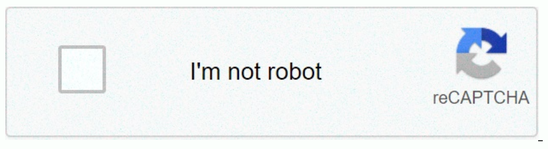

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Arteriovenous malformation hand ultrasound

Due to the fact that vascular malformations are rare and because of the precision required to treat them, our interventional radiologists within the Vascular Anomalies Center are working to offer patients individualized treatment plans. Ask an appointment phone410-614-5276 Vascular malformation is a general term that includes congenital vascular abnormalities of only veins, only lymph vessels, both veins and lymph vessels, or both arteries and veins. Why do these vascular deformities occur? These are all present at birth and become apparent at different ages. We're just beginning to understand how deformities occur. The pulmonary arteriovenous malformation, when associated with hereditary hemorrhagic Telangiectasia (HHT), is genetically inherited. A lot of work is currently being done on the possible genetics of other deformities. Most are known only as something that occurs during the development of arteries, veins, and/or lymph vessels, but without specific cause. What are the symptoms of a vascular deformity? These vascular deformities can cause a variety of symptoms, depending on the location in the body: Venous malformation can cause pain wherever they are located. Venous and lymph deformities can cause a lump under the skin. There may be an upper birthmark on the skin. Bleeding or lymph fluid leaking from skin lesions. Lymphomas tend to become infected, requiring repeated antibiotic treatments. Venous and lymph deformities can be associated with a syndrome called Klippel-Trenaunay Syndrome.Arteriovenous malformations can cause pain. They are also more stressful on the heart due to the rapid shunting of blood from arteries to veins. Depending on their location, they can also bleed (e.g. from the intestines, from the uterus or from the bladder). Hemangioma is another common term used for vascular abnormalities. However, this name actually applies to a childhood vascular anomaly that has a rapid growth phase between birth and 3 months of age. These will disappear completely by the age of 7. The main reason for us to treat these is for low platelets that do not respond to medical treatment, or in the liver due to mass shunting with a pressure on the heart. Lung arteriovenous malformations (PAVMs) are somewhat different during that time they shunt blood from the right heart system to the left heart system without picking up oxygen in the lungs. This results in symptoms of low oxygen, shortness of breath, fatigue. These deformities can also bleed, resulting in coughing up blood or blood in the chest. Also, this pulmonary artery to pulmonary vein shunts can allow clots through the lungs and travel to the arteries in the body, at risk of stroke or Abscess. This is a very important reason to block these shunts in anyone who has a non-hereditary lung arteriovenous deformity (PAVM) or anyone who has inherited pavms with HHT syndrome [HHT has another name: OWR - - Weber Rendu]. Watch: Embolization of Longarteriovenous Deformities ©2015 Caitlin Mock These are often seen on physical examination. Deeper vascular deformities can be diagnosed on MRI (magnetic resonance imaging). Although surgery is sometimes useful, it is usually difficult for surgeons to completely remove vascular deformities, which return if they are not completely removed. A non-surgical method of shutting down the blood or lymphatic flow in the deformity is done by interventional radiologists, who treat patients with image-guided procedures. Vascular deformities are treated by embolization. The AVMs and hemangiomas can be closed by promoting a small plastic tube, no larger than a pencil point, in the feeding artery to the deformity. This can be done without incisions or stitches, and with only mild sedation. Medical glue or alcohol or small beads are then floated into the deformity until it is full and no longer has blood flowing through it. For Long AVMs platinum coils are used to block the flow through the feeding artery to the deformity. The VMs and LMs are closed by injecting alcohol into the sachens filled with venous blood or lymph until these sacs collapse and no longer fill. The arteriovenous malformations can be treated with a one-night stay in the hospital. There is usually minimal discomfort for one to three days. The venous and lymph deformities also require a night in the hospital. These deformities swell after treatment with alcohol, and the swelling and pain can last for three to five days. During this time we give patients medication for any pain or swelling they may have. The complete shrinkage of these deformities can last four to six weeks. Pulmonary arteriovenous malformations are treated very effectively by embolization that blocks only the abnormal artery feeder, and preserves the normal pulmonary arteries. Patients usually notice an immediate improvement in symptoms with the