

NEOPLASTIC VASCULAR TUMORS (ISSVA: *Vascular Tumors*)

[1] ♦ **[NVT]**

Vascular tumors — neoplastic proliferations (benign, borderline, malignant). These may be congenital, perinatal, or acquired in adulthood, depending on subtype. Endothelial cells divide abnormally, creating new tissue — the definition of neoplastic.

Benign *
[1.1] ♦ **[NVT-BE]**

Infantile hemangioma	[1.1.01] ♦ [NVT-BE-IH]
Congenital hemangioma	[1.1.02] ♦ [NVT-BE-CH]
Tufted angioma	[1.1.03] ♦ [NVT-BE-TA]
Cherry angioma	[1.1.04] ♦ [NVT-BE-CA]
Epithelioid hemangioma	[1.1.05] ♦ [NVT-BE-EH]
Cutaneous epithelioid angiomatous nodule	[1.1.06] ♦ [NVT-BE-CN]
Pyogenic granuloma — Lobular Capillary Hemangioma	[1.1.07] ♦ [NVT-BE-PG]
Spindle-cell hemangioma	[1.1.08] ♦ [NVT-BE-SH]
Hobnail hemangioma	[1.1.09] ♦ [NVT-BE-HH]
Microvenular hemangioma	[1.1.10] ♦ [NVT-BE-MH]
Anastomosing hemangioma	[1.1.11] ♦ [NVT-BE-AH]
Glomeruloid hemangioma	[1.1.12] ♦ [NVT-BE-GH]
Papillary hemangioma	[1.1.13] ♦ [NVT-BE-PH]
Acquired elastotic hemangioma	[1.1.14] ♦ [NVT-BE-AE]
Intravascular papillary endothelial hyperplasia — Masson tumor	[1.1.15] ♦ [NVT-BE-IP]
Littoral cell hemangioma of the spleen	[1.1.16] ♦ [NVT-BE-LH]
Placental chorioangioma	[1.1.17] ♦ [NVT-BE-PL]
Eccrine angiomatous hamartoma	[1.1.18] ♦ [NVT-BE-EC]
Reactive angioendotheliomatosis	[1.1.19] ♦ [NVT-BE-RA]

Bacillary angiomatosis [1.1.20] ♦ [NVT-BE-BA]

* Reactive proliferative vascular lesions are listed with benign vascular tumors.

Borderline

[1.2] ♦ [NVT-B0]

Kaposiform hemangioendothelioma [1.2.01] ♦ [NVT-B0-KH]

Retiform hemangioendothelioma [1.2.02] ♦ [NVT-B0-RH]

Papillary intralymphatic angioendothelioma (PILA), Dabska Tumor [1.2.03] ♦ [NVT-B0-PI]

Pseudomyogenic hemangioendothelioma [1.2.04] ♦ [NVT-B0-PS]

Polymorphous hemangioendothelioma [1.2.05] ♦ [NVT-B0-P0]

Kaposi's sarcoma [1.2.06] ♦ [NVT-B0-KS]

Composite hemangioendothelioma [1.2.07] ♦ [NVT-B0-C0]

Multifocal lymphangioendotheliomatosis with thrombocytopenia (MLT) [1.2.08] ♦ [NVT-B0-ML]

Malignant

[1.3] ♦ [NVT-MA]

Angiosarcoma [1.3.01] ♦ [NVT-MA-AN]

Epithelioid hemangioendothelioma [1.3.02] ♦ [NVT-MA-EP]

STRUCTURAL VASCULAR MALFORMATIONS (ISSVA: Vascular

[2] ♦ [SVM]

Malformations)

Vascular malformations involve **abnormal vascular architecture** — vessels formed incorrectly during development. The structure is wrong; the endothelial cells are not proliferating abnormally. ISSVA: “errors of vascular morphogenesis” — structural, not proliferative.

FAST-FLOW

[2.1] ♦ [SVM-FF]

Isolated

[2.1.01] ♦ [SVM-FF-IS]

AVM [2.1.01.01] ♦ [SVM-FF-IS-AM]

Intramuscular Fast-flow Vascular Anomaly [2.1.01.02] ♦ [SVM-FF-IS-IN]

