CARE4-RARE — ISSVA × HAMBURG CROSS-REFERENCE MAPPING TABLE

A Patient-Synthesized Model | Version 1.0 | 2026.06 | For reference and patient advocacy use only

This document maps every Care4-Rare ISSVA alpha catalog code to its Hamburg Classification cross-reference. The two systems answer different clinical questions and are complementary, not competing. ***ISSVA alpha code*** *identifies structural anatomy and vessel type.* ***Hamburg code*** *identifies embryological origin and predicts clinical behavior — particularly recurrence risk and treatment approach. Where one Hamburg code maps to multiple ISSVA entries, this reflects Hamburg's broader biological buckets. Where mapping is not applicable (NVT — vascular tumors), this is noted explicitly.*

HAMBURG CLASSIFICATION — COMPLETE CODE REFERENCE

The Hamburg Classification organizes congenital vascular malformations by ***embryological origin*** *— the stage of fetal development at which the vascular defect occurred. Its defining contribution is the* ***Truncular / Extratruncular*** *distinction: truncular lesions arise later in development from formed vascular trunks and carry minimal recurrence risk; extratruncular lesions arise earlier from primitive mesenchymal tissue and retain the potential to grow, proliferate, and recur after any intervention. This distinction answers the question patients ask most:* ***"Why does mine keep coming back?"*** *The 19 codes below are the complete Hamburg alpha catalog as adapted for the Care4-Rare Patient-Synthesized Model.*

ARTERIAL (AR)

AR–TR–AP	Truncular · Aplasia/obstruction of arterial trunk
AR–TR–OD	Truncular · Obstruction/dilation (aneurysm)
AR–EX–LI	Extratruncular · Limited/localized
AR–EX–IN	Extratruncular · Infiltrating/diffuse

VENOUS (VE)

VE–TR–AP	Truncular · Aplasia/hypoplasia of venous trunk
VE–TR–OD	Truncular · Obstruction/dilation/valvular incompetence
VE–EX–LI	Extratruncular · Limited/localized
VE–EX–IN	Extratruncular · Infiltrating/diffuse

LYMPHATIC (LY)

LY–TR–AP	Truncular · Aplasia/obstruction · lymphedema
LY–TR–OD	Truncular · Ectasia/dilation of lymphatic trunk
LY–EX–LI	Extratruncular · Limited · macrocystic/microcystic
LY–EX–IN	Extratruncular · Infiltrating · diffuse complex LM

ARTERIOVENOUS (AV)

AV–TR–DE	Truncular · Deep AV fistula of named vessel
AV–TR–SU	Truncular · Superficial AV fistula
AV–EX–LI	Extratruncular · Limited AVM with nidus
AV–EX–IN	Extratruncular · Infiltrating/diffuse AVM

COMBINED / HEMOLYMPHATIC (CO)

CO–TR–AV	Truncular · Combined with AV component
CO–TR–HS	Truncular · Hemolymphatic · Superficial
CO–TR–HN	Truncular · Hemolymphatic · No AV shunting
CO–EX–HI	Extratruncular · Hemolymphatic · Infiltrating · high recurrence
CO–EX–HL	Extratruncular · Hemolymphatic · Limited

ISSVA CLASSIFICATION — COMPLETE CODE REFERENCE

> *The Care4-Rare ISSVA alpha catalog code identifies each anomaly by its **structural anatomy and vessel type** — what the malformation is and where it sits in the classification hierarchy. Each entry is cross-referenced to its Hamburg code, which adds the embryological lens. Together the two codes form a complete clinical picture: **what it is** (ISSVA) and **how it behaves** (Hamburg). The **Overlap** column distinguishes entries with a one-to-one Hamburg relationship (**discrete**) from those sharing a Hamburg code with related entries (**shared**) — a reflection of Hamburg's intentionally broader biological groupings.*

