Congenital Arteriovenous Fistula and the Klippel-Trenaunay Syndrome

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Congenital vascular anomalies of the extremities are poorly understood and are the source of much confusion. The striking findings present on physical examination have centered attention on the more obvious manifestations of these abnormalities and further investigation of the underlying vascular system often is not undertaken. In order to attempt to understand congenital vascular anomalies one must perform angiography (and in certain instances lymphangiography) in order to assess the status of the underlying arterial, venous, and lymphatic circulation. All too often assassments are made and opinions rendered merely on the basis of physical signs. Furthermore, angiography should include the entire affected limb and utilize sequential filming technics since the vascular abnormalities may not only be morphologic, but dynamic as well. While angiography is essential for an understanding of congenital vascular anomalies, technical factors sometimes prohibit elucidating all of the desired information.

The classification of congential vascular anomalies of the extremities has been difficult because of the many common manifestations. The most comprehensive and yet useful classification has been recently outlined by Malan and Puglionisi.[6] They classify four groups of anomalies as follows: venous dysplasias, arterial dysplasias, arterial and associated venous dysplasias, and mixed angiodysplasias. While at times it has seemed difficult to separate certain anomalies into sub-groups under these four general headings, the classification otherwise has merit. Arteriovenous fistulas are considered under the heading of combined arterial and venous dysplasias. An arteriovenous fistula is defined as an abnormal communication between an artery and vein that bypasses a capillary bed. Arteriovenous fistulas may be either acquired or congenital.

Acquired fistulas are usually traumatic in origin and single. Congenital arteriovenous fistulas are almost always multiple. While the connections are multiple they may also be localized or diffuse and active or hypoactive.

Congenital arteriovenous fistula of the extremity is not a common vascular lesion. It is marked by enlargement of the affected extremity, dilated superficial veins that are sometimes pulsatile, increased warmth of the part, the presence of a thrill and bruit and often a cutaneous hemangioma. While Branham's sign may be positive, other systemic manifestations are uncommon. Cardiac enlargement and high output cardiac failure are rare.

In such a lesion, the presence of many large, abnormal arteriovenous fistulous connections can be readily demonstrated by arteriography. These large fistulous connections correlate with the presence of a

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thrill and bruit, pulsatile veins and increased venous oxygen saturation. However, when the fistulous connections are small or hypoactive they may not be seen by arteriography and the thrill and bruit may be absent. The veins may be dilated but do not pulsate and the venous oxygen saturation may not be increased significantly. Such a fistula may be exceedingly difficult to demonstrate by arteriography. In such a lesion one may see indirect arteriographic evidence that suggests the presence of multiple small fistulas. Such evidence may include early venous return, dilated or tortuous proximal arteries, abnormal arterial branches, and delay in the fill of distal arteries.

Arteriovenous fistulas are sometimes associated with a pulsatile vascular mass. This is usually referred to as an arteriovenous aneurysm when the fistula is traumatic in origin and a cirsoid aneurysm or arteriovenous angioma when associated with a congenital fistula. A non-pulsatile vascular mass may be associated with several different congenital vascular anomalies and its presence is compatible with but by no means diagnostic of a congenital arteriovenous fistula.

The association of varicose veins, soft tissue and bony hypertrophy and cutaneous hemangioma of the "port-wine" variety confined to one extremity was reported in 1900 by Klippel and Trenaunay.[4] In 1907 Parkes-Weber described three similar cases and subsequently described several other cases associated with arteriovenous fistulas.[11, 12] Since then the terms Klippel-Trenaunay Syndrome, Parkes-Weber Syndrome, and Klippel-Trenaunay-Weber Syndrome have been used extensively in the literature to describe patients with the triad of varicose veins, bony and soft tissue hypertrophy, and cutaneous hemangioma. Some of these patients have had no evidence of arteriovenous fistula while others have had clear evidence of arteriovenous fistula. Some authors have suggested that all patients with the Klippel-Trenaunay triad have an arteriovenous fistula.[1,2] However, this does not always appear to be the case. Certainly patients with the Klippel-Trenaunay triad may have arteriovenous fistulas which may represent the basic underlying pathology. In the presence of a pulsatile vascular mass, pulsