STRUCTURAL VASCULAR MALFORMATIONS [SVM]

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

SLOW-FLOW [SVM–SF]

CAPILLARY [SVM–SF–CM]

Nevus simplex, Salmon Patch	[SVM–SF–CM–NS] ◈	[VE–EX–LI]
Port Wine CM — Port Wine Birthmark, Nevus flammeus	**[SVM–SF–CM–PW]** ◈	**[VE–EX–LI]**
Isolated *(incl. Phacomatosis Pigmentovascularis)*	[SVM–SF–CM–PW–IS] ◈	[VE–EX–LI]
Syndromic	[SVM–SF–CM–PW–SY] ◈	[VE–EX–IN]
With hypertrophy or extracutaneous disease	[SVM–SF–CM–PW–SY–HYP] ◈	[CO–EX–HI]
Sturge-Weber Syndrome	[SVM–SF–CM–PW–SY–SWS] ◈	[VE–EX–IN]
DCMO	[SVM–SF–CM–PW–SY–DCM] ◈	[VE–EX–IN]
Reticulate / Telangiectatic CM	**[SVM–SF–CM–RT]** ◈	**[VE–EX–LI]**
Isolated	[SVM–SF–CM–RT–IS] ◈	[VE–EX–LI]
Syndromic	[SVM–SF–CM–RT–SY] ◈	[VE–EX–IN]
M-CM	[SVM–SF–CM–RT–SY–MC] ◈	[CO–EX–HI]
MIC-CAP	[SVM–SF–CM–RT–SY–MI] ◈	[CO–EX–HL]
DCMO	[SVM–SF–CM–RT–SY–DC] ◈	[VE–EX–IN]
Geographic Pattern CM	**[SVM–SF–CM–GP]** ◈	**[CO–EX–HI]**
Isolated	[SVM–SF–CM–GP–IS] ◈	[VE–EX–LI]
Syndromic	[SVM–SF–CM–GP–SY] ◈	[CO–EX–HI]
Klippel-Trenaunay Syndrome (KTS)	[SVM–SF–CM–GP–SY–KT] ◈	[CO–EX–HI]
Associated with CLOVES / Disorders of PROS	[SVM–SF–CM–GP–SY–CL] ◈	[CO–EX–HI]

Low-resistance CM / CM with faster flow	**[SVM–SF–CM–LR] ◈ [AV–EX–LI]**
Isolated	[SVM–SF–CM–LR–IS] ◈ [AV–EX–LI]
Syndromic	[SVM–SF–CM–LR–SY] ◈ [AV–EX–IN]
CM-AVM 1 and 2	[SVM–SF–CM–LR–SY–AV] ◈ [AV–EX–IN]
Parkes-Weber Syndrome	[SVM–SF–CM–LR–SY–PW] ◈ [AV–EX–IN]
Cutis Marmorata Telangiectatic Congenita	[SVM–SF–CM–CC] ◈ [VE–TR–OD]
Telangiectasias and Spider Angiomas	**[SVM–SF–CM–TS] ◈ [VE–EX–LI]**
Isolated	[SVM–SF–CM–TS–IS] ◈ [VE–EX–LI]
Syndromic	[SVM–SF–CM–TS–SY] ◈ [AV–EX–LI]
CM-AVM 1 and 2	[SVM–SF–CM–TS–SY–AV] ◈ [AV–EX–LI]
HHT 1 & 2 and JPHT (see also AVM category)	[SVM–SF–CM–TS–SY–HJ] ◈ [AV–EX–IN]
LYMPHATIC	**[SVM–SF–LM]**
Isolated	**[SVM–SF–LM–IS] ◈ [LY–EX–LI]**
LM (Discrete)	[SVM–SF–LM–IS–LD] ◈ [LY–EX–LI]
Macrocystic	[SVM–SF–LM–IS–LD–MA] ◈ [LY–EX–LI]
Microcystic	[SVM–SF–LM–IS–LD–MI] ◈ [LY–EX–LI]
Mixed Macromicrocystic	[SVM–SF–LM–IS–LD–MX] ◈ [LY–EX–LI]
Angiokeratoma	[SVM–SF–LM–IS–AN] ◈ [LY–EX–LI]
Complex	**[SVM–SF–LM–CO] ◈ [LY–EX–IN]**
GLA	[SVM–SF–LM–CO–GL] ◈ [LY–EX–IN]
KLA	[SVM–SF–LM–CO–KL] ◈ [LY–EX–IN]
GSD	[SVM–SF–LM–CO–GS] ◈ [LY–EX–IN]
GLD	[SVM–SF–LM–CO–GD] ◈ [LY–EX–IN]
CCLA	[SVM–SF–LM–CO–CC] ◈ [LY–EX–IN]
Isolated	[SVM–SF–LM–CO–CC–IS] ◈ [LY–EX–IN]
Syndromic (RASopathy)	[SVM–SF–LM–CO–CC–SY] ◈ [CO–EX–HI]

