Klippel-Trenaunay Syndrome

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Learning Objectives: After studying this article, the participant should be able to: 1. Define the triad of signs and symptoms that describe Klippel-Trenaunay syndrome. 2. Speculate on the various theories regarding its pathogenesis. 3. Discuss the necessary radiologic workup required to pursue appropriate management. 4. Restate the most common reasons for surgically treating this syndrome. 5. Formulate a concise and appropriate management protocol for patients with Klippel-Trenaunay syndrome.

The association of three physical findings including capillary malformation, varicosities, and hypertrophy of bony and soft tissues corresponds to Klippel-Trenaunay syndrome. This triad of findings, described by the two French physicians Klippel and Trenaunay in 1900, differs from Parkes-Weber syndrome, in that Klippel-Trenaunay syndrome does not incorporate significant hemodynamic arteriovenous fistulas. Generally, management of this disease process should be individualized. Surgery should be considered in cases where skin ulcerations lead to persisting and recurrent bleeding, or where digital deformities lead to functional disabilities or where significant limb overgrowth leads to both functional and psychological impairment. Persistent hematochezia, hematuria, and vaginal and esophageal bleeding are considered indications for surgical intervention. Recurrent attacks of thrombophlebitis and cellulitis are treated medically with anti-inflammatory agents and antibiotics. Otherwise, management of this syndrome is generally conservative, consisting of psychological encouragement, reassurance, and the continued use of graduated compressive stockings for varicosities and intermittent pneumatic compression pumps for lymphatic edema. (Plast. Reconstr. Surg. 109: 2052, 2002.)

In 1900, two French physicians described a congenital abnormality that consisted of a triad of clinical findings localized to one or more extremities. Initially called naevus variqueus osteo hypertrophique (varicose osteohypertrophic nevus), and subsequently renamed after the two investigators who had recognized it as “a syndrome,” Klippel-Trenaunay syndrome consists of a constellation of anomalies, including capillary malformation (most commonly portwine stains), varicose veins or venous malformations, and hypertrophy of both bony and soft tissues primarily involving limbs and, to a much lesser degree, intraabdominal and thoracic structures.

Although theoretical explanations have been proposed for its pathophysiologic characteristics, the etiology remains largely obscure and unresolved. The prognosis is generally benign and the management is primarily supportive unless symptoms of hematuria, hematochezia, or esophageal varices arise. In these circumstances, improper treatment may lead to subsequent fatalities. Ligation and stripping of varicosities is rarely recommended and should be performed only after careful invasive radiologic review of the venous outflow of the extremities. Hemodynamically insignificant arteriovenous malformations do not preclude the diagnosis of this syndrome. We present a case of Klippel-Trenaunay syndrome with a review of the pathophysiologic characteristics, diagnostic management, and objective findings and both conservative and surgical treatments of this rare and perplexing disease.

CASE REPORT

A healthy 25-year-old white man was referred with complaints of increasing varicosities and discomfort of the right lower extremity. A few days after his birth, a prominent asymmetric discoloration of his right hip, buttocks, and lower extremity was observed. As a toddler, during the process of standing and walking, obvious signs of venous varicosities and asymmetric enlargement of the right lower extremity emerged. Within a few years, he developed episodes of tiredness, achiness, and occasional pain along with a prominent limp of the right lower extremity. By age 12, after biyearly
scanograms (a radiographic technique depicting true dimensions of the long bone), he was found to have a maximum length discrepancy of 3.2 cm of the right lower extremity. Estimated growth length was calculated according to growth prediction charts. The patient then underwent epiphysiodesis (stapling of the epiphyseal plate) of the right distal femur and proximal tibia at the age of 14. Since the epiphysiodesis, the patient no longer limps and has a length discrepancy of less than 1 cm. However, in the past 2 years, he has observed an increasing number of venous varicosities and complains of intermittent aching, tiredness, and heaviness of the right lower extremity, primarily at the end of a long day of standing.

