Pelvic Vascular Malformations

Brian M. Christenson, MD¹ Matthew G. Gipson, MD¹ Mitchell T. Smith, MD, MS¹

¹ Division of Interventional Radiology, Department of Radiology, University of Colorado School of Medicine, Denver Anschutz Medical Campus, Aurora, Colorado

Address for correspondence Mitchell T. Smith, MD, MS, Division of Interventional Radiology, Department of Radiology, University of Colorado, Denver Anschutz Medical Campus, Aurora, CO 80045 (e-mail: Mitchell.Smith@ucdenver.edu).

Semin Intervent Radiol 2013;30:364–371

Abstract

Keywords

- interventional radiology
- vascular malformations
- endovascular treatment
- arteriovenous malformations
- lymphatic malformations
- venous malformations

Vascular malformations (VMs) comprise a wide spectrum of lesions that are classified by content and flow characteristics. These lesions, occurring in both focal and diffuse forms, can involve any organ and tissue plane and can cause significant morbidity in both children and adults. Since treatment strategy depends on the type of malformation, correct diagnosis and classification of a vascular lesion are crucial. Slow-flow VMs (venous and lymphatic malformations) are often treated by sclerotherapy, whereas fast-flow lesions (arteriovenous malformations) are generally managed with embolization. In addition, some cases of VMs are best treated surgically. This review will present an overview of VMs in the female pelvis as well as a discussion of endovascular therapeutic techniques.

Objectives: Upon completion of this article, the reader will be able to discuss the evaluation and endovascular treatment of patients with low- and high-flow vascular malformations in the pelvis.

Accreditation: This activity has been planned and implemented in accordance with the Essential Areas and policies of the Accreditation Council for Continuing Medical Education (ACCME) through the joint sponsorship of Tufts University School of Medicine (TUSM) and Thieme Medical Publishers, New York. TUSM is accredited by the ACCME to provide continuing medical education for physicians.

Credit: Tufts University School of Medicine designates this journal-based CME activity for a maximum of **1 AMA PRA Category 1 Credit™**. Physicians should claim only the credit commensurate with the extent of their participation in the activity.

Vascular malformations (VMs) can occur in any organ or tissue, including the female pelvis, and can often cause significant morbidity. Several classification systems have been proposed for VMs; the most widely accepted classifica-

tion system was described by Mulliken and Glowacki¹ in 1982. It is a biologic classification based on cellular turnover, histologic features, natural history, and physical findings.¹ In this classification scheme, malformations are categorized as venous, lymphatic, capillary, arterial, or combined. In 1993, Jackson et al² proposed a re-structuring of the radiologic classification formulated in combination with the biologic classification of Mulliken and Glowacki.¹ Jackson et al² sub-categorized VMs according to their flow dynamics as low-flow or high-flow malformations, as these two categories have varied therapeutic approaches.

Traditionally, ultrasound (US) has been the imaging modality of choice for initial assessment and characterization of soft-tissue lesions of presumed vascular origin.³ However, US has the disadvantages of a limited field of view, restricted three-dimensional (3D) imaging, and operator dependency.⁴ Magnetic resonance imaging (MRI) has now become the most valuable modality for classification of vascular anomalies because it accurately demonstrates the extension of a lesion and its anatomic relationship to adjacent structures.⁵ In most patients, diagnosis and proper classification of pelvic VMs are

achieved on the basis of an accurate clinical history and physical examination results, as well as imaging findings.

Venous Malformations

Venous malformations are defined as slow flowing abnormal venous networks.⁶ Typically, these anomalies are caused by germline or somatic mutations in the *TIE2* gene, which is involved in signaling between endothelial and mesenchymal cells during vasculogenesis and angiogenesis.^{7,8} Histologically, these lesions are composed of large and small dysplastic, postcapillary, thin-walled vascular channels with sparse smooth muscle and variable amounts of hamartomatous stroma, thrombi, and phleboliths.^{9,10} The dysplastic venous channels usually connect with adjacent normal physiologic veins via narrow tributaries.¹¹ An intramural muscular anomaly is likely responsible for the gradual expansion of these lesions.^{6,12}

Venous malformations are the most common VM^{6,11,13,14} and may be isolated anomalies or part of an underlying syndrome, such as blue rubber bleb nevus, Proteus, and Maffucci syndromes.⁵ They are usually located in the head and neck (40% of cases), trunk (20%), and extremities (40%).¹⁰ Venous anomalies involving the female pelvis are rare. The most common site of a venous malformation in the female pelvis is the perineum, usually extending to involve the labia majora.¹⁵ Less commonly, the rectal wall, vagina, uterus, or bladder may be affected.

