Adjusted classification for venous malformations

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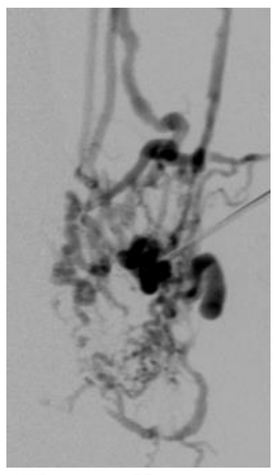
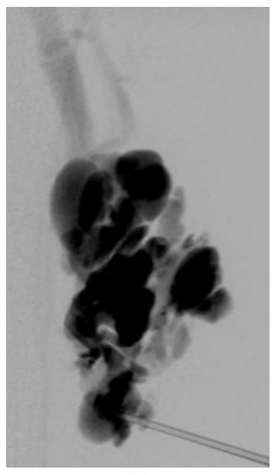
Aim of this study is to validate an adjusted classification for congenital venous malformations (VM). The current classification as proposed by Puig et al. in 2003 is based on the type of drainage (type I, II, III and IV), but does not take into count the exact anatomic features of the VM nidus itsself. As the angioarchitecture of the VM plays an important role in interventional treatment planning and reduction of potential complications of embolo-sclerotherapy, we aim to validate the adjustment of the existing classification by separating a non-lacunar (a) and a lacunar (b) type of venous nidus as new element for classification.

Exemplary direct puncture phlebograms



Intramuscular VM of the thigh

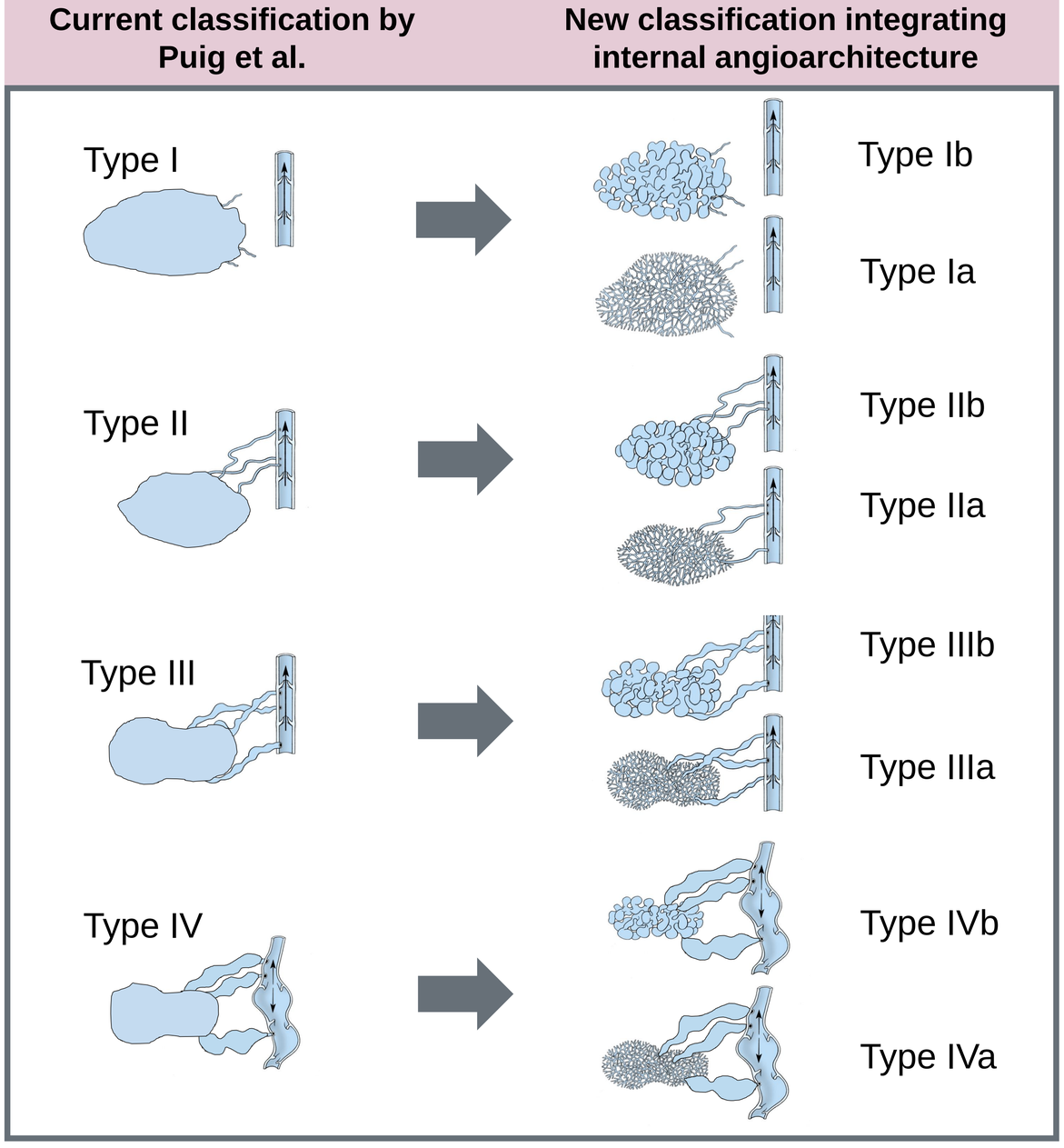
Type II a
(non-lacunar > 80% by visual estimation)