SVM — SLOW-FLOW · CAPILLARY MALFORMATIONS

ISSVA Alpha Code	Hamburg	Overlap	Clinical behavior note
SVM–SF–CM–NS	**VE–EX–LI**	*shared*	Nevus simplex / Salmon Patch — superficial capillary, fades spontaneously. Extratruncular limited.
SVM–SF–CM–PW	**VE–EX–LI**	*shared*	Port Wine CM (isolated) — extratruncular, limited. PDL best evidence. Recurrence common.
SVM–SF–CM–PW–IS	**VE–EX–LI**	*shared*	Isolated PWS incl. Phacomatosis Pigmentovascularis. Same Hamburg behavior as parent.
SVM–SF–CM–PW–SY	**VE–EX–IN**	*shared*	Syndromic PWS — venous pressure driven, deeper infiltration. Extratruncular, infiltrating.
SVM–SF–CM–PW–SY–HYP	**CO–EX–HI**	*shared*	With hypertrophy — combined hemolymphatic involvement. High recurrence potential.
SVM–SF–CM–PW–SY–SWS	**VE–EX–IN**	*discrete*	Sturge-Weber — venous pressure pathophysiology, leptomeningeal involvement. Infiltrating.
SVM–SF–CM–PW–SY–DCM	**VE–EX–IN**	*shared*	DCMO — diffuse capillary-venous pattern. Extratruncular infiltrating.
SVM–SF–CM–RT	**VE–EX–LI**	*shared*	Reticulate / Telangiectatic CM — superficial capillary network. Limited extratruncular.
SVM–SF–CM–RT–IS	**VE–EX–LI**	*shared*	Isolated reticulate CM.
SVM–SF–CM–RT–SY	**VE–EX–IN**	*shared*	Syndromic reticulate CM — deeper involvement.
SVM–SF–CM–RT–SY–MC	**CO–EX–HI**	*shared*	M-CM — macrocephaly-CM. Combined hemolymphatic, infiltrating. High recurrence.
SVM–SF–CM–RT–SY–MI	**CO–EX–HL**	*shared*	MIC-CAP — microcephaly-capillary. Combined, limited.
SVM–SF–CM–RT–SY–DC	**VE–EX–IN**	*shared*	DCMO syndromic reticulate variant.
SVM–SF–CM–GP	**CO–EX–HI**	*shared*	Geographic Pattern CM — broad distribution. Combined hemolymphatic. Infiltrating.
SVM–SF–CM–GP–IS	**VE–EX–LI**	*shared*	Isolated geographic CM — limited, no systemic involvement.
SVM–SF–CM–GP–SY	**CO–EX–HI**	*shared*	Syndromic geographic CM — combined infiltrating.
SVM–SF–CM–GP–SY–KT	**CO–EX–HI**	*discrete*	**KTS** — hemolymphatic, extratruncular, infiltrating. Explains PDL recurrence. Genetic program ongoing.
SVM–SF–CM–GP–SY–CL	**CO–EX–HI**	*shared*	CLOVES/PROS — PIK3CA driven. Same hemolymphatic infiltrating behavior as KTS.
SVM–SF–CM–LR	**AV–EX–LI**	*shared*	Low-resistance CM / faster flow — transitional to AV territory. Extratruncular limited.
SVM–SF–CM–LR–IS	**AV–EX–LI**	*shared*	Isolated low-resistance CM.
SVM–SF–CM–LR–SY	**AV–EX–IN**	*shared*	Syndromic low-resistance CM — AV shunting present. Infiltrating.
SVM–SF–CM–LR–SY–AV	**AV–EX–IN**	*shared*	CM-AVM 1 & 2 — capillary + AV component. High recurrence. Not laser appropriate.

SVM–SF–CM–LR–SY–PW	**AV–EX–IN**	*discrete*	**Parkes-Weber** — high-flow AV shunting. Extratruncular infiltrating. Embolization, not laser.
SVM–SF–CM–CC	**VE–TR–OD**	*discrete*	Cutis Marmorata Telangiectatic Congenita — truncular, often resolves partially. Less recurrence.
SVM–SF–CM–TS	**VE–EX–LI**	*shared*	Telangiectasias / Spider Angiomas — superficial extratruncular limited.
SVM–SF–CM–TS–IS	**VE–EX–LI**	*shared*	Isolated telangiectasias.
SVM–SF–CM–TS–SY	**AV–EX–LI**	*shared*	Syndromic telangiectasias — AV component emerging.
SVM–SF–CM–TS–SY–AV	**AV–EX–LI**	*shared*	CM-AVM 1 & 2 telangiectatic variant.
SVM–SF–CM–TS–SY–HJ	**AV–EX–IN**	*shared*	HHT 1 & 2 / JPHT — hereditary hemorrhagic, AV infiltrating. Systemic involvement.

