ISSVA classification for vascular anomalies ©

(Approved at the 20th ISSVA Workshop, Melbourne, April 2014)

Overview table

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*	CVM, CLM LVM, CLVM CAVM* CLAVM* others	See details	See list

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

* high-flow lesions

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.

Abbreviations used

For more details, click on the underlined links

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ISSVA classification of vascular tumors ©

Benign vascular tumors
Infantile hemangioma / Hemangioma of infancy — see details
Congenital hemangioma
Rapidly involuting (RICH) *
Non-involuting (NICH)
Partially involuting (PICH)
Tufted angioma * °
Spindle-cell hemangioma
Epithelioid hemangioma
Pyogenic granuloma (aka lobular capillary hemangioma)
Others
Locally aggressive or borderline vascular tumors
Kaposiform hemangioendothelioma * °
Retiform hemangioendothelioma
Papillary intralymphatic angioendothelioma (PILA), Dabska tumor
Composite hemangioendothelioma
Kaposi sarcoma
Others
Malignant vascular tumors
Angiosarcoma
Epithelioid hemangioendothelioma
Others

* some lesions may be associated with thrombocytopenia and/or consumptive coagulopathy see details

° many experts believe that these are part of a spectrum rather than distinct entities

N.B. reactive proliferative vascular lesions are listed with benign tumors

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ISSVA classification for vascular anomalies©

Simple vascular malformations I	
Capillary malformations (CM)	
Cutaneous and/or mucosal CM (aka “port-wine” stain)	G
CM with bone and/or soft tissue overgrowth	
CM with CNS and/or ocular anomalies (Sturge-Weber syndrome)	
CM of CM-AVM	G
CM of MICCAP (microcephaly-capillary malformation)	
CM of MCAP (megalencephaly-capillary malformation-polymicrogyria)	
Telangiectasia	
Hereditary hemorrhagic telangiectasia (HHT) (different types)	G
Others	
Cutis marmorata telangiectatica congenita (CMTC)	
Nevus simplex / Salmon patch / “angel kiss”, “stork bite”	
Others	

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ISSVA classification for vascular anomalies©

Simple vascular malformations II	
Lymphatic malformations (LM)	
Common (cystic) LM	
Macrocystic LM	
Microcystic LM	
Mixed cystic LM	
Generalized lymphatic anomaly (GLA)	
LM in Gorham-Stout disease	
Channel type LM	
Primary lymphedema (different types)	G
Others	

some lesions may be associated with thrombocytopenia and/or consumptive coagulopathy see details

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ISSVA classification for vascular anomalies©

Simple vascular malformations IIb

Primary lymphedema	
Nonne-Milroy syndrome	G
Primary hereditary lymphedema	G
Lymphedema-distichiasis	G
Hypotrichosis-lymphedema-telangiectasia	G
Primary lymphedema with myelodysplasia	G
Primary generalized lymphatic anomaly (Hennekam lymphangiectasia-lymphedema syndrome)	G
Microcephaly with or without chorioretinopathy, lymphedema, or mental retardation syndrome	G
Lymphedema-choanal atresia	G

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Simple vascular malformations III	
Venous malformations (VM)	
Common VM	G
Familial VM cutaneo-mucosal (VMCM)	G
Blue rubber bleb nevus (Bean) syndrome VM	
Glomuvenous malformation (GVM)	G
Cerebral cavernous malformation (CCM) (different types)	G
Others	

some lesions may be associated with thrombocytopenia and/or consumptive coagulopathy see details

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ISSVA classification for vascular anomalies©

Simple vascular malformations IV	
Arteriovenous malformations (AVM)	
Sporadic	
In HHT	G
In CM-AVM	G
Others	
Arteriovenous fistula (AVF) (congenital)	
Sporadic	
In HHT	G
In CM-AVM	G
Others	

Abbreviations used

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ISSVA classification for vascular anomalies©

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 m.	CLVAVM

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

Abbreviations used

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ISSVA classification for vascular anomalies©

Anomalies of major named vessels

(aka "channel type" or "truncal" vascular malformations)

Affect

- lymphatics
- veins
- arteries

Anomalies of

- origin
- course
- number
- length
- diameter (aplasia, hypoplasia, stenosis, ectasia / aneurysm)
- valves
- communication (AVF)
- persistence (of embryonal vessel)

Abbreviations used

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ISSVA classification for vascular anomalies©