Intramuscular VM of th calf

Type II b
(lacunar > 80% by visual estimation)

Current classification by Puig et al. → **New classification integrating internal angioarchitecture**



Methods

We retrospectively analyzed all patients with confirmed diagnosis of VM and direct contrast enhanced phlebogram with sufficient image quality to reliably assess morphology and flow characteristics of the VM, seen in the division of Angiology between 2009 and 2018.
Phlebograms were read by two experienced readers in a three-step process: 1. non-lacunar (a) or lacunar (b) nidus of VM (defined as > 80% non-lacunar or lacunar type by visual estimation), 2. assignment according to the Puig classification (type I-IV), 3. adjusted classification combining both. Both readers were blinded to clinical data and reading was performed independently.
Primary endpoint was the inter-reader-reliability for the new classification.

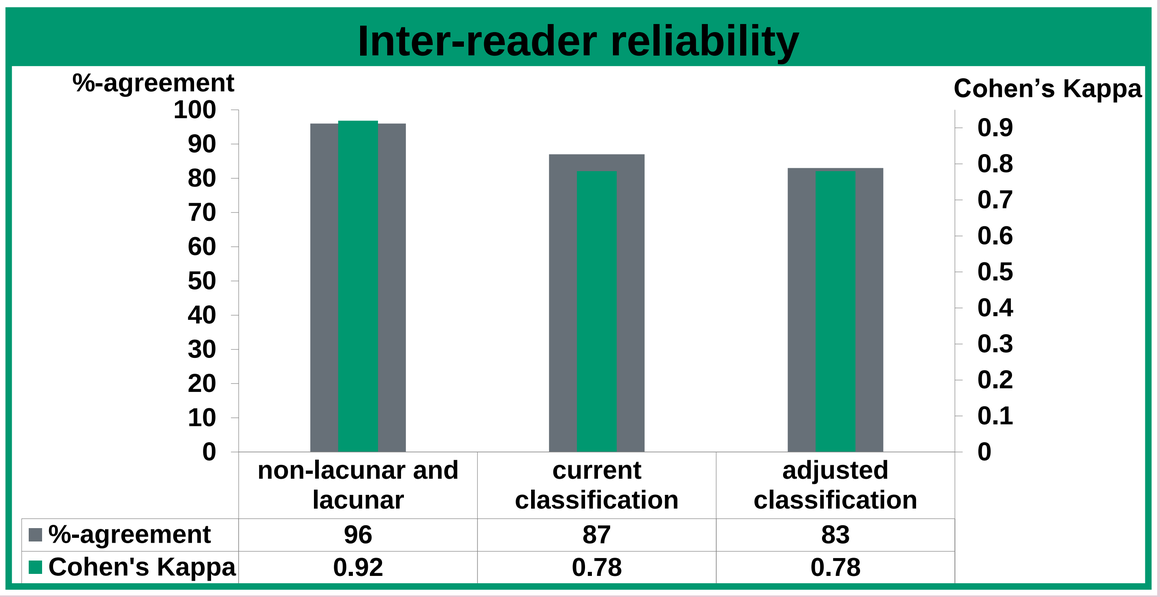
Results

Patient characteristics

	Total		Type A (non-lacunar)		Type B (lacunar)		
	n	median [lq, uq] or n (%)	n	median [lq, uq] or n (%)	n	median [lq, uq] or n (%)	P-value
Sex	**67**		**26**		**41**		**0.48**
Female		40 (59.7%)		18 (69.2%)		22 (53.7%)	
Male		27 (40.3%)		8 (30.8%)		19 (46.3%)	
Age at diagnosis	**67**	**23.0 [18.0, 37.0]**	**26**	**26.0 [18.0, 40.0]**	**41**	**22.0 [18.0, 35.0]**	**0.15**
Localization	**67**		**26**		**41**		**0.44**
Extremities		51 (76.1%)		23 (88.5%)		29 (70.7%)	
Trunk		8 (11.9%)		2 (7.7%)		7 (17.1%)	
Head		5 (7.5%)		1 (3.8%)		5 (12.2%)	
Other		3 (4.5%)		0 (0.000%)		2 (4.35%)	
Extension	**67**		**26**		**41**		**0.81**
Localized		38 (56.7%)		16 (61.5%)		22 (53.7%)	
Infiltrative		26 (43.3%)		10 (38.5%)		19 (46.3%)	
D-dimere levels	**64**	**348 [208, 1047]**	**25**	**300 [208, 594]**	**39**	**477 [208, 1236]**	**0.18**
≤ 500 µg/l		39 (58.2%)		17 (68.0%)		22 (56.4%)	
> 500 µg/l		28 (41.8%)		8 (32.0%)		17 (44.6%)	