immediate increase in oxygen levels. Other arteriovenous malformations are harder to treat because they tend to pull into new artery feeders from time to time. However, embolization is very effective in blocking abnormal artery feeders while maintaining normal arteries. AVMs may need a range of treatments to block all abnormal feeders. Venous and lymph deformities respond well to alcohol metabolism. These can also take a series of treatments about six weeks apart to block all abnormal blood vessels. All vascular deformities require prolonged surveillance so that if there is a change, such as a growth spurt with puberty or pregnancy or menopause, they can be monitored for symptoms that may warrant re-checking and possibly retreating. We check PAVMs over time to make sure they collapse and do not refill. Embolization techniques have been widely used around the world for 30 years. They have been well established for many years and have proved invaluable in the treatment of vascular vascular However, since congenital vascular malformations are relatively rare, it is likely advisable to obtain treatment in an important center that sees many patients. We can treat any age, from newborn to adult. The best age of treatment depends on the specific vascular deformity and its symptoms, and is best individualized to each person. Vascular malformatiyons and hemangiomas can lead to significant morbidity and even mortality in both children and adults. For a number of reasons, doctors often confuse these lesions. The nomenclature for classifying these lesions is often used interchangeably and inappropriately. Clinically significant deformities are uncommon, and patients with these deformities are rarely found in primary medical facilities, leaving most doctors inexperienced in providing optimal care. Radiologists may become involved in the care of these patients when imaging or imaging-guided therapy is requested; therefore, knowledge of the imaging and treatment of these patients is essential. This article discusses the clinical and imaging approaches of vascular malformations and hemangiomas used in our institution's multidisciplinary clinic, with an emphasis on a multidisciplinary approach, a practical categorization schedule, characteristic imaging findings and common clinical scenarios. Caring for children and adults with hemangiomas and vascular malformations requires the expertise of multiple subspecialties. The clinic for the treatment of vascular malformations at our institution has representatives of pediatric hematology-oncology, pediatric radiology, pediatric surgery, pediatric dermatology, pediatric otolaryngology, and orthopedic surgery. The combined skills and knowledge of these subspecialists help to provide the wide range of services these children may need. These services include medical therapy, tailored imaging studies, imaging-led intervention procedures, surgical resection, laser therapy, and monitoring for short- and long-term complications. Because expertise in multiple areas is needed to effectively treat these patients, patients with hemangiomas and vascular malformations are often best served in a tertiary center with a multidisciplinary clinic. Treatment is complicated by the relative rarity of these lesions [1], resulting in inexperience with diagnosis and treatment. Many of the patients we see in our clinic have previously seen several doctors and are frustrated with the inexperience and lack of answers they have encountered [1]. Often the families of the patients have obtained information from special websites and are more educated than the doctors from whom they have medical Search. The patients in our clinic have often undergone multiple surgical procedures, usually resulting in little improvement or even a worsening of symptoms. Doctors have recommended radical surgery for many patients. Conversely, many children who may benefit from other therapeutic therapeutic have been treated with watchful waiting. Many patients with vascular malformations have been misinformed that their lesions are hemangiomas and will eventually disappear. In addition, deformities related to areas such as the face can cause difficult social or emotional problems for patients and their families. Because of these setbacks, one of the most important team members in our clinic is our patient advocate. The lawyer helps families by providing educational material (such as a hemangioma-vascular deformity newsletter), addressing psychosocial problems, and putting families in touch with others who have been through similar experiences. Both hemangiomas and vascular deformities are endothelial deformities. The classification of and nomenclature used to describe endothelial deformities has been a source of confusion. Historically, lesions have been mentioned based on the size of the ducts in the lesions and the type of liquid that the lesions contain. Blood-containing lesions were called hemangiomas and were