Lymphedemas	**[SVM–SF–LM–LY] ◈ [LY–TR–AP]**
Primary	[SVM–SF–LM–LY–PR] ◈ [LY–TR–AP]
Isolated	[SVM–SF–LM–LY–PR–IS] ◈ [LY–TR–AP]
Syndromic	[SVM–SF–LM–LY–PR–SY] ◈ [CO–TR–HN]
Secondary	[SVM–SF–LM–LY–SE] ◈ [LY–TR–OD]

VENOUS	**[SVM–SF–VM]**
Isolated	**[SVM–SF–VM–IS] ◈ [VE–EX–LI]**
VM (Discrete)	[SVM–SF–VM–IS–VD] ◈ [VE–EX–LI]
Phlebectatic	[SVM–SF–VM–IS–PH] ◈ [VE–TR–OD]
Spongiform	[SVM–SF–VM–IS–SP] ◈ [VE–EX–IN]
VVM	[SVM–SF–VM–IS–VV] ◈ [VE–TR–OD]
FAVA	[SVM–SF–VM–IS–FA] ◈ [VE–EX–IN]
Multifocal	**[SVM–SF–VM–MU] ◈ [VE–EX–IN]**
VMCM	[SVM–SF–VM–MU–CM] ◈ [VE–EX–IN]
MSVM	[SVM–SF–VM–MU–MS] ◈ [VE–EX–IN]
BRBNS	[SVM–SF–VM–MU–BR] ◈ [VE–EX–IN]
GVM	[SVM–SF–VM–MU–GV] ◈ [VE–EX–LI]
HCCVM / CCM	[SVM–SF–VM–MU–HC] ◈ [VE–EX–IN]
VMOS	[SVM–SF–VM–MU–VO] ◈ [VE–EX–IN]
Syndromic	**[SVM–SF–VM–SY] ◈ [CO–TR–HN]**
PHTS	[SVM–SF–VM–SY–PH] ◈ [CO–EX–HI]
CLOVES	[SVM–SF–VM–SY–CL] ◈ [CO–EX–HI]
Mafucci Syndrome	[SVM–SF–VM–SY–MS] ◈ [CO–TR–HN]
Sinus Pericranii	[SVM–SF–VM–SY–SP] ◈ [VE–TR–OD]

COMBINED	[SVM–SF–CO]
Isolated	**[SVM–SF–CO–IS] ◈ [CO–TR–HN]**
CLVM	[SVM–SF–CO–IS–CLV] ◈ [CO–EX–HI]
LVM	[SVM–SF–CO–IS–LVM] ◈ [CO–EX–HI]
CLM	[SVM–SF–CO–IS–CLM] ◈ [CO–EX–HL]
CVM	[SVM–SF–CO–IS–CVM] ◈ [CO–EX–HL]
HCCVM / VVM	[SVM–SF–CO–IS–HCC] ◈ [CO–TR–HN]
Syndromic	**[SVM–SF–CO–SY] ◈ [CO–EX–HI]**
PROS	[SVM–SF–CO–SY–PR] ◈ [CO–EX–HI]
KTS (CLVM with hypertrophy)	[SVM–SF–CO–SY–KT] ◈ [CO–EX–HI]
CLOVES	[SVM–SF–CO–SY–CV] ◈ [CO–EX–HI]
CLAPO	[SVM–SF–CO–SY–CL] ◈ [CO–EX–HL]
Proteus Syndrome	[SVM–SF–CO–SY–PS] ◈ [CO–EX–HI]

FAST-FLOW	[SVM–FF]
Isolated	**[SVM–FF–IS] ◈ [AV–EX–LI]**
AVM	[SVM–FF–IS–AM] ◈ [AV–EX–IN]
Intramuscular Fast-flow Vascular Anomaly	[SVM–FF–IS–IN] ◈ [AV–EX–IN]
AVF	[SVM–FF–IS–AF] ◈ [AV–TR–SU]
Multifocal	**[SVM–FF–MU] ◈ [AV–EX–IN]**
CM-AVM 1 & 2	[SVM–FF–MU–CA] ◈ [AV–EX–IN]
HHT 1 & 2 / JPHT	[SVM–FF–MU–HJ] ◈ [AV–EX–IN]
PHTS	[SVM–FF–MU–PH] ◈ [CO–EX–HI]
Syndromic	**[SVM–FF–SY] ◈ [AV–EX–IN]**
PHOST (PHTS)	[SVM–FF–SY–PH] ◈ [CO–EX–HI]
Parkes-Weber Syndrome	[SVM–FF–SY–PW] ◈ [AV–EX–IN]
SAMS	[SVM–FF–SY–SA] ◈ [AV–EX–IN]
CAMS	[SVM–FF–SY–CA] ◈ [AV–EX–IN]

