Surgical Implications of Klippel–Trenaunay Syndrome

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Between January 1956 and July 1981, 40 patients with Klippel–Trenaunay syndrome were seen at the Mayo Clinic. Twenty male and 20 female patients presented with the classic triad of soft tissue and bony hypertrophy of the extremity, hemangioma, and varicosity without evidence of functional arteriovenous fistulae. The lower extremity was involved in 38 patients (95%), the upper extremity in six patients (15%). In four of these cases (10%), both the upper and lower extremities were affected. The disease was unilateral in 34 patients (85%), bilateral in five cases (12.5%), and crossed-bilateral in one case (2.5%). Surgery was done in 13 patients (32.5%), nine of whom were operated on at the Mayo Clinic and four of whom had had previous surgery elsewhere. Excision and stripping of varices were performed in three patients; of these three, a deterioration of symptoms was observed in one, but the procedure was beneficial in the other two. Partial varicectomy was performed in four cases, while resection of the angioma was attempted in eight cases, with good results in three cases of small angiomas. Femoral and tibial epiphysiodesis stopped the overgrowth and produced an excellent result in one case. In four cases of epiphysiodesis at the foot level and two cases of derotational tibial osteotomy, moderate improvement was achieved. The indication for vascular or orthopedic surgery should be carefully considered in each patient who has this syndrome. One patient (2.5%) died of a severe form of the disease, but the relatively benign course is documented by the 21 patients (52.5%) who are free of complaints without any treatment or with elastic support only.

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IN THEIR BASIC work, "Du Naevus Variquex Osteo-Hypertrophique," published in 1900 in the *Archives Generales de Medecine*, French physicians Klippel and Trenaunay described a clinical syndrome with three major symptoms: 1) hemangiomas, 2) hypertrophy of the soft tissue and bone with overgrowth of the extremity, and 3) varicose veins.[1] This syndrome is often mentioned together with the "hemiangiectatic hypertrophy" described by F. Parkes Weber in 1907[2] and 1918.[3] However, the differences in hemodynamic pathology, in prognosis, and in the treatment of these two diseases seem to be significant. In their original paper, Klippel and Trenaunay do not mention any sign of arterial dilatation, bruit, or localized murmur of pulsating veins, although the clinical picture of arteriovenous (A-V) fistula was already well known at the turn of the century. To the contrary, Parkes Weber called the syndrome of hemiangiectatic hypertrophy a congenital "phlebarterectasy," which is a "dilatation of arterial and venous trunks . . . with tumor-like haemangiomatous overgrowth in the vascular system." He clearly describes the signs of arteriovenous fistula, in which "sometimes . . . a definite kind of thrill or pulsation . . . is transmitted to the veins."

Several classifications of the congenital vascular anomalies were published in the literature,[4–9] but the confusion about the two syndromes still persists. The terms congenital angiectatic hypertrophy,[10] infantile angiectatic osteohyperplastic syndrome,[11] osteoangiohypertrophic syndrome,[12] and Klippel–Trenaunay–Weber syndrome[12–14] are all used. However, it is widely accepted that the existence of functional arteriovenous shunts determines not only the treatment, but the outcome of the disease.

Therefore, this paper reserves the term Klippel–Trenaunay syndrome, or osteoangiohypertrophy-type "Klippel–Trenaunay," for cases with limb hypertrophy and varicosity associated with hemangiomas and/or lymphangiomas, but without A-V fistula. Anomalies of the deep veins of the extremity (hypoplasia, atresia, aplasia, valvular agenesis) are frequently seen. The historic, hemodynamic, therapeutic, and prognostic differences between the Klippel–Trenaunay and the Parkes Weber syndromes are emphasized by Servelle,[15] Vollmar,[9] May,[7] Lindenauer,[16] Schobinger,[8] and several other authors.[17–20]

In recent years the development of isotopic diagnostic procedures allowed detection of "hypoactive" or "inactive" arteriovenous fistulae, in some cases with Klippel–Trenaunay syndrome.[17] However, the shunt volume is significantly lower than that observed in Parkes Weber

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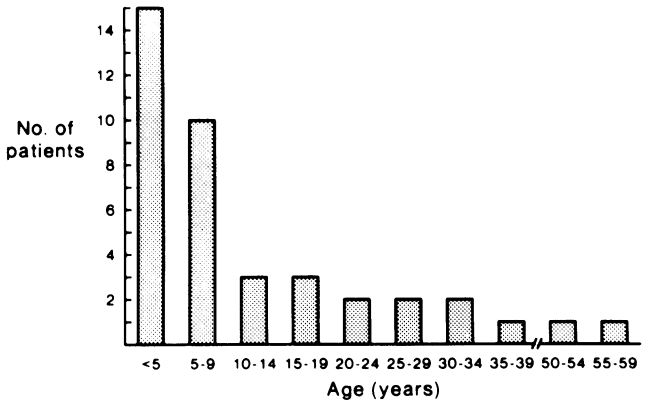

FIG. 1. Age distribution of patients with Klippel–Trenaunay syndrome.

syndrome, and the inactive A-V fistula has no hemodynamic effect and no surgical or prognostic consequences.

This paper is a review of patients treated at the Mayo Clinic in the last 25 years for Klippel–Trenaunay syndrome, with analysis of the clinical picture and the results of different methods of conservative and surgical management. All of the patients who presented any evidence of functioning arteriovenous fistulae proven by physical examination, arteriography or venous blood oxygen saturation measurements were excluded from this study.