separated into capillary hemangiomas, strawberry hemmangiomas and hollow hemangiomas based on channel size. Lymphatic lesions were referred to as lymphangiomas or cystic hygromes. This classification system has been replaced by a system described by Mulliken and Glowacki in 1982 [2]. This newer classification system is an important tool in determining and separating the diagnoses of these two lesions. This system separates endothelial deformities into two large groups, hemangiomas and vascular malformations, based on their natural history, cellular turnover, and histology [2] (Table 1). Infantile hemangiomas indicate endothelial proliferation and go through a two-stage process of growth and regression. Hemangiomas are usually small or absent at birth and are often not initially noticed by parents and caregivers. Shortly after birth, they undergo a proliferative phase, with rapid growth that can last several months. They then undergo a stationary period, followed by a period of involution. View larger version TABLE 1 Distinctive features of Hemangiomas and Vascular Malformations Conversely, vascular malformations are always present at birth and increase in proportion to the child's growth. They are not involute and remain present throughout the patient's life [2]. Vascular malformations are classified as lymph, capillary, venous, arteriovenous and mixed deformities based on their histological makeup [2,3,4,5]. Although MR imaging has been used to classify vascular malformations in one of these categories [4, 6,7,8,9,10], a more relevant problem classifies vascular malformations as low-flow or high-flow lesions [3, 5]. Deformities with arterial components are considered high-flow lesions and arterial components are considered low-flow lesions. The two non-invasive imaging techniques most useful in the study of vascular deformities deformities MR imaging and sonography. In our clinic, MR imaging is the primary imaging technique in the evaluation of suspected vascular malformations. The primary objectives of imaging vascular malformations or hemangiomas include characterizing the lesion and discovering the anatomical extent of the disease [3]. Knowing which tissues include vascular deformity and whether adjacent vital structures, such as neurovascular bundles, are involved in the lesion is important (Fig. 1A,1B,1C). Such information is vital for planning surgery or imaging-guided procedures. In physical examination, it is difficult to determine whether the subcutaneous tissue, the underlying deep muscle tissues or both are involved (fig. 2A,2B). Check out the larger version (138K) Fig. 1A. — 4-month-old female infant with extensive spread of infantile hemangioma revealed on MR imaging. The photo shows hemangioma from right perirectal area, which was extent of illness presented on physical inspection. Due to foot drop during physical examination, MR imaging of lumbar spine was performed. Check out the larger version (141K) Fig. 1B. — 4-month-old female infant with extensive distribution of infantile hemangioma revealed on MR imaging. Coronal (B) and axial (C) T2-weighted fat-saturated rapid spin-echo MR images (3000/98 [TR/TE]) show abnormally elevated signal intensity (long arrow, B) in subcutaneous area of right buttock. Extended hemangioma during retroperitoneum of pelvis and abdomen is considered abnormally high signal intensity (short arrows, B and C). Mass appeared to engulf sacrum, rectum, uterus and vagina. Note prominent veins that appear as signal pupils. Check out the larger version (154K) Fig. 1C. — 4 month old female infant with extensive distribution of infantile hemangioma revealed on MR imaging. Coronal (B) and axial (C) T2-weighted fat-saturated rapid spin-echo MR images (3000/98 [TR/TE]) show abnormally elevated signal intensity (long arrow, B) in subcutaneous area of right buttock. Extended hemangioma during retroperitoneum of pelvis and abdomen is considered abnormally high signal intensity (short arrows, B and C). Mass appeared to engulf sacrum, rectum, uterus and vagina. Note prominent veins that appear as signal pupils. View larger version (161K) Fig. 2A. — Venous malformation involving the posterior abdominal wall in 3-year-old boy with pain and gradually enlarge lesion. Photo shows involvement of the skin with red discoloration and expansion of underlying soft tissues. Check out the larger version (154K) Fig. 2B. — Venous malformation involving the posterior abdominal wall in 3-year-old boy with pain and gradually enlarge lesion. Axial T2-weighted fat-saturated rapid spin echo MR image (3500/72 [TR/TE]) shows a mass with a high which mainly involves skin and subcutaneous tissue. Note involvement of the underlying abdominal wall musculature (large arrow) and prominent bleeding veins (small arrows). In characterizing suspected vascular malformations, important important the presence or absence of a discrete mass, excessive skin thickening and the presence of prominent vessels [3] (fig. 2A,2B). When physical examination and clinical history are diagnostic or highly suggestive of a vascular deformity, the main characteristic is whether the lesion is a high- or low-flow vascular deformity. Most of the information needed to investigate the lesion is available from a combination of T1-weighted, fat-saturated T2-weighted, and gradient-echo (flow-weighted) MR images. Our protocols include each of these sequences obtained in the axial plane, with the addition of coronal and sagittal rapid spin-echo T2-weighted images as needed. We find the axial plane most useful in depicting the relationship between lesion, neurovascular structures and tissue surfaces. Gadolinium-enhanced images have been advocated as useful in distinguishing between low-flow vascular malformations such as lymphangiomas and venous malformations [6, 11]. We have not found this distinction useful in our decision-making and do not use gadolinium in the evaluation of vascular malformations. Other potential imaging sequences that have been advocated as useful in evaluating vascular malformations include MR angiography, venography, and lymph nodes [3,4,5,6,7,8,9,10]. Sonography has been advocated as useful in examining soft tissue masses that indicate hemangiomas or vascular malformations [4, 12, 13]. The density of the vessel as depicted on Doppler sonography has been used in distinguishing other types of masses of vascular malformations [14]. Certainly, the Doppler characteristics of vascular deformities are useful in distinguishing low- from high-flow vascular deformities. We also found the sonographic representation of abundant low-flow vascular channels as a predictor of the potential success of percutaneous sclerosis [15], and we use sonography to guide needle placement during percutaneous sclerosis. Hemangiomas are benign neoplasms of endothelial cells. As previously discussed, they undergo a characteristic process in two stages of growth and regression [2]. At birth, the lesions are often small and unobtrusive, with 60% absent at birth [5, 16]. The lesions may appear as an erythematous macule, a blanching, or an area of localized telangiectasia [5, 16]. Shortly after birth, the phase of rapid proliferation occurs, which can last several months. This proliferation phase corresponds to a rapid period of growth of endothelial cells that form syncytial masses with and without vascular lumen. This phase is also defined by a high expression of angiogenic factors such as vascular endothelial growth factor and fundamental fibroblast growth factor. The typical hemangioma will begin to involuted about 10 months after birth and 50% of the lesions are complete in 5 years [5]. Hemangiomas are the most common childhood tumor, occurring in 12% of infants [4, 5, 17, 18]. 18]. more commonly found in girls, whites, premature babies and twins [4, 5, 19,20,21]. During the proliferative phase, hemangiomas are high-flow lesions [4] that are often revealed by bruit, pulsatitility, and increased heat. Hemangiomas can have deep, superficial or mixed components. The clinical appearance of hemangiomas varies with the degree of skin involvement and depth of lesions [5, 18]. A characteristic strawberry appearance is present when the lesions relate to the skin (Fig. 3). Deep hemangiomas, which are not accompanied by the subcutaneous tissues, may have a blue appearance [3] (fig. 4A,4B,4C). Check out the larger version (135K) Fig. 3. —Kaposiform hemangioendothelioma involving lip and left face in 8 month old female baby who had been treated with steroids for Kasabach-Merritt syndrome. Photo shows superficial involvement that makes the skin look red. Note deep component deforming region inferiro to left ear. Region inferior to left lip developed crevices. Check out the larger version (125K) Fig. 4A. — Infantile hemangioma in 21-day-old male neonate. Photo shows lobulated mass extending from area of knee. Lack of superficial involvement makes lesion bluish instead of strawberry red. Check out the larger version (143K) Fig. 4B. — Infantile hemangioma in 21-day-old male neonate. Axial T1-weighted MR image (500/14 [TR/TE]) shows mass (arrows) with signal intensity similar to that of skeletal muscles. Note low signal intensity and prominent veins. Check out the larger version (156K) Fig. 4C. — Infantile hemangioma in 21 days old male neonate. Axial T2-weighted fat-saturated rapid spin-echo MR image (4000/98) shows heterogeneous high signal-intensity mass (arrows) confined to subcutaneous tissue. Because lesion did not have a classical temporal growth pattern during physical examination, biopsy was performed to confirm the diagnosis of hemangioma. Most hemangiomas do not require therapy. Even many large lesions are treated conservatively because of the characteristic pattern of involution. Although hemangiomas are usually benign, a percentage of them develop life-threatening complications. Possible complications include Kasabach-Merritt