> ** Hamburg capillary malformations are classified within the venous or combined categories — Hamburg predates the ISSVA capillary-specific classification and treats capillary lesions as capillary-venous in nature.*

SVM — SLOW-FLOW · LYMPHATIC MALFORMATIONS

ISSVA Alpha Code	Hamburg	Overlap	Clinical behavior note
SVM–SF–LM–IS	**LY–EX–LI**	*shared*	Isolated LM — extratruncular limited. Most common presentation.
SVM–SF–LM–IS–LD	**LY–EX–LI**	*shared*	LM Discrete — macrocystic / microcystic / mixed. All extratruncular limited.
SVM–SF–LM–IS–LD–MA	**LY–EX–LI**	*shared*	Macrocystic — best sclerotherapy response. Limited extratruncular.
SVM–SF–LM–IS–LD–MI	**LY–EX–LI**	*shared*	Microcystic — more resistant to treatment. Still limited extratruncular.
SVM–SF–LM–IS–LD–MX	**LY–EX–LI**	*shared*	Mixed — variable response. Extratruncular limited.
SVM–SF–LM–IS–AN	**LY–EX–LI**	*shared*	Angiokeratoma — superficial lymphatic-capillary. Limited extratruncular.
SVM–SF–LM–CO	**LY–EX–IN**	*shared*	Complex LM — infiltrating, systemic involvement. High recurrence.
SVM–SF–LM–CO–GL	**LY–EX–IN**	*shared*	GLA — Generalized Lymphatic Anomaly. Diffuse extratruncular infiltrating.
SVM–SF–LM–CO–KL	**LY–EX–IN**	*shared*	KLA — Kaposiform Lymphangiomatosis. Aggressive infiltrating. High morbidity.
SVM–SF–LM–CO–GS	**LY–EX–IN**	*shared*	GSD — Gorham-Stout Disease. Bone involvement. Infiltrating.
SVM–SF–LM–CO–GD	**LY–EX–IN**	*shared*	GLD — diffuse complex lymphatic anomaly. Infiltrating.
SVM–SF–LM–CO–CC	**LY–EX–IN**	*shared*	CCLA — central conducting lymphatic anomaly. Truncal, infiltrating.
SVM–SF–LM–CO–CC–IS	**LY–EX–IN**	*shared*	CCLA Isolated.
SVM–SF–LM–CO–CC–SY	**CO–EX–HI**	*shared*	CCLA Syndromic (RASopathy) — combined hemolymphatic, infiltrating.
SVM–SF–LM–LY	**LY–TR–AP**	*shared*	Lymphedemas — truncular aplasia/obstruction. Less recurrence post-treatment than extratruncular.
SVM–SF–LM–LY–PR	**LY–TR–AP**	*shared*	Primary lymphedema — truncular developmental arrest.
SVM–SF–LM–LY–PR–IS	**LY–TR–AP**	*shared*	Primary lymphedema isolated.
SVM–SF–LM–LY–PR–SY	**CO–TR–HN**	*shared*	Primary lymphedema syndromic — combined truncular, no AV shunting.
SVM–SF–LM–LY–SE	**LY–TR–OD**	*shared*	Secondary lymphedema — acquired truncular obstruction/dilation.

