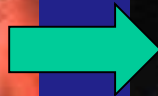
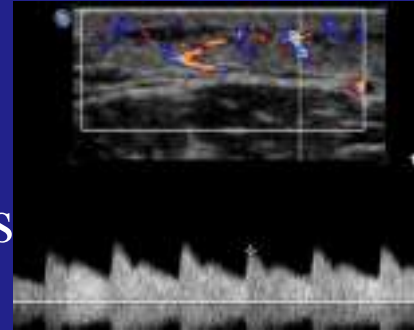
C=capillary; V=venous; L=lymphatic; AV=arteriovenous; M=malformation. RICH=rapidly involuting congenital hemangioma; NICH=noninvoluting congenital hemangioma.

Vascular tumors are not malformation but :

- Infantile hemangiomas (very frequent!!!)

Must be known because :

- differential diagnosis with vascular malformations
- benign , spontaneously involuting by 3-5 ears

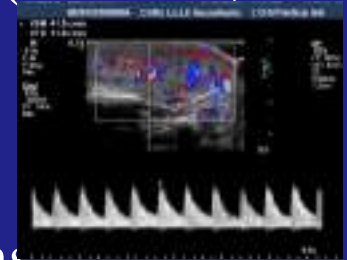


- but may be dangerous according to its location and requires specific treatment : Cortisone, Beta-blockers



Vascular tumors are not malformation but :

- Rapidly Congenital hemangiomas (RICH)
- Non involuting Congenital hemangiomas (NICH)
- Tufted angioma
- Kaposiform hemangioendothelioma



- differential diagnosis with vascular malformations
- benign BUT may be dangerous when combined with

Kasabach-Merrit syndrome (**Hemangioma with thrombocytopenia**) with disseminated intravascular coagulation and even death. **CORTISONE+++++**



CLASSIFICATION PHYSIOPATHOLOGIQUE

ENCYCLOPEDIE PRATIQUE D'ECHOTOMOGRAPHIE ET DE DOPPLER VASCULAIRE

Les malformations vasculaires congénitales

C. Franceschi*

1994

Les malformations vasculaires congénitales constituent, à plus d'un titre, un sujet particulier. En effet, ce sont des affections mal connues, non seulement en raison de leur rareté, mais aussi parce que leur classification nosologique fait encore l'objet de débats. Elles ont été décrites depuis

peuvent apparaître isolément ou associées, réalisant alors des formes mixtes.

Leur localisation, souvent métamérique, locorégionale et superficielle, peut aussi être profonde, diffuse ou plurifocale. Elle peut atteindre aussi bien les membres que le tronc, la tête et le cou.

Les signes d'appel sont habituellement cliniques, soit dès le premier

sables d'aggravations. Pour ces raisons, les traitements sont devenus moins ambitieux quant aux résultats mais plus efficaces et moins risqués grâce à une meilleure évaluation de chaque malformation selon sa nature, sa topographie et ses caractéristiques hémodynamiques, permettant de mesurer le meilleur compromis risque-bénéfice. L'éventail thérapeutique s'étend de la

Modified Hamburg Classification

Main classification based on its predominant vascular component

- Arterial defects
- Venous defects
- A/V (arteriovenous) shunting defects
- Lymphatic defects
- Capillary defects
- Combined vascular defects

1993-2012

*Based on the consensus on CVM through the international workshop in Hamburg, Germany, 1988

Embryological subclassification based on its embryological stage of the defect developed

- (1) Extratruncular forms – developmental arrest at the earlier stages of embryonal life
 - Diffuse, infiltrating
 - Limited, localized
- (2) Truncular forms – developmental arrest at the later stages of embryonal life
 - Aplasia or obstruction
 - Hypoplasia; aplasia; hyperplasia
 - Stenosis; membrane; congenital spur
 - Dilation
 - Localized (aneurysm)
 - Diffuse (ectasia)

*Developmental arrest at the different stages of embryonal life: earlier stage – extratruncular form; latter stage – truncular form

*Both forms may exist together; may be combined with other various malformations (e.g. capillary, arterial, A/V shunting, venous, hemolymphatic and/or lymphatic); and/or may exist with haemangioma

CVM, congenital vascular malformation

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or
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Regarding the early angiogenesis

COMBINED MALFORMATIONS

Vascular

Artérielles

Artério-veineuses

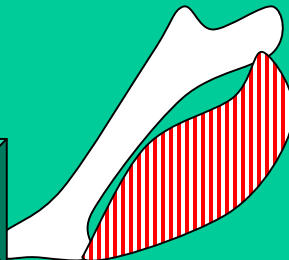
Veineuses

Artério-veino-lymphatiques

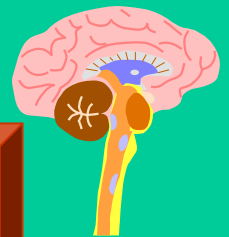
Veino-lymphatiques

Lymphatiques

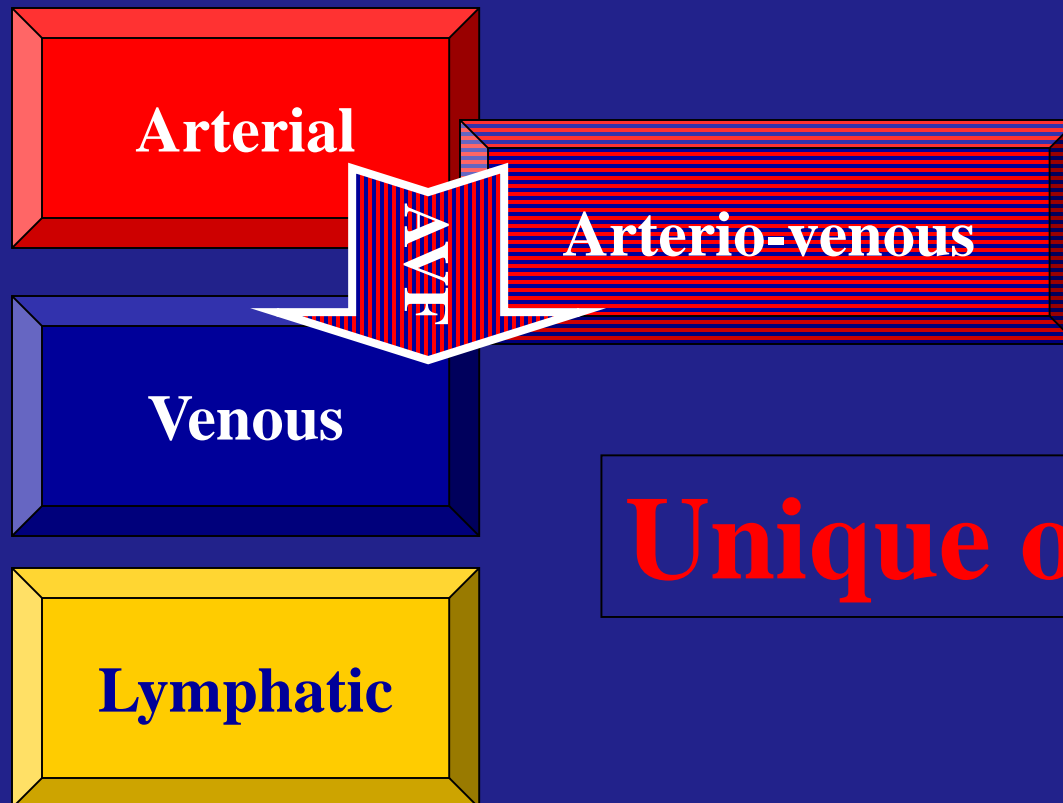
OSTEO-MUSCULAR
DYSTROPHY



NEURO-CUTANEOUS
DYSTROPHY



ANGIODYSPLASIAS or VASCULAR MALFORMATIONS



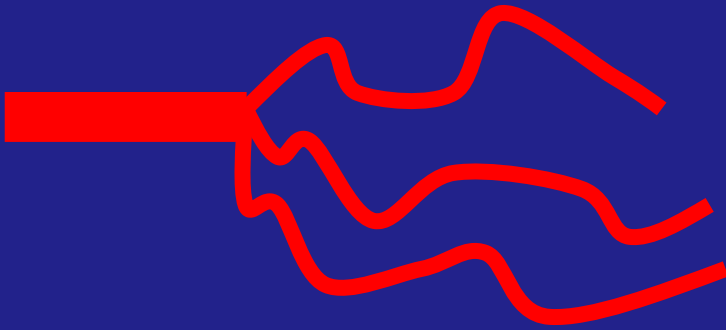
Unique or associated



Arterial Malformations

**Arterial
Capillary**

Mega arterial capillaries



**Low
Flow**



Arterial Truncular

Hypoplasia,
Coarctatio

Aplasia ,

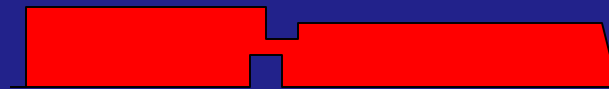
Megadolichoarteries

Vestigila aneurism

Fibro-muscular dysplasia



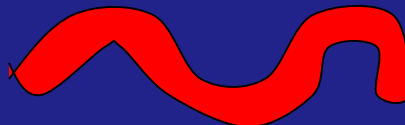
AORTA



CERVICO-



CEREBRAL ARTERIES



RENAL ARTERIES



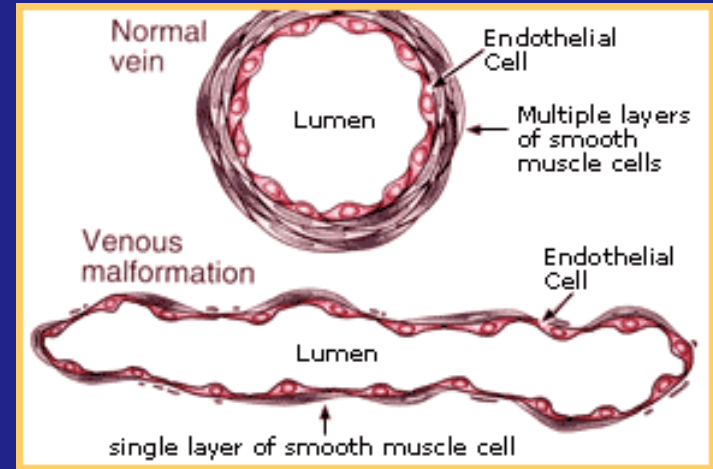
LIMBS ARTERIES





Venous Malformations

**Venous
Capillary
Extra-truncular**



Simple Venous Mega capillary

Nodular Venous Mega capillary

Cavernous Venous Mega capillary

Risk of Kasabach-Merrit syndrome

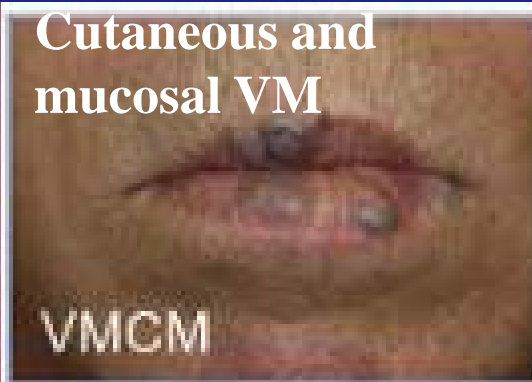


Rare but important venous capillary M to know

Glomuvenous M



Cutaneous and mucosal VM



Hyperkeratotic cutaneous capillaryvenous M



VMCM



MAFFUCCI SDR

Blue Rubber Bleb Nevus Sd (Bean)

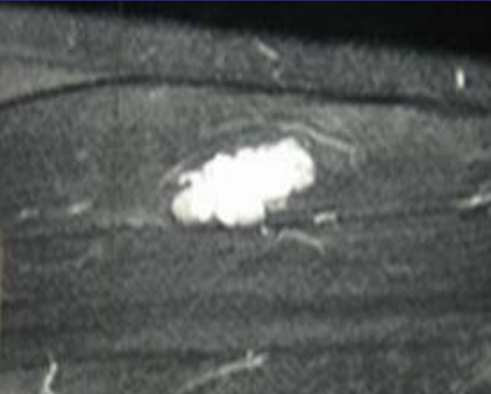
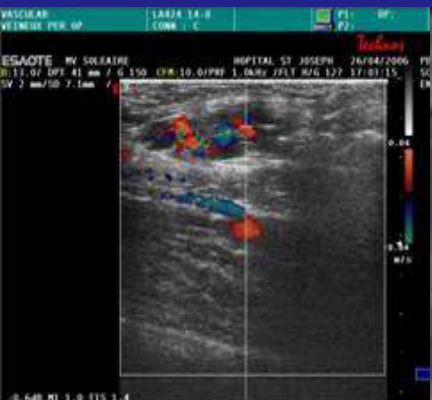


BRBN

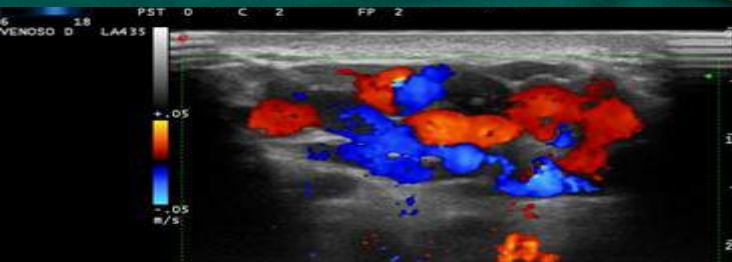
+Cartilage benign tumor + malignant T risk!

Vital risk! Intestinal bleeding location

Usual venous capillary deep /superficial VM



Deep intramuscular VM

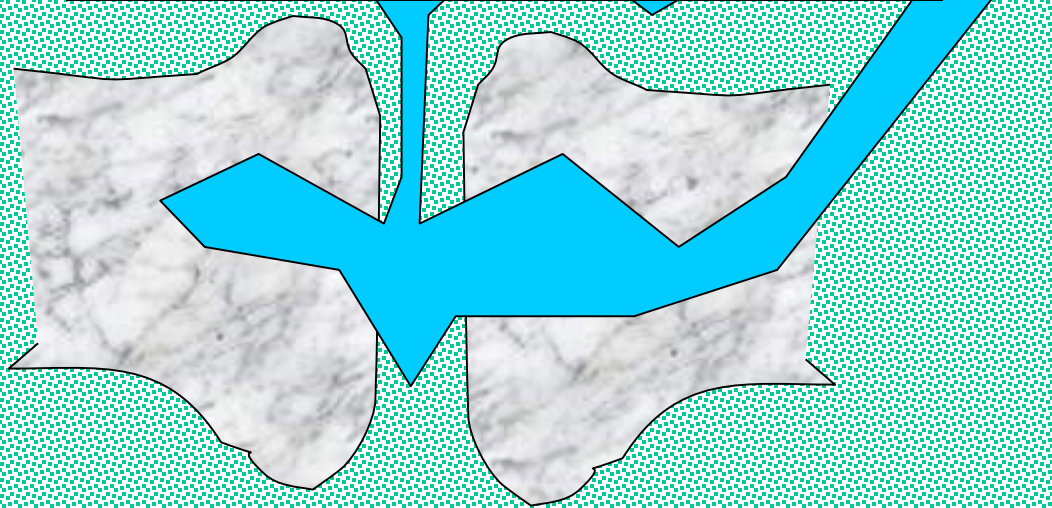


Deep extra_muscular VM



Deep/Superficial extra_muscular VM

Knee synovial
location: bleeding,
hemartrosis then
joint destruction



NON DRAINING VM Treatment


- Indication

- Pain, Edema, cosmetic
- Complications
 - Thrombosis
 - PE
 - Knee hemartrosis

- Means:

- Compression, aspirin
- Ablative Surgery whenever feasible
- Laser, sclerotherapy in addition to surgery or when surgery is not feasible . Ethanol must be prohibited (DANGEROUS !!!!!)

Bleeding control



Delacroix
Chevalier

Take now **ALL THE TIME** you need for vascular suture, even in alarm condition of the patient

Prevenir les saignements. Réduire le temps de suture vasculaire. Réduire les situations critiques.