AVF	[2.1.01.03] ♦ [SVM-FF-IS-AF]
Multifocal	[2.1.02] ♦ [SVM-FF-MU]
CM-AVM 1 & 2	[2.1.02.01] ♦ [SVM-FF-MU-CA]
HHT 1 & 2 / JPHT	[2.1.02.02] ♦ [SVM-FF-MU-HJ]
PHTS	[2.1.02.03] ♦ [SVM-FF-MU-PH]
Syndromic	[2.1.03] ♦ [SVM-FF-SY]
PHOST (PHTS)	[2.1.03.01] ♦ [SVM-FF-SY-PH]
Parkes-Weber Syndrome	[2.1.03.02] ♦ [SVM-FF-SY-PW]
SAMS	[2.1.03.03] ♦ [SVM-FF-SY-SA]
CAMS	[2.1.03.04] ♦ [SVM-FF-SY-CA]
SLOW-FLOW	[2.2] ♦ [SVM-SF]
CAPILLARY	[2.2.01] ♦ [SVM-SF-CM]
Nevus simplex, Salmon Patch	[2.2.01.01] ♦ [SVM-SF-CM-NS]
Port Wine CM — Port Wine Birthmark, Nevus flammeus	[2.2.01.02] ♦ [SVM-SF-CM-PW]
Isolated (<i>incl. Phacomatosis Pigmentovascularis</i>)	[2.2.01.02.01] ♦ [SVM-SF-CM-PW-IS]
Syndromic	[2.2.01.02.02] ♦ [SVM-SF-CM-PW-SY]
With hypertrophy or extracutaneous disease	[2.2.01.02.02.01] ♦ [SVM-SF-CM-PW-SY-HYP]
Sturge-Weber Syndrome	[2.2.01.02.02.02] ♦ [SVM-SF-CM-PW-SY-SWS]
DCMO	[2.2.01.02.02.03] ♦ [SVM-SF-CM-PW-SY-DCM]
Reticulate / Telangiectatic CM	[2.2.01.03] ♦ [SVM-SF-CM-RT]
Isolated	[2.2.01.03.01] ♦ [SVM-SF-CM-RT-IS]
Syndromic	[2.2.01.03.02] ♦ [SVM-SF-CM-RT-SY]
M-CM	[2.2.01.03.02.1] ♦ [SVM-SF-CM-RT-SY-MC]
MIC-CAP	[2.2.01.03.02.2] ♦ [SVM-SF-CM-RT-SY-MI]
DCMO	[2.2.01.03.02.3] ♦ [SVM-SF-CM-RT-SY-DC]

Geographic Pattern CM	[2.2.01.04] ♦ [SVM-SF-CM-GP]
Isolated	[2.2.01.04.1] ♦ [SVM-SF-CM-GP-IS]
Syndromic	[2.2.01.04.2] ♦ [SVM-SF-CM-GP-SY]
Klippel-Trenaunay Syndrome (KTS)	[2.2.01.04.2.1] ♦ [SVM-SF-CM-GP-SY-KT]
Associated with CLOVES / Disorders of PROS	[2.2.01.04.2.2] ♦ [SVM-SF-CM-GP-SY-CL]
Low-resistance CM / CM with faster flow	[2.2.01.05] ♦ [SVM-SF-CM-LR]
Isolated	[2.2.01.05.1] ♦ [SVM-SF-CM-LR-IS]
Syndromic	[2.2.01.05.2] ♦ [SVM-SF-CM-LR-SY]
CM-AVM 1 and 2	[2.2.01.05.2.1] ♦ [SVM-SF-CM-LR-SY-AV]
Parkes-Weber Syndrome	[2.2.01.05.2.2] ♦ [SVM-SF-CM-LR-SY-PW]
Cutis Marmorata Telangiectatic Congenita	[2.2.01.06] ♦ [SVM-SF-CM-CC]
Telangiectasias and Spider Angiomas	[2.2.01.07] ♦ [SVM-SF-CM-TS]
Isolated	[2.2.01.07.1] ♦ [SVM-SF-CM-TS-IS]
Syndromic	[2.2.01.07.2] ♦ [SVM-SF-CM-TS-SY]
CM-AVM 1 and 2	[2.2.01.07.2.1] ♦ [SVM-SF-CM-TS-SY-AV]
HHT 1 & 2 and JPHT (<i>see also AVM category</i>)	[2.2.01.07.2.2] ♦ [SVM-SF-CM-TS-SY-HJ]
LYMPHATIC	[2.2.02] ♦ [SVM-SF-LM]
Isolated	[2.2.02.01] ♦ [SVM-SF-LM-IS]
LM (Discrete)	[2.2.02.01.1] ♦ [SVM-SF-LM-IS-LD]
Macrocystic	[2.2.02.01.1.1] ♦ [SVM-SF-LM-IS-LD-MA]
Microcystic	[2.2.02.01.1.2] ♦ [SVM-SF-LM-IS-LD-MI]
Mixed Macromicrocystic	[2.2.02.01.1.3] ♦ [SVM-SF-LM-IS-LD-MX]
Angiokeratoma	[2.2.02.01.2] ♦ [SVM-SF-LM-IS-AN]
Complex	[2.2.02.02] ♦ [SVM-SF-LM-CO]
GLA	[2.2.02.02.1] ♦ [SVM-SF-LM-CO-GL]
KLA	[2.2.02.02.2] ♦ [SVM-SF-LM-CO-KL]
GSD	[2.2.02.02.3] ♦ [SVM-SF-LM-CO-GS]
GLD	[2.2.02.02.4] ♦ [SVM-SF-LM-CO-GD]