Venous malformations are present at birth, but symptoms from these lesions are often delayed until late childhood or early adulthood. The clinical presentation of venous malformations depends on both the depth and extent of the lesion.⁵ Patients with venous malformations in the pelvic region commonly present with swelling and pain, especially during prolonged standing, walking, or exercising, and these symptoms often worsen during menses or pregnancy. Venous malformations involving the external genitals may be both aesthetically and functionally disabling.¹⁶ On physical examination, the overlying skin often has a bluish-tinged color, and subcutaneous or cutaneous varices may also be evident. They are easily compressible, nonpulsatile masses that increase in size with maneuvers that increase venous pressure.¹⁷ Although these venous anomalies may be limited to the labia majora, they are more commonly an extension of a diffuse venous malformation of the lower extremity, perineum, and buttock.^{18,19}

Venous malformations of the rectum are frequently associated with varicosities due to insufficiency of the hemorrhoidal, mesenteric, and portal veins.^{20–22} Morbidity involved with rectal venous malformations can be significant. In addition to the presence of rectal bleeding, which may be severe, affected patients may develop thrombosis of the hemorrhoidal veins with subsequent portal vein thromboembolism and possible hepatic infarction.^{20–22} Venous malformations of the uterus and ovaries are typically associated with insufficiency of the ovarian vein and may coexist with pelvic congestion syndrome.²³ An isolated venous malformation of the uterus is rare. Venous malformations can also

involve the urinary system, which can lead to severe problems with hematuria.^{21,22} Diffuse venous malformations of the lower extremities with extension into the perineum and buttock are typically associated with painful swelling, and patients are at increased risk for venous thrombosis.^{19,24}

On MRI evaluation, venous malformations are generally septated lesions with intermediate to decreased signal intensity on T1-weighted images and increased signal intensity on T2-weighted images. Hemorrhage or thrombosis can cause heterogeneous signal intensity on T1-weighted images, and occasionally hemorrhage or high protein content may cause internal fluid–fluid levels.⁵ A helpful clue for identification of a venous malformation is the presence of phleboliths,²⁵ which appear as small low-signal-intensity foci on all sequences. Although venous malformations can be associated with surrounding fibrofatty stroma or edema, they rarely appear mass-like.^{9,26,27} Accordingly, lesions with unusual presentation or imaging features should be evaluated with biopsy.^{26,27} Therapeutic options for the treatment of pelvic venous malformations include lower extremity graded elastic compression stockings, endovascular ablative techniques, and resection.

Endovascular Treatment of Pelvic Venous Malformations

Utilization of MRI has been helpful in guiding therapy. Together with lack of evident venous drainage, well-defined lesion margins at MRI have been shown to be a predictor of good outcome after percutaneous sclerotherapy.^{25,28,29} The length of skin involvement and the absence of deep muscle involvement on MRI are additional features that can predict outcome. Patients with more skin involvement and less deep muscle involvement are more likely to develop skin burns when treated with sclerotherapy.³⁰ Direct cannulation and sclerosant injection, typically using either 3% sodium tetradecyl sulfate (STS) foam or absolute ethanol, constitutes the main endovascular approach to treatment of external genitalia venous malformations.^{15,24,31} Ethanol and STS typically cause significant soft tissue swelling, with the associated risk of skin breakdown and possible infection.¹⁷

An additional available treatment modality is endovenous laser therapy, which is performed by inserting a bare laser fiber directly through an intralesional cannula. This therapy can be combined with sclerosant injection and often results in less swelling than sclerotherapy alone due to less sclerosant being injected.¹⁷ In some larger venous malformations, surgical resection of a grossly enlarged labium majora is often an effective way of reducing the bulk. These procedures all result in good palliation, but recurrence is common. Some rectal venous malformations can be treated by sclerotherapy, either endoscopically or percutaneously³²; those that involve the rectum circumferentially are usually managed surgically.^{33,34}

Lymphatic Malformations

Lymphatic malformations (LMs) are the second most common type of VM after venous malformations.³⁵ They are usually located in the neck (70–80%) or in the axillary region

(20%) and are rarely found in the pelvis or extremities.^{4,10} Like venous malformations, they can be focal or part of a diffuse anomaly.