SVM — SLOW-FLOW · VENOUS MALFORMATIONS

ISSVA Alpha Code	Hamburg	Overlap	Clinical behavior note
SVM–SF–VM–IS	**VE–EX–LI**	*shared*	Isolated VM — extratruncular limited. Sclerotherapy first line.
SVM–SF–VM–IS–VD	**VE–EX–LI**	*shared*	VM Discrete — localized. Limited extratruncular. Good sclerotherapy response.
SVM–SF–VM–IS–PH	**VE–TR–OD**	*shared*	Phlebectatic VM — truncular dilation of venous trunk. Hemodynamic consequences.
SVM–SF–VM–IS–SP	**VE–EX–IN**	*shared*	Spongiform VM — diffuse infiltrating. High recurrence. Difficult to treat completely.
SVM–SF–VM–IS–VV	**VE–TR–OD**	*shared*	VVM — venous/vascular malformation. Truncular dilation pattern.
SVM–SF–VM–IS–FA	**VE–EX–IN**	*discrete*	FAVA — fibro-adipose vascular anomaly. Deeply infiltrating. Pain predominant. Surgical.
SVM–SF–VM–MU	**VE–EX–IN**	*shared*	Multifocal VM — multiple sites, infiltrating. Systemic monitoring required.
SVM–SF–VM–MU–CM	**VE–EX–IN**	*shared*	VMCM — cutaneomucosal VM, familial. Infiltrating, multiple lesions.
SVM–SF–VM–MU–MS	**VE–EX–IN**	*shared*	MSVM — multisystemic VM. Infiltrating.
SVM–SF–VM–MU–BR	**VE–EX–IN**	*discrete*	BRBNS — Blue Rubber Bleb Nevus Syndrome. GI involvement. Infiltrating, systemic.
SVM–SF–VM–MU–GV	**VE–EX–LI**	*shared*	GVM — glomuvenous malformation. Limited, less aggressive recurrence.
SVM–SF–VM–MU–HC	**VE–EX–IN**	*shared*	HCCVM / CCM — cerebral cavernous malformation. CNS infiltrating. Neurological monitoring.
SVM–SF–VM–MU–VO	**VE–EX–IN**	*shared*	VMOS — VM with osteoporosis/skeletal involvement. Infiltrating.
SVM–SF–VM–SY	**CO–TR–HN**	*shared*	Syndromic VM — combined truncular, no significant AV shunting.
SVM–SF–VM–SY–PH	**CO–EX–HI**	*shared*	PHTS (PTEN) — combined hemolymphatic infiltrating. Tumor suppressor involvement.
SVM–SF–VM–SY–CL	**CO–EX–HI**	*shared*	CLOVES — complex combined. PIK3CA driven. High recurrence. Infiltrating.
SVM–SF–VM–SY–MS	**CO–TR–HN**	*shared*	Mafucci Syndrome — combined truncular. Enchondroma association.
SVM–SF–VM–SY–SP	**VE–TR–OD**	*discrete*	Sinus Pericranii — truncular dilation of dural venous sinus. Hemodynamic consequences.

SVM — SLOW-FLOW · COMBINED MALFORMATIONS

ISSVA Alpha Code	Hamburg	Overlap	Clinical behavior note
SVM–SF–CO–IS	**CO–TR–HN**	*shared*	Isolated combined — truncular combined, no AV shunting. Structural.
SVM–SF–CO–IS–CLV	**CO–EX–HI**	*shared*	CLVM — capillary-lymphatic-venous. Hemolymphatic infiltrating. High recurrence.
SVM–SF–CO–IS–LVM	**CO–EX–HI**	*shared*	LVM — lymphatic-venous combined. Infiltrating.
SVM–SF–CO–IS–CLM	**CO–EX–HL**	*shared*	CLM — capillary-lymphatic combined. Limited.
SVM–SF–CO–IS–CVM	**CO–EX–HL**	*shared*	CVM — capillary-venous combined. Limited.
SVM–SF–CO–IS–HCC	**CO–TR–HN**	*shared*	HCCVM / VVM — combined truncular. No AV shunting.
SVM–SF–CO–SY	**CO–EX–HI**	*shared*	Syndromic combined — hemolymphatic infiltrating. Genetic driver present in most.