Dr Franceschi's
HEMOSTATOR

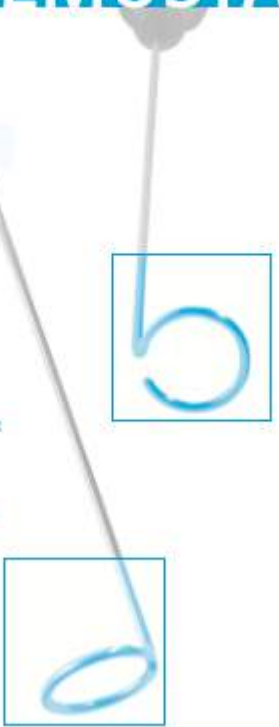
forme rectangulaire

The ideal Hemostasis Safety tool:

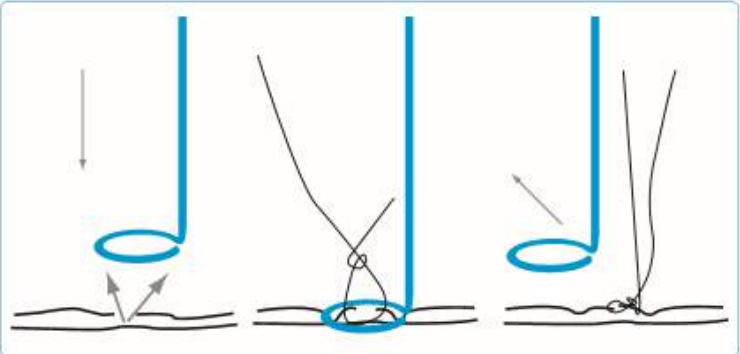
- > Immediate control of haemorrhage
- > Dramatic drop down of blood loss
- > Drying of operative field
- > Usable in every operative field even on non clamping zones (thoracic, pelvic vessels, sclerosed tissues...)

L'outil idéal d'hémostase de sauvetage :

- > Contrôle immédiat de l'hémorragie
- > Réduction majeure des pertes sanguines
- > Assèchement du champ opératoire
- > Utilisable partout où il y a des vaisseaux (thoraciques, pelviens, en zone de tissus sclérosés...)



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Les faits :


- > Une suture vasculaire accidentelle provoque une situation stressante et haut risque
- > Les veines sont plus fragiles, moins visibles et plus difficiles à disséquer et clamer que les artères
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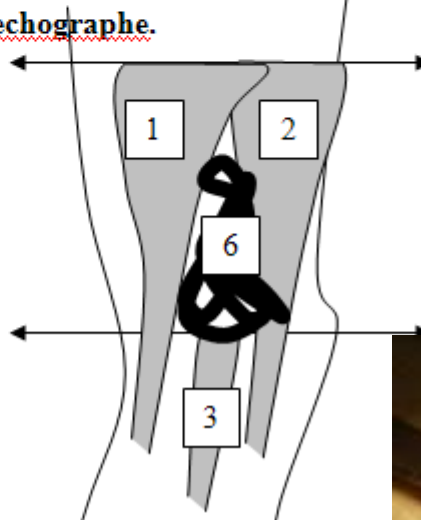
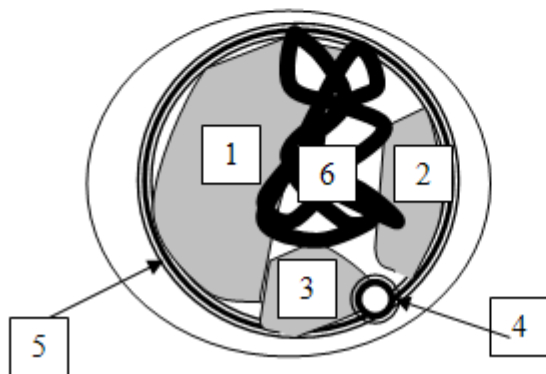
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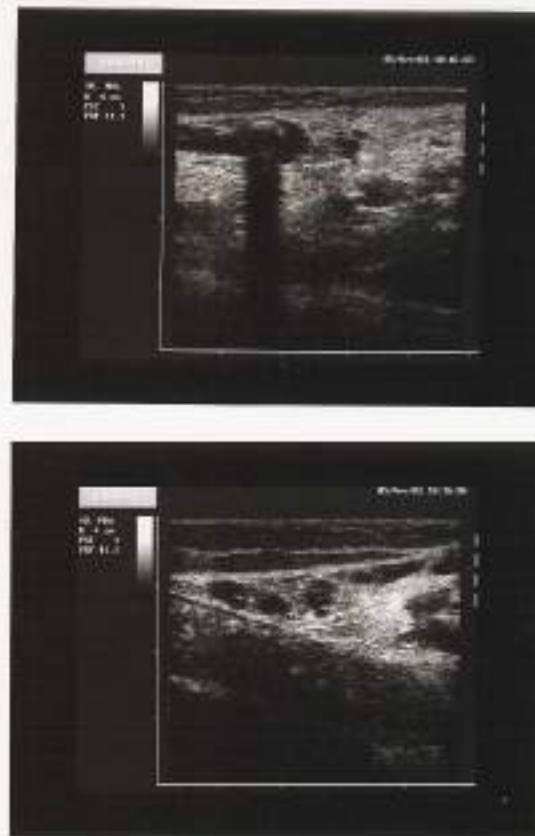
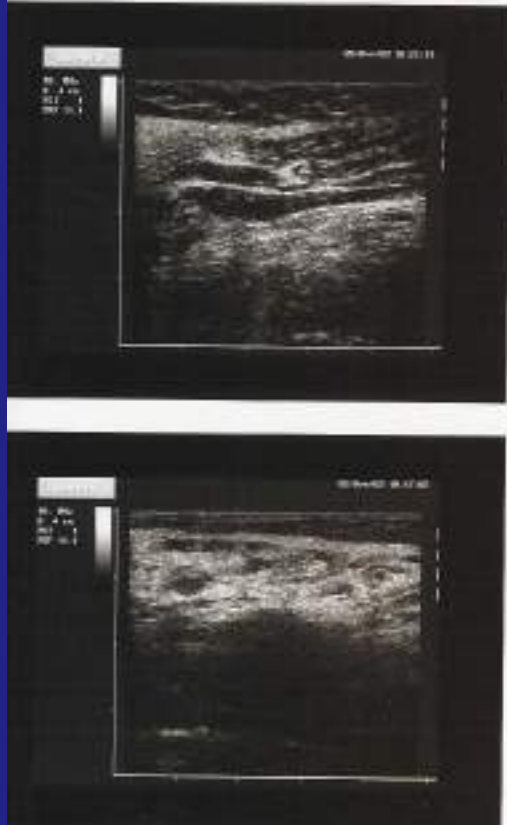
Les TRONCS VEINEUX PROFONDS des deux membres inférieurs sont normaux, sans incontinence, ni thrombose ni altération pariétale et sans séquelles post-phlébitiques.

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Les TRONCS VEINEUX PROFONDS des deux membres inférieurs sont normaux, sans incontinence, ni thrombose ni altération pariétale et sans séquelles post-phlébitiques. SAUF: ANGIOME VEINEUX 6 capillaire intrant partiellement caverneux (partiellement compressible et liquide), sous-aponevrotique 5, large de 17 mm et profond de 16 mm au 1/4 inf de la loge ant de jambe gauche (78 mm de haut) limité en dedans par le muscle et tendon jambier ant. 1, en dehors par le muscle et tendon extenseur commun 2 et en arrière par l'extenseur propre du GO 3 et le paquet vasculo nerveux tibial ant 4 qui ne semble pas être infiltré.

Dessin fait sur la peau: et photos sauvegardées dans echographie.



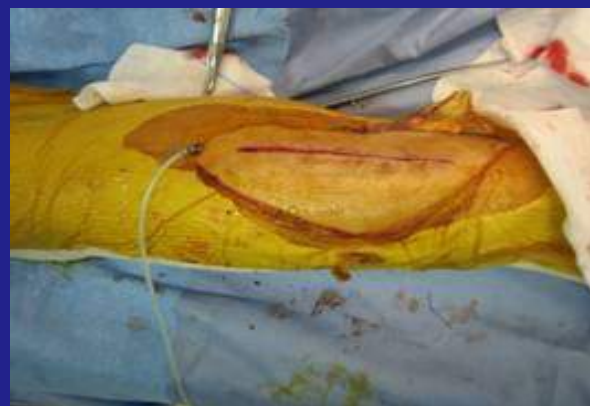
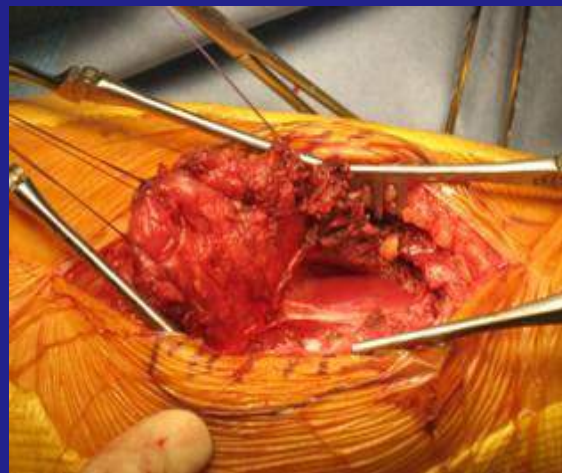
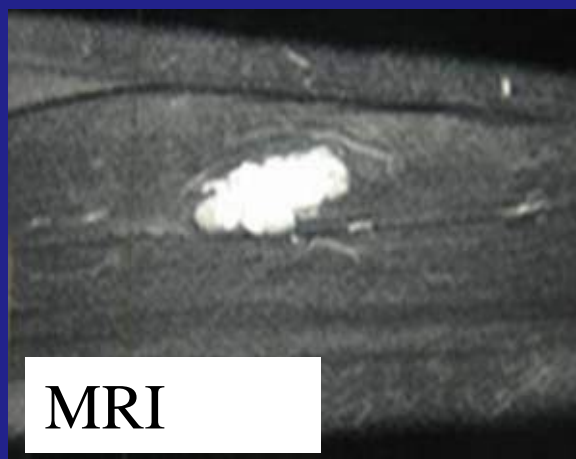
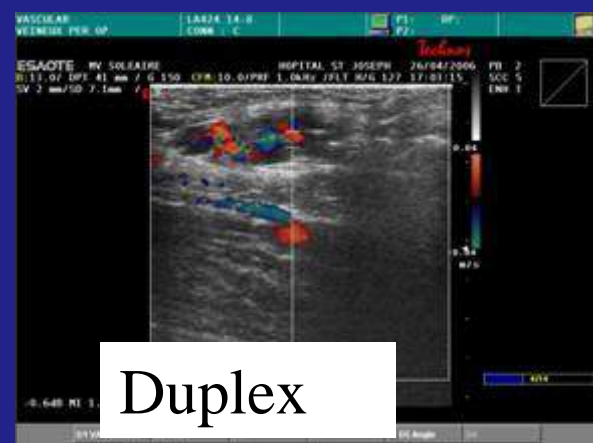


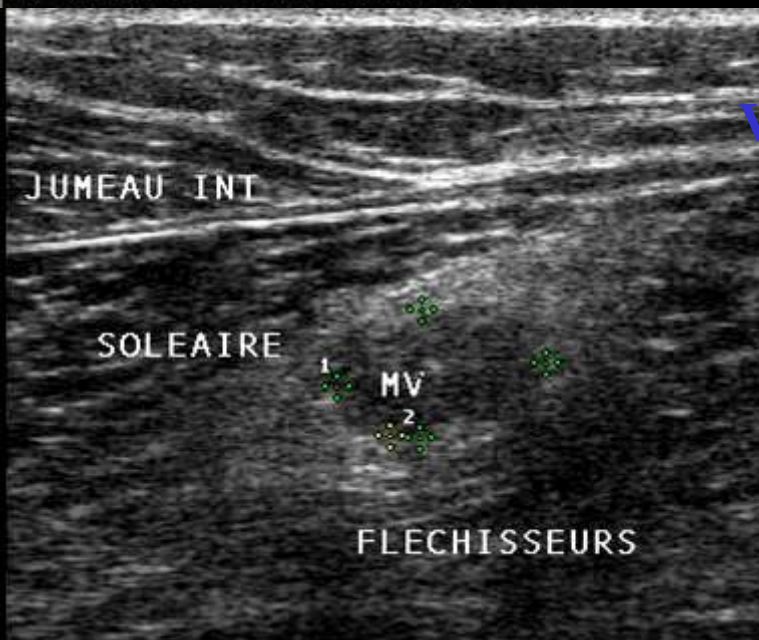
**VM of gastrocnemius
(caput mediale)**



Medial gastrocnemius excision





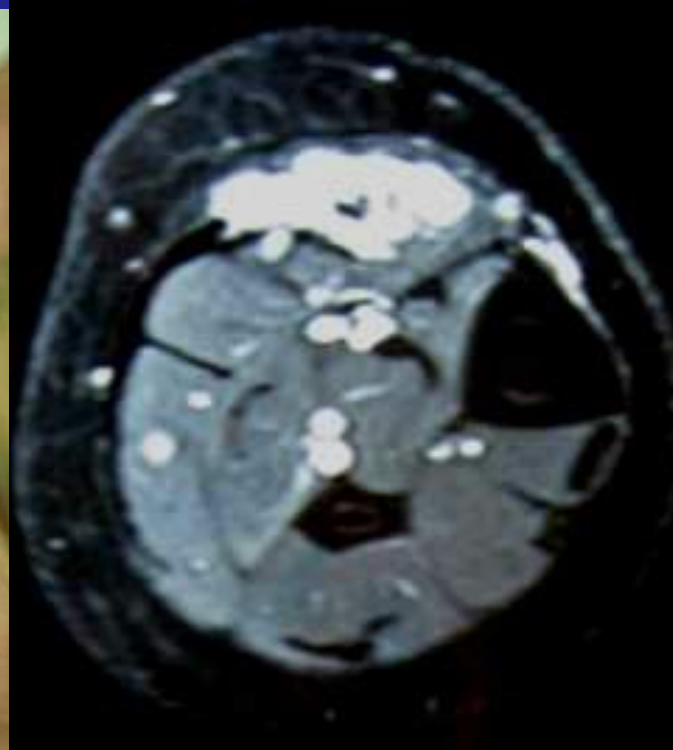
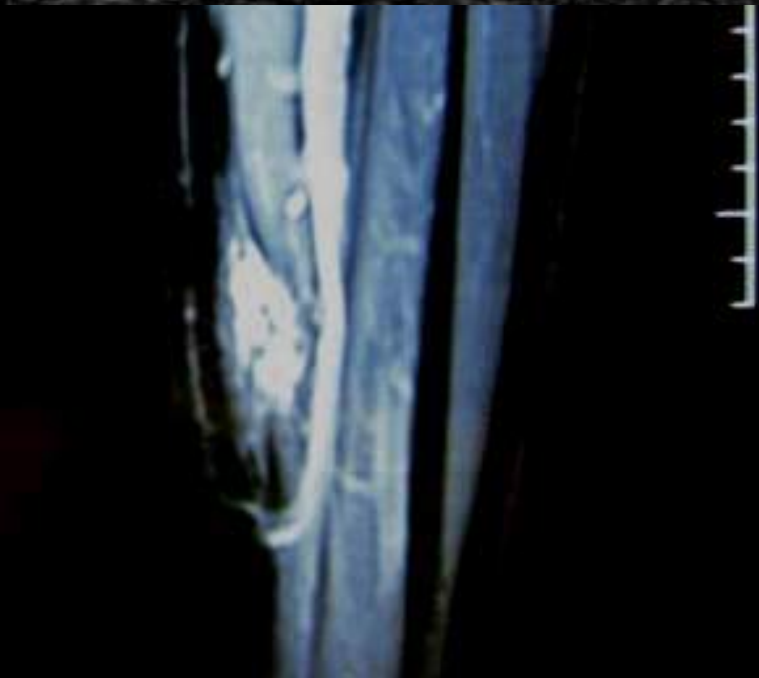
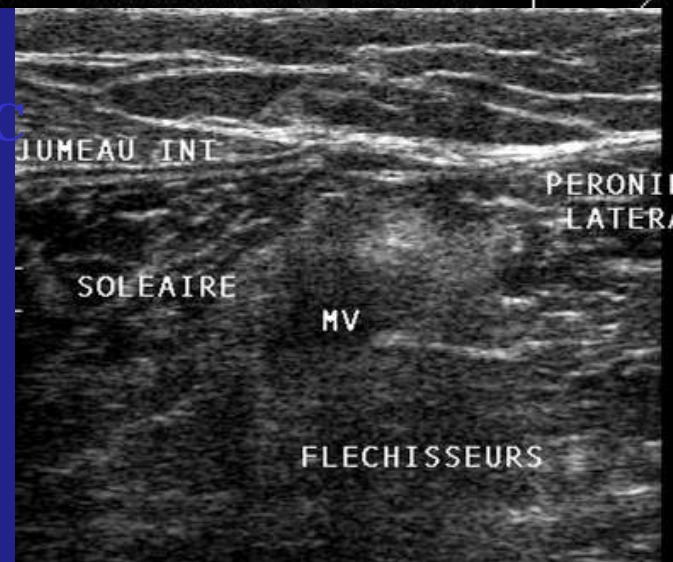