syndrome (consumer clotting), compression of vital structures (for example, airways, orbital structures), tear formation, ulceration, and bleeding [3]. These complications usually occur in the rapid multiplication phase and can be associated with a mortality rate of up to 20-30% [5]. Kasabach-Merritt syndrome consists of thrombocytopenia, anemia, and consumer coagulopathy associated with a proliferative hemangioma. The syndrome has recently been shown to be associated with two specific subtypes of lesions, kaposiform hemangioendotheliomas and tuft angiomes [22, 23] (Fig. 3). These lesions are not typical infantile hemangiomas and poor on standard therapy [22, 23]. Their time to resolution is also much longer than that of typical infantile hemangiomas. Numerous therapies have been used in an attempt to complications develop during the proliferative phase. These regimens include high-dose steroids, α interferon, and chemotherapeutic drugs. The long-term use of systemic agents during the period of endothelial proliferation is often associated with increased side effects. The current first line of treatment is systemic administration of corticosteroids [20, 24]. Approximately 30% of hemangiomas will respond dramatically to corticosteroids and another 40% will have some response [20, 24]. Unfortunately, the doses of corticosteroids needed to treat hemangiomas are often associated with multiple side effects. These include severe irritability, weight gain, cushingoid appearance, growth retardation, hypertension, diabetes, gastroesophageal reflux, and susceptibility to infections. Patients should be closely monitored for these complications. When corticosteroid therapy does not improve symptoms, other antiloensy drugs, such as α interferon, may be used [24, 25]; however, α-interferon therapy has been associated with irreversible neurological spastic diplegia [26] and is now much less commonly used to treat hemangiomas. Chemotherapy with vincristine sulfate, surgical excision and embolization can be used in refractory cases [4, 24, 27]. Laser therapy can be used to treat complications associated with the superficial parts of the lesions such as ulceration, bleeding, and marked discoloration of the skin [18, 28, 29]. In most cases, the diagnosis of hemangioma can be made on the basis of the temporal growth history and the appearance of physical inspection; therefore, imaging is usually not required. In atypical cases, imaging can be performed to characterize the lesion and to investigate the anatomical extent of the disease. MR imaging of proliferating hemangiomas often shows a discrete lobulated mass that is hyperintense for muscles on T2-weighted images and isointense to muscle on T1-weighted images [6]. Typically, prominent draining veins will be identified as both central and peripheral high current vessels [6] (fig. 4A,4B,4C). Hemangiomas usually improve diffusely with gadolinium [6]. Involution hemangiomas may indicate areas of fibrous tissue with associated high signal intensity on T1-weighted images and less contrast improvement than those of proliferating hemangiomas [6]. The density of blood vessels as seen on Doppler sonography is also advocated as useful in making the diagnosis of hemangioma during the proliferative phase [14]. Unfortunately, many of the malignancy of the soft tissue of infancy, such as fibrosarcocoma or rhabdomyosarcoma, may have an imaging appearance similar to that of usury atoms [30, 31]. Therefore, cases that do not show the typical appearance and growth patterns for hemangioma are often biopsy to exclude it. Low-flow vascular deformities mainly include venous, lymphatic, and mixed malformations. Venous malformations are dysplasia of small and large venous channels associated with a variable amount of hamartomatous hamartomatous [3]. The venous channels connect to adjacent veins. Many venous malformations cause pain. Often patients will suffer from increasing symptoms in late childhood or early adulthood. Other clinical problems associated with venous malformations include reduced range of motion and deformity. Venous malformations rarely fall back [32, 33]. Treatment for venous malformations includes elastic compression clothing, percutaneous sclerosis and surgical excision [32, 33]. Lymphoma formations consist of chylefilled cysts coated with endothelial [34]. The most common locations for lymphoma formations are the neck (about 75%) and armpit (approximately 25%), with less common locations including the mediastinum, retroperitoneum, pelvis and groin [3, 35, 36]. When lymphphiles occur in the neck and armpit, they are often called cystic hygromes. Most lymphoma formations occur early in childhood, with 65% present at birth and 90% seen by age 2 years [3, 34,35,36,37]. The primary therapy for lymphoma formations that occur in infancy is surgical excision [35, 38,39,40,41,42]. Another therapeutic option is percutaneous sclerotherapy using means containing absolute