On examination, a red to purplish flat discoloration covering the right lower abdominal quadrant extending in a patchy distribution throughout the right lower extremity is present. The discoloration blanched on pressure. There was a notable abrupt termination of the skin discoloration at the midline appreciated physically by a distinct sharp linear border. There was no evidence of atrophy, eczema, or hyperhidrosis. Varicosities were localized primarily to the right lower extremity and buttock region. There was a large lateral vein and several tortuous posterior veins originating at the mid leg, which demonstrated multiple venous aneurysms (Fig. 1). Arterial pulses were palpable and intact, without detectable bruits, thrills, or pulsating vessels.

The patient has had several diagnostic tests throughout his life. A barium enema and sigmoidoscopy were both found to be negative for the presence of venous varicosities. Venous sonography depicted findings suggestive of an abrupt occlusion of the superior popliteal vein and the absence of the mid to distal superficial femoral vein. In addition, the greater saphenous vein was not identified. Venography revealed complete occlusion of the popliteal vein above the knee and multiple incompetent venous perforators in the calf, with filling of multiple large calf and thigh varices (Fig. 2). An impressive large varicoid collateral vein was observed throughout the thigh region, which reconstituted the external iliac vein at the level of the iliac crest. The lower extremity arteriogram depicted a hemodynamically insignificant small arteriovenous shunt at the arteriolar level.

The patient was treated conservatively with simple extremity elevation and by wearing graduated compressive stockings. Significant improvement of his symptoms was noted within a 2-month period.

**DISCUSSION**

Klippel-Trenaunay syndrome is a rare disease occurring in all ethnic groups with equal frequency, resulting in capillary malformation, abnormally large varicose veins or venous malformations, and enlargement of both soft and bony tissues. Unlike Parkes-Weber syndrome, which consists of hemodynamically large arteriovenous communications, small insignificant arteriovenous malformations do not preclude the diagnosis of this syndrome. The vascular abnormalities commonly occur in the lower extremity but have also been observed in other areas as well (head, neck, buttocks, abdomen, chest, and oral cavity). Klippel-Trenaunay syndrome has no known direct hereditary factors but may be inherited on a multifactorial basis. As a result, the etiology of this syndrome remains hypothetical, obscure, and controversial.

![Fig. 1. (Left) Multiple tortuous veins of the posterior and lateral right lower extremity. In addition, note capillary malformation at the foot extending to the right lower flank and hypertrophy of the entire extremity. (Right) Typical large lateral vein, demonstrating the “lumbar-to-foot” pattern.](image-url)
In 1985, Servelle theorized that the symptomatic triad observed in Klippel-Trenaunay syndrome was attributable to venous hypertension that develops as a result of a congenital deep vein anomaly consisting of atresia, hypoplasia, or vascular agenesis. Servelle also postulated the cause of the syndrome to be the result of an external deep venous compression by abnormally placed muscles or fibrovascular cords. In 1984, Servelle submitted an article reporting on 768 patients with Klippel-Trenaunay syndrome who underwent operation and who were found to have a malformation of the deep venous system that was thought to be responsible for the elongation and edema of the affected limb.6 Others such as Bourde have suggested that Klippel-Trenaunay syndrome is an abnormality of the embryologic vascular system that partially persists in the fetus.6 Young suggested that the soft-tissue and bony hypertrophy seen in the syndrome are mesodermal developmental abnormalities. The mesodermal abnormality theory postulated by Baskerville et al. would explain all three symptomatic features of Klippel-Trenaunay syndrome. A partial persistence of the embryologic vascular system would cause an augmentation of blood flow, resulting in an increased temperature to the involved limb that in turn would increase the circumferential size and elongation of the limb. Furthermore, this may cause the histologic vascular changes that later in life become apparent as varicosities and capillary malformations. Teratogens have not been implicated in the etiology of Klippel-Trenaunay syndrome. The classic triad of findings of Klippel-Trenaunay syndrome consists of varicose veins, capillary malformation, and hypertrophy of the affected limbs. The diagnosis can be made on at least two of the three cardinal features. Findings that may be associated with Klippel-Trenaunay syndrome but do not necessarily constitute the syndrome are hypoplasia, aplasia, venous and valvular incompetence, and aneurysmal dilatation of the deep venous system. Furthermore, lymphatic malformations are not uncommon. According to Jacob’s study of 252 patients with Klippel-Trenaunay syndrome, 63 percent had three clinical findings and the remaining 37 percent had two clinical findings. In the majority of cases (94 percent), at least one of the findings is noted shortly after birth, whereas the remaining features become evident as the child begins to stand and ambulate. In Lindenauer’s study of 18 patients with Klippel-Trenaunay syndrome, cutaneous capillary malformations were noted first, followed by varicosities and limb hypertrophy. Although older studies have shown elongation of the limb as being the most common clinical finding, more recent studies with a larger number of patients have shown that capillary malformations occur with a greater frequency, followed by venous malformations (72 percent) and limb hypertrophy (67 percent).