Vascular malformations associated with other anomalies		
Klippel-Trenaunay syndrome:	CM + VM +/- LM + limb overgrowth	
Parkes Weber syndrome:	CM + AVF + limb overgrowth	G
Servelle-Martorell syndrome:	limb VM + bone undergrowth	
Sturge-Weber syndrome:	facial + leptomeningeal CM + eye anomalies +/- bone and/or soft tissue overgrowth	G
Limb CM + congenital non-progressive limb hypertrophy		
Maffucci syndrome:	VM +/- spindle-cell hemangioma + enchondroma	
Macrocephaly - CM (M-CM / MCAP)		G
Microcephaly - CM (MICCAP)		G
CLOVES syndrome:	LM + VM + CM +/- AVM + lipomatous overgrowth	G
Proteus syndrome:	CM, VM and/or LM + asymmetrical somatic overgrowth	G
Bannayan-Riley-Ruvalcaba sd:	AVM + VM +macrocephaly, lipomatous overgrowth	G

Abbreviations used

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ISSVA classification for vascular anomalies©

Provisionally unclassified vascular anomalies	
Verrucous hemangioma	
Angiokeratoma	
Multifocal lymphangioendotheliomatosis with thrombocytopenia / cutaneovisceral angiomatosis with thrombocytopenia (MLT/CAT)	
Kaposiform lymphangiomatosis (KLA)	
PTEN (type) hamartoma of soft tissue / "angiomatosis" of soft tissue	G

some lesions may be associated with thrombocytopenia and/or consumptive coagulopathy see details

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Appendix 1
abbreviations used
(excluding gene names)

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AVF	arteriovenous fistula
AVM	arteriovenous malformation
CAT	cutaneovisceral angiomatosis with thrombocytopenia
CAVM	capillary arteriovenous malformation
CCM	cerebral cavernous malformation
CLAVM	capillary lymphatic arteriovenous malformation
CLOVES	congenital lipomatous overgrowth, vascular malformations, epidermal nevi, skeletal/scoliosis and spinal abnormalities
CLM	capillary lymphatic malformation
CLVAVM	capillary lymphatic venous arteriovenous malformation
CLVM	capillary lymphatic venous malformation
CM	capillary malformation
CM-AVM	capillary malformation-arteriovenous malformation
CMTC	cutis marmorata telangiectatica congenita
CNS	central nervous system
CVAVM	capillary venous arteriovenous malformation
CVM	capillary venous malformation
DIC	disseminated intravascular coagulopathy
GLA	generalized lymphatic anomaly
GSD	Gorham-Stout disease
GVM	glomuvenous malformation
HHT	hereditary hemorrhagic telangiectasia

HI	hemangioma of infancy / infantile hemangioma
IH	infantile hemangioma / hemangioma of infancy
INR	international normalized ratio
JPHT	juvenile polyposis hemorrhagic telangiectasia
KHE	kaposiform hemangioendothelioma
KLA	kaposiform lymphangiomatosis
KMP	Kasabach-Merritt phenomenon,
LM	lymphatic malformation
LVM	lymphatic venous malformation
MCAP	megalencephaly-capillary malformation-polymicrogyria
M-CM	macrocephaly-capillary malformation
MICCAP	microcephaly-capillary malformation
MLT	Multifocal lymphangioendotheliomatosis with thrombocytopenia
NICH	non-involuting congenital hemangioma
PHACE	posterior fossa malformations, hemangioma, arterial anomalies, cardiovascular anomalies, eye anomalies
PILA	papillary intralymphatic angioendothelioma
PICH	partially involuting congenital hemangioma
RICH	rapidly involuting congenital hemangioma
TA	tufted angioma
VM	venous malformation
VMCM	venous malformation cutaneo mucosal

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Appendix 2-a
causal genes of vascular anomalies

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Capillary malformations (CM)	
Cutaneous and/or mucosal CM (aka “port-wine” stain)	GNAQ
CM with bone and/or soft tissue hyperplasia	
CM with CNS and/or ocular anomalies (Sturge-Weber syndrome)	GNAQ
CM of CM-AVM	RASA1
Telangiectasia	
Hereditary hemorrhagic telangiectasia (HHT)	
HHT1	ENG
HHT2	ACVRL1
HHT3	
JPHT (juvenile polyposis hemorrhagic telangiectasia)	SMAD4
Others	
Cutis marmorata telangiectatica congenita (CMTC)	
Nevus simplex / Salmon patch	
Others	

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Appendix 2-b
causal genes of vascular anomalies

