ANGIODYSPLASIAS
or
VASCULAR MALFORMATIONs

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

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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.)

### 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)
  (glomerangioma)
- 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

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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+++++
Les malformations vasculaires congénitales

C. Franceschi

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 1994.

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

Leur localisation, souvent métagérique, 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...
Modified Hamburg Classification

Main classification based on its predominant vascular component

- Arterial defects
- Venous defects
- AV (arteriovenous) shunting defects
- Lymphatic defects
- Capillary defects
- Combined vascular defects

*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, AV shunting, venous, hemolymphatic and/or lymphatic); and/or may exist with haemangioma

CVM, congenital vascular malformation
Regarding the early angiogenesis

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

Regarding the early angiogenesis
COMBINATED MALFORMATIONS

Vascular

Artérielles

Veineuses

Veino-lymphatiques

Lymphatiques

OSTEOMUSCULAR DYSTROPHY

NEURO-CUTANEOUS DYSTROPHY
ANGIODYSPLASIAS
or
VASCULAR MALFORMATIONS

Arterial

Venous

Lymphatic

Arterio-venous

Unique or associated
Arterial Malformations
Mega arterial capillaries
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
- Hyperkeratotic cutaneous capillary venous M
- Cutaneous and mucosal VM
- Blue Rubber Bleb Nevus Sd (Bean)

+ 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, hemarthrosis 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
Les TRONCS VEINEUX PROFONDS des deux membres inférieurs sont normaux, sans incontinence, ni thrombose ni altération pariétale et sans séquelles post-phlébitiques.

Les TRONCS VEINEUX SUPERFICIELS 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 echographe.
VM of gastrocnemius (caput mediale)
Medial gastrocnemius excision
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
Venous
Troncular
Hypoplasia
Aplasia
Varicose veins
Avalvulation.

CAVA VEIN
DEEP and SPRFICIAL VEINS
CAVA VEIN

DEEP and SUPERFICIAL VEINS

Asymptomatic, Cosmetic
Pain
Limited function
Thrombosis, Pulmonary Embolism (chronic....)
Duplex anatomic and hemodynamic mapping BECAUSE most malformative veins are draining pathways which are to be respected.
DRAINING VM
Treatment

• **Indication**
  • Pain, Edema, cosmetic
  • Complications
    • Thrombosis
    • PE

• **Means:**
  - Compression, aspirin
  - CHIVA whenever feasible
  - Laser, sclerotherapy in addition to surgery or when surgery is not feasible. Ethanol must be prohibited (DANGEROUS !!!!!)
Bleeding control
Angoma, angiokeratoma

Superf. Vein = eg Marginal vein

perforators

Deep veins

Suppressing the superficial vein leads to angioma aggravation and varicose collaterals

Cutaneous angioma: normal draining venous network
Cutaneous angioma: incompetent superficial vein: **Closed shunt**

Overloaded: Angioma flow + Deep diverted retrograde flow (closed circuit)
Pattern 2 treatment

Disconnected the closed shunts drains (relieves) the angioma and suppresses the deep overloading flow.

Cutaneous angioma: incompetent superficial vein: Closed shunt.

Overloaded: Angioma flow + Deep diverted retrograde flow (closed circuit)
When angima or angiokeratoma is absent or small, Marginal vein destruction doesn’t lead to recurrence and the outcome is roughly similar to disconnection.

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

Overloaded: Angioma flow + Deep diverted retrograde flow (closed circuit)
Closed shunts disconnections stage by stage
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
Dis-connecting 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.
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 prosed 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)

<table>
<thead>
<tr>
<th>Not compensatory varices</th>
<th>Compensatory varices</th>
</tr>
</thead>
<tbody>
<tr>
<td>37 limbs</td>
<td>Varicose territory: 21 limbs</td>
</tr>
<tr>
<td>Varicose territory:</td>
<td>Saphena (10) Marginal (3)</td>
</tr>
<tr>
<td>Saphena (17) Marginal (14)</td>
<td>Saphena+Marginal (8)</td>
</tr>
<tr>
<td>Saphena+Marginal (6)</td>
<td>Deep veins:</td>
</tr>
<tr>
<td>Deep veins:</td>
<td>- aplasia and agenesis</td>
</tr>
<tr>
<td>32 limbs: ectasia, avalvulation, extra-truncular (muscle, nerve and interfascial infiltration)</td>
<td>leg veins and/or Popliteal V and/or Femoral V and/or Iliac V and/or Cava V.</td>
</tr>
</tbody>
</table>
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.
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.
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.
Closed shunts disconnections stage by stage
Preserved open vicarious shunt varicose (marginal) shunts
Cutaneous angioma: competent 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 fistula
High flow
Low resistances

- AVF Capillary
- AVF Arteriolo-venular
- AVF Troncular
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
Lymphatic Malformations
Lymphatic Capillaries

Cutaneous Verrucous nodules and papilloma (angiokeratoma)

Simple cyst

Cavernous cyst
Lymphatic Truncular Hypoplasia, Aplasia Avalvulation
AV Fistula

Parkes-Weber Syndrome (P.W.):
- Truncular AVF + or – capillary (bones and joints)
- Very high flow

CONSEQUENCES:
Venous Hypertension
Varicose draining veins
Arm (hypertrophic hémangiectasia)
AV Fistula

*Cirsoid 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
Klippel-Trenaunay-Weber Syndrome
Definition still disputed…….
- Metameric Topography
- More or less malformations lymphatiques
- Deep veins agenesia (hypoplasia)
- Limb hypertrophy (angio-ostéo-hypertrophic syndrome)
- Capillary cutaneous angioma + or less angiokeratoma1
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**
Le Syndrome de Proteus

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

- Capillary various angiomas.
- Lymphangiomes
- Lymphatic M.
- Venous M
- Limbs, hands, feet hypertrophy
Sd de Protée = ML + hémihypertrophie corporelle + hypertrophie osseuse
PRIMUM
NON NON NOCERE