There are no reports in the literature proposing a gender difference or a preference in frequency between the various limbs. The most common limbs involved are the lower extremities (88 to 95 percent), followed by the upper extremities (5 percent). Unilateral involvement occurs in 85 percent of cases. Unilateral or bilateral involvement occurs in 15 percent. Uncommonly, one may observe upper extremities (unilateral or bilateral) and crossed bilateral involvement (crossed dissociation). Very rarely, four-extremity involvement has been reported.

Capillary Malformation

The capillary malformation found in Klippel-Trenaunay syndrome is usually red to purple in color, flat, and irregular in distribution.
Whereas malformations of the extremities take on a patchy configuration, truncal malformations demonstrate a clear, sharp border, which rarely crosses the midline. The capillary malformations seen in Klippel-Trenaunay syndrome have been described as a capillary or port-wine type of malformation, which may or may not significantly blanch on pressure. Although erroneously called hemangiomas in the past, suggesting proliferation of endothelial cells, the dermatopathologic characteristics reveal them to be malformations represented by vascular channels lined by a single layer of endothelial cells localized to the superficial dermis. Skin changes usually occur on the same side as the affected limb (85 percent); however, it is not uncommon to have additional capillary malformations occurring on the contralateral limb or in another area of the body such as the trunk or head and neck. Although the capillary malformation is usually confined to the skin, Gloviczki et al. showed involvement of subcutaneous tissues, muscles, and abdominal and thoracic cavities. Some patients may undergo depigmentation or “fading” of the malformation as a result of capillary thromboses, whereas others progress to additional skin changes such as atrophy, eczema, verrucae, hyperhidrosis, bleeding, and infections. Although complete disappearance of the capillary malformation does not occur, aging may result in significant “fading” of the malformation. Puberty and pregnancy may exacerbate the capillary malformation.

Limb Hypertrophy

Even though limb hypertrophy may be seen at birth, such findings become more evident later, when the individual begins to ambulate. Although some cases have shown the affected limb to be of equal length or shorter than the contralateral normal limb, the sine qua non of Klippel-Trenaunay syndrome is a limb that undergoes both hypertrophy and elongation. Generally, the average limb discrepancy as measured by scanograms is greater than 2 cm, with the largest discrepancy reported being 11.8 cm. The discrepancy in limb length is secondary to long-bone growth and, to a lesser extent, muscular hypertrophy accompanying by increasing vascular tissue and skin thickness. The limb enlargement is generally disproportionately larger distally, and the rate of progression in limb growth is not progressively consistent and predictable.

Varicosities and Venous Anomalies

Unlike the more common varicosities, Klippel-Trenaunay syndrome varicosities differ in that they are extensive, atypically very large, take an erratic course, and begin to manifest in the early childhood years. Without supportive care, these varicosities may be painful, tender, and worsen symptomatically with increasing age. At times, ulcerations develop that may result in a superficial thrombophlebitis or cellulitis. In addition, patches of lipodermatosclerosis may also occur in the advanced untreated cases.