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Lymphatic malformations (LM)	
Primary lymphedema	
Nonne-Milroy syndrome	FLT4 / VEGFR3
Primary hereditary lymphedema	VEGFC
Primary hereditary lymphedema	GJC2 / Connexin 47
Lymphedema-distichiasis	FOXC2
Hypotrichosis-lymphedema-telangiectasia	SOX18
Primary lymphedema with myelodysplasia	GATA2
Primary generalized lymphatic anomaly (Hennekam lymphangiectasia-lymphedema syndrome)	CCBE1
Microcephaly with or without chorioretinopathy, lymphedema, or mental retardation syndrome	KIF11
Lymphedema-choanal atresia	PTPN14

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Appendix 2-c
causal genes of vascular anomalies

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Venous malformations (VM)	
Common VM	TIE2 somatic
Familial VM cutaneo-mucosal (VMCM)	TIE2
Blue rubber bleb nevus (Bean) syndrome VM	
Glomuvenous malformation (VM with glomus cells)	Glomulin
Cerebral cavernous malformation (CCM)	
CCM1	KRIT1
CCM2	Malcavernin
CCM3	PDCD10

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Appendix 2-d
causal genes of vascular anomalies

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Arteriovenous malformations (AVM)	
Sporadic	
In HHT	
HHT1	ENG
HHT2	ACVRL1
JPHT (juvenile polyposis hem. telangiect.)	SMAD4
In CM-AVM	RASA1
Arteriovenous fistulas (AVF)	
Sporadic	
In HHT	
HHT1	ENG
HHT2	ACVRL1
JPHT (juvenile polyposis hemorrhagic telangiectasia)	SMAD4
In CM-AVM	RASA1

Abbreviations used

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Appendix 2-e
causal genes of vascular anomalies

Vascular malformations associated with other anomalies	
Klippel-Trenaunay syndrome	
Parkes Weber syndrome	RASA1
Servelle-Martorell syndrome	
Sturge-Weber syndrome	GNAQ
Limb CM + congenital non-progressive limb overgrowth	
Maffucci syndrome	
Macrocephaly - CM (M-CM or MCAP)	PIK3CA
Microcephaly - CM (MICCAP)	STAMBP
CLOVES syndrome	PIK3CA
Proteus syndrome	AKT1
Bannayan-Riley-Ruvalcaba syndrome	PTEN

Abbreviations used

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Appendix 2 -f
causal genes of vascular anomalies

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Provisionally unclassified vascular anomalies	
Verrucous hemangioma	
Multifocal lymphangioendotheliomatosis with thrombocytopenia / cutaneovisceral angiomatosis with thrombocytopenia (MLT/CAT)	
Kaposiform lymphangiomatosis (KLA)	
PTEN (type) hamartoma of soft tissue / "angiomatosis" of soft tissue	PTEN

some lesions may be associated with thrombocytopenia and/or consumptive coagulopathy see details

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Appendix 3
infantile hemangioma

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Pattern
- focal - multifocal - segmental - indeterminate

Different types
- superficial - deep - mixed (superficial + deep) - reticular / abortive / minimal growth - others

Association with other lesions	
PHACE association / syndrome	Posterior fossa malformations, Hemangioma, Arterial anomalies, Cardiovascular anomalies, Eye anomalies, sternal clefting and/or supraumbilical raphe
LUMBAR (SACRAL, PELVIS) association / syndrome	Lower body hemangioma, Urogenital anomalies, Ulceration, Myelopathy, Bony deformities, Anorectal malformations, Arterial anomalies, and Renal anomalies

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Appendix 4
vascular anomalies
possibly associated with platelet count / coagulation disorders

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Anomalies	Hematological disorders
Tufted angioma Kaposiform hemangioendothelioma	Profound and sustained thrombocytopenia with profound hypofibrinogenemia, consumptive coagulopathy and elevated D-dimer (Kasabach-Merritt phenomenon)
Rapidly involuting congenital hemangioma	Transient mild/moderate thrombocytopenia, +/- consumptive coagulopathy and elevated D-dimer
Venous malformations / Lymphatic-venous malformations	Chronic localized intravascular coagulopathy with elevated D-dimer, +/- hypofibrinogenemia, and +/- moderate thrombocytopenia (may progress to DIC after trauma or operation)
Lymphatic malformations	Chronic localized intravascular coagulopathy with elevated D-dimer and +/- mild to moderate thrombocytopenia *(consider Kaposiform lymphangiomatosis)* (may progress to DIC after trauma or operation)
Multifocal lymphangioendotheliomatosis with thrombocytopenia / Cutaneovisceral angiomatosis with thrombocytopenia	Sustained, fluctuating, moderate to profound thrombocytopenia with gastrointestinal tract bleeding or pulmonary hemorrhage
Kaposiform lymphangiomatosis	Mild to Moderate thrombocytopenia, +/- hypofibrinogenemia, and D-dimer elevation