ethanol, bleomycin, cyclofosphamide, doxycycline, alcohol solution of zein, and OK-432 [37, 43,44,45,46,47,48,49]. Chemotherapy, such as cyclophosffamide, has also been used for life-threatening lesions. A high percentage of vascular malformations, referred to as mixed vascular malformations, have different tissue characteristics in different parts of the lesion. Recognizing that the lesion is a low-flow vascular deformity is more important than determining whether the lesion is predominantly venous or lymph when making treatment decisions. The appearance of a low-flow vascular deformity on MR imaging depends on the composition of lymph and venous components. The venous parts of a deformity will appear as a collection of serpentine structures separated by septations. These serpentine structures represent slow-flowing blood in the venous channels and appear as high signal intensity on T2-weighted images and intermediate signal intensity on T1-weighted images [9] (Fig. 5A,5B). Flanges may be present and appear as round, low signal-intensity lesions on MR imaging [4, 6, 9, 50]. Gadolinium-enhanced T1-weighted images can show improvement in slow-flowing venous channels [6]. Lymph components of the deformity may contain cystic structures of different sizes, ranging from macrocystic to microcystic [5] (fig. 6A,6B). These cystic structures usually appear as high signal intensity on T2-weighted MR images and show no central improvement with gadolinium [5]. Liquid-liquid levels are often present [51]. Characteristic imaging findings of vascular malformations include tendency to infiltrate, disrespect for facial surfaces, and involvement of multiple tissue types such as muscle and subcutaneous fat [3] (Fig. 1A,1B,1C). 1A.1B,1C). larger version (173K) Fig. 5A. — Venous deformity of left anterior pelvis in 10-year-old girl. Axial T1-weighted MR image (500/8 [TR/TE]) shows mass (arrows) limited to subcutaneous tissues. Mass is isointense in signal intensity to adjacent muscle. Note prominent draining veins. View larger version (180K) Fig. 5B. — Venous deformity of left anterior pelvis in 10-year-old girl. Axial T2-weighted fat-saturated rapid spin-echo MR image (4000/98) shows mass (arrows) consisting of multiple high signal-intensity serpentine structures. Mass is limited to subcutaneous tissue. Note prominent draining veins. View larger version (102K) Fig. 6A. — Lymphthing involving arm and chest wall of 4-month-old female baby. Photo shows extension and multilobulated contour of left upper extremity. View larger version (170K) Fig. 6B. — Lymphemisformation involving the arm and chest wall of 4-month-old female baby. Coronal T2-weighted fat-saturated rapid spin-echo MR image (4316/98 [TR/TE]) shows multilocular cystic-appearing mass (m) involving subcutaneous tissues of left upper extremity. Note chest wall involvement (arrow). Low-flow vascular deformities can be difficult to treat. Surgical resection, medical therapy and transarterial embolization have all had limited success [1, 15, 27, 32, 52, 53]. Multiple studies have advocated the use of percutaneous sclerosis of low-flow vascular malformations and it is currently treating choice [1, 15, 27, 32, 52, 53]. Ethanol is the most commonly used sclerosing agent. Other means include sodiumtetradecylsulphate, ethibloc, bleomycin, cyclofosfdiade, doxycycline, alcohol solution of zein and OK-432 [37, 43,44,45,46,47,48,49]. Percutaneous sclerotherapy is usually performed under general anesthesia. Direct puncture of the vascular channels is performed using a combination of sonographic and fluoroscopic guidance [15]. Swelling of soft tissue generally increases in the area of deformity immediately after the procedure (fig. 7A,7B). If the necrosis and inflammation caused by the sclerosis begin to pull with fibrous scarring, the lesion will decrease in size. The full clinical effect of sclerosis may not be visible for several months (fig. 8A,8B,8C). Patients and their families should be informed of this expected course so that they do not have false expectations. In our experience with percutaneous sclerosis for low-flow vascular malformations, about 20% of patients will have complete relief from symptoms after a sclerotherapy procedure. An equal amount will have no therapeutic benefit. Approximately 60% of patients will have a decrease in their clinical symptoms that are significant enough to improve their quality of life; therefore, approximately 80% of patients will percutaneous sclerosis. View larger version (105K) Fig. 7A. — Change in appearance after percutaneous ethanol sclerosis of venous malformation in 7-year-old girl with pain. Photo for for shows bluish discoloration of the skin with underlying fullness. Check out the larger version (127K) Fig. 7B. — Change in appearance after percutaneous ethanol sclerosis of venous malformation in 7-year-old girl with pain. Photo 4 days after sclerosis with only 7 ml of ethanol shows a marked increase in swelling, hematoma