Of all the varicose abnormalities, the most impressive is the lateral venous anomaly known as the “vein of Servelle,” which was first described by Trelat and Monod in the late nineteenth century. This large lateral vein found in approximately 68 to 80 percent of patients initiates as a plexus of veins in the dorsum of the foot, it extends proximally and laterally for a variable distance, before penetrating the deep system. In the majority (33 percent) of cases, the lateral vein will extend to the full length of the leg, terminating into the internal iliac system through the gluteal veins. In the rest of the patients, the lateral vein is variable in distance, terminating in decreasing frequency in the profundus femoris vein, superficial femoral vein, popliteal vein, and external iliac vein. Although the varicosities are primarily specific to the lower extremity, they may also radiate to the buttock or abdominal region. In addition to the lateral vein, there are numerous other venous anomalies at various levels occurring with variable frequencies that may be appreciated in Klippel-Trenaunay patients. These include hypoplasia, agenesis, valvular incompetence, and aneurysmal dilatation of the deep venous system. Customarily, the most common deep veins involved with these anomalies are the superficial femoral and popliteal veins. The most frequent abnormality is attributable to either hypoplasia or possibly external compression by a fibrous band.

Associated Abnormality

The major associated malformations seen in Klippel-Trenaunay syndrome are generally divided into vascular, skeletal, cutaneous, and
lymphatic. The specific findings are numerous and beyond the scope of this article. However, it is interesting to note that these congenital anomalies may be seen within, adjacent to, or at various distances from the involved limb, suggesting “a more generalized mesodermal and/or ectodermal dysplasia.”

With regard to the skeletal findings, the most commonly (29 percent) associated abnormalities found were dislocation of the hip and syndactyly. Hematochezia and hematuria, and esophageal variceal bleeding, although rare, are very serious findings that may result in mortalities. Vaginal and vulvar hemorrhaging are not as serious, but may be problematic cases to treat. In an article by Servelle et al. reporting on 588 patients with Klippel-Trenaunay syndrome, six children were found to have significant symptoms of rectal bleeding and hematuria. Baskerville et al. observed similar findings, although at a higher frequency.

Hematochezia is considered the most common form of bleeding in Klippel-Trenaunay patients, and its pathophysiologic characteristics are intriguing. Servelle reported that the majority of Klippel-Trenaunay patients had a deep venous anomaly resulting in a compensatory venous pathway, that is, the frequently observed lateral vein of the lower extremity, and the sciatic vein (embryologic remnant vein). Both of these veins make up the posterior venous system of the lower extremity. The lateral and sciatic vein unite superiorly in the thigh to later bifurcate and penetrate the pelvis through the sciatic and gluteal notches, terminating in an extremely dilated and overloaded internal iliac vein. The internal iliac vein accepts venous drainage from a variety of other vessels such as rectal, pudendal, vesicular, and genital veins. When these veins are no longer capable of effectively draining into the dilated internal iliac vein, because of the overload drainage from the posterior or compensatory venous pathway of the limb, dilated varicosities with concomitant bleeding may occur, resulting in hematochezia and hematuria. Vulvar bleeding occurs by means of a different mechanism of shunting blood from the affected to the normal limb secondary to underdeveloped femoral and external iliac veins. Esophageal variceal bleeding has been shown to be secondary to a hypoplastic portal vein.

Lymphatic abnormalities are common and can be extremely disabling. It has been shown that even patients without outward physical signs or symptoms of lymphatic abnormalities have lymphatic anomalies after appropriate studies (e.g., lymphangiography). The lymphatic anomalies most commonly observed are lymphatic aplasia, hypoplasia, and reduction of both lymphatic trunks and nodes. The resulting signs appear as either lymphedema or cutaneous lymphatic vesicles that occur secondary to backflow from an obstructed or congested deep lymphatic system.

**Diagnosis and Treatment**

The pathophysiologic characteristics and etiology of Klippel-Trenaunay syndrome are uncertain. For this reason, management and treatment of this disease is generally supportive unless the patient becomes overly symptomatic or disabled. Although the majority of Klippel-Trenaunay patients will complain of physical disturbances, approximately 25 percent will approach the physician for cosmetic reasons.

**Capillary Malformation**

In general, capillary malformations are left alone unless they become progressively symptomatic. Skin breakdown and ulcerations with subsequent bleeding may lead to surgical excision of the overlying skin. Although this may be a direct approach to the malformation, it is important to note that the affected skin heals rather poorly. Hence, any resection of the malformation should be performed with caution in trying to minimizing excessive scar formation and the likelihood of wound-healing complications.