Clinical Material

Forty patients with Klippel–Trenaunay syndrome were seen at the Mayo Clinic between January 1956 and July 1981. The mean age of the patients admitted for treatment was 16.8 years, with a range of 1 month to 55 years (Fig. 1). There were 20 men and 20 women, a male-to-female ratio of exactly 1:1. All of the patients were Caucasians.

The onset of the first symptom, hemangioma in the majority of cases, varied from being present at birth to detection at the age of six years. Thirty patients had symptoms at birth, three before the age of 1, and four between 1 and 6 years of age. The time of onset was not specified in the charts of three patients (Fig. 2). The mean follow-up period was 4.52 years, and this varied from one week to 24 years.

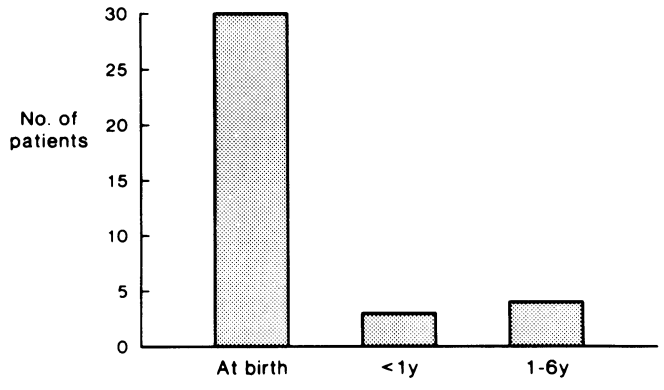

FIG. 2. Age at onset of first symptom in patients with Klippel–Trenaunay syndrome.

There was no history of associated birth trauma, and no evidence of genetic transmission of the disease could be statistically established, although in one patient's family history the mother had an angioma of the forehead and one sister had an angioma of the right lower extremity. The most common concomitant congenital anomalies were syndactylia, clinodactylia, spina bifida, atresia of the ear canal, congenital dislocation of a hip, and metatarsal and phalangeal agenesis.

Patients with clinically evident arteriovenous fistulae and the so-called "formes frustes" (patients with hemangioma without overgrowth or varicosity and patients with overgrowth without hemangioma and varicosity) were not considered to belong in the category of Klippel–Trenaunay syndrome. However, if the varicosity was mild or not significant and soft tissue, bony hypertrophy and hemangioma were present (15 cases), the patients were classified as having Klippel–Trenaunay syndrome. Nine of these 14 patients were under 9 years of age and six were under 3 years of age; thus the future development of a clinically significant varicosity might still be expected to occur.

The lower extremity was involved in 38 patients (95%) and the upper extremity in six patients (15%). Four patients (10%) had symptoms on both the upper and lower extremities. The disease was unilateral in 34 patients (85%), bilateral (Fig. 3) in five (12.5%), and crossed-bilateral (involving the right upper and left lower extremities of a 5-year-old boy) in one case (2.5%). Of the five patients with bilateral disease, the lower extremities were involved in four, while all four extremities were affected in one patient (Table 1).

The affected extremity was longer in every case. The difference in length of the extremities was determined with x-ray scanograms in 29 cases. The average difference in documented length was 2.39 cm, with the largest difference being 12 cm. The follow-up of nine patients with scanograms taken during periods of two to ten years showed significant differences in the progression of the disease (Fig. 4). However, there were only two patients who had an increase of more than 1 cm during a period of two years. The most significant change appeared in a female who had a longer left lower extremity. The differences in length of her lower extremities were 2.9 cm at 22 months, 4.8 cm at three years, 7.5 cm at four years, and 12 cm at six years. The circumference of the affected extremity was larger in 30 patients (75%). Two

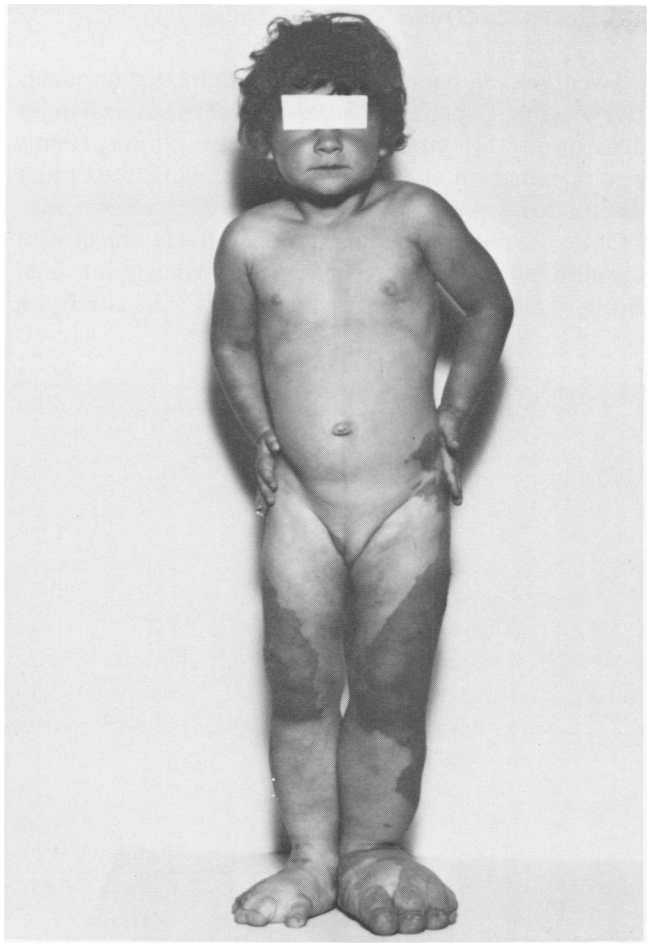

FIG. 3. Two-year-old girl with bilateral Klippel-Trenaunay syndrome. Note: bilateral hemangiomas and extensive varicosities, overgrowth of both feet and left lower extremity, and enlargement of left breast and left labium majus.