LMs can be divided into microcystic types (cysts smaller than 2 cm) and macrocystic types.^{4,36} LMs are also commonly associated with other VMs.³⁷ Most LMs are discovered at the age of infancy. Clinically, they manifest as soft-tissue masses with a rubbery consistency and, unlike venous malformations, are noncompressible. Diffuse soft-tissue thickening and surrounding lymphedema may occur locally with these lesions.⁴

On MRI, LMs are usually septated, lobulated masses that have intermediate to decreased signal intensity on T1-weighted images and increased signal intensity on T2-weighted images. LMs tend to be infiltrative masses that involve multiple tissues and permeate across fat planes.^{4,9} Internal fluid–fluid levels may be present. Macrocystic LMs generally exhibit only rim and septal enhancement, whereas there is usually no significant enhancement of microcystic LMs.^{4,38} Occasionally, microcystic LMs or combined veno-lymphatic malformations may demonstrate diffuse enhancement, which is due to septal enhancement of the small, imperceptible cysts in microcystic LMs or enhancement of the venous component in mixed malformations.^{11,14}

Endovascular Treatment of Pelvic Lymphatic Malformations

Macrocystic LMs can be treated by aspiration or drainage with sclerosant injection. Sclerosant agents that have shown effectiveness in treatment of LMs include ethanol, STS, doxycycline, OK-432 (picibanil), acetic acid, and bleomycin. Seventy-five to 90% of patients with a simple macrocystic LM have an excellent response to sclerotherapy, with a very low rate of recurrence, and a reduced morbidity compared with resection.^{31,39–41} Microcystic or combined mixed LMs tend to demonstrate a less predictable response to treatment, however. In patients with cutaneous vesicles, sclerotherapy or superficial laser treatment can be helpful in decreasing the seeping of chylous fluid, but the results are temporary and multiple treatments are often needed. Patients with diffuse tubular lymphatic anomalies fare the least well in general,

although some patients can get temporary relief of leaking of chylous fluid following treatment.⁴²

Fast-Flow Vascular Malformations

High-flow malformations include both arteriovenous malformations (AVMs) and arteriovenous fistulas (AVFs). AVMs consist of feeding arteries, draining veins, and a nidus composed of multiple dysplastic vascular channels that connect the arteries and veins, whereas AVFs are formed by a single vascular channel between an artery and a vein.^{9,11} AVMs and AVFs can affect the pelvic wall and/or viscera, with AVFs being less common.^{43,44} AVFs are most often acquired and are mostly the consequence of an iatrogenic or traumatic penetrating injury. AVMs are already present at birth, although they do not usually become evident until childhood or early adulthood.²⁵ Like other VMs, they generally increase proportionally in size as the child grows. AVM growth may be exacerbated due to hormonal changes during puberty or pregnancy⁹ or as a result of thrombosis, infection, or trauma (►Fig. 1).¹¹ These lesions may also evolve from an asymptomatic lesion with minimal shunting to one with active shunting, venous engorgement, and venous hypertension. Owing to their high blood flow, they generally manifest as a pulsatile, warm mass with a thrill, and they may lead to bone overgrowth, arterial steal phenomenon, or cutaneous ischemia (►Fig. 2).²⁵ Ulceration and hemorrhage may be seen in later stages. Symptoms of pelvic AVMs range from mild discomfort and a feeling of pressure to significant vaginal or rectal bleeding to cardiac volume overload and resultant congestive cardiac failure.

Although most AVMs are sporadic, numerous genetic mutations have been identified. These include ALK-1 and endoglin in hereditary hemorrhagic telangiectasia, RASA-1 in familial capillary malformation-AVM, and phosphatase and tensin (PTEN) in patients with Bannayan–Riley–Ruvalcaba syndrome or Cowden syndrome.^{45–50}

Many fast-flow VMs are initially evaluated by pelvic US.^{51–54} Doppler interrogation of AVMs typically shows high velocity, low resistance flow in the feeding arteries and draining veins (►Fig. 3). MRI findings include high-flow serpentine and enlarged feeding arteries and draining

Figure 1 (A) Grayscale ultrasound image demonstrating a sagittal view of the uterus with a hypoechoic cesarean section scar (arrow). (B) Color ultrasound image demonstrating increased vascularity in the region of the cesarean section scar (arrow).

Figure 2 (A) Grayscale ultrasound images demonstrate a hypoechoic lesion in the right adnexa (asterisk). (B) Color ultrasound image demonstrates a vessel (arrow) extending into the lesion with both arterial and venous color Doppler signal characteristics. (C) Coronal CT image demonstrates contrast filling a PSA in the right adnexa (asterisk) with early draining veins (arrow). (D) Axial CT image demonstrates contrast filling a PSA in the right adnexa (asterisk) with early draining veins (arrow). (E) Angiographic image with a base catheter in the proximal uterine artery and a microcatheter advanced into the PSA. Note early venous drainage (arrow). (F) Postembolization angiography via the base catheter in the uterine artery demonstrates multiple coils in the aneurysm sac and feeding uterine artery (arrow). Note no further venous drainage. CT, computed tomography; PSA, pseudoaneurysm.