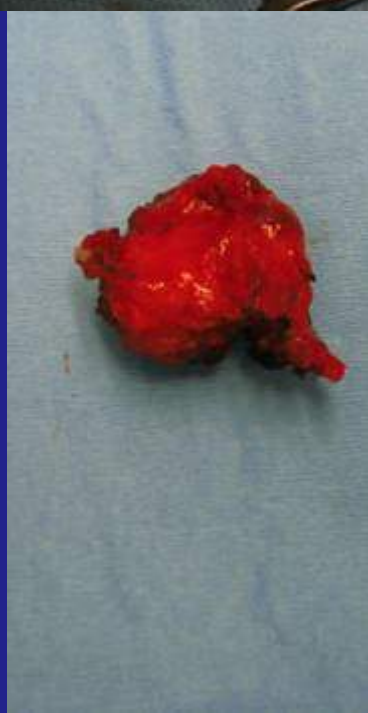
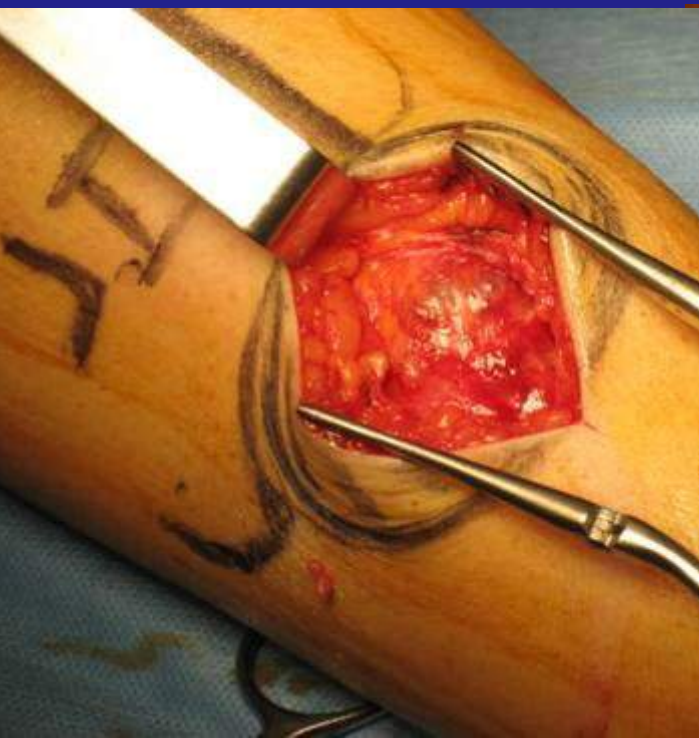
MALFORMATION VEINEUSE DU MUSC

SEchodoppler

IRM

**Marquage
pré_opératoire**





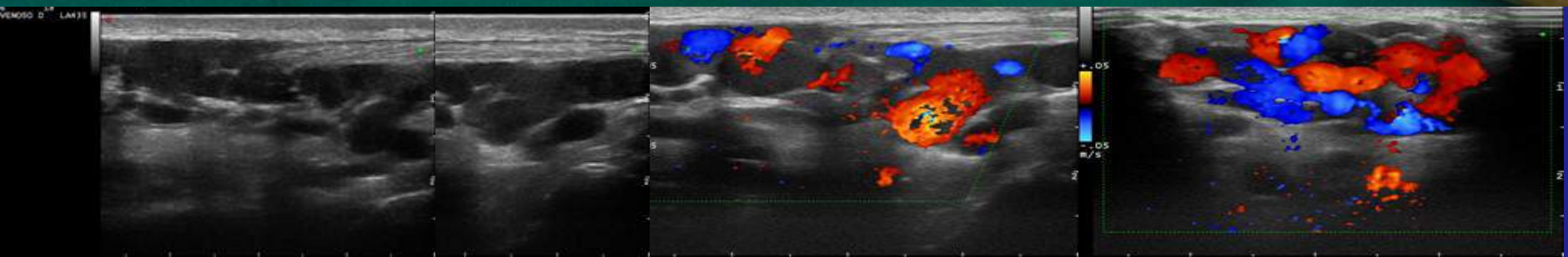
**MALFORMATION
VEINEUSE DU
MUSCLE SOLEAIRE**

**EXERESE
ECHOGUIDEE**



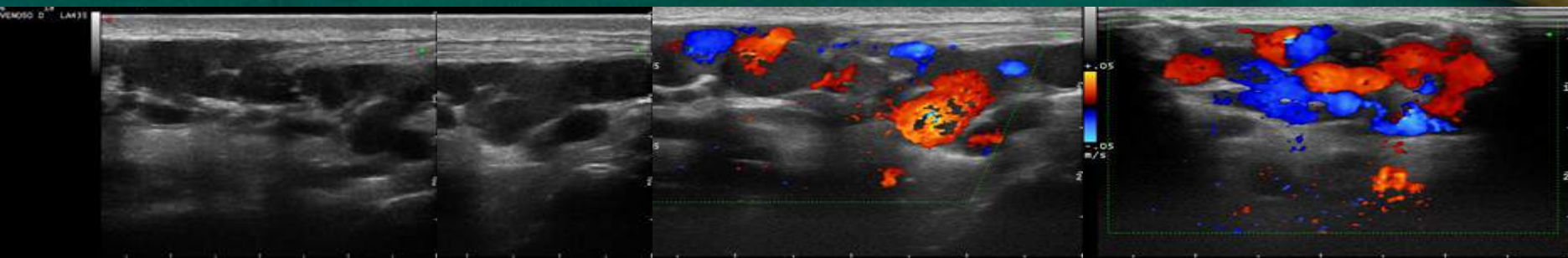


VM sub aponeurotic Ant. Tibial compartment. Previous foam treatment. Pain increased after foam (1 year) . VM unchanged



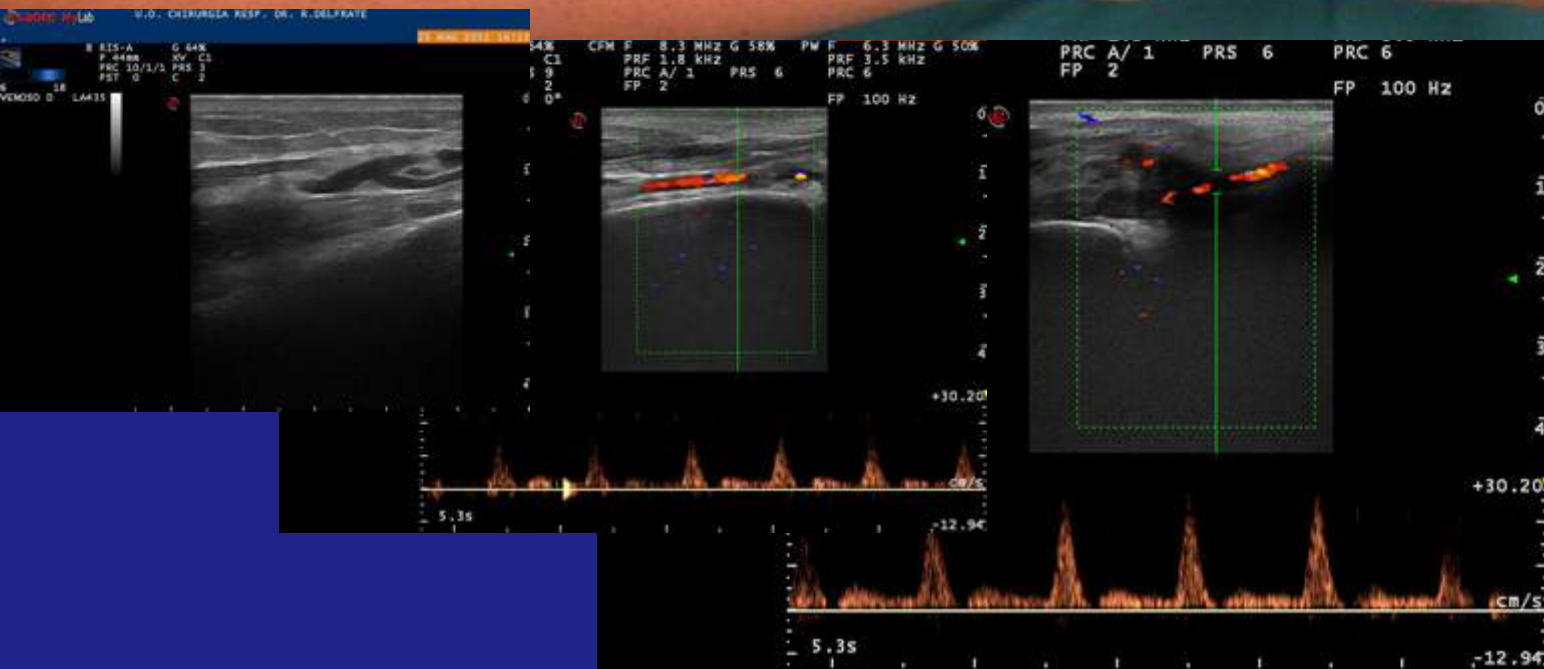


Echo guided surgical exhaustive ablation





30 days post Op. Swelling
dramatically reduced. No pain.





CAVA VEIN

DEEP and SPRFICIAL VEINS

Venous Troncular

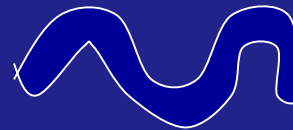
Hypoplasia



Aplasia



Varicose veins



Avalvulation.



Venous Troncular

CAVA VEIN

DEEP and SPRFICIAL VEINS

Asymptomatic,

Cosmetic

Pain

Limited function

Thrombosis,

Pulmonar Emboly (chronic....)



Venous Truncular

Duplex anatomic and
hemodynamic
mapping BECAUSE
most malformative
veins are draining
pathways which are
to be respected

CAVA VEIN

DEEP and SPRFICIAL VEINS



DRAINING VM

Treatment

- Indication


- Pain, Edema, cosmetic
- Complications
 - Thrombosis
 - PE

- Means:

- Compression, aspirin
- CHIVA whenever feasible
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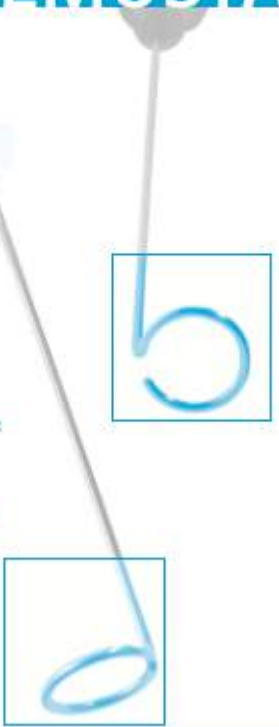
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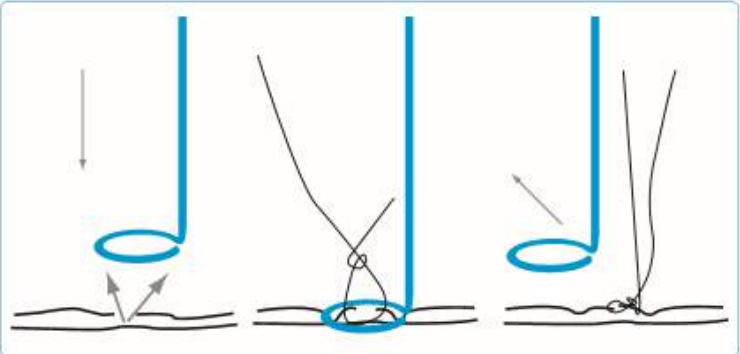
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Réf. DC53200 - 20



