Classification Systems of Arteriovenous Malformations

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Introduction

Vascular anomalies are **rare** conditions, which are based on deviations in vasculogenesis and angiogenesis, resulting in vascular tumors or malformations.

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This leads to a **great variety** of diseases, locations, degree of symptoms and therapies.

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The great variety represents a **challenge** to a classification, affecting treatment standardization and scientific evaluation.

ISSVA Classification – Tumors

- Infantile Hemangioma
- Rapidly involuting congenital hemangioma (RICH)
- Non-involuting hemangioma (NICH)
- Tufted Angioma
- Kaposiform Hemangioendothelioma
- Epitheloid Hemangioendothelioma (various subtypes)
- Dermatologic acquired vascular tumors (various subtypes)

Wassef M, Blei F, Adams D et al. Vascular Anomalies Classification: Recommendations from the International Society for the Study of Vascular Anomalies. Pediatrics 2015; 136:e203-e214

ISSVA Classification – Vascular Malformations

- **C**apillary Malformation
- **V**enous Malformation
- **L**ymphatic Malformation
- **A**rterial or Arteriovenous Malformations
- Arteriovenous Fistula
- Combined:
  - CVM, CLM, LVM, CLVM
  - AVM-LM, CM-AVM

70-80% venous, 5-15% lymphatic, 2-10% arteriovenous

Classification based on angiography

Type	Vascular Anomaly	Characterization
Type I	Arteriovenous fistula	Up to 3 direct fistulae without circumscribed nidus
Type II	Arteriolovenous Malformation	Many feeding arteries and one dominant draining vein
Type IIIa	Arteriolovenoulous Malformation	Non-dilated, microfistulous nidus
Type IIIb	Arteriolovenoulous Malformation	Dilated nidus

Classification based on angiography

Type I		Up to 3 direct fistulae without circumscribed nidus
Type II		Many feeding arteries and one dominant draining vein
Type IIIa		Non-dilated, microfistulous nidus
Type IIIb		Dilated nidus

Cho SK, Do YS, Shin SW, et al. Arteriovenous Malformations of the Body and Extremities: Analysis of Therapeutic Outcomes and Approaches According to a Modified Angiographic Classification. J Endovasc Ther 2006;13:527-538.

Categorization based on flow speed

Category[1]	Vascular Anomaly	Possible Rx Options[2]
Fast-Flow Malformation	Arteriovenous Malformation	Transarterial, direct, transvenous embolization
	Arteriovenous Fistula	Embolization
Slow-Flow Malformation	Venous Malformation	Sclerotherapy, Lasertherapy, Resection
	Lymphatic Malformation	Sclerotherapy, Lasertherapy, Resection, (mTOR inhibitors)
	Capillary Malformation	Lasertherapy

1. There is no V_{max} threshold; A flow void in T2 is proposed as differentiation
2. Based on: Wohlgemuth WA, Meyer L, Sadick M. Gefäßanomalien: Eine interdisziplinäre Herausforderung. Dtsch Arztebl 2017;114:51-52

Schobinger Classification of clinical presentation

Type	Characterization	Description
Grade 1	Quiescence	Asymptomatic, blue-skin blush, skin warmth
Grade 2	Expansion	Swelling, thrill/bruit, pain
Grade 3	Destruction	Skin dystrophy, persistent pain, bleeding, ulceration,
Grade 4	Decompensation	Cardiac failure

Based on Eigth Meeting of the International Workshop on Vascular Malformations. Amsterdam, Netherlands, 1990. Found in: Leghien GM, Heran MKS: Classification, Diagnosis, and Interventional Radiologic Management of Vascular Malformations. Orthop Clin N Am 2006;37:435-474.

Which classification should be used clinically?

Dignity?	Rx Indicated?	Which Rx?	Success likely?
ISSVA	ISSVA	Flow Speed	Angiography
	Schobinger	Angiography	

Last Words

Almost everything relevant I have learned about AVMs I have picked up from Walther Wohlgemuth, so please excuse me if you see many parallels. I hope I will someday get anywhere close to his understanding of complex disease.

This work will further be enhanced by own angiographies, which still must be optimized.

I would be grateful for thoughts and corrections: giovanni-federico.torsello@charite.de