veins, which appear as large flow voids on spin echo images or high-signal-intensity structures on gradient echo images, with absence of a well-defined mass.⁵ Areas of high signal intensity on T1-weighted images may be representative of hemorrhage, intravascular thrombosis, or flow-related enhancement.³⁸ The dynamic enhancement of AVMs is well assessed by using time-resolved dynamic 3D MR angiography, with typical early venous filling.⁵

Endovascular Treatment of Pelvic Fast-Flow Vascular Malformations

Intrauterine AVFs are generally acquired, and arise after pregnancy or uterine trauma.^{51–55} For the majority of asymp-

tomatic women diagnosed by ultrasonography, the AVF resolves spontaneously, and therefore, conservative management is recommended.⁵¹ Symptomatic women presenting with hemorrhage are best treated with embolization. Embolization of uterine AVFs is usually performed through standard arterial microcatheter techniques (►Fig. 2). This treatment is generally effective in eliminating the lesion, with a very low reported recurrence rate.^{52,54–58}

AVMs of the pelvic wall and viscera are often supplied by multiple arteries in the pelvis, including the anterior branches of the internal iliac arteries, the inferior mesenteric arteries, and the median sacral artery.^{43,44,59} Given this extensive arterial supply, surgical resection of AVM lesions

Figure 3 (A) Grayscale ultrasound image demonstrating two large hypoechoic regions (arrows) at the lateral aspect of the uterus. (B) Color ultrasound image demonstrating previous hypoechoic regions have color flow (arrows). (C) Doppler interrogation showing simultaneous arterial and venous waveforms in the uterus. (D) Axial CT image showing contrast in the vascular spaces identified on ultrasound (arrow). A dermoid is incidentally noted (asterisk). (E) Pre-embolization angiogram from the uterine artery (arrow) showing increased vascularity in the left lateral uterus. (F) Delayed phase from previous angiogram demonstrating early venous filling (arrow) and drainage from the uterus. CT, computed tomography.

is difficult and carries the risk of a massive intraoperative hemorrhage, incomplete removal of the AVM nidus, surrounding organ injury, and high recurrence rates. Therefore, endovascular therapy with various embolic and sclerosing materials, independently or combined with surgical treatment, has become an accepted therapeutic option for patients with AVMs. Many embolic materials have been used to treat AVM lesions, including coils, sclerosants (e.g., alcohol), rapidly polymerizing acrylic adhesives (e.g., *n*-butyl cyanoacrylate or isobutyl cyanoacrylate), polyvinyl alcohol foam particles, detachable silicon balloons, and ethylene-vinyl alcohol copolymers (Onyx) (Covidien, Plymouth, MN).^{18,22,23,31,51,60–63}

Many pelvic AVMs represent arteriovenous fistulae in which many feeding arteries drain into a single draining vein,

often a pudendal or obturator vein.⁶⁴ The access routes or complications related with endovascular treatment vary according to the location of the AVM lesion. Transarterial, transvenous, and direct percutaneous approaches have all been described. Combined approaches have been used as well. Combined embolosclectomy of pelvic AVMs with coils and ethanol has been shown to be efficacious at achieving complete remission.^{61,63} To prolong the contact time between the sclerosing agent and the endothelium of the target vascular lesion, the draining vein can be embolized via a transvenous approach before ethanol sclerotherapy via a transarterial or direct percutaneous approach. Additional methods to prolong the contact time between the sclerosing agent and the endothelium of the AVM include using a lower

extremity tourniquet, pneumatic cuff, or an intravascular occlusion balloon.^{63,65,66}

Combined Vascular Malformations

Klippel-Trenaunay syndrome and Parkes-Weber syndrome are combined vascular overgrowth malformations that typically affect a lower extremity and may affect the ipsilateral pelvis.¹² Klippel-Trenaunay syndrome is a congenital disorder classically characterized by findings of a port-wine stain (nevus flammeus), abnormal venous structures (such as varicosities and slow-flow venous malformations), and osseous and soft-tissue hypertrophy. This syndrome was initially described in 1900 by Klippel and Trenaunay.⁶⁷ In 1907, Weber⁶⁸ observed similar findings in association with high-flow AVMs; this combined entity is referred to as Parkes-Weber syndrome.

Individuals with Klippel-Trenaunay syndrome have lymphatic and venous anomalies of variable severity.^{69–75} The venous anomalies typically consist of persistent embryonic veins that course in the subcutaneous tissue of the lateral calf and thigh. These veins communicate with the deep venous circulation in a variety of ways, often coursing through the pelvis. The deep veins of the leg can be hypoplastic or interrupted and will often demonstrate abnormal function on noninvasive imaging.⁷⁶ Lymphatic involvement may be manifested as the presence of small vesicles in the nevus flammeus, or may be more severe, with microcystic and macrocystic components in the lower extremity and pelvis. Patients with venous anomalies may present with painful limb swelling, venous thromboembolism, stasis dermatitis, and cellulitis, as well as bleeding from the vagina or rectum.⁷⁷ Symptoms typically worsen during pregnancy, so those who do become pregnant must be vigilant in wearing graded elastic stockings and may require anticoagulation.