Categorical data are presented as n (%) and compared using Fisher exact test. Continuous data are presented as median [lower quartile, upper quartile] and compared using Wilcoxon-Mann-Whitney test.

Inter-reader reliability



	non-lacunar and lacunar	current classification	adjusted classification
%-agreement	96	87	83
Cohen's Kappa	0.92	0.78	0.78

Discussion

There were no significant differences in the patients characteristics, but patients with lacunar VM nidus showed slightly higher D-dimer levels and were younger at the time of diagnosis. This observation further underlines the clinical and interventional importance of the differentiation into lacunar and non-lacunar nidus of venous malformations. There seems to be a more pronounced low-flow in lacunar lesions resulting in an increased localized coagulation leading to earlier clinical manifestation and impacting flow and distribution patterns during embolo-sclerotherapy. The addition of the internal angioarchitecture of the VM nidus itself to the existing classification did not change the level of agreement.

Conclusion

In the majority of patients with VM a reliable phlebographic differentiation of venous drainage (Puig classification) and nidus structure (non-lacunar/lacunar) can be achieved. We recommend to adjust the existing VM classification by differentiating “type a” for non-lacunar and “type b” for lacunar VM nidus to facilitate interventional treatment planning and reduce potential complications of embolo-sclerotherapy.

References
[1] Puig S, Aref H, Chigot V, Bonin B, Brunelle F. Classification of venous malformations in children and implications for sclerotherapy. Pediatr Radiol. 2003 Feb;33(2):99-103. Epub 2002 Nov 19.
[2] Puig S, Casati B, Staudenherz A, Paya K. Vascular low-flow malformations in children: current concepts for classification, diagnosis and therapy. Eur J Radiol. 2005 Jan;53(1):35-45.