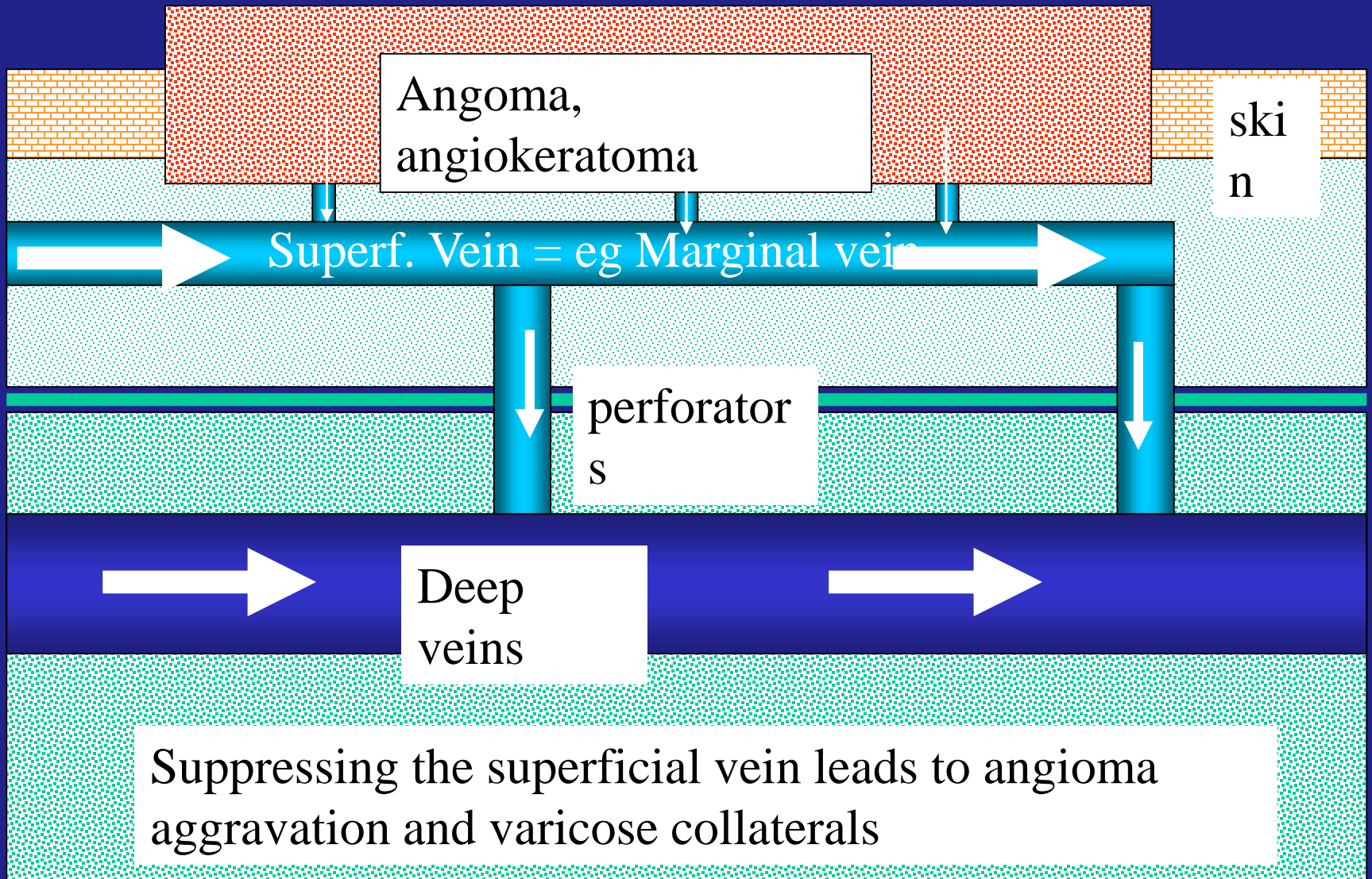
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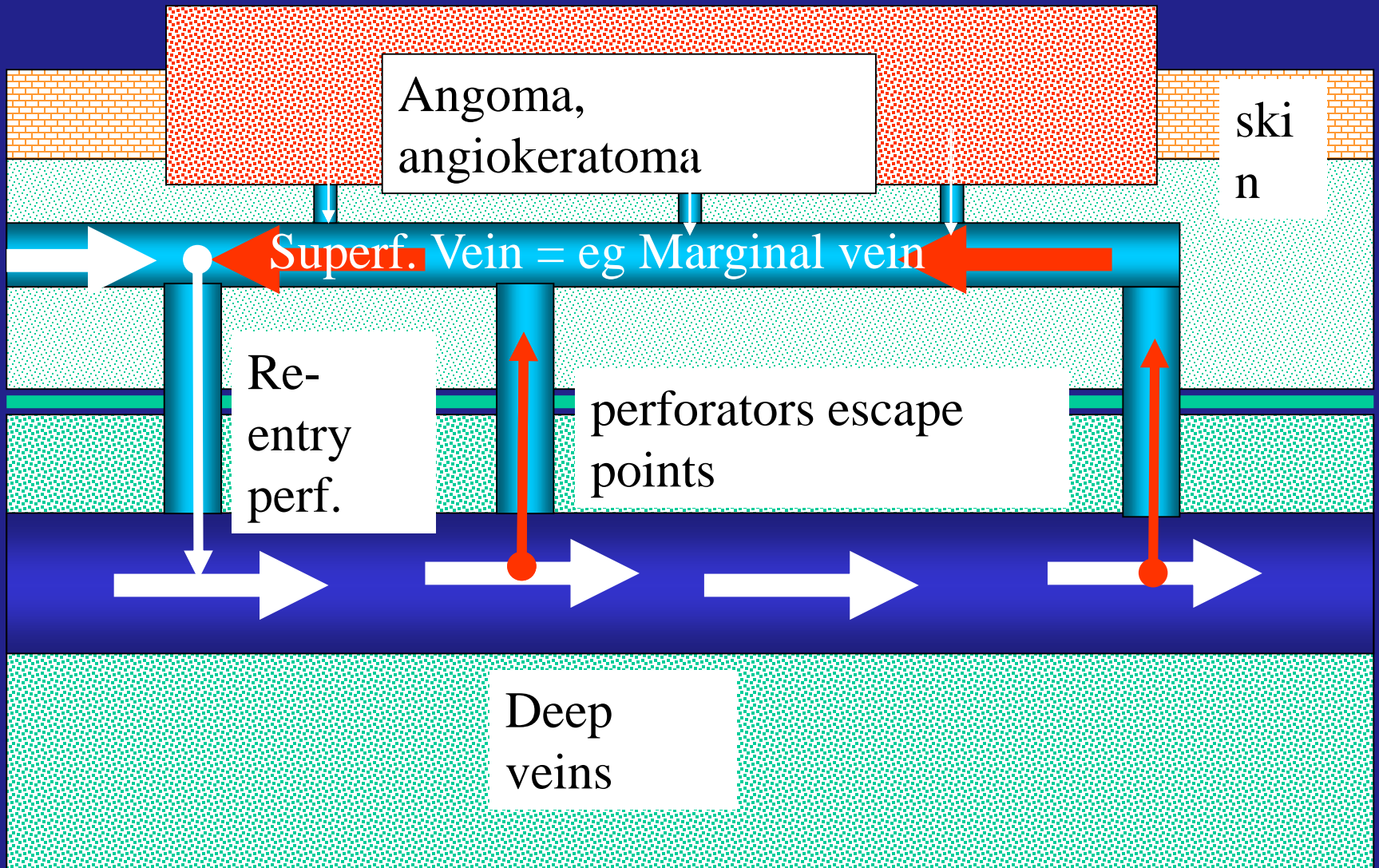
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Pattern 1



Cutaneous angioma : normal draining venous network

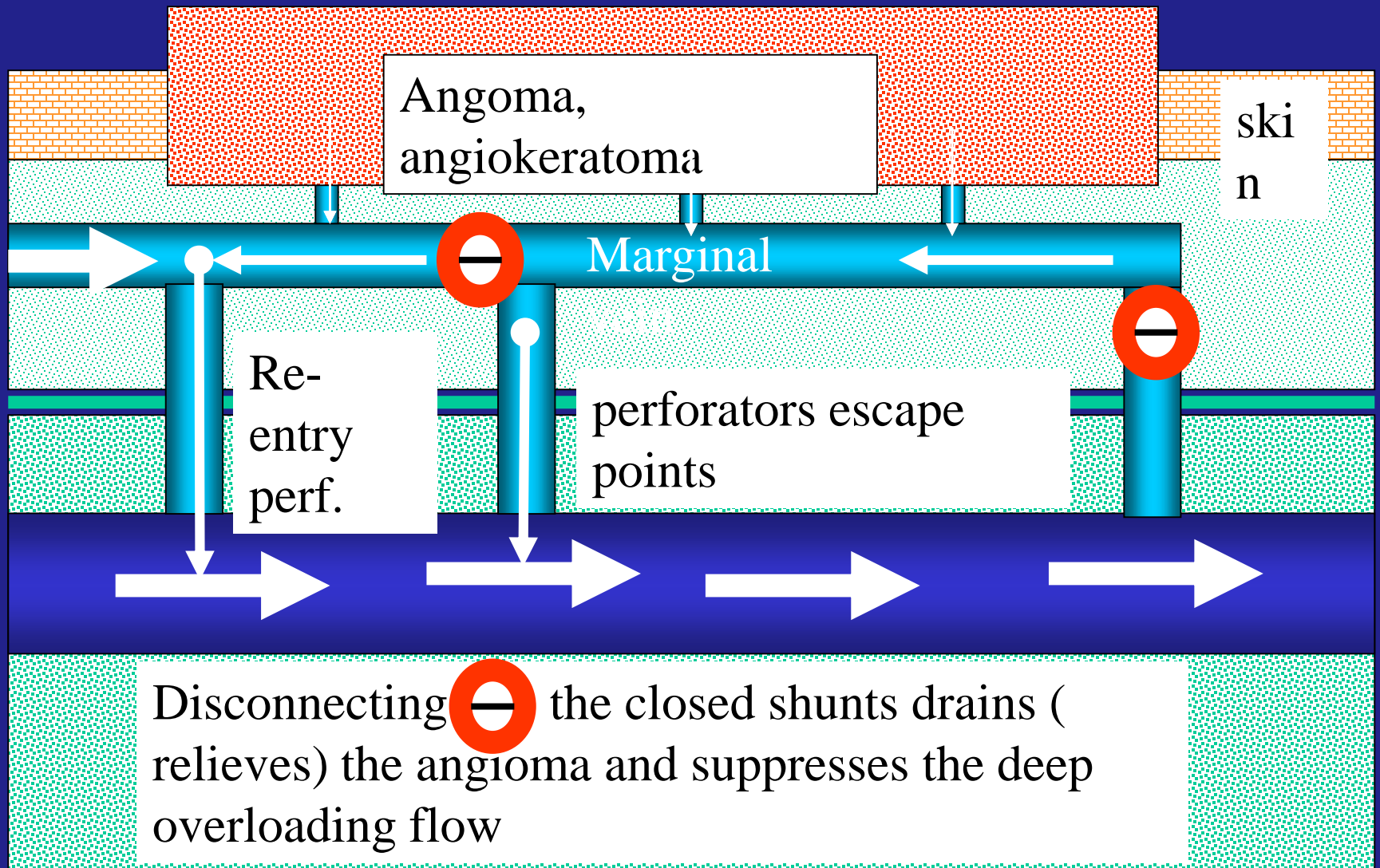
Pattern 2



Cutaneous angioma : incompetent superficial vein: **Closed shunt**

Overloaded: Angioma flow + Deep diverted retrograde flow (closed circuit)