patients (5%) had marked lymphedema with Klippel-Trenaunay syndrome. The greatest difference in the circumference of their two calves was 15 cm; the mean increase was 4.7 cm.

Hemangioma was present in 39 cases (97.5%), while lymphangioma was the histologic diagnosis in one case (2.5%). Lymphangioma and hemangioma together were found in five additional cases. The typical port wine flat cutaneous hemangioma was present in 30 cases (75%). Seven from this group, as well as nine more cases, also had involvement of the deeper layers (subcutaneous tissue, muscles, and the abdominal and thoracic cavities). Cavernous hemangioma was present in 16 patients (40%), with pelvic involvement in six cases and thoracic and pleural involvement in two cases. Rectal bleeding was found in five patients and the presence of pelvic angioma was confirmed in four patients. Three patients had macroscopic hematuria at least once during their life and pelvic hemangioma was found in two of the

TABLE 1. *Klippel-Trenaunay Syndrome: Localization of Symptoms*

Location	No. of Pts.
Lower extremity	38 (95%)
Upper extremity	6 (15%)
Upper and lower extremities	4 (10%)
Unilateral disease	34 (85%)
Bilateral disease:	5 (12.5%)
Both lower extremities	4 (10%)
All four extremities	1 (2.5%)
Crossed bilateral:	
Right upper and left lower extremities	1 (2.5%)

three cases. Hemangioma was located on the extremity and the face in four cases. A 27-year-old male had several bleeding episodes from a large angioma of his left lower extremity, hematuria due to pelvic hemangioma, hemothorax from pleural hemangioma, and hemorrhage from spinal angioma that finally led to paraparesis. In 17 patients (42.5%), lower extremity hemangiomas extended to the buttock and, in ten patients, to the genitalia as well.

Varicosities were marked and significant, with incompetent perforators in 25 cases (62.5%). Seven of the 25 had persistent large embryonic veins on the lateral aspect of the thigh (17.5%). Mild or nonsignificant (mostly reticular) varicosities were noted in 15 cases (37.5%).

The most significant additional skin changes were eczema, atrophy, verrucae and hyperhydrosis. Two patients had ulceration of the leg, while five had recurrent thrombophlebitis and four presented with cellulitis several times in the course of the disease.

Arteriography was performed on nine patients, and in eight patients no arteriovenous fistula was present. In the remaining case, the early filling of the veins of the pelvic hemangioma suggested an extremely minimal degree of A-V shunting, but lower extremity angiography of the same patient revealed no arteriovenous

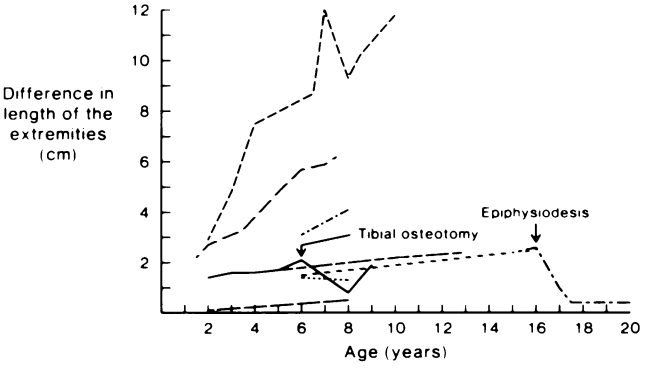

FIG. 4. Differences in length of the extremities in nine patients with Klippel-Trenaunay syndrome.

shunts. Venography was performed in only four cases. An extrinsic obstruction of the superficial femoral vein could be observed in one patient, while large dilated deep tibial veins without valves and a lack of filling of the femoropopliteal deep veins were observed in another patient. Venography performed for the third patient showed deep venous insufficiency with incompetent perforators, while venography for the fourth patient revealed anomalous dilated lateral superficial veins, but good filling of the deep venous system (Figs. 5a and b).

Treatment and Results

Twenty-seven patients (67.5%) were treated nonoperatively, while 13 patients had some type of surgical treatment during the course of the disease. Nine patients were operated on at the Mayo Clinic, while four more patients had previously undergone surgery elsewhere.