Parkes-Weber syndrome typically consists of an extensive cutaneous capillary malformation, diffuse small-vessel AVM, and limb and buttock overgrowth.^{18,45,73} These patients have similar presentations as those with Klippel-Trenaunay syndrome, and may also develop leg length discrepancy and cardiac overload. A small percentage of these patients have a lymphatic component that can result in chylous leakage and cellulitis.

Various imaging techniques can be used in the diagnosis of suspected combined VMs. Both plain radiography and computed tomography can detect phleboliths, suggesting an underlying venous malformation. Doppler sonography can be used to assess the venous system within an affected limb, and MRI can evaluate for both extremity hypertrophy and VMs in these patients. Specifically, MRI allows differentiation of low-flow (venous) from high-flow (arteriovenous) malformations. Occasionally, however, conventional angiography or venography is needed to define the vascular anomalies associated with these conditions.⁷⁸

Endovascular Treatment of Pelvic Combined Vascular Malformations

In patients with Klippel-Trenaunay syndrome, ablation of venous malformations in the lower extremities can be pur-

sued, using a combination of endovenous laser therapy, microcatheter embolization, and sclerotherapy. However, this may not result in clinical improvement, particularly if the deep veins are inadequate.¹⁷ LMs in patients involving the extremity or pelvis are often amenable to sclerotherapy, particularly the macrocystic types. Patients who undergo partial resection of LMs may develop worsening of the residual lesion, leading to severe bleeding. Sclerotherapy is often helpful in managing these recurrences.¹⁷ In patients with Parkes-Weber syndrome, the high-flow VMs are often treated in a similar fashion to AVMs elsewhere with a combination of embolization or occlusion of the outflow vein and sclerotherapy of the vascular nidus.⁷⁹ In general, a multidisciplinary approach to these complex patients is the optimum choice for management.

Summary

Pelvic VMs can be focal or diffuse, and characterization of the specific type of VM based on clinical and imaging findings is essential for guiding therapy. Endovascular treatment with or without resection can often relieve significant morbidity associated with these lesions, and it remains the mainstay for therapy.

References

- Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. *Plast Reconstr Surg* 1982;69(3):412–422
- Jackson IT, Carreño R, Potparic Z, Hussain K. Hemangiomas, vascular malformations, and lymphovenous malformations: classification and methods of treatment. *Plast Reconstr Surg* 1993;91(7):1216–1230
- Dubois J, Alison M. Vascular anomalies: what a radiologist needs to know. *Pediatr Radiol* 2010;40(6):895–905
- Moukaddam H, Pollak J, Haims AH. MRI characteristics and classification of peripheral vascular malformations and tumors. *Skeletal Radiol* 2009;38(6):535–547
- Flors L, Leiva-Salinas C, Maged IM, et al. MR imaging of soft-tissue vascular malformations: diagnosis, classification, and therapy follow-up. *Radiographics* 2011;31(5):1321–1340, discussion 1340–1341
- Dubois J, Soulez G, Oliva VL, Berthiaume MJ, Lapierre C, Therasse E. Soft-tissue venous malformations in adult patients: imaging and therapeutic issues. *Radiographics* 2001;21(6):1519–1531
- Vikkula M, Boon LM, Mulliken JB. Molecular genetics of vascular malformations. *Matrix Biol* 2001;20(5-6):327–335
- Boon LM, Mulliken JB, Enjolras O, Vikkula M. Glomuvenous malformation (glomangioma) and venous malformation: distinct clinicopathologic and genetic entities. *Arch Dermatol* 2004;140(8):971–976
- Donnelly LF, Adams DM, Bisset GS III. Vascular malformations and hemangiomas: a practical approach in a multidisciplinary clinic. *AJR Am J Roentgenol* 2000;174(3):597–608
- Dubois J, Garel L. Imaging and therapeutic approach of hemangiomas and vascular malformations in the pediatric age group. *Pediatr Radiol* 1999;29(12):879–893
- Fayad LM, Hazirolan T, Bluemke D, Mitchell S. Vascular malformations in the extremities: emphasis on MR imaging features that guide treatment options. *Skeletal Radiol* 2006;35(3):127–137
- Mulliken JB, Fishman SJ, Burrows PE. Vascular anomalies. *Curr Probl Surg* 2000;37(8):517–584