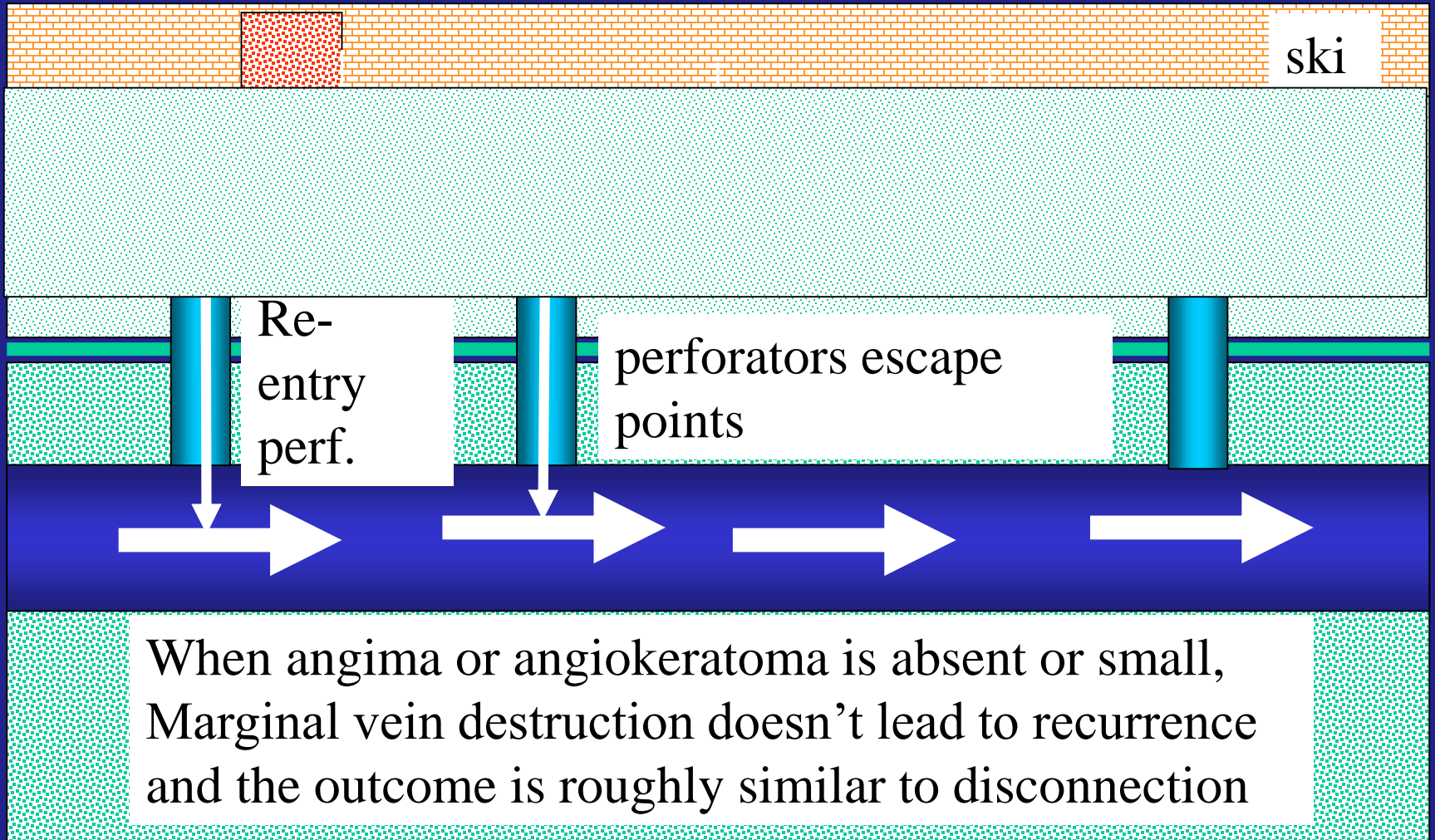
Pattern 2 treatment



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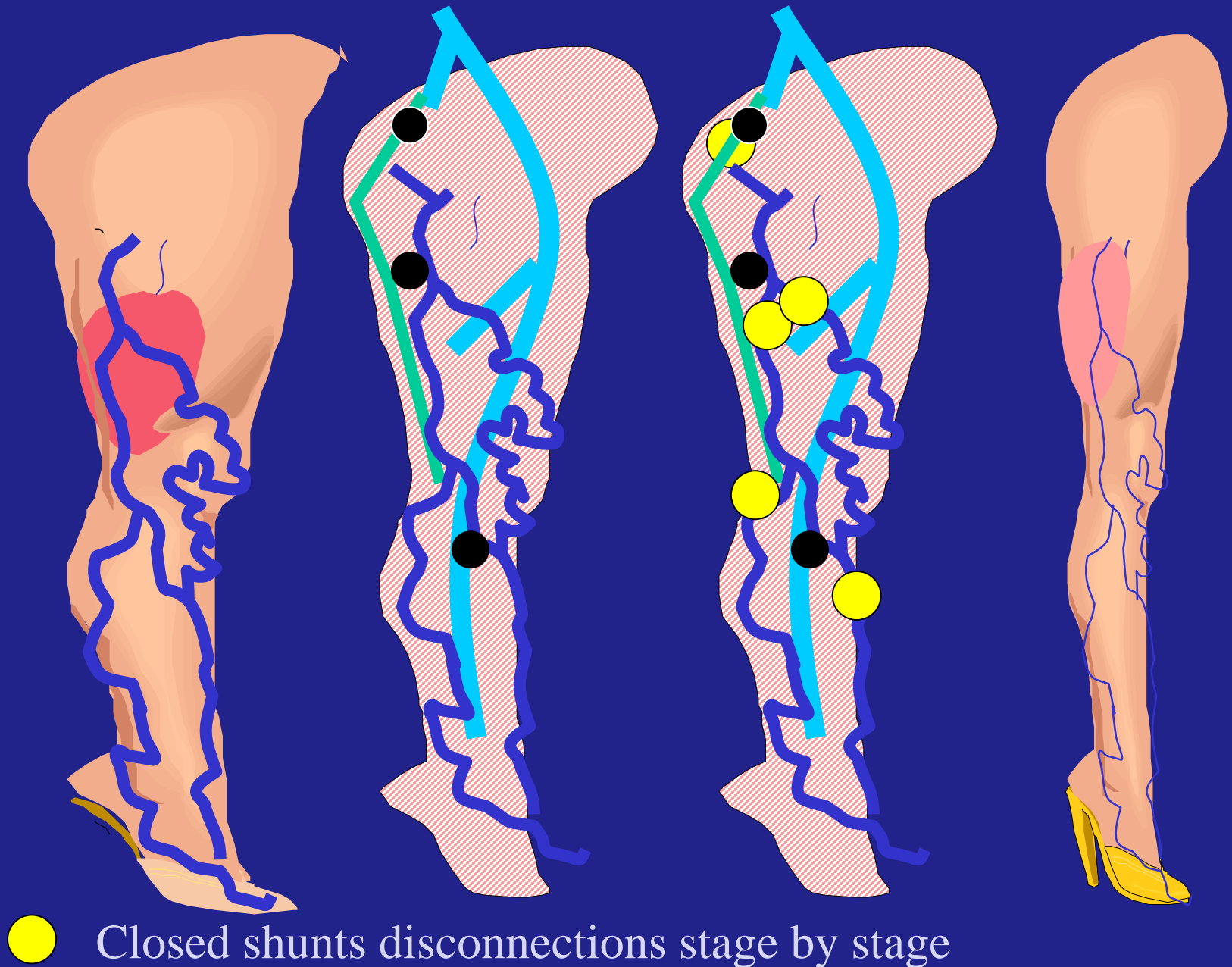
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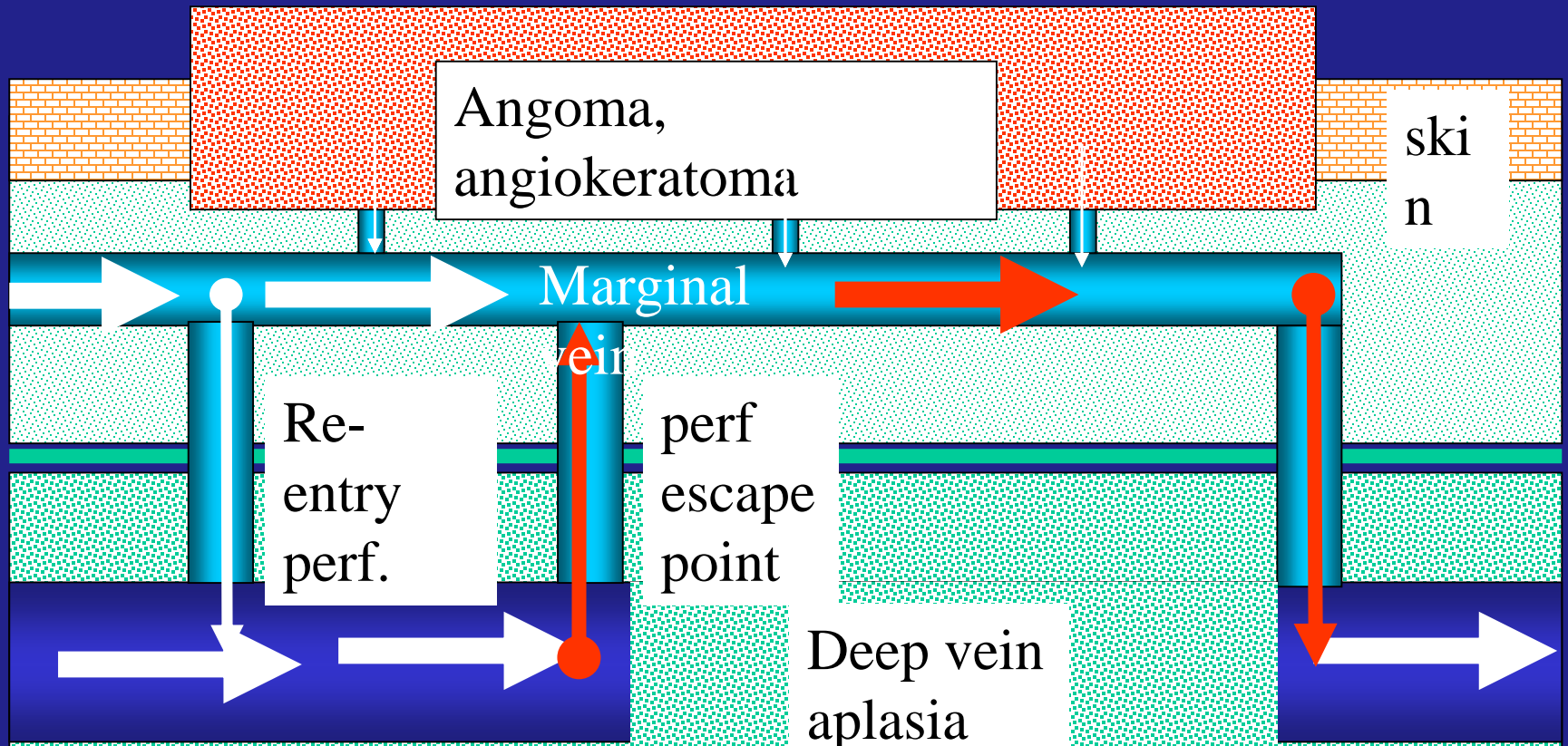
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Overloaded: Angioma flow + Deep diverted retrograde flow (closed circuit)

Pattern 2 treatment



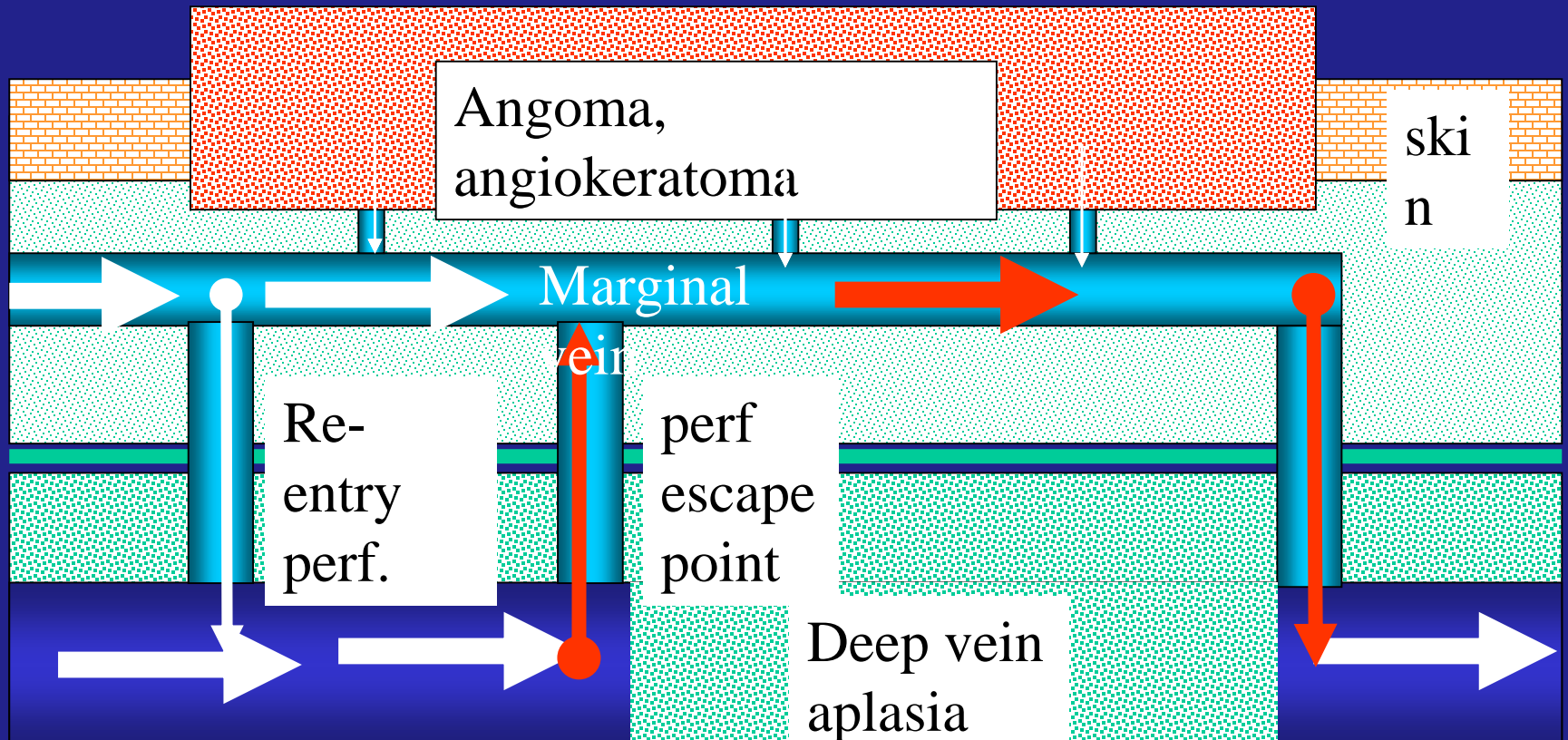
Pattern 3



Open Vicarious shunt is preserved in order to avoid the dangerous block of the deep and superficial draining flow and further varicose collaterals

Cutaneous angioma : incompetent superficial vein: **Open Vicarious Shunt**
Overloaded: Angioma flow + Deep antegrade flow + segmental Deep veins aplasia (open circuit)

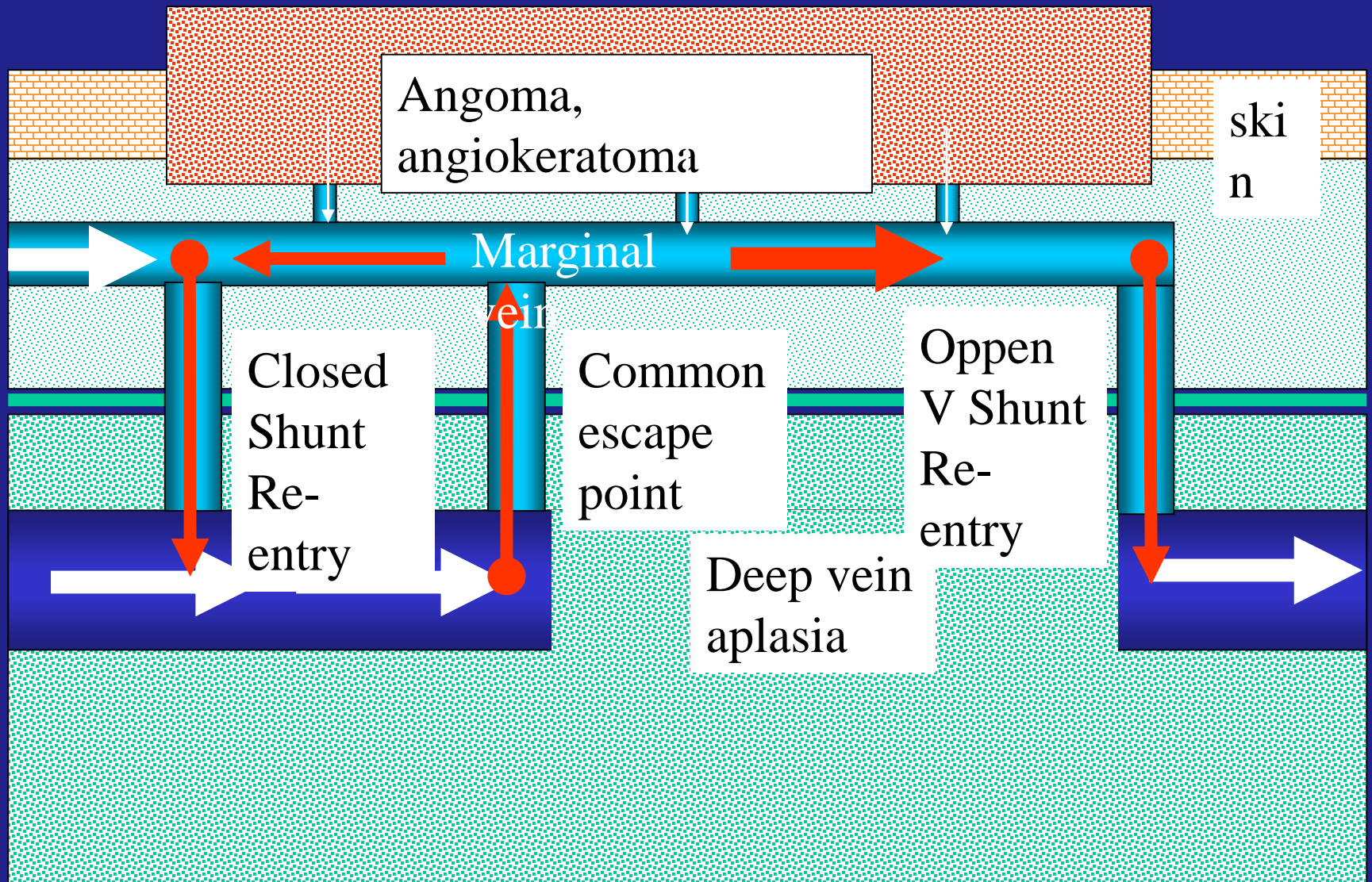
Pattern 3 treatment =0



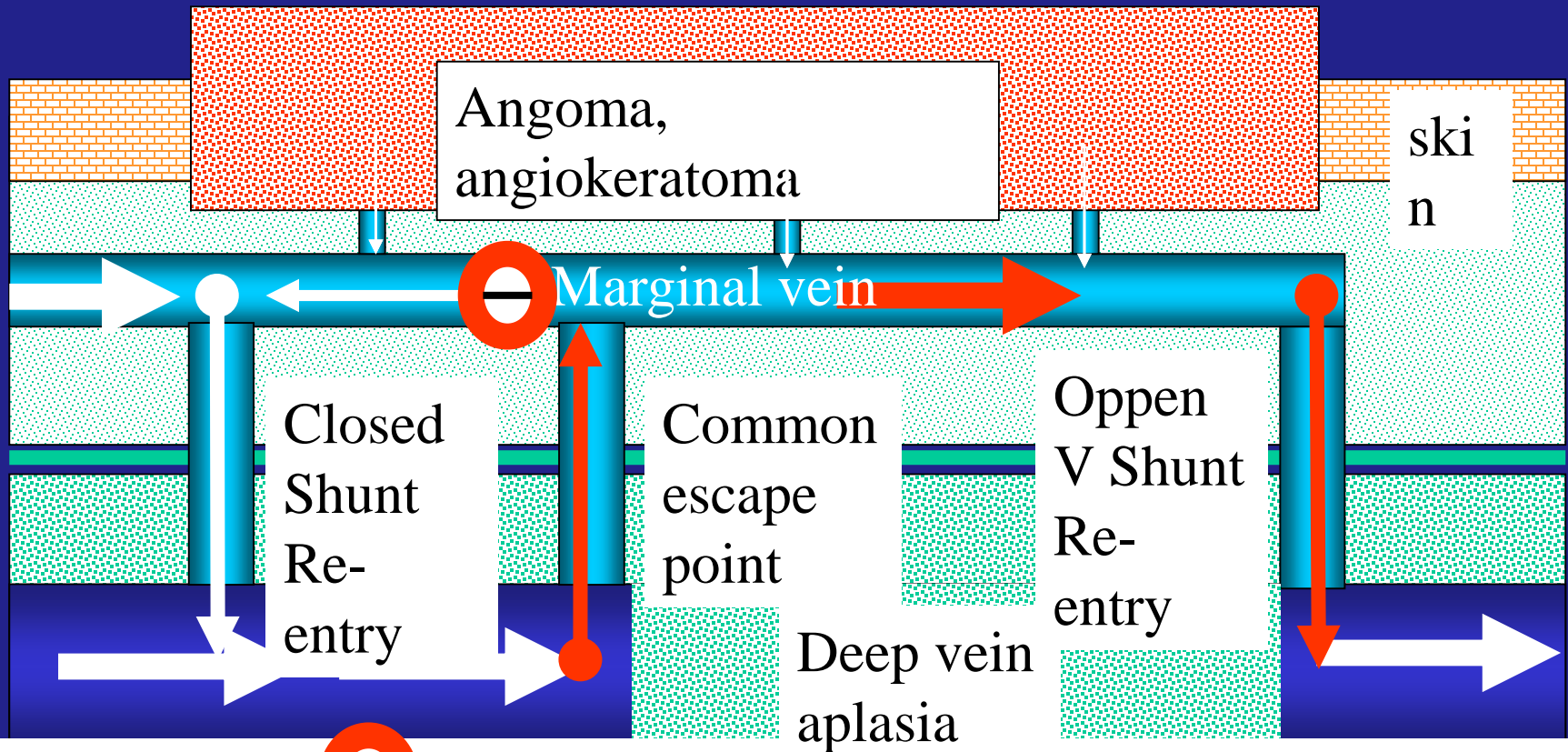
Open Vicarious shunt is preserved in order to avoid the dangerous block of the deep and superficial draining flow and further varicose collaterals


Cutaneous angioma : incompetent superficial vein: **Open Vicarious Shunt**
Overloaded: Angioma flow + Deep antegrade flow + segmental Deep veins aplasia (open circuit)