SVM–SF–CO–SY–PR	**CO–EX–HI**	*shared*	PROS — PIK3CA spectrum. Hemolymphatic infiltrating. High recurrence. mTOR pathway.
SVM–SF–CO–SY–KT	**CO–EX–HI**	*discrete*	**KTS (CLVM with hypertrophy)** — hemolymphatic infiltrating. Explains all recurrence patterns. Core audience diagnosis.
SVM–SF–CO–SY–CV	**CO–EX–HI**	*shared*	CLOVES — complex combined. PIK3CA. Infiltrating. High recurrence.
SVM–SF–CO–SY–CL	**CO–EX–HL**	*shared*	CLAPO — capillary-lymphatic-AVM with PROS features. Limited combined.
SVM–SF–CO–SY–PS	**CO–EX–HI**	*shared*	Proteus Syndrome — PTEN/AKT1 driven. Hemolymphatic infiltrating. Progressive.

SVM — FAST-FLOW MALFORMATIONS

ISSVA Alpha Code	Hamburg	Overlap	Clinical behavior note
SVM–FF–IS	**AV–EX–LI**	*shared*	Isolated fast-flow — extratruncular limited. Embolization/surgery first line.
SVM–FF–IS–AM	**AV–EX–IN**	*shared*	AVM — nidus present. Extratruncular infiltrating. High recurrence without complete excision.
SVM–FF–IS–IN	**AV–EX–IN**	*shared*	Intramuscular fast-flow anomaly — deep infiltrating AV component.
SVM–FF–IS–AF	**AV–TR–SU**	*shared*	AVF — direct AV fistula, no nidus. Truncular superficial. Better surgical outcomes.
SVM–FF–MU	**AV–EX–IN**	*shared*	Multifocal fast-flow — multiple infiltrating AV lesions. Systemic hemodynamic impact.
SVM–FF–MU–CA	**AV–EX–IN**	*shared*	CM-AVM 1 & 2 — capillary + AV multifocal. RASA1/ EPHB4. Infiltrating.
SVM–FF–MU–HJ	**AV–EX–IN**	*shared*	HHT 1 & 2 / JPHT — hereditary hemorrhagic telangiectasia. Multifocal AV infiltrating. Pulmonary/ hepatic risk.
SVM–FF–MU–PH	**CO–EX–HI**	*shared*	PHTS — PTEN hamartoma. Combined hemolymphatic infiltrating.
SVM–FF–SY	**AV–EX–IN**	*shared*	Syndromic fast-flow — AV infiltrating, genetic driver.
SVM–FF–SY–PH	**CO–EX–HI**	*shared*	PHOST (PHTS) — combined hemolymphatic infiltrating.
SVM–FF–SY–PW	**AV–EX–IN**	*discrete*	**Parkes-Weber Syndrome** — AV infiltrating, extratruncular. Hemodynamically significant. High cardiac risk.
SVM–FF–SY–SA	**AV–EX–IN**	*shared*	SAMS — AV infiltrating. Spinal involvement.
SVM–FF–SY–CA	**AV–EX–IN**	*shared*	CAMS — cerebral AV malformation syndromic. CNS infiltrating.

SVM — DEVELOPMENTAL ANOMALIES OF NAMED VESSELS

ISSVA Alpha Code	Hamburg	Overlap	Clinical behavior note
SVM–NV–VC	**VE–TR–AP**	*discrete*	Vena Cava anomaly — truncular aplasia/stenosis. Major hemodynamic consequences.
SVM–NV–AO	**AR–TR–AP**	*discrete*	Aortic anomaly — truncular arterial. Hemodynamic. Surgical intervention often required.
SVM–NV–VG	**AV–TR–DE**	*discrete*	Vein of Galen malformation — deep truncular AV. High-flow. Cardiac and neurological risk.
SVM–NV–OT	**VE–TR–OD**	*shared*	Other named vessel anomalies — truncular obstruction/ dilation pattern.