- 13 Breugem CC, Maas M, Reekers JA, van der Horst CM. Use of magnetic resonance imaging for the evaluation of vascular malformations of the lower extremity. *Plast Reconstr Surg* 2001;108(4):870-877
- 14 Laor T, Burrows PE. Congenital anomalies and vascular birthmarks of the lower extremities. *Magn Reson Imaging Clin N Am* 1998;6(3):497-519
- 15 Herman AR, Morello F, Strickland JL. Vulvar venous malformations in an 11-year-old girl: a case report. *J Pediatr Adolesc Gynecol* 2004;17(3):179-181
- 16 Marrocco-Trischitta MM, Nicodemi EM, Nater C, Stillo F. Management of congenital venous malformations of the vulva. *Obstet Gynecol* 2001;98(5 Pt 1):789-793
- 17 Burrows PE. Vascular malformations involving the female pelvis. *Semin Intervent Radiol* 2008;25(4):347-360
- 18 Enjolras O, Chapot R, Merland JJ. Vascular anomalies and the growth of limbs: a review. *J Pediatr Orthop B* 2004;13(6):349-357
- 19 Enjolras O, Ciabrin D, Mazoyer E, Laurian C, Herbreteau D. Extensive pure venous malformations in the upper or lower limb: a review of 27 cases. *J Am Acad Dermatol* 1997;36(2 Pt 1):219-225
- 20 Azizkhan RG. Life-threatening hematochezia from a rectosigmoid vascular malformation in Klippel-Trenaunay syndrome: long-term palliation using an argon laser. *J Pediatr Surg* 1991;26(9):1125-1127, discussion 1128
- 21 Azouz EM. Hematuria, rectal bleeding and pelvic phleboliths in children with the Klippel-Trenaunay syndrome. *Pediatr Radiol* 1983;13(2):82-88
- 22 Servelle M, Bastin R, Loygue J, et al. Hematuria and rectal bleeding in the child with Klippel and Trenaunay syndrome. *Ann Surg* 1976;183(4):418-428
- 23 Ashour MA, Soliman HE, Khougeer GA. Role of descending venography and endovenous embolization in treatment of females with lower extremity varicose veins, vulvar and posterior thigh varices. *Saudi Med J* 2007;28(2):206-212
- 24 Krokidis M, Venetucci P, Hatzidakis A, Iaccarino V. Sodium tetradecyl sulphate direct intralesional sclerotherapy of venous malformations of the vulva and vagina: report of five cases. *Cardiovasc Intervent Radiol* 2011;34(Suppl 2):S228-S231
- 25 Ernemann U, Kramer U, Miller S, et al. Current concepts in the classification, diagnosis and treatment of vascular anomalies. *Eur J Radiol* 2010;75(1):2-11
- 26 Dobson MJ, Hartley RW, Ashleigh R, Watson Y, Hawnaur JM. MR angiography and MR imaging of symptomatic vascular malformations. *Clin Radiol* 1997;52(8):595-602
- 27 Konec O, Burrows PE, Mulliken JB, Fishman SJ, Kozakewich HP. Angiographic features of rapidly involuting congenital hemangioma (RICH). *Pediatr Radiol* 2003;33(1):15-19
- 28 Goyal M, Causer PA, Armstrong D. Venous vascular malformations in pediatric patients: comparison of results of alcohol sclerotherapy with proposed MR imaging classification. *Radiology* 2002;223(3):639-644
- 29 Yun WS, Kim YW, Lee KB, et al. Predictors of response to percutaneous ethanol sclerotherapy (PES) in patients with venous malformations: analysis of patient self-assessment and imaging. *J Vasc Surg* 2009;50(3):581-589, e1
- 30 Fayad LM, Hazirolan T, Carrino JA, Bluemke DA, Mitchell S. Venous malformations: MR imaging features that predict skin burns after percutaneous alcohol embolization procedures. *Skeletal Radiol* 2008;37(10):895-901
- 31 Burrows PE, Mason KP. Percutaneous treatment of low flow vascular malformations. *J Vasc Interv Radiol* 2004;15(5):431-445
- 32 Keljo DJ, Yakes WF, Andersen JM, Timmons CF. Recognition and treatment of venous malformations of the rectum. *J Pediatr Gastroenterol Nutr* 1996;23(4):442-446
- 33 Fishman SJ, Shamberger RC, Fox VL, Burrows PE. Endorectal pull-through abates gastrointestinal hemorrhage from colorectal venous malformations. *J Pediatr Surg* 2000;35(6):982-984