Light-colored malformations can be successfully camouflaged with appropriate cosmetic products, whereas more intense and dark-colored malformations require a more aggressive approach. With the advent of lasers in the past decade, pulse dye lasers have made an impact in the treatment of malformations. The new pulse dye lasers have been adjusted with a longer wavelength, which has provided a greater depth of dermal penetration, without compromising vascular specificity. Thus, the heat produced by the laser is targeted toward vascular structures and not to adjacent normal elements. Managing malformations by means of this method requires a half dozen or more treatments with 6-week intervals. Malformations of the bladder in patients with Klippel-Trenaunay syndrome resulting in hematuria have also been successfully treated with a...
neodymium:yttrium-aluminum-garnet laser, resulting in excellent tissue coagulation.\textsuperscript{27}

**Varicosities**

Klippel-Trenaunay patients complain most commonly about their prominent and painful varicosities. Continued dilatation of venous vessels may result in ulceration and phlebitis, which results in bleeding. Servelle et al.\textsuperscript{5,28} reported deep venous anomalies were frequently secondary to fibrous bands or anomalous vessels which, if properly divided, would improve the varicosities. Conversely, Lindenauer\textsuperscript{4} and others\textsuperscript{9} have reported that ligation and stripping of varicosities worsened symptoms (90 percent) by the rapid development of new varicosities that subsequently made edema and extremity discomfort worse. The reason for Lindenauer’s findings was that the anomalies of the deep venous system in his experience were secondary to agenesis of the venous system and not secondary to compression by abnormally placed fibrous bands, as found by Servelle.

Generally, varicosities are managed supportive with intermittent rest, elevation, reassurance, and continued reinforcement in wearing graduated compressive stockings. Thrombophlebitis or cellulitis should be treated nonoperatively with analgesics and antibiotics, respectively. When conservative measures become realistically resistant, both aggressive and invasive forms of treatment may be used. These consist of two forms: resection or sclerosing of involved varicosities.

The management of these varicosities must be dealt on an individual basis. Before surgical treatment of any varicosities, radiographic examination is required. The most effective imaging techniques used are venography and magnetic resonance imaging. Duplex venograms should be obtained in all cases before surgical interventions to visualize the entire venous anatomy for venous anomalies, valvular incompetence, and persistent embryologic veins. Large varicosities should never be ligated or stripped, especially when the deep venous system is found to be either partially or completely atretic. The supra-pubic veins should never be resected because they serve as a substitute channel for the atretic iliac vein.\textsuperscript{50} In fact, most surgical procedures performed with the intention of improving varicosities are temporary and short-lived and frequently worsen the condition.

An alternative method for the management of vascular disorders seen in Klippel-Trenaunay syndrome is percutaneous injection of sclerosing agents (sodium tetradecyl sulfate, sodium morrhuate, ethanolamine oleate, and absolute alcohol).\textsuperscript{30} This method promotes irritation of the endothelium, obliterating the vessel lumen and resulting in fibrosis. Sclerotherapy may be facilitated by color-duplex imaging.\textsuperscript{31} Because some veins recanalize (18 percent),\textsuperscript{31} multiple injections (up to 30 in some cases) may be required at appropriate intervals (4 to 6 weeks) allowing the inflammatory responses to subside. Eventually, with an appropriate sclerosing protocol, almost half (44 percent) of venous malformations disappeared and greater than a quarter (28 percent) diminished.\textsuperscript{31}

