**ISSVA classification of Vascular anomalies**

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Laurence M. Boon, MD, PhD
Center for Vascular Malformations
Division of Plastic Surgery
Cliniques universitaires St-Luc
Brussels
laurence.boon@uclouvain.be

**ISSVA classification of Vascular anomalies**

<table>
<thead>
<tr>
<th>Vascular Tumors</th>
<th>Vascular Malformations</th>
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<tbody>
<tr>
<td>Benign</td>
<td>Simple</td>
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<td>Combined* of major named vessels associated with other anomalies</td>
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* Defined as rare or mixed vascular malformations (have in arteries and veins)

**Vascular anomalies**

**The 2014 updated ISSVA* classification of vascular anomalies**

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**N.B. The classification tables do not list exhaustively all known vascular anomalies.**

Some rare "dermatologic" vascular anomalies will be found in dermatology textbooks.

The tumor or malformation nature or precise classification of some lesions is still unclear.

These lesions appear in a separate provisional list.

See [details](http://www.issva.org/classification)

**Venous malformations**

**Lymphatic malformations**

CVM, CLM

LVM, CLVM

CAVM

CLVAM

Others

**Arteriovenous malformations**

* Defined as rare or mixed vascular malformations (have in arteries and veins)

See [list](http://www.issva.org/classification)

**Arteriovenous fistula**

**Veinomalytasia**

**Veinopatia**

**Veinopathy**

**Vascular anomalies**

**Vascular tumors**

**Vascular malformations**

**Simple Combined**

of major named vessels associated with other anomalies

**Benign**

**Locally aggressive or borderline**

**Malignant**

**Capillary malformations**

**Lymphatic malformations**

**Venous malformations**

**Arteriovenous malformations**

**Arteriovenous fistula**

** Others**

See [details](http://www.issva.org/classification)

See [list](http://www.issva.org/classification)
### ISSVA classification of vascular tumors

**Non-involuting (NICH)**
- Infantile hemangioma / Hemangioma of infancy
- Composite hemangioendothelioma
- Kaposiform hemangioendothelioma
- Pyogenic granuloma (aka lobular capillary hemangioma)
- Epithelioid hemangioma
- Spindle-cell hemangioma
- Tufted angioma
- Congenital hemangioma
- Reactive proliferative vascular lesions

**Partially involuting (PICH)**
- Rapidly involuting (RICH) *

**Rapidly involuting (RICH) **
- Angiosarcoma
- Kaposi sarcoma
- Composite hemangioendothelioma
- Papillary intralymphatic angioendothelioma (PILA), Dabska tumor
- Retiform hemangioendothelioma
- Kaposiform hemangioendothelioma
- Other

*N.B.*

Some lesions may be associated with thrombocytopenia and/or consumptive coagulopathy.

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**ISSVA classification of vascular tumors**

- Site-like capillaries surrounded by spindle cells
- Packed capillaries in cannonball pattern

**ISSVA classification of vascular tumors**

- Slite-like capillaries surrounded
- Invasive lobules

**ISSVA classification of vascular tumors**

- 1 week
- 1 month
- 2 weeks

**ISSVA classification of vascular tumors**

- 5 months
ISSVA classification of vascular malformations

### Vascular anomalies

<table>
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<tr>
<th>Tumor Type</th>
<th>Malformation Type</th>
<th>Associated with Other Anomalies</th>
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<tr>
<td>Simple</td>
<td>Capillary malformations</td>
<td>CM, LCM, LVM, CVM</td>
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* Defined as two or more vascular malformations found in one lesion

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**Vascular Tumors**

- Capillary malformations (CM)
  - CM with CNS and/or eye anomalies (Sturge-Weber syndrome)
  - CM with bone and/or soft tissue hypertrophy
  - CM with CNS and/or eye anomalies (Sturge-Weber syndrome)
  - CM of CCMCAP (capillary-cavernous malformation)
  - CM of CCM (capillary-cavernous malformation polyphlebitis)

**Vascular Malformations**

- Lymphatic malformations (LM)
  - Common (cystic) LM
  - Macrocystic LM
  - Microcystic LM
  - Mixed cystic LM
  - Generalized lymphatic anomaly (GLA)

- Venous malformations
  - CVM, CLM, LVM, CLVM

- Arteriovenous malformations
  - CAVM, CLAVM

- Others

**ISSVA classification of vascular malformations**

- Some lesions may be associated with thrombocytopenia and/or consumptive coagulopathy
  - See details
  - See list

- http://www.issva.org/classification

**ISSVA classification of vascular malformations II**

- Lymphatic malformations (LM)
  - Common (cystic) LM
  - Macrocystic LM
  - Microcystic LM
  - Mixed cystic LM

- Generalized lymphatic anomaly (GLA)

- LM in Gorham syndrome

- Channel type LM

- Primary lymphedema (different types)

- Others

- http://www.issva.org
Some lesions may be associated with thrombocytopenia and/or consumptive coagulopathy.