DEVELOPMENTAL ANOMALIES OF NAMED VESSELS	[SVM–NV]
Vena Cava	[SVM–NV–VC] ◈ [VE–TR–AP]
Aorta	[SVM–NV–AO] ◈ [AR–TR–AP]
Vein of Galen	[SVM–NV–VG] ◈ [AV–TR–DE]
Others	[SVM–NV–OT] ◈ [VE–TR–OD]

◈ — a clickable hot link — go to a full Classification Description (Handout)

HOW TO READ THE CATALOG CODES

Each entry presents three elements: a descriptive name, followed by two Care4-Rare catalog codes separated by the ◈ navigation symbol. The **left code** represents the **ISSVA taxonomy** — identifying structural anatomy and vessel type. The **right code** represents the **Hamburg taxonomy** — identifying embryological origin and predicting clinical behavior, particularly recurrence risk.

These catalog codes are not expressly issued by either ISSVA or Hamburg. Care4-Rare created them as a cross-referencing method — a common anchor for moms, dads, those of us affected, and doctors as we work together to understand and communicate the one hundred individual classifications that define our community.

One important boundary: the Hamburg model does not apply to Neoplastic Vascular Tumors. Entries in that section carry the ISSVA catalog code only.

KEY TERMS

`Truncular` — malformation arising later in fetal development from formed vascular trunks. Minimal recurrence risk.

`Endothelial cell` — the cells lining the inside of every blood and lymphatic vessel; the "wallpaper" of the vascular system.

`Congenital` — present at birth; the condition began during fetal development even if diagnosed later.

`Isolated` — a single malformation with no broader syndrome or systemic involvement.

`Syndromic` — a malformation within a recognized pattern of features: classic clinical syndrome (e.g., KTS, SWS) or genetic diagnosis (e.g., PIK3CA, GNAQ) where the gene is the defining feature.

`Extratruncular` — malformation arising early from primitive tissue. Retains growth potential; high recurrence risk.

`Neoplastic` — abnormal, uncontrolled cell growth creating new tissue. Distinct from malformation, which is structural.

`Perinatal` — occurring in the period just before, during, or shortly after birth.

`Multifocal` — the same malformation type at multiple body sites, often with a shared genetic driver.

NEOPLASTIC VASCULAR TUMORS [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 *	**[NVT–BE]**
Infantile hemangioma	[NVT–BE–IH]
Congenital hemangioma	[NVT–BE–CH]
Tufted angioma	[NVT–BE–TA]
Cherry angioma	[NVT–BE–CA]
Epithelioid hemangioma	[NVT–BE–EH]
Cutaneous epithelioid angiomatous nodule	[NVT–BE–CN]
Pyogenic granuloma — Lobular Capillary Hemangioma	[NVT–BE–PG]
Spindle-cell hemangioma	[NVT–BE–SH]
Hobnail hemangioma	[NVT–BE–HH]
Microvenular hemangioma	[NVT–BE–MH]
Anastomosing hemangioma	[NVT–BE–AH]
Glomeruloid hemangioma	[NVT–BE–GH]
Papillary hemangioma	[NVT–BE–PH]
Acquired elastotic hemangioma	[NVT–BE–AE]
Intravascular papillary endothelial hyperplasia — Masson tumor	[NVT–BE–IP]
Littoral cell hemangioma of the spleen	[NVT–BE–LH]
Placental chorioangioma	[NVT–BE–PL]
Eccrine angiomatous hamartoma	[NVT–BE–EC]
Reactive angioendotheliomatosis	[NVT–BE–RA]
Bacillary angiomatosis	[NVT–BE–BA]

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

Borderline	[NVT–BO]
Kaposiform hemangioendothelioma	[NVT–BO–KH]
Retiform hemangioendothelioma	[NVT–BO–RH]
Papillary intralymphatic angioendothelioma (PILA), Dabska Tumor	[NVT–BO–PI]
Pseudomyogenic hemangioendothelioma	[NVT–BO–PS]
Polymorphous hemangioendothelioma	[NVT–BO–PO]
Kaposi's sarcoma	[NVT–BO–KS]
Composite hemangioendothelioma	[NVT–BO–CO]
Multifocal lymphangioendotheliomatosis with thrombocytopenia (MLT)	[NVT–BO–ML]

Malignant	[NVT–MA]
Angiosarcoma	[NVT–MA–AN]
Epithelioid hemangioendothelioma	[NVT–MA–EP]

POTENTIALLY UNIQUE VASCULAR ANOMALIES [PUV]

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

NVT — Neoplastic Vascular Tumors | SVM — Structural Vascular Malformations |

PUV — Potentially Unique Vascular Anomalies