- 34 de la Torre L, Carrasco D, Mora MA, Ramírez J, López S. Vascular malformations of the colon in children. *J Pediatr Surg* 2002;37(12):1754-1757
- 35 Marler JJ, Mulliken JB. Current management of hemangiomas and vascular malformations. *Clin Plast Surg* 2005;32(1):99-116, ix
- 36 Sanlialp I, Karnak I, Tanyel FC, Senocak ME, Büyükpamukçu N. Sclerotherapy for lymphangioma in children. *Int J Pediatr Otorhinolaryngol* 2003;67(7):795-800
- 37 Navarro OM, Laffan EE, Ngan BY. Pediatric soft-tissue tumors and pseudo-tumors: MR imaging features with pathologic correlation: part 1. Imaging approach, pseudotumors, vascular lesions, and adipocytic tumors. *Radiographics* 2009;29(3):887-906
- 38 Abernethy LJ. Classification and imaging of vascular malformations in children. *Eur Radiol* 2003;13(11):2483-2497
- 39 Alomari AI, Karian VE, Lord DJ, Padua HM, Burrows PE. Percutaneous sclerotherapy for lymphatic malformations: a retrospective analysis of patient-evaluated improvement. *J Vasc Interv Radiol* 2006;17(10):1639-1648
- 40 Molitch HI, Unger EC, Witte CL, vanSonnenberg E. Percutaneous sclerotherapy of lymphangiomas. *Radiology* 1995;194(2):343-347
- 41 Perkins JA, Manning SC, Tempero RM, et al. Lymphatic malformations: review of current treatment. *Otolaryngol Head Neck Surg* 2010;142(6):795-803, e1
- 42 Maeda Y, Arima K, Matsuura H, Hayashi N, Yanagawa M, Kawamura J. [Percutaneous sclerotherapy for chyluria with leg edema: a case report]. *Hinyokika Kiyo* 1998;44(1):25-27
- 43 Mitty HA, Baron MG, Jacobson JH II. Pelvic arteriovenous malformations. *Am J Roentgenol Radium Ther Nucl Med* 1968;102(2):427-430
- 44 Calligaro KD, Sedlacek TV, Savarese RP, Carneval P, DeLaurentis DA. Congenital pelvic arteriovenous malformations: long-term follow-up in two cases and a review of the literature. *J Vasc Surg* 1992;16(1):100-108
- 45 Revencu N, Boon LM, Mulliken JB, et al. Parkes Weber syndrome, vein of Galen aneurysmal malformation, and other fast-flow vascular anomalies are caused by RASA1 mutations. *Hum Mutat* 2008;29(7):959-965
- 46 Zhou XP, Marsh DJ, Hampel H, Mulliken JB, Gimm O, Eng C. Germline and germline mosaic PTEN mutations associated with a Proteus-like syndrome of hemihypertrophy, lower limb asymmetry, arteriovenous malformations and lipomatosis. *Hum Mol Genet* 2000;9(5):765-768
- 47 Takaya N, Iwase T, Maehara A, et al. Transcatheter embolization of arteriovenous malformations in Cowden disease. *Jpn Circ J* 1999;63(4):326-329
- 48 Guttmacher AE, Marchuk DA, White RI Jr. Hereditary hemorrhagic telangiectasia. *N Engl J Med* 1995;333(14):918-924
- 49 Bourdeau A, Cymerman U, Paquet ME, et al. Endoglin expression is reduced in normal vessels but still detectable in arteriovenous malformations of patients with hereditary hemorrhagic telangiectasia type 1. *Am J Pathol* 2000;156(3):911-923
- 50 Eerola I, Boon LM, Mulliken JB, et al. Capillary malformation-arteriovenous malformation, a new clinical and genetic disorder caused by RASA1 mutations. *Am J Hum Genet* 2003;73(6):1240-1249
- 51 Timmerman D, Van den Bosch T, Peeraer K, et al. Vascular malformations in the uterus: ultrasonographic diagnosis and conservative management. *Eur J Obstet Gynecol Reprod Biol* 2000;92(1):171-178
- 52 Yang JJ, Xiang Y, Wan XR, Yang XY. Diagnosis and management of uterine arteriovenous fistulas with massive vaginal bleeding. *Int J Gynaecol Obstet* 2005;89(2):114-119
- 53 Jain KA, Gerscovich EO. Sonographic spectrum of pelvic vascular malformations in women. *J Clin Ultrasound* 1999;27(9):523-530
- 54 Kwon JH, Kim GS. Obstetric iatrogenic arterial injuries of the uterus: diagnosis with US and treatment with transcatheter arterial embolization. *Radiographics* 2002;22(1):35-46