**Hypertrophy and Elongation**

Hypertrophy and elongation of the extremity should also be dealt with on an individual basis. Although the rate of overgrowth in Klippel-Trenaunay syndrome is unpredictable,\textsuperscript{12,32} the method of predicting extremity growth has been well described by Anderson and Green.\textsuperscript{33} Furthermore, serial scanograms and computed tomography may be used to measure limb length and to determine the best time for length equalization procedures such as epiphyseal stapling and femoral shortening. Extremity discrepancy of 2 cm or less may be easily managed by using a shoe lift on the contralateral side to compensate for the discrepancy and the possible development of scoliosis. Extremity discrepancy of 2 to 3 cm or more is likely to result in significant ambulatory difficulties, abnormal posturing, and contralateral compensatory changes, resulting in an unphysiologic gait. These discrepancies should be addressed by means of limb-shortening procedures before they cause permanent and irreversible consequences. Although technically challenging and plagued with various significant complications, arresting epiphyseal growth is performed by either stapling or epiphysiodesis. Stapling is considered unreliable, unpredictable, and fraught with numerous complications. Epiphysiodesis as described by Mulliken and Young\textsuperscript{18} is considered to be reliable and permanent, provided that measurements and predictions are made accurately. Both femoral and tibial shortening are additional methods effective in shortening lower limbs and at the same time reducing the period of immobilization that is so frequently required with these procedures.
Another method used to compensate for extremity discrepancy, which is not accepted and recommended, is to ligate the contralateral deep veins. Servelle hypothesized that the elongation, unlike Parkes-Weber syndrome, is not secondary to an arteriovenous malformation, but rather is a result of venous hypertension. Servelle showed in dog studies that by ligating the veins at the groin or popliteal region, elongation of an extremity ranged between 2.6 and 7.6 percent within 1 to 1.5 years. Hutchinson and Burdeaux reported similar findings. Servelle performed ligation of the popliteal vein in the normal limb on 48 children, with differences in lengths between the two limbs significantly reduced or even absent by adult life.

Upper extremity disparity is rarely severe enough to be noticed by both patients and others and thus rarely requires surgical intervention. Uncommon major digital deformities resulting in significant functional disabilities and unmanageable skin complications may be treated by amputation. This is not the case when major grotesque and functionally disabling extremities are a concern. In these cases, careful selection in debulking procedures may be beneficial.

**Bleeding**

Surgically correcting hematuria and hematochezia involves releasing the superficial femoral and deep femoral veins from their adherence to nearby muscles. This allows a free flow within these veins that, in turn, allows decompression of the inferior limb. This can only be achieved if the abnormality does not also lie on the agenesis of the anterior venous system. Furthermore, one should keep in mind that the retroadductor vein forms an anastomosis with the sciatic vein. This in turn facilitates decompression of the internal iliac system by reducing these overload phenomena. If the above surgical treatment is not feasible, one may need to rely on performing a rectosigmoidectomy or a hemorrhoidectomy, the latter of which usually does not have adequate long-term benefits. Hematuria may be managed by a partial or total cystectomy. Esophageal variceal bleeding caused by portal vein hypoplasia may be surgically corrected by performing a splenorenal shunt.

**Lymphedema**

Lymphedema therapy can be divided into conservative and operative management, the former being the most common. Initial management of lymphedema begins by extensively educating both patient and family on the basics of skin hygiene, with the end goal of minimizing skin infections. Combination of physical therapies, which include manual lymphedema treatment, remedial exercises, and compression applied with multilayered bandage wrapping, can be extremely beneficial. Additional therapy is performed in the way of intermittent pneumatic compression, thermal therapy, elevation of the involved extremity, and drug therapy, which involves the use of diuretics, oral benzopyrones, and antibiotics. Operative management involves “debulking” of excess skin and subcutaneous tissue performing reconstructive microsurgical procedures with which interpositional vein segments to restore lymphatic continuity are achieved, or creation of lymphovenous and lymph-nodal shunts to improve lymphatic transport. Suffice it to say that lymphedema may be simple or complex, but should not be neglected in view of the numerous complications that can result. Thus, clinical lymphologists, physicians, nurses, and physiotherapists trained to deal with lymphedema are mandatory in all circumstances. Fortunately, although lymphedema is seen to an extent in all Klippel-Trenaunay patients, very few cases result in debilitating findings that require operative treatments.

**SUMMARY**

The association of three physical findings including capillary malformation, varicosities, and hypertrophy of bony and soft tissues correspond to Klippel-Trenaunay syndrome. This triad of findings, described by the two French physicians Klippel and Trenaunay in 1900, differs from Parkes-Weber syndrome, in that Klippel-Trenaunay syndrome does not incorporate significant hemodynamic arteriovenous fistulas.