Pattern 4



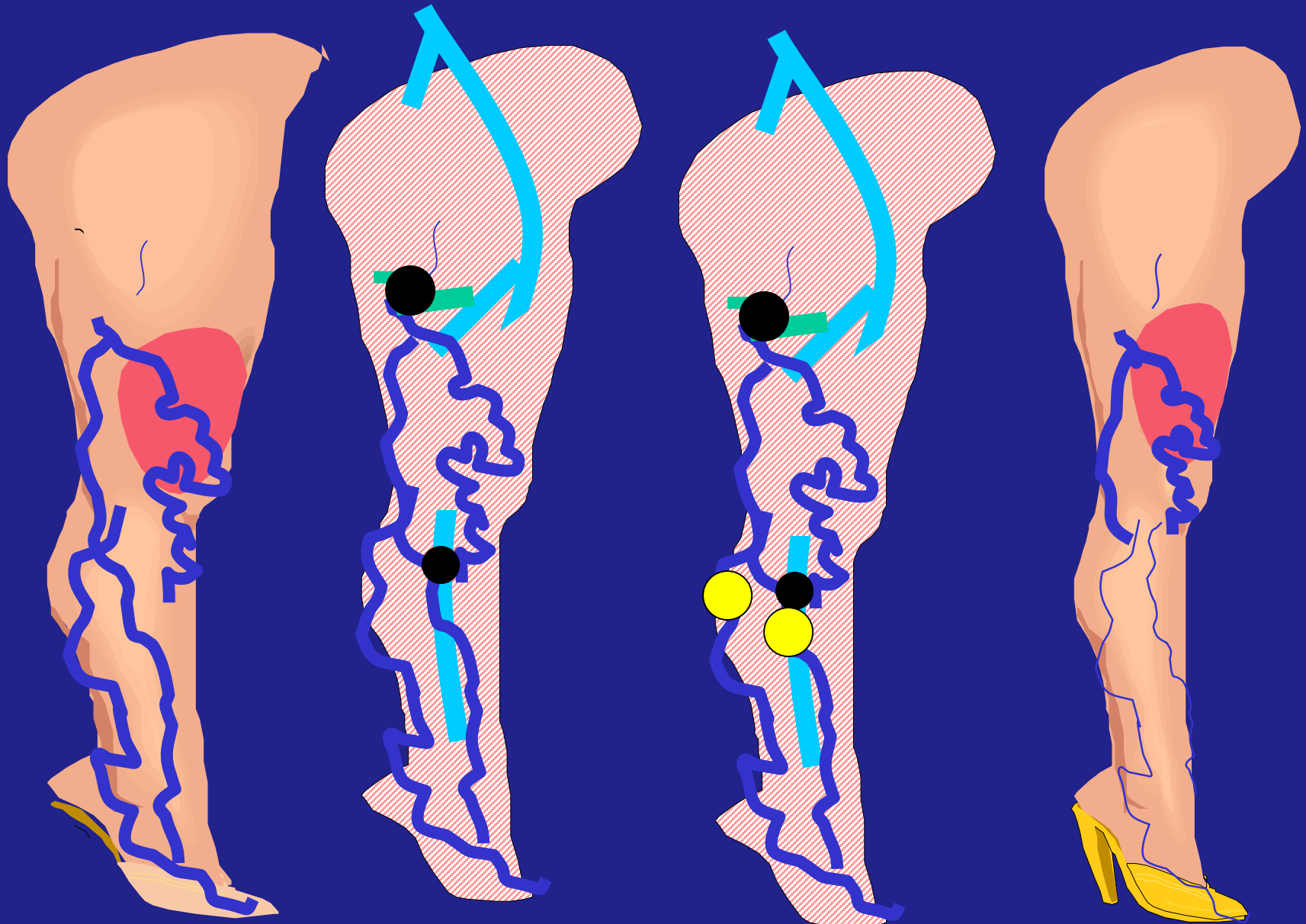
Cutaneous angioma : incompetent superficial vein: **Mixed shunt : Closed + Open Vicarious Shunt: common escape point, different re-entries**



Disconnecting  the closed shunts drains (relieves) the angioma and suppresses the deep overloading flow and preserving the Open Vicarious avoids the dangerous block of the deep and superficial draining flow and further varicose collaterals

Cutaneous angioma : incompetent superficial vein: **Mixed shunt : Closed + Open Vicarious Shunt: common escape point, different re-entries**

Pattern 4 treatment



Closed shunts disconnections stage by stage

Preserved open vicarious shunt varicose (marginal) shunts

Prospective study (2012 et 2013) of venous malformations with varicose veins assessed by Duplex US : 56 patients (58 limbs).

Objective: identify the varicose veins necessary to the limb drainage in order to avoid their ablation which could increase the obstructive syndrome.

Method: Probes: 14, 7 and 3 Mhz. Assessment of aplasia, hypoplasia and incompetence of the deep and superficial veins of the lower limbs, iliac and cava veins.

Compensatory Vein Test proposed by C. Franceschi.

Compensatory Vein Test (Franceschi)

While the patient rocks (oscillates) , one hand compresses and occludes the varicose vein with the US probe and the other hand assesses its tension (pression) just below.

Compensatory vein: Tension increases

Not compensatory vein: Tension decreases



Varices are compensatory in

35% of varices located in the marginal vein territory

37% of varices located in the saphena vein territory (in absence of marginal vein)

Not compensatory varices

37 limbs

Varicose territory:

Saphena (17) Marginal (14)

Saphena+Marginal (6)

Deep veins:

32 limbs: ectasia, avalvulation,
extra-truncular (muscle, nerve
and interfascial infiltration)

Compensatory varices

Varicose territory: 21 limbs

Saphena (10) Marginal (3)

Saphena+Marginal (8)

Deep veins:

aplasia and agenesis

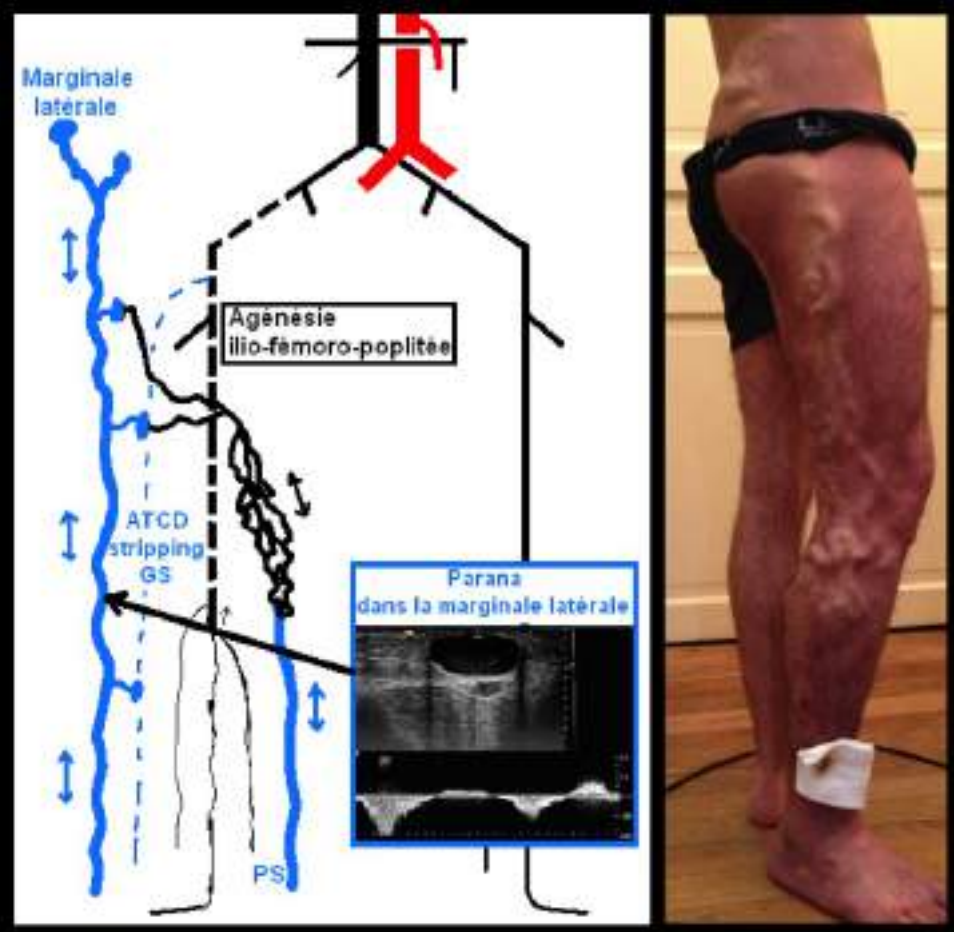
leg veins and/or Popliteal V and/or
Femoral V and /or Iliac V and /or
Cava V.

Example of Duplex Mapping

Not compensatory Varice
(No Open vicarious shunt)



Compensatory Varice
(Open vicarious shunt)



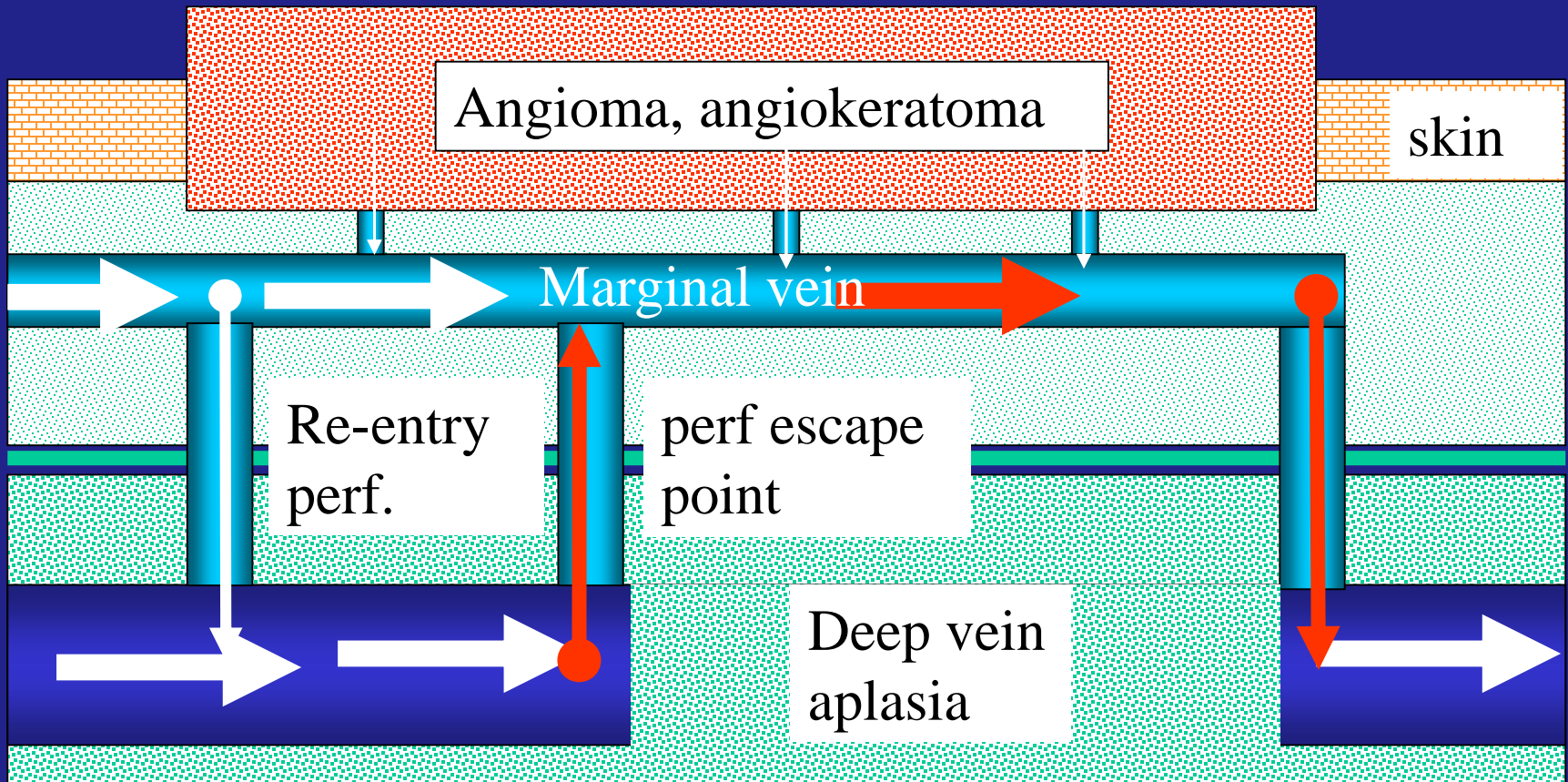
Blue = Superficial veins **Black** = Deep veins **Dotted black** = aplasia

In 35-37 % of Venous Malformations including superficial varicose veins one or more of them are compensatory .

Their ablation could increase the obstructive syndrome

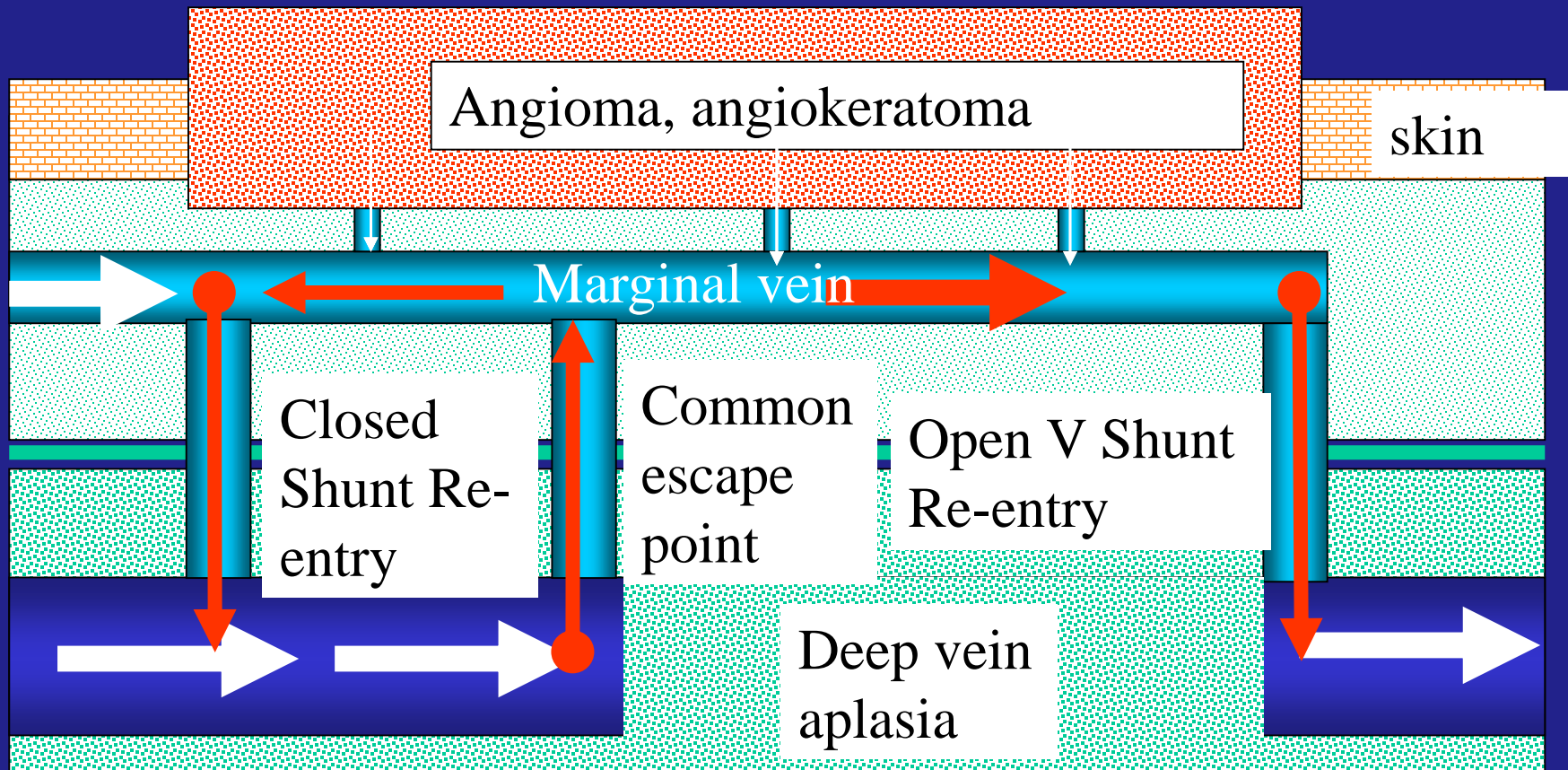
An exhaustive topographic and hemodynamic mapping with compensatory vein test prevents these mistakes.