- 55 Maleux G, Timmerman D, Heye S, Wilms G. Acquired uterine vascular malformations: radiological and clinical outcome after transcatheter embolotherapy. *Eur Radiol* 2006;16(2):299–306
- 56 Kelly FW. Forceps delivery after molar malignancy in a woman with arteriovenous malformation. A case report. *J Reprod Med* 2001;46(11):1013–1016
- 57 Ghai S, Rajan DK, Asch MR, Muradali D, Simons ME, TerBrugge KG. Efficacy of embolization in traumatic uterine vascular malformations. *J Vasc Interv Radiol* 2003;14(11):1401–1408
- 58 Lim AK, Agarwal R, Seckl MJ, Newlands ES, Barrett NK, Mitchell AW. Embolization of bleeding residual uterine vascular malformations in patients with treated gestational trophoblastic tumors. *Radiology* 2002;222(3):640–644
- 59 Jacobowitz GR, Rosen RJ, Rockman CB, et al. Transcatheter embolization of complex pelvic vascular malformations: results and long-term follow-up. *J Vasc Surg* 2001;33(1):51–55
- 60 Bae S, Do YS, Shin SW, et al. Ethanol embolotherapy of pelvic arteriovenous malformations: an initial experience. *Korean J Radiol* 2008;9(2):148–154
- 61 Mallios A, Laurian C, Houbballah R, Gigou F, Marteau V. Curative treatment of pelvic arteriovenous malformation—an alternative strategy: transvenous intra-operative embolisation. *Eur J Vasc Endovasc Surg* 2011;41(4):548–553
- 62 Messina ML, Carnevale FC, Barakat EC. Percutaneous embolization of large vulvar vascular malformation. *Int J Gynaecol Obstet* 2008;100(1):85–86
- 63 Do YS, Kim YW, Park KB, et al. Endovascular treatment combined with embolosclectomy for pelvic arteriovenous malformations. *J Vasc Surg* 2012;55(2):465–471
- 64 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(4):527–538
- 65 Yakes WF, Rossi P, Odink H. How I do it. Arteriovenous malformation management. *Cardiovasc Intervent Radiol* 1996;19(2):65–71
- 66 Jackson JE, Mansfield AO, Allison DJ. Treatment of high-flow vascular malformations by venous embolization aided by flow occlusion techniques. *Cardiovasc Intervent Radiol* 1996;19(5):323–328
- 67 Klippel M, Trénaunay P. Du naevus variqueux osteohypertrophique. *Arch Genet Med Paris* 1900;3:611–672
- 68 Weber FP. Angioma formation in connection with hypertrophy of limbs and hemihypertrophy. *Br J Dermatol* 1907;19:231–235
- 69 Berry SA, Peterson C, Mize W, et al. Klippel-Trenaunay syndrome. *Am J Med Genet* 1998;79(4):319–326
- 70 Kanterman RY, Witt PD, Hsieh PS, Picus D. Klippel-Trenaunay syndrome: imaging findings and percutaneous intervention. *AJR Am J Roentgenol* 1996;167(4):989–995
- 71 Baskerville PA, Ackroyd JS, Lea Thomas M, Browse NL. The Klippel-Trenaunay syndrome: clinical, radiological and haemodynamic features and management. *Br J Surg* 1985;72(3):232–236
- 72 Oduber CE, van der Horst CM, Hennekam RC. Klippel-Trenaunay syndrome: diagnostic criteria and hypothesis on etiology. *Ann Plast Surg* 2008;60(2):217–223
- 73 Ziyeh S, Spreer J, Rössler J, et al. Parkes Weber or Klippel-Trenaunay syndrome? Non-invasive diagnosis with MR projection angiography. *Eur Radiol* 2004;14(11):2025–2029
- 74 Jacob AG, Driscoll DJ, Shaughnessy WJ, Stanson AW, Clay RP, Gloviczki P. Klippel-Trénaunay syndrome: spectrum and management. *Mayo Clin Proc* 1998;73(1):28–36
- 75 Cherry KJ, Gloviczki P, Stanson AW. Persistent sciatic vein: diagnosis and treatment of a rare condition. *J Vasc Surg* 1996;23(3):490–497
- 76 Delis KT, Gloviczki P, Wennberg PW, Rooke TW, Driscoll DJ. Hemodynamic impairment, venous segmental disease, and clinical severity scoring in limbs with Klippel-Trenaunay syndrome. *J Vasc Surg* 2007;45(3):561–567
- 77 Walder B, Kapelanski DP, Auger WR, Fedullo PF. Successful pulmonary thromboendarterectomy in a patient with Klippel-Trenaunay syndrome. *Chest* 2000;117(5):1520–1522
- 78 Elsayes KM, Menias CO, Dillman JR, Platt JF, Willatt JM, Heiken JP. Vascular malformation and hemangiomatosis syndromes: spectrum of imaging manifestations. *AJR Am J Roentgenol* 2008;190(5):1291–1299
- 79 Hyodoh H, Hori M, Akiba H, Tamakawa M, Hyodoh K, Hareyama M. Peripheral vascular malformations: imaging, treatment approaches, and therapeutic issues. *Radiographics* 2005;25(1, Suppl 1):S159–S171