Of the 27 patients treated nonoperatively, the disease was mild in ten of them. Their overgrowth was minimal and they did not require elastic support. The condition

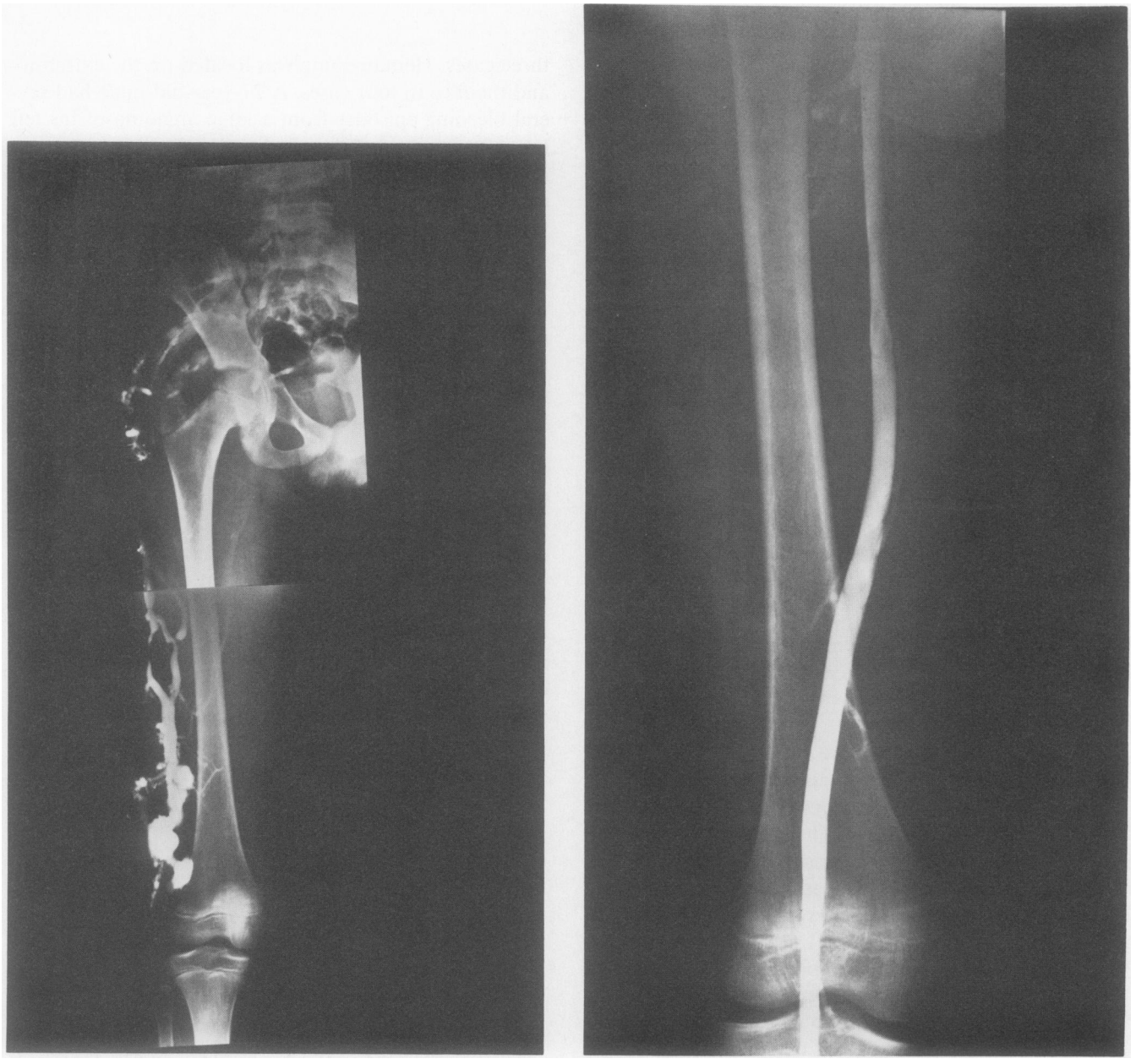

FIG. 5a and b. (a, left) Large embryonic veins on the lateral aspect of the thigh of a four-year-old patient with Klippel–Trenaunay syndrome. (b, right) Note patent deep femoral vein on the same patient.

of 11 patients was considered stable because, with the use of elastic support, they had no complaints. Therefore, 21 patients (52.5%) without treatment or with elastic support only were in good general condition and without any major complaint. Elastic support and medical treatment (antibiotics, diuretics, iron) were given to five patients whose complaints were pain, recurrent cellulitis, thrombophlebitis, recurrent bleeding and anemia, contractures, or paralysis. Four of this group remained active, while one patient (who had spinal hemorrhage and paraplegia) was in poor general condition. Another required irradiation of the hemangiomatous tissue, elastic support and repeated steroid and antibiotic treatment (Fig. 6). (The clinical condition of this female was considered to be hopelessly poor. She had severe involvement of her entire trunk and left lower extremity. She had developed severe edema, cachexia, walking disability, gluteal decubitus, and enlarging abdominal and thoracic hemangiomas and died at the age of 11 years.)