and area of skin ulcers. Findings all resolved in the coming weeks; patient's pain resolved and fullness decreased. Check out the larger version (136K) Fig. 8A. — Percutaneous sclerosis of venous deformity of an 18-year-old dancer's foot with pain. Sagittal T2-weighted fat-saturated rapid spin-echo MR image (4000/98 [TR/TE]) obtained before procedure shows serpentine areas of high signal intensity (arrows). View larger version (139K) Fig. 8B. — Percutaneous sclerosis of venous deformity of an 18-year-old dancer's foot with pain. Image of percutaneous venogram obtained during sclerosis shows tangle of venous structures and draining veins. Watch for angiocatheter (arrow). Check out the larger version (166K) Fig. 8C. — Percutaneous sclerosis of venous deformity of an 18-year-old dancer's foot with pain. Sagittal T2-weighted fat-saturated rapid spin-echo MR image (4000/98) obtained 7 months later than A and B shows resolution of serpentine high signal-intensity structures and replacement by low signal-intensity structures (arrows), likely fibrotic scars. The procedure is not without potential risks. Complications have been reported in as many as 10-15% of cases and include skin osteoarthritis, nerve damage (sensory or motor), pain and swelling, muscle atrophy or contracture, deep vein thrombosis, pulmonary metabolism, spread intravascular clotting, and cardiopulmonary collapse [1, 15]. In our experience, 31% of patients will develop temporary side effects that will prolong recovery. It is important that the patient understands this before the procedure. Because of these potential risks, we monitor the patients in the hospital for 24 hours after the procedure. Any lesion that has arterial components is considered a high-flow deformity. These include arteriovenous malformations (AVM) and arteriovenous fistulas. During the proliferating phase, infantile hemangiomas can also be considered high-flow lesions. AVMs form a direct link between arterial and venous systems [1, 3, 4]. The lesions can occur in childhood or adulthood and are often aggravated during puberty or pregnancy [4, 54]. Presenting symptoms include congestive heart failure, embolism, pain, bleeding, and ulceration [1]. During physical examination, the lesions may appear blue and can feel warm with pulsating and sensation due to the increased blood flow. Lesions tend to grow with the child, but can increase rapidly due to thrombosis, infection, or hormonal stimulation [4, 54]. High-flow vascular deformities are rare.MR and Doppler sonography can be used for both diagnosis and follow-up of AVMs after therapy [1]. On MR MR the lesions appear as a tangle of multiple flow cavities [1] indicating a high current on gradient echo images [1]. Although lesions may be associated with surrounding edema or fibrofatty stroma, usually no focal discrete soft tissue mass is found (Fig. 9A,9B,9C) [3]. Color Doppler sonography gives a direct connection between the arterial and venous systems and resistance indexes indicate a low resistance flow [1] (Fig. 10A,10B,10C,10D). The most effective treatment for AVMs is transarterial embolization [1] (fig. 10A,10B,10C,10D). Check out the larger version (194K) Fig. 9A. — High-flow vascular deformity of the foot in 12-year-old boy. Sagitttal T1-weighted MR image (750/12 [TR/TE]) shows multiple tubular flow cavities (arrows). Note absence of discrete mass. Check out the larger version (147K) Fig. 9B. — High-flow vascular deformity of the foot in 12-year-old boy. Short-axis T2-weighted fat-saturated rapid spin-echo MR image (3200/76) shows multiple tubular flow voids (arrow) with surrounding edema. Check out the larger version (122K) Fig. 9C. — High-flow vascular deformity of the foot in 12-year-old boy. Arteriogram shows abnormal increase in arterial flow to the center of the foot area. View the larger version (127K) Fig. 10A. — Embolization of arteriovenous deformity of the liver in female neonate that has severe congestive heart failure requiring tracheal intubation and arterial presses. Color Doppler echo of the liver for embolization shows large feeding artery (long arrow) communicating with large draining vein (short arrow). View larger version (159K) Fig. 10B. — Embolization of arteriovenous deformity of the liver in female neonate that has severe congestive heart failure requiring tracheal intubation and arterial presses. Arteriogram before embolization performed with catheter in liver artery shows tangle of arterial structures in the liver and large draining vein (arrows). View the larger version (153K) Fig. 10C. — Embolization of arteriovenous deformity of the liver in female neonate that has severe congestive heart failure requiring tracheal intubation and arterial presses. Arteriogram after embolization shows elimination of current by arteriovenous malformation. The patient's congestive heart failure dissolved immediately and it is currently going well 1 year later. Check out the larger version (157K) Fig. 