SUMMARY — HAMBURG CODE DISTRIBUTION ACROSS CARE4-RARE CATALOG

Hamburg Code	Meaning	ISSVA entries	Primary associations
CO—EX—HI	Combined · Extratruncular · Hemolymphatic · Infiltrating	**16**	KTS, CLOVES, PROS, Proteus, PHTS, M-CM — the high-recurrence combined malformation cluster
VE—EX—LI	Venous · Extratruncular · Limited	**12**	Isolated PWS, isolated CM, telangiectasias, discrete VM — common limited lesions
AV—EX—IN	Arteriovenous · Extratruncular · Infiltrating	**11**	AVM, Parkes-Weber, HHT, CM-AVM, CAMS — high-flow infiltrating
VE—EX—IN	Venous · Extratruncular · Infiltrating	**9**	SWS, syndromic PWS, spongiform VM, BRBNS, FAVA
LY—EX—IN	Lymphatic · Extratruncular · Infiltrating	**7**	GLA, KLA, GSD, complex LM — aggressive lymphatic anomalies
LY—EX—LI	Lymphatic · Extratruncular · Limited	**6**	Macrocystic, microcystic, mixed LM, angiokeratoma
CO—EX—HL	Combined · Extratruncular · Hemolymphatic · Limited	**4**	CLM, CVM, CLAPO, MIC-CAP — limited combined
AV—EX—LI	Arteriovenous · Extratruncular · Limited	**4**	Low-resistance CM, isolated fast-flow, CM-AVM limited
VE—TR—OD	Venous · Truncular · Obstruction/Dilation	**4**	Phlebectatic VM, VVM, Sinus Pericranii, CMTC
CO—TR—HN	Combined · Truncular · Hemolymphatic · No AV shunting	**4**	Syndromic VM, Mafucci, primary lymphedema syndromic, HCCVM
LY—TR—AP	Lymphatic · Truncular · Aplasia	**3**	Primary lymphedemas — truncular developmental arrest
AR—TR—AP	Arterial · Truncular · Aplasia	**1**	Aortic anomaly
AV—TR—DE	Arteriovenous · Truncular · Deep	**1**	Vein of Galen malformation
AV—TR—SU	Arteriovenous · Truncular · Superficial	**1**	AVF — direct arteriovenous fistula
VE—TR—AP	Venous · Truncular · Aplasia	**1**	Vena Cava anomaly
LY—TR—OD	Lymphatic · Truncular · Obstruction/Dilation	**1**	Secondary lymphedema
N/A	Not applicable — neoplastic, outside Hamburg scope	**32**	All NVT entries — Benign, Borderline, Malignant

> *Total SVM entries with Hamburg cross-reference: 87 | NVT entries (Hamburg N/A): 32 | PUV: pending individual assignment | Hamburg codes used: 16 of 19 defined codes | Unused codes: AR—EX—LI, AR—EX—IN, CO—TR—AV, CO—TR—HS (arterial extratruncular and truncular AV combined — not represented in current ISSVA catalog entries)*

On overlapping classes: Hamburg categories are intentionally broader than ISSVA entries. One Hamburg code commonly maps to multiple ISSVA entries — this reflects Hamburg's classification of biological behavior rather than structural identity. Where multiple ISSVA entries share a Hamburg code, they share the same embryological origin story and the same clinical behavior predictions. This overlap is a feature, not an ambiguity.

NVT — NEOPLASTIC VASCULAR TUMORS · HAMBURG NOT APPLICABLE

The Hamburg Classification was designed specifically for **congenital vascular malformations (CVM)** — structural errors of vascular development. It does not apply to neoplastic vascular tumors, which involve abnormal endothelial cell proliferation rather than structural malformation. Hamburg explicitly excludes hemangiomas and lymph node abnormalities from its scope. The NVT entries below are therefore annotated **N/A — Hamburg** and remain classified solely by the Care4-Rare ISSVA alpha code. This boundary is clinically important: it reinforces the fundamental distinction between malformation and neoplasm.

ISSVA Alpha Code	Hamburg	Note
`NVT–BE – all entries`	N/A	Benign vascular tumors — endothelial proliferative. Outside Hamburg scope.
`NVT–BO – all entries`	N/A	Borderline tumors — proliferative. Outside Hamburg scope.
`NVT–MA – all entries`	N/A	Malignant tumors — outside Hamburg scope. ISSVA alpha code only.