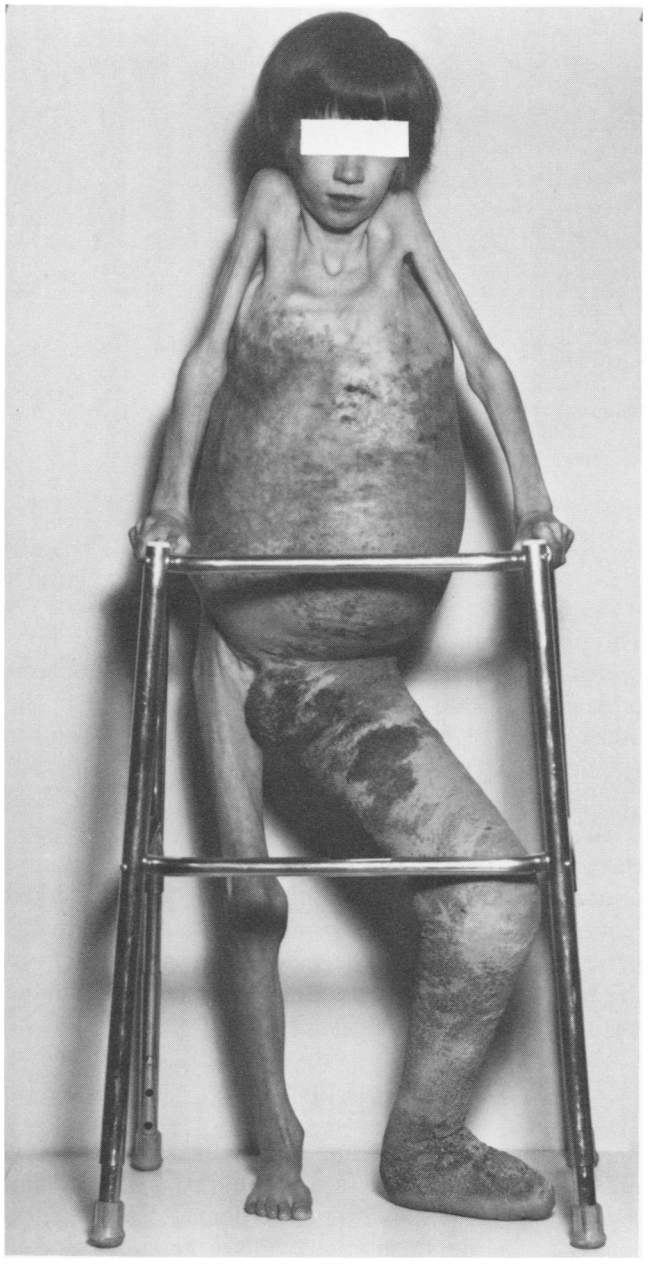

FIG. 6. Ten-year-old girl with severe progression of Klippel–Trenaunay syndrome. Hemangiomatous involvement of chest, abdomen, pelvis, and left lower extremity, with a length discrepancy of 11.8 cm.

TABLE 2. *Conservative Treatment of Klippel–Trenaunay Syndrome*

Type of Treatment	Complaints	Results	No. of Pts.
No treatment	None	Good	10
Elastic support	None	Good	11
Elastic support + medical treatment: (antibiotic, diuretic, iron, etc.)	Pain, recurrent cellulitis, thrombophlebitis, recurrent bleeding, anemia, functional impairment	Fair	4
	Paraplegia, bleeding from angioma	Poor	1
Irradiation, elastic support, steroids, antibiotics	Severe involvement of whole trunk and left lower extremity	Died	1

n = 27: 13 male, 14 female.

Table 2 summarizes the results of the nonoperative treatment. At present, 21 patients (52.5%) are in good condition, four patients (10%) are in fair condition, and one patient (2.5%) is in poor condition. One patient (2.5%) died from a progressive severe form of the disease. Another patient, a 10-year-old female who did not require treatment for her mild Klippel–Trenaunay syndrome, died in the follow-up period due to pneumonia and glomerulonephritis, causes unrelated to her vascular anomaly.

Thirteen patients received surgical treatment. Orthopedic surgery was performed on six patients. Rotational tibial osteotomy, as well as epiphysiodesis on several levels of the extremity, were performed on five of these six patients in an attempt to reduce the overgrowth of the extremity. In the sixth case, first and second metatarsal amputations were performed. A partial or total excision of the hemangioma was carried out in eight cases, while varicectomy with greater saphenous stripping was done in three cases and varicectomy without stripping in three more cases. In one patient only thrombosed varices were removed, and two patients with concomitant lymphedema had excisions of subcutaneous tissue. The overall results of the surgical treatment are presented in Table 3.

Good functional and aesthetic results could be doc-

TABLE 3. *Surgical Treatment of Klippel–Trenaunay Syndrome*

Case No.	Age	Sex	Operation	Improved	Worse	None	F/U
1	3	F	Varicectomy + v. saph. magna stripping + lat. vein excision + cavernous angioma excision.	X			4 y
2	2	F	Amputation 4° L toe + "cyst" removal from groin + epiphysiodesis + partial transmetatarsal amp. left foot.	X			5 y
3	3	F	Lateral vein excision + derotational tibial osteotomy + cast.	X			5 y
4	20	M	Angioma excision.	X			4 y
5	15	M	Epiphysiodesis (lt. distal femur + prox. tibia + prox. fibula).	X			2 y
6	33	M	Excisional operations for concomitant lymphedema (scrotum + rt. lower extremity).			X	1 y
7	4	M	Removal of clotted varices.	X			11 y
8	7	M	Varicectomy (stripping) + perforant ligature + excision hemangioma. Varicectomy of recurrent varices.		X		7 y
9	6	M	Excision angioma (3 operations), + excision lateral vein.	X			11 y
10	15	M	Excision hemangioma + amputation of 1°–2° metatarsus.	X			15 y
11	6 mo.	F	Excision angioma. Epiphysiodesis all phalanges both feet. Bilat. transmetatarsal amp. + Thompson procedure for concomitant lymphedema (6 operations).	X			8 y
12	6 mo.	M	Excision hemangioma, midtarsal amp. both feet, lateral vein excision, epiphysiodesis, derotational osteotomy (10 op. by age 4).	X			4 y
13	51	F	Varicectomy + stripping of saphenous vein + conservative treatment.	X			1 y

n = 13: 8 male, 5 female.