ISSVA classification of vascular malformations

**Primary lymphedema**
- Nonne-Milroy syndrome: FLT4/VEGFR3
- Primary hereditary lymphedema: VEGFC
- Primary hereditary lymphedema: GJC2

**Lymphedema-distichiasis** FOXC2

**Hypertrichosis-lymphedema-telangiectasia** SOX18

**Primary lymphedema with myelodysplasia** GATA2

**Primary generalized lymphatic anomaly** del(12q)(pl-q21.1)
- Lymphedema, limb-girdle hypoplasia, and mental retardation syndrome: CCBE1
- Lymphedema with or without chondroectodysplasia, lymphedema, or mental retardation syndrome: KIF11
- Lymphedema-chondal atresia: PTPN14

**Simple vascular malformations III**

**Venous malformations (VM)**
- Common VM
- Familial VM cutaneo-mucosal (VMCM)
- Blue rubber bleb nevus (Bean) syndrome VM
- Glomuvenous malformation (GVM)
- Cerebral cavernous malformation (CCM)
- Others

**Arteriovenous malformations (AVM)**
- Sporadic
- In HHT
  - HHT1: ENG
  - HHT2: ACVRL1
- In juvenile polyposis hem. telangiectasia
- In CM-AVM
  - SMAD4
  - RASA1
- Arteriovenous fistulas (AVF)
- Sporadic
- In HHT
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- In juvenile polyposis hemangiocytosis telangiectasia
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**ISSVA classification of vascular malformations**

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    - SMAD4
    - RASA1
Combined vascular malformations*:
- CM + VM = capillary-venous malformation (CVM)
- CM + LM = capillary-lymphatic malformation (CLM)
- CM + AVM = capillary-arteriovenous malformation (CAVM)
- LM + VM = lymphatic-venous malformation (LVM)
- CM + LM + VM = capillary-lymphatic-venous malformation (CLVM)
- CM + LM + AVM = capillary-lymphatic-arteriovenous malformation (CLAVM)
- CM + VM + AVM = capillary-venous-arteriovenous malformation (CVAVM)
- CM + LM + VM + AVM = capillary-lymphatic-venous-arteriovenous malformation (CLVAVM)

* defined as two or more vascular malformations found in one lesion

Vascular malformations associated with other anomalies:
- Klippel-Trenaunay syndrome: PIK3CA
- Parkes-Weber syndrome: RASA1
- Servelle-Martorell syndrome: GNAQ
- Sturge-Weber syndrome: AKT1, PTEN
- Limb CM + congenital non-progressive limb overgrowth
- Maffucci syndrome: VM +/- spindle-cell hemangioma + enchondroma
- Macrocephaly - CM (M-CM / MCAP)
- Microcephaly - CM (MICCAP)
- CLOVES syndrome: LM + VM + CM +/- AVM + lipomatous overgrowth
- Proteus syndrome: CM, VM and/or LM + asymmetrical somatic overgrowth
- Bannayan-Riley-Ruvalcaba syndrome: AVM + VM + macrocephaly, lipomatous overgrowth

Vascular malformations associated with other anomalies:
- Malattia syndrome: VM = spindle cell hemangioendothelioma

Limitations:
- The images provided are in the form of diagrams and tables, which may be difficult to interpret without the corresponding text.
- The text is overlaid on the images, making it challenging to extract natural text from the images.
ISSVA classification of vascular malformations

Provisionally unclassified vascular anomalies

- Venous hemangiomata
- Arteriovenous
- Multilocular lymphangioendothelial malformation with thrombocytopenia / cutaneous or ocular angiomatosus with thrombocytopenia (MLT/CAT)
- Kaposisiform lymphangiomatosis (KLA)
- PTEN (type) hamartomas of soft tissue / "angiomatosus" of soft tissue

Vascular anomalies

- Vascular anomalies is a complex field with various entities
- ISSVA classification is essential to speak a common language to get the proper diagnosis (www.ISSVA.org)
- Active classification with update every 2 yrs
- Provisionally unclassified lesions
- Interdisciplinary approach

Thank you!