Pattern 3



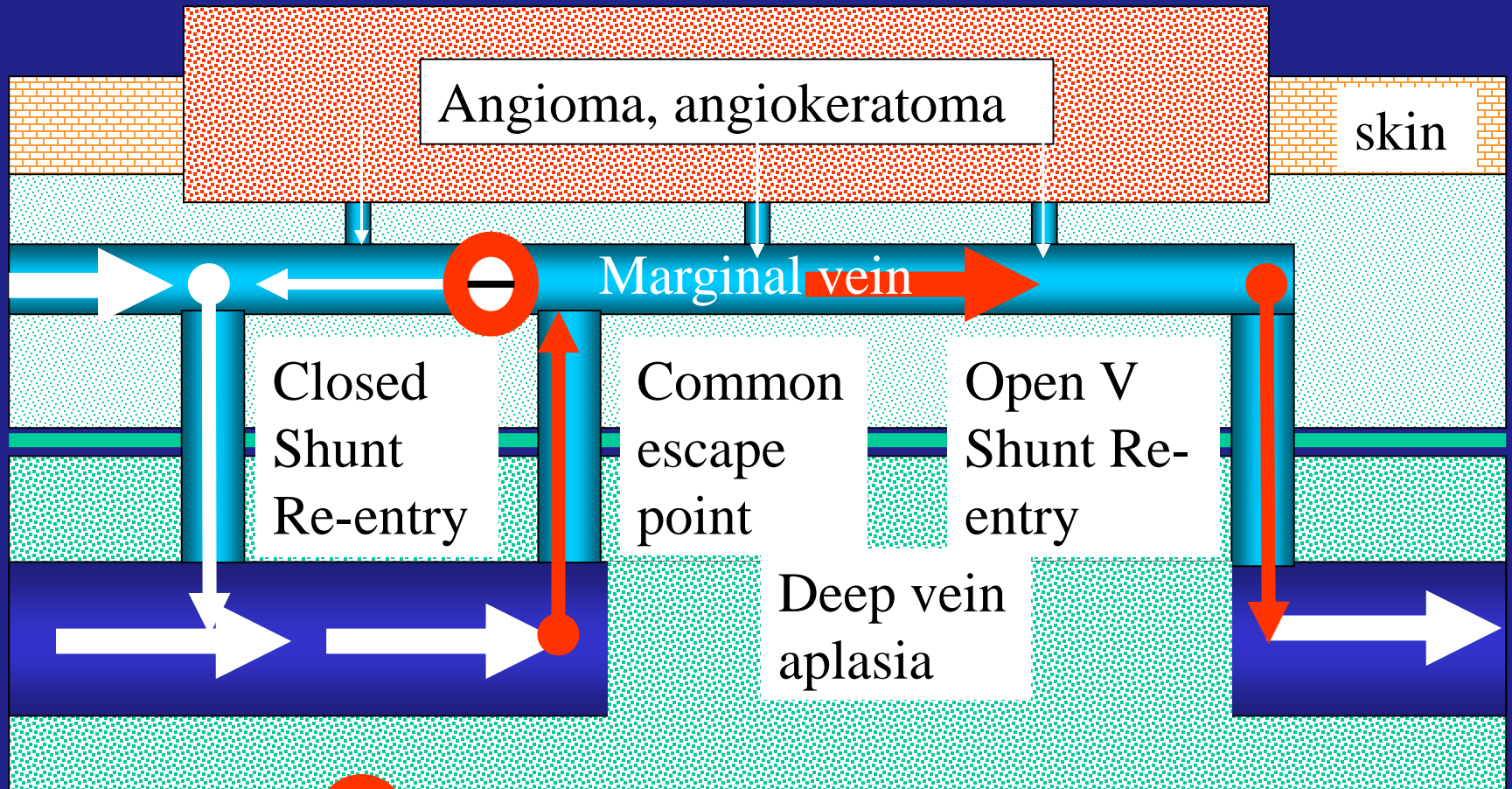
When the deep veins (femoral, popliteal) are hypoplastic and the Marginal vein plays the role of “natural” by-pass, ablating the vicarious segment is too dangerous for the leg drainage.


Pattern 4



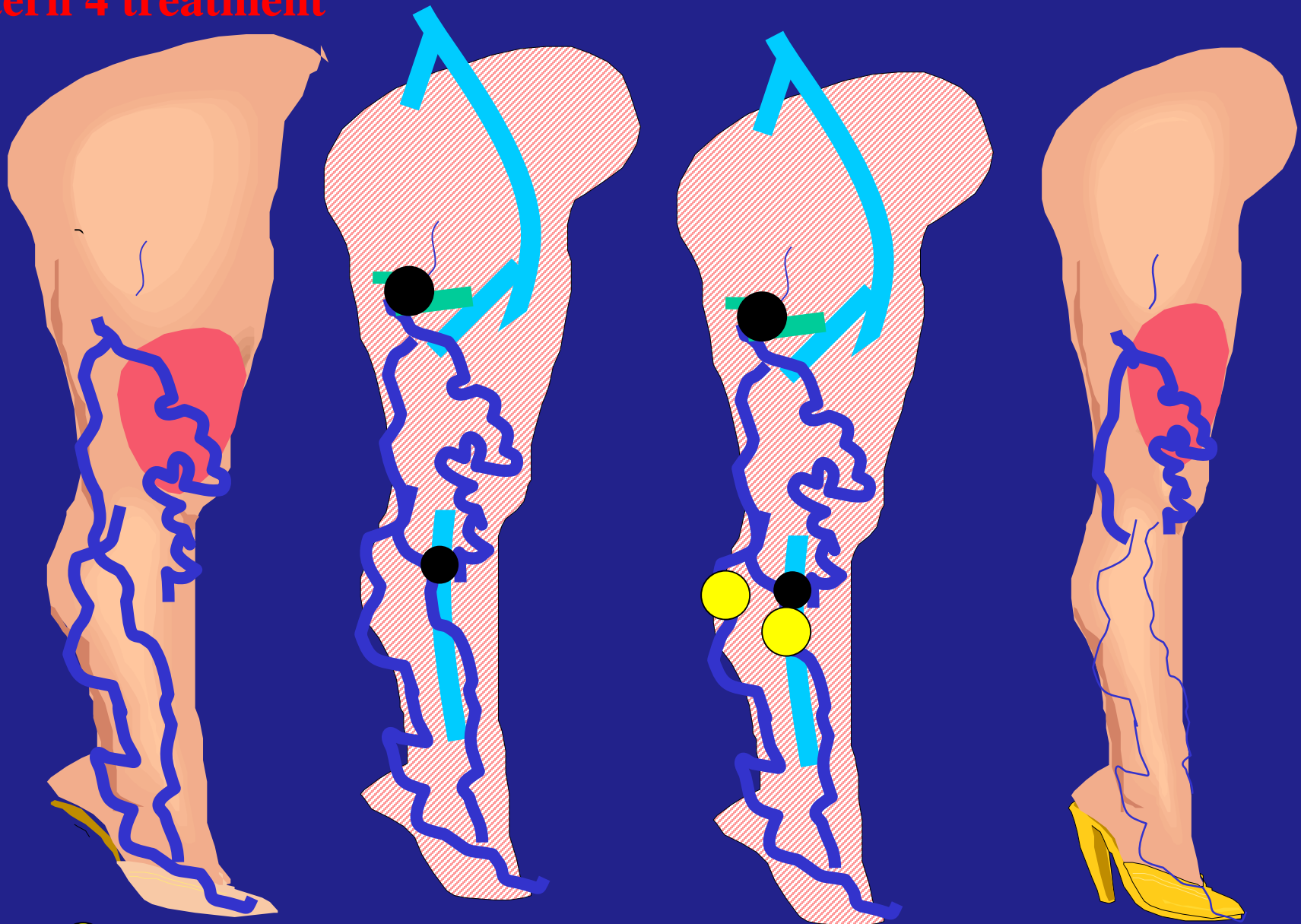
On the other hand, the Marginal vein segment is disconnected when its not involved in the vicarious effect but responsible for a closed shunt.

Pattern 4 treatment



Disconnecting  the closed shunts segment drains and relieves the angioma and suppresses the deep overloading flow whilst preserving the Open Vicarious segment avoids the dangerous stop of the deep and superficial draining flow and further varicose collaterals

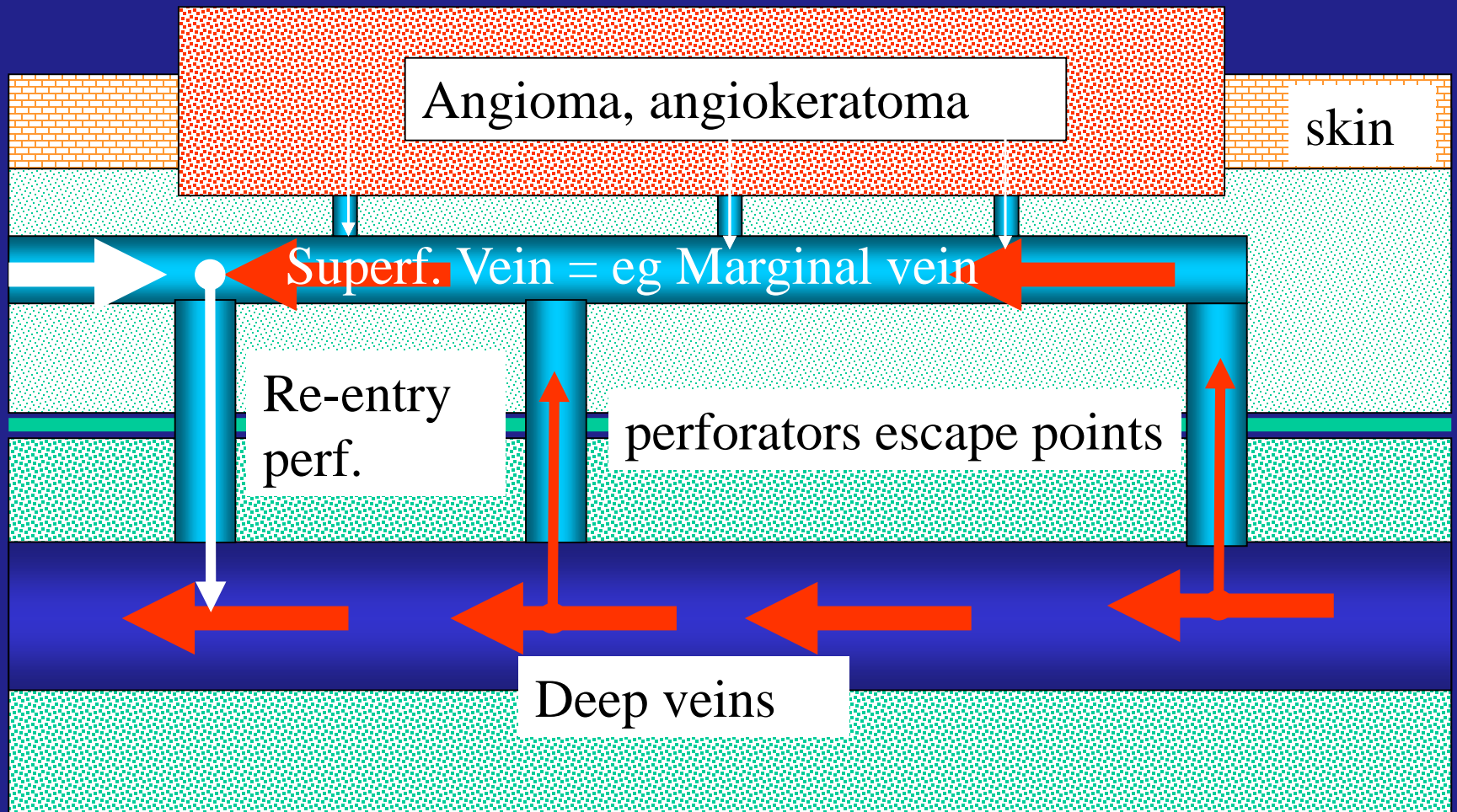
Pattern 4 treatment



Closed shunts disconnections stage by stage

Preserved open vicarious shunt varicose (marginal) shunts

Pattern 5



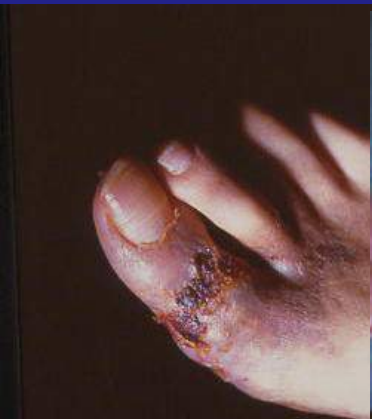
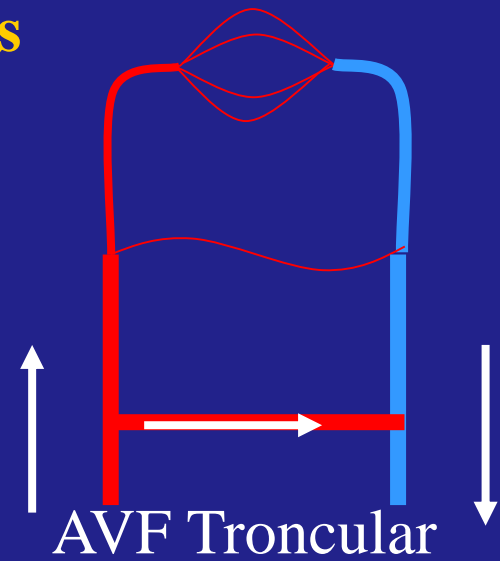
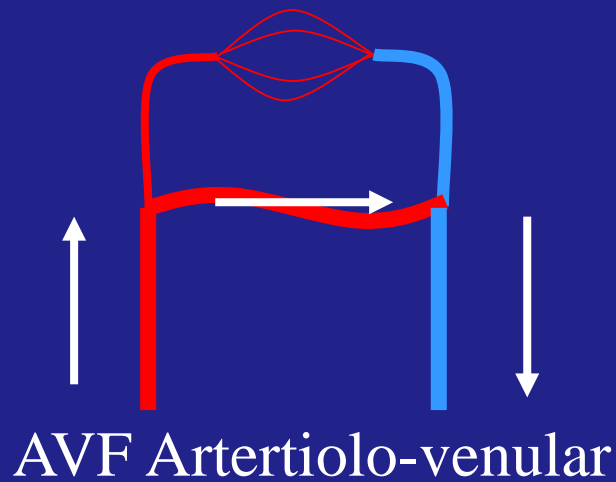
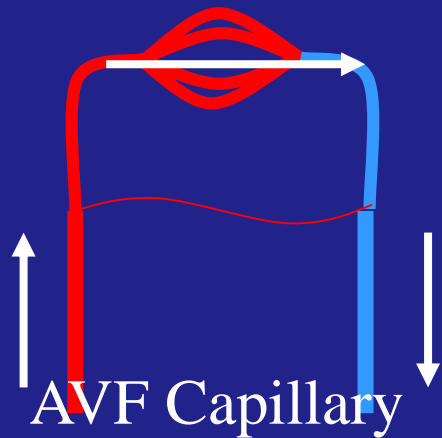
Cutaneous angioma : **incompetent superficial and deep veins**: Despite the deep incompetence, if the Marginal vein shows a diastolic reflux ; the treatment is = pattern **2 and 4**.