CCLA	[2.2.02.02.5] ♦ [SVM-SF-LM-CO-CC]
Isolated	[2.2.02.02.5.1] ♦ [SVM-SF-LM-CO-CC-IS]
Syndromic (<i>RASopathy</i>)	[2.2.02.02.5.2] ♦ [SVM-SF-LM-CO-CC-SY]
Lymphedemas	[2.2.02.03] ♦ [SVM-SF-LM-LY]
Primary	[2.2.02.03.1] ♦ [SVM-SF-LM-LY-PR]
Isolated	[2.2.02.03.1.1] ♦ [SVM-SF-LM-LY-PR-IS]
Syndromic	[2.2.02.03.1.2] ♦ [SVM-SF-LM-LY-PR-SY]
Secondary	[2.2.02.03.2] ♦ [SVM-SF-LM-LY-SE]
VENOUS	[2.2.03] ♦ [SVM-SF-VM]
Isolated	[2.2.03.01] ♦ [SVM-SF-VM-IS]
VM (Discrete)	[2.2.03.01.1] ♦ [SVM-SF-VM-IS-VD]
Phlebectatic	[2.2.03.01.2] ♦ [SVM-SF-VM-IS-PH]
Spongiform	[2.2.03.01.3] ♦ [SVM-SF-VM-IS-SP]
VVM	[2.2.03.01.4] ♦ [SVM-SF-VM-IS-VV]
FAVA	[2.2.03.01.5] ♦ [SVM-SF-VM-IS-FA]
Multifocal	[2.2.03.02] ♦ [SVM-SF-VM-MU]
VMCM	[2.2.03.02.1] ♦ [SVM-SF-VM-MU-CM]
MSVM	[2.2.03.02.2] ♦ [SVM-SF-VM-MU-MS]
BRBNS	[2.2.03.02.3] ♦ [SVM-SF-VM-MU-BR]
GVM	[2.2.03.02.4] ♦ [SVM-SF-VM-MU-GV]
HCCVM / CCM	[2.2.03.02.5] ♦ [SVM-SF-VM-MU-HC]
VMOS	[2.2.03.02.6] ♦ [SVM-SF-VM-MU-VO]
Syndromic	[2.2.03.03] ♦ [SVM-SF-VM-SY]
PHTS	[2.2.03.03.1] ♦ [SVM-SF-VM-SY-PH]
CLOVES	[2.2.03.03.2] ♦ [SVM-SF-VM-SY-CL]
Mafucci Syndrome	[2.2.03.03.3] ♦ [SVM-SF-VM-SY-MS]
Sinus Pericranii	[2.2.03.03.4] ♦ [SVM-SF-VM-SY-SP]

COMBINED		[2.2.04] ♦ [SVM-SF-C0]
Isolated		[2.2.04.01] ♦ [SVM-SF-C0-IS]
CLVM		[2.2.04.01.1] ♦ [SVM-SF-C0-IS-CLV]
LVM		[2.2.04.01.2] ♦ [SVM-SF-C0-IS-LVM]
CLM		[2.2.04.01.3] ♦ [SVM-SF-C0-IS-CLM]
CVM		[2.2.04.01.4] ♦ [SVM-SF-C0-IS-CVM]
HCCVM / VVM		[2.2.04.01.5] ♦ [SVM-SF-C0-IS-HCC]
Syndromic		[2.2.04.02] ♦ [SVM-SF-C0-SY]
PROS		[2.2.04.02.01] ♦ [SVM-SF-C0-SY-PR]
KTS (CLVM with hypertrophy)		[2.2.04.02.02] ♦ [SVM-SF-C0-SY-KT]
CLOVES		[2.2.04.02.03] ♦ [SVM-SF-C0-SY-CV]
CLAPO		[2.2.04.02.04] ♦ [SVM-SF-C0-SY-CL]
Proteus Syndrome		[2.2.04.02.05] ♦ [SVM-SF-C0-SY-PS]

DEVELOPMENTAL ANOMALIES OF NAMED VESSELS		[2.3] ♦ [SVM-NV]
Vena Cava		[2.3.01] ♦ [SVM-NV-VC]
Aorta		[2.3.02] ♦ [SVM-NV-A0]
Vein of Galen		[2.3.03] ♦ [SVM-NV-VG]
Others		[2.3.04] ♦ [SVM-NV-OT]

POTENTIALLY UNIQUE VASCULAR ANOMALIES [3] ♦ [PUV]

As new or unique vascular anomalies are identified and confirmed, they will be documented here. Updated: 2026.06

Note: This compendium is an adaptation by the Care4-Rare Project. The ISSVA Classification & Glossary for Vascular Anomalies 2025 is owned and copyrighted by ISSVA. Non-commercial use and reproduction are permitted without prior approval. This is for personal reference and patient advocacy use only. Diagnosis is the realm of qualified medical professionals.

Color key: ■ NVT — Neoplastic Vascular Tumors | ■ SVM — Structural Vascular Malformations | ■ PUV — Potentially Unique Vascular Anomalies

◆ **Navigation:** The ◆ symbol between each ISSVA number and Catalog ID is a link to that entry's dedicated page. Pages are added as content is developed.