Generally, management of this disease process should be individualized. Surgery should be considered in cases were skin ulcerations lead to persisting and recurrent bleeding, or where digital deformities lead to functional disabilities or where significant limb overgrowth leads to both functional and psychological impairment. Persistent hematochezia, he-
maturia, and vaginal and esophageal bleeding are considered indications for surgical intervention. Recurrent attacks of thrombophlebitis and cellulitis are treated medically with antiinflammatory agents and antibiotics. Otherwise, management of this syndrome is generally conservative, consisting of psychological encouragement, reassurance, and the continued use of graduated compressive stockings for varicosities and intermittent pneumatic compression pumps for lymphatic edema.

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REFERENCES

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Self-Assessment Examination follows on the next page.
Klippel-Trenaunay Syndrome
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1. ALL OF THE FOLLOWING ARE FINDINGS ASSOCIATED WITH KLIPPEL-TRENAUNAY SYNDROME EXCEPT:
   A) Varicose veins
   B) Hypertrophy of bony and soft tissues
   C) Significant arteriovenous malformations
   D) Hypoplasia and atresia of the deep venous system
   E) Cutaneous capillary malformation

2. WHICH OF THE FOLLOWING IS TRUE REGARDING KLIPPEL-TRENAUNAY SYNDROME?
   A) Klippel-Trenaunay syndrome and Parkes-Weber syndrome are synonymous diseases
   B) Associated with hereditary factors
   C) Well-described etiology
   D) More frequently seen in the right lower extremity
   E) May involve all four extremities

3. WHICH OF THE FOLLOWING IS NOT A THEORY OF THE PATHOGENESIS OF KLIPPEL-TRENAUNAY SYNDROME?
   A) Increase in intrauterine capillary and venous blood flow
   B) Persistence of arterial hypertension resulting in limb elongation
   C) Congenital mesodermal developmental abnormality
   D) Abnormality of the embryologic vascular system

4. THE MOST COMMON EXTREMITY SYMPTOMS SEEN IN KLIPPEL-TRENAUNAY SYNDROME ARE:
   A) Capillary malformation and limb hypertrophy
   B) Varices and edema
   C) Elongation and trophic skin changes
   D) Edema and hemangioma
   E) Trophic changes and edema

5. ALL OF THE FOLLOWING REGARDING THE HEMANGIOMA SEEN IN KLIPPEL-TRENAUNAY SYNDROME ARE TRUE EXCEPT:
   A) Blanches with pressure
   B) May occur on the extremity not affected by the varicosity and hypertrophy
   C) Usually red to purple in color with an irregular and patchy distribution
   D) Involvement of the subcutaneous tissue and muscle does not occur
   E) A distinct clear, sharp border is usually appreciated at the midline on the abdomen

6. ALL OF THE FOLLOWING REGARDING ELONGATION, HYPERTROPHY, AND VARICOSITIES SEEN IN KLIPPEL-TRENAUNAY SYNDROME ARE TRUE EXCEPT:
   A) The “lumbar-to-foot” pattern vein that is seen takes a medial tortuous pathway through the extremity
   B) Varicosities may be tender
   C) Discrepancy in extremity length is because of excessive growth of the femur and/or tibia
   D) Hypertrophy of the extremity is associated with pitting edema
   E) Ulceration of superficial varicosities leading to cellulitis is common

7. PRIOR TO THE SURGICAL TREATMENT OF THE SYMPTOMATIC KLIPPEL-TRENAUNAY PATIENT, VENOGRAMS SHOULD BE OBTAINED IN ALL CASES.
   A) True
   B) False
8. ALL OF THE FOLLOWING ARE TRUE REGARDING THE MANAGEMENT OF KLIPPEL-TRENAUNAY SYNDROME EXCEPT:
   A) Varicosities are managed supportively with rest, elevation, and graduated compressive stockings
   B) Ligation, stripping, or resection of the large and prominent varicosity leads to improvement of the syndrome
   C) Extremity discrepancy of greater than 2 cm is usually treated by epiphysiodesis
   D) Extremity discrepancy of less than 2 cm is treated nonsurgically
   E) The capillary malformation is usually not treated unless persistent bleeding from superficial ulcerations occurs

To complete the examination for CME credit, turn to page 2182 for instructions and the response form.