Arterio-venous Malformations



Arterio-venous fistula
High flow
Low resistances



Arterio-venous fistula

High flow

Low resistances



DANGEROUS according to:

Flow rate: **Cardiac failure**

Location: **Bleeding**, pain ,function

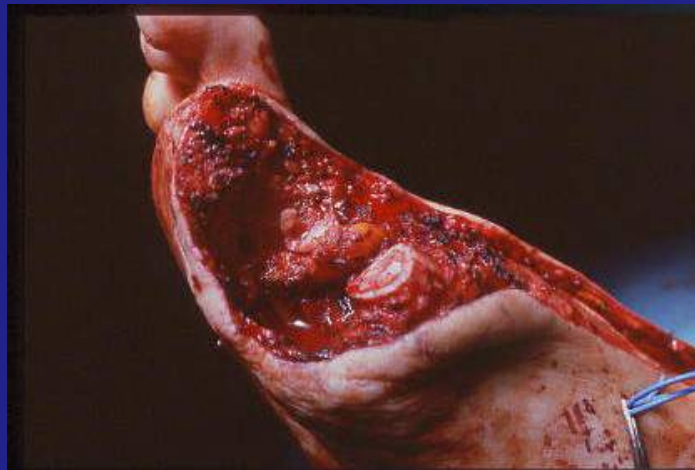
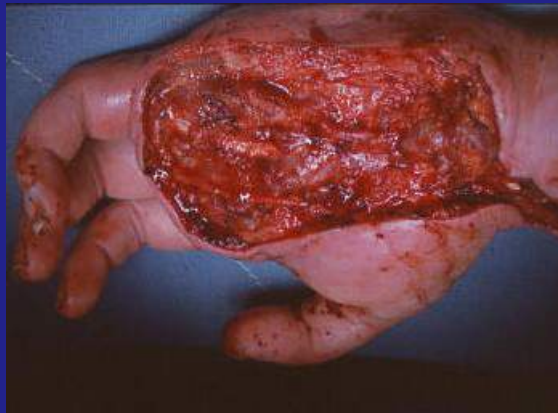
Treatment:

Compression++++


ONLY If complications:

Surgery whenever possible

Ebolization when surgery not possible



Bleeding control



Delacroix
Chevalier

Take now **ALL THE TIME** you need for vascular suture, even in alarm condition of the patient

Prevenir les saignements. Réduire le temps de suture vasculaire. Réduire les situations critiques.

Dr Franceschi's
HEMOSTATOR

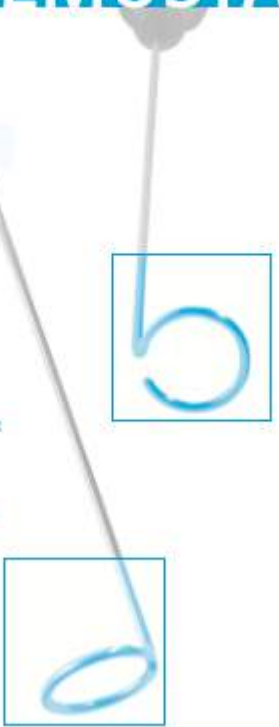
forme rectangulaire

The ideal Hemostasis Safety tool:

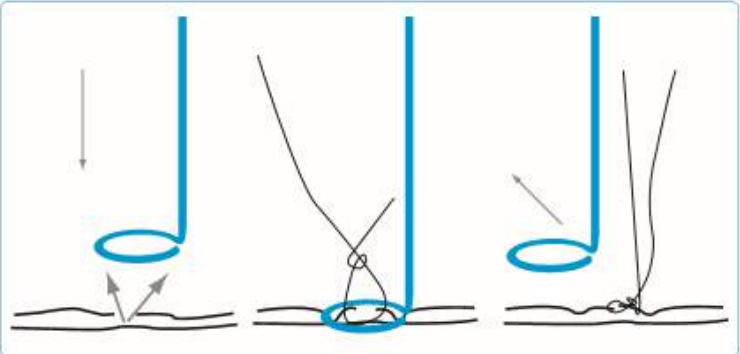
- > Immediate control of haemorrhage
- > Dramatic drop down of blood loss
- > Drying of operative field
- > Usable in every operative field even on non clamping zones (thoracic, pelvic vessels, sclerous tissues...)

L'outil idéal d'hémostase de sauvetage :

- > Contrôle immédiat de l'hémorragie
- > Réduction majeure des pertes sanguines
- > Assèchement du champ opératoire
- > Utilisable partout où il y a des vaisseaux (thoraciques, pelviens, en zone de tissus sclérotés...)



Brand of LANDANGER Group



The facts:

- > Emergency vascular suture is a very risky and stressful situation
- > Veins are fragile, less visible, and more difficult to dissect and clamp than arteries
- > Simply using the finger or dressing gauze that stop the bleeding hide the vessel
- > Pressure required to compress an aorta : only 1,5 kg

Les faits :


- > Une plaie vasculaire accidentelle provoque une situation stressante à haut risque
- > Les veines sont plus fragiles, moins visibles et plus difficiles à disséquer et clamer que les artères
- > L'utilisation du doigt ou d'une compresse pour arrêter le saignement cache le vaisseau
- > La pression nécessaire pour comprimer efficacement une aorte est seulement de 1,5 kg

The Specifications:


- > 3 diameters for ideal fitting to the vessel size
- > The ring is lightly opened to allow the clearing of suture thread
- > The handle is oriented for a perfect control of the ring position

Les spécifications :


- > 3 diamètres pour s'adapter à la taille du vaisseau
- > Anneau discrètement ouvert pour permettre le dégagement du fil de suture
- > Poignée orientée pour un contrôle parfait de la position de l'anneau



Réf. DC53200 - 10



Réf. DC53200 - 20



Réf. DC53200 - 30

Brand of LANDANGER Group

9 Boulevard du Maréchal de Lattre de Tassigny | B.P. 2152 | 52000 CHALUMONT
Tel. : +33 (0)3 25 02 10 10 / Fax : +33 (0)3 25 02 10 20
E-mail : client@delacroix-chevalier.fr | www.delacroix-chevalier.fr

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A 3D rectangular box with a yellow face and darker yellow sides, set against a dark blue background. The box is centered and contains the text "Lymphatic Malformations".

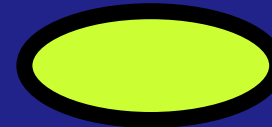
Lymphatic Malformations

Lymphatic Capillaries

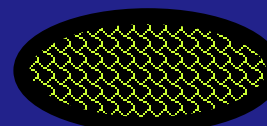
Cutaneous Verrucous nodules
and papilloma (angiokeratoma)



Simple cyst



Cavernous cyst



**Lymphatic
Truncular**

Hypoplasia,

Aplasia

Avalvulation



COMBINED MALFORMATIONS

Vascular

Artérielles

Artério-veineuses

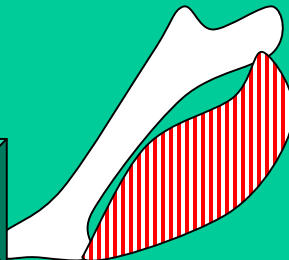
Veineuses

Artério-veino-lymphatiques

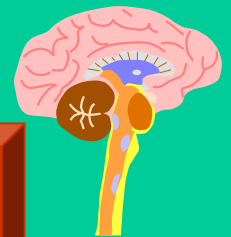
Veino-lymphatiques

Lymphatiques

OSTEO-MUSCULAR
DYSTROPHY



NEURO-CUTANEOUS
DYSTROPHY



AV Fistula

Parkes-Weber Syndrome (P.W.) :

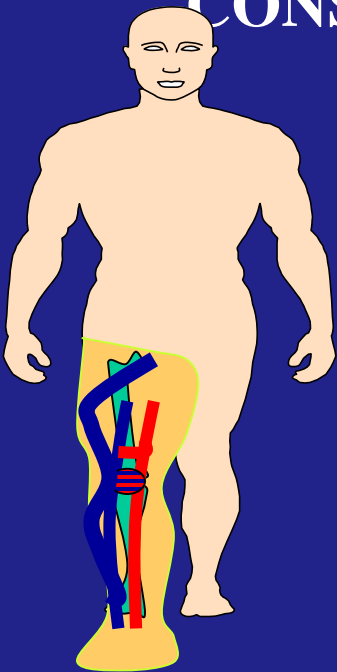
- Truncular AVF + or – capillar (bones and joints)
- Very high flow

CONSEQUENCES :

Venous Hypertension

Varicose draining veins

Arm (hypertrophic hémangiectasia)



AV Fistula

Cirroid AVM: extratruncular

Scalp



Hands



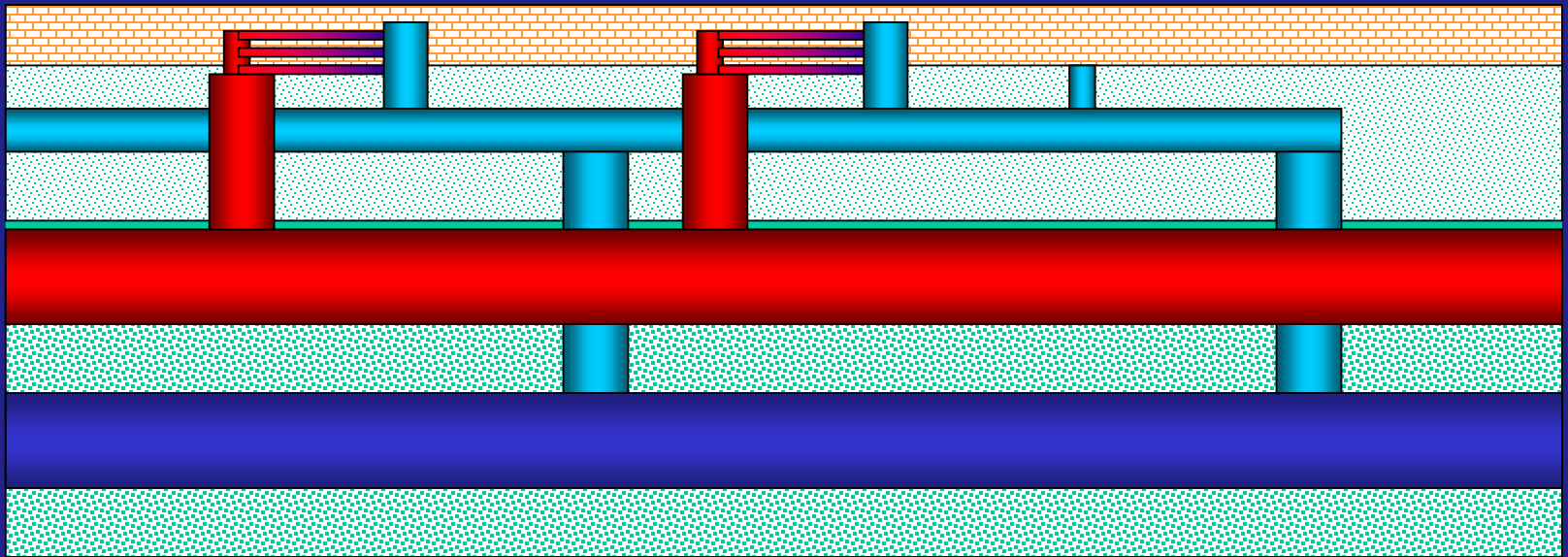
Feet



AV Fistula

Cutaneous capillary angioma :

May be low flow...nevertheless higher than normal skin
Which is *a good reason for sparing the draining veins*

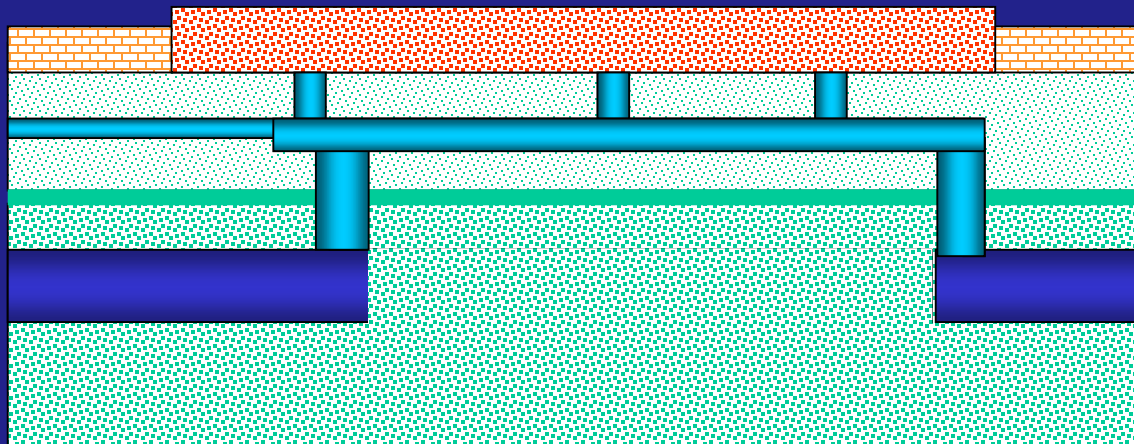


Combined VM

Klippel-Trenaunay-Weber Syndrome

Definition still disputed.....

- **Metameric Topography**
- More or less **malformations lymphatiques**
- **Deep veins agenesis (hypoplasia)**
- *Limb hypertrophy (angio-ostéo-hypertrophicsyndrome)*
- *Capillary cutaneous angioma + or less angiokeratoma*



MALFORMATIONS VASCULAIRES COMPLEXES :
« ANGIODYSPLASIES OSTEO-
HYPERTROPHIQUES » → Syndrome de KLIPPEL-
TRENAUNAY (= MCVL)



Combined VM

KTW signs without:

Deep venous aplasia

And/or Capillary Cutaneous Angioma

And/or limb hypertrophy

*FREQUENTLY interesting the
MARGINAL VEIN*

Combined VM

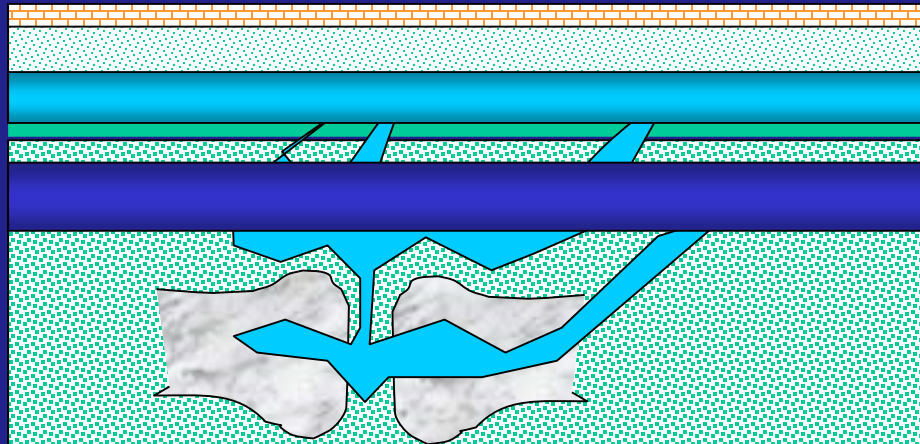
KTW signs without:

Deep venous aplasia

And/or limb hypertrophy

BUT

KNEE SYNOVIAL VM



SYNDROMES MULTI-MALFORMATIFS

Le Syndrome de Proteus

association de malformations essentiellement veineuses ,capillaires et lymphatiques dans lesquels dominant :

- *Capillary various angiomas.*
- *Lymphangiomes*
- *Lymphatic M.*
- *Venous M*
- *Limbs , hands, feet hypertrophy*

Sd de Protée = ML + hémihypertrophie corporelle
+hypertrophie osseuse





PRIMUM
NON NOCERE