umented in five cases. In one case, the procedure was minor removal of large thrombosed superficial veins. In two cases, extensive varicectomy and vein strippings were performed. This produced a good result in a 3-year-old girl, with almost no recurrences after a followup of four years. In the second case, a 51-year-old female, deep vein insufficiency and stasis ulcer were present and were treated by repeat vein stripping (after a 7-year interval) and continued elastic support. Repeated excisions of angioma with varicectomy produced a good result in a 6-year-old male. Distal femoral, proximal tibial and proximal fibular epiphysiodesis produced a good result in a 15-year-old male. The discrepancy between the lengths of his two extremities was 2.4 cm before operation, 1 cm at the age of 17, and 0.4 cm at the age of 17½. Aesthetic improvement was achieved in six cases. Transmetatarsal amputation and epiphysiodesis at the foot level were performed to decrease the size of the affected foot in two cases. Toe amputation and partial metatarsal amputation were done for the same reason in two other patients. Derotational tibial osteotomy resulted in some improvement in two cases. Partial excision of the subcutaneous tissue and a modified Thompson procedure to diminish concomitant lymphedema were performed in two cases, with some improvement in one (Case 11) and no real change in the other (Case 6). One patient underwent six, and another patient ten different operative procedures (Cases 11, 12) (Table 3).

Stripping and ligation of perforators produced good early results in a 7-year-old boy (Figs. 7a and b) who underwent another varicectomy at age 14 for marked recurrent varicosity. The operation did not improve this patient's condition, and he suffers increasing pain from the scars and recurrent episodes of hemorrhage from the angiomatous tissue.

Discussion

The etiology of the Klippel–Trenaunay syndrome is unknown. No hereditary factors could be confirmed and the exact time of the embryonal damage is also questionable. The hemangiomas of the different angiodysplasias are never real tumors, but only the result of faulty autogenesis.[6,21] According to de Takats,[21] the differences in vascular anomalies are due to the stage of develop-