10D. — Embolization of arteriovenous deformity of the liver in female neonate that has severe congestive heart failure that compresses tracheal intubation and arterial. Color Doppler echo after procedure shows thrombosis of large draining vein (arrow). Transarterial embolization is performed under general anesthesia in children. Coaxial systems are used to achieve cannulation of subselective arteries. It is important to embolize subselectively because when the embolization proximal feeder arteries are carried out, the recruitment of other feeding vessels to AVM can occur and access to these vessels may have been eliminated [4]. Materials used for ethanol, coils and particles [1]. After embolization of each feed vessel, a second arteriogram is obtained to investigate for parasitization by other feed vessels. If sub-selective arterial access to the AVM is not possible due to previous operations, previous embolization or difficult anatomy, direct puncture of the AVM's feeding vessels can be achieved with sonographic guidance [1]. Both hemangiomas and vascular deformities can be seen in combination with certain syndromes. Knowledge of these associations helps in obtaining appropriate imaging studies to investigate for additional vascular malformations or other lesions. Lymphomaformations may be associated with Turner syndrome, Down syndrome, trisomy 13 and 18, and Noonan syndrome [55]. In blue rubber bleb nevus syndrome, venous malformations can be seen with regard to the skin, musculoskeletal system and gastrointestinal tract [4, 5] (fig. 11A,11B,11C). Skin lesions are often dome-shaped and painful [5]. Maffucci syndrome refers to venous malformations and multiple enchondromatosis [4]. Hemangiomas may be associated with a number of abnormalities. A cluster of abnormalities has been referred to as PHACE syndrome: posterior fossa abnormalities, facial hemangiomas, arterial abnormalities, cardiovascular abnormalities, and eye abnormalities [56, 57] (Fig. 12A,12B,12C). The syndrome is also associated with a supraumbilical midline raphe [56, 57] (Fig. 12A,12B,12C). Klippel-Trénaunay syndrome is a combined capillary-lymph-venous deformity of the stem or limbs in combination with limb overgrowth [5]. Sturgeon-Weber syndrome is a trigeminal nerve distribution capillary deformity with intracranial abnormalities [5]. Proteus syndrome includes cutaneous and visceral vascular deformities with pigmented nevi, hemihypertrophy, hand or foot overgrowth, exostosis, and lipomatosis [5]. Check out the larger version (138K) Fig. 11A. — Blue rubber bleb nevus syndrome in 11-year-old boy. Photo of tongue shows mass (arrows) in back tongue. View larger version (192K) Fig. 11B. — Blue rubber bleb nevus syndrome in 11-year-old boy. Axial T2-weighted fat-saturated rapid spin-echo MR image (4550/84 [TR/TE]) shows venous deformity as lobulated, high signal-intensity mass (arrows). Patient also suffered bleeding from multiple gastrointestinal sources due to other venous malformations of the

gastrointestinal tract. View larger version (170K) Fig. 11C. — Blue rubber bleb nevus syndrome in 11-year-old boy. Photo of plantar surface of the feet shows multiple venous deformities. View the larger version (157K) Fig. 12A. —PHACE (posterior fossa abnormalities, facial hemangiomas, arterial abnormalities, cardiovascular abnormalities, and eye abnormalities) syndrome in 1 month old female baby. Photo of face shows hemangioma of right job and ear. Eye is closed due to mass effect of Check out the larger version (182K) Fig. 12B. —PHACE (rear fossa fossa facial hemangiomas, arterial abnormalities, cardiovascular abnormalities, and eye abnormalities) syndrome in 1-month-old female baby. Axial T2-weighted rapid spin-echo MR image (2800/100 [TR/TE]) with fat saturation through orbits shows lobulated high signal-intensity hemangioma (large arrow) around right ball. Note abnormally high signal intensity in subcutaneous area around the right ear (small arrows). View the larger version (147K) Fig. 12C. —PHACE (posterior fossa abnormalities, facial hemangiomas, arterial abnormalities, cardiovascular abnormalities, and eye abnormalities) syndrome in 1 month old female baby. Photo shows supraumbilical midline raphe. Hemangiomas and vascular malformations are endotalizations that can show a number of serious medical problems. Knowledge of the distinctive clinical characteristics and characteristic imaging findings of these lesions is essential for providing appropriate monitoring and therapy. We have presented the approach used to examine these patients when they are seen in a multidisciplinary clinic. This is the third in a series of Centennial Dissertations that the AJR publishes this year in honor of former presidents of the American Roentgen Ray Society, two of whom are pictured above. Address correspondence to L.F. Donnelly.1. Yakes WF, Rossi P, Odink H. Arteriovenous deformity management. 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