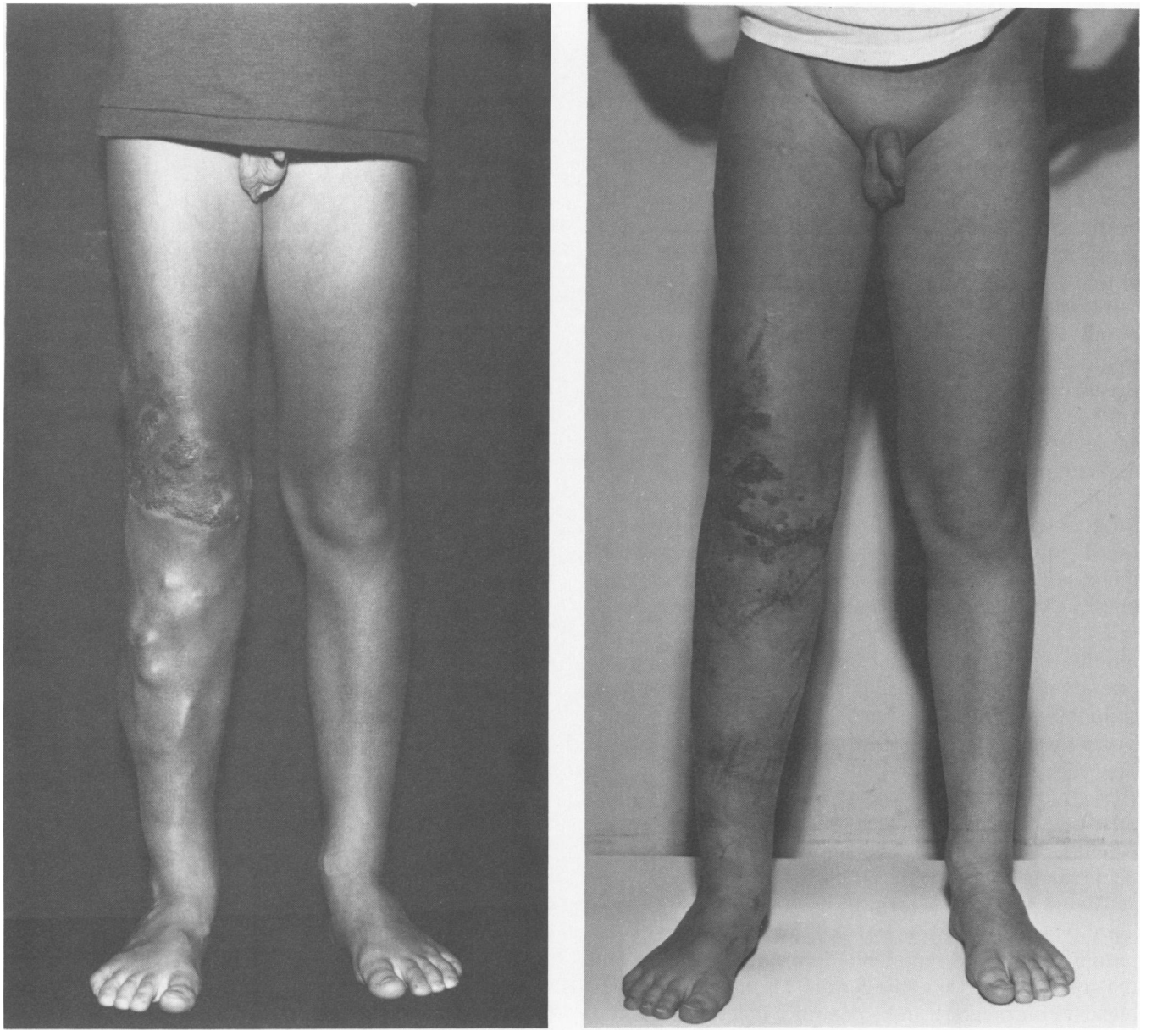

FIG. 7a and b. (a, left) Seven-year-old male patient with Klippel–Trenaunay syndrome of the right lower extremity. (b, right) Excision of angioma and resection of varices with stripping in the same patient. Good postoperative result six months following surgical intervention.

ment when aberration from the normal occurred. An excellent review of the etiologic and morphogenetic hypotheses was published by Malan and Puglionisi.[6] Most differences, in the authors' opinion, concern the relationship between the vascular and skeletal anomalies. Is the second a consequence of the altered circulation, or does the same teratogenetic agent cause angiodysplasia and hypertrophy as well? Vollmar[22] states that an "inborn error" of tissue composition and distribution results in vascular anomalies and skeletal overgrowth, and the latter is not a secondary phenomenon of the vascular disorder, as it is in the case of an arteriovenous fistula.[7,23] His hypothesis is supported by the fact that, in Klippel–Trenaunay syndrome, the intraosseous involvement by hemangiomas is rare. There is usually no bone anomaly, except its hypertrophy in diameter and length. In Parkes Weber syndrome, the existence of intraosseous A-V hemangiomas, especially next to the epiphyseal plate, is frequent.[7] Horton, who already in 1934 had reported on 38 cases of congenital A-V fistulae, emphasized that an increased blood flow to the epiphyseal line causes abnormal growth of bone.[24] Also, from

the Mayo Clinic, Janes and Musgrove in 1949 succeeded in creating overgrowth of the hind leg of young dogs by artificial A-V fistulae.[25] Servelle[26] and, later, Hutchinson and Burdeaux[23] were able to produce overgrowth of the hind limb in dogs when they created venous stasis by ligating veins or applying a tourniquet. Nevertheless, it still has not been proven whether the increased skeletal growth is caused by an increased arterial flow in the area of the epiphyseal plate, by the venous hyperemia and stasis caused by the A-V fistula, or by an inborn error in tissue development.

To establish the diagnosis of Klippel–Trenaunay syndrome, the presence of the three main symptoms—varicosity, hemangioma, and extremity hypertrophy—should be mandatory. Involvement is usually unilateral, but the rare bilateral or simultaneous upper and lower extremity involvement have also been mentioned by the first authors.[7]

The varicosity is usually atypical, often being significant on the lateral aspect of the extremity, with the so-called "lumbar-to-foot" pattern. Dodd and Cockett[5] call this syndrome the "lateral venous anomaly," to emphasize the persistence of the embryonic dorsal or sciatic vein system that normally disappears in the second month of intrauterine life. The persistence of lateral or primary marginal veins can be found in upper and lower extremity deep vein hypoplasias or aplasias in Klippel–Trenaunay syndrome, or in venous hemangiomatosis (Type Servelle–Martorell or Kasabach–Meritt[27,28]). However, this pattern occurs with congenital A-V fistulae also.[29] In this study, it was found in only seven patients. The posterolateral lumbar-to-foot pattern of the hemangiomas was much more frequent.

The most common form of hemangioma is the capillary type, or port wine nevus,[30] which has a pink-to-purplish color and represents diffuse telangiectasias of the superficial vessels of the dermis. The flat purple patch blanches on pressure and sometimes fades, but usually becomes darker as the patient gets older. The existence of port wine nevus is much less frequent in congenital arteriovenous fistulae, and is only present in about one third of the cases.[29] Cavernous hemangioma or lymphangioma can also be found. The hemangiomatous involvement of the internal organs can lead to severe complications in some very rare cases.

Besides the three main symptoms, additional common features are hyperhydrosis, skin atrophy, verrucae, dermatitis, thrombophlebitis, and cellulitis. In Klippel–Trenaunay syndrome, edema of the extremity is often present (seen in 75% of the patients). Anomalies of the lymphatic circulation in Klippel–Trenaunay syndrome have been reported by Servelle.[31] Additional congenital anomalies such as syndactylia, polydactylia, spina bifida, and equinovarus can occur in some cases.[7]

Rectal bleeding and hematuria are rare, but serious, complications of the Klippel–Trenaunay syndrome. In some cases they occur due to pelvic or abdominal hemangioma. This can also occur because the dysplastic posterolateral veins of the extremity drain the blood to the hypogastric vein. The overload of the hypogastric system prevents normal drainage of the pelvis, which results in the dilatation of hemorrhoidal veins and bladder veins and can finally cause rectal bleeding or hematuria. Five patients were observed with rectal bleeding. Earlier, Servelle[32] reported on six patients with similar symptoms.

Among the noninvasive diagnostic procedures, doppler flow measurements and venous occlusion plethysmography are useful in the exclusion of A-V fistulae. However, they can be used as a screening method only.[33] Venous blood oxygen saturation measurements have been widely used to diagnose A-V fistulae,[24,29] but are of little practical use today. Cormier[34] mentions several factors, such as temperature, anesthesia, and local vasodilatation, that can easily change these values; therefore, they should be interpreted with care. Quantitative values of arteriovenous shunt volume can be given by isotope diagnostic methods, using radioactive-labeled small microspheres.[13] The authors' opinion concerning arteriography and venography has changed in recent years. Arteriography is particularly useful in demonstrating large caliber A-V fistulae and thus is important in separating Klippel–Trenaunay from Parkes Weber syndrome. The absence of A-V fistulae in the former allows one to contemplate surgery more readily in patients with symptomatic varices. If vein stripping is to be undertaken, venography should also be done to delineate the status of the deep venous system prior to extirpative surgery.

If the varicosity is minimal, the hemangioma is a port wine superficial nevus, and the hypertrophy of the extremity is under 1 cm, the patient usually does well without treatment. If the varicosity is more significant, elastic support should be suggested to the patient. If leg length discrepancy is more than 1.5 cm, a lift placed in the shoe on the healthy foot can help to avoid the compensatory scoliosis of the vertebral column. The concomitant skin changes and the rare cases of leg ulcer might also need local care. Drug therapy should be restricted to treatment of inflammatory changes, cellulitis or anemia due to recurrent bleeding from the angiomatous tissue. Patients who develop recurrent cellulitis should be placed on long-term, prophylactic antibiotic therapy. Steroids and irradiation will not stop the development of visceral hemangiomas, and their use in the treatment of this disease is not indicated.

Varicectomy and vein stripping are usually not indicated in the treatment of Klippel–Trenaunay syn-

drome, unless specific symptoms are present. Lindenauer[16] reported on 12 patients who were operated on for varicosity, undergoing multiple ligations and stripping. Eleven of these patients had an increase in ankle edema and a rapid recurrence of the varicosity associated with leg discomfort. Martorell[35] calls attention to rare cases with iliac atresia in which excision of the large suprapubic veins is contraindicated. If agenesis of the deep veins is established, extensive varicectomy should not be performed.[17,27] If the compression of the deep veins by fibrotic bands can be found at surgical exploration, liberation of these veins can be attempted, as suggested by Servelle.[15] According to Vollmar,[27] in every fifth case of Klippel-Trenaunay syndrome there is aplasia or hypoplasia of the femoral or popliteal veins, and excision of the superficial bypassing veins leads to an increase in venous insufficiency in these patients. However, if physical examination and venography show patent deep veins, local varicectomy or even stripping can be performed after careful evaluation in each case. Deterioration was observed in one of three cases. In the second case, a 51-year-old woman with deep venous insufficiency and leg ulcer (Case 14), the Klippel-Trenaunay syndrome was mild and, after repeated varicectomy and conservative therapy, her ulcer healed rapidly. The third patient (Case 1) had good results with minimal recurrence, but the follow-up period lasted only four years. Varicectomy of large sinusoid superficial veins should be considered under the same principles of evaluation. Out of 19 patients at the Mayo Clinic who were operated on for cavernous hemangioma and varicosity without A-V fistula, Myers and Janes performed varicectomy and stripping in five who had the Klippel-Trenaunay syndrome, with good results.[36]

In three cases of smaller angiomas, the authors observed good results after cosmetic excision (Cases 1,3,4). However, complete excision of hemangiomatous tissue is seldom possible and can lead to scar tissue formation, repeated bleeding, pain, and a walking impairment. Surgery is usually not recommended in cases of large hemangiomas, even if bleeding is a major problem for the patient. Percutaneous injection of thrombotic material, obliterative polymers, or sclerosing agents may be of greater benefit in these patients.

Surgical intervention is indicated in some cases to inhibit extreme overgrowth of the extremity. Although a discrepancy of not more than 2 cm can easily be corrected by placing a lift in the heel of the shoe on the healthy foot,[37] when a marked discrepancy is noted epiphysiodesis or tibial osteotomy should be considered. In 1933, Phemister[38] first performed successful epiphysiodesis with the insertion of a bone graft across the epiphyseal plate on the medial and lateral sides following curettage of the plate. In 1945, Haas was the first to use the wire loop,[39] while Blount and Clarke[40] introduced staples that were removable at any age if further growth of the extremity was required. The lower femoral, upper tibial, and upper fibular epiphyses play the most important roles in the overgrowth of the lower extremity. The rate of overgrowth in Klippel-Trenaunay syndrome is unpredictable.[18] It is essential to obtain serial scanograms at regular intervals (usually every six months) and leg lengths should be plotted on the Green-Anderson charts so that accurate predictions can be made as to the need for epiphysiodesis and the proper time during the growth period to perform the procedure.[41]

Therapy for Klippel-Trenaunay syndrome is mostly symptomatic. Elastic compression and, in selected cases, varicectomy or transection of fibrous bands are available methods of treatment. The ideal operation in cases of hypoplasia or aplasia of the deep veins would be venous reconstruction, but this is a task for the future. Severe forms of the Klippel-Trenaunay syndrome rarely occur. Fortunately, because of the benign course of this disease, the majority of these patients do not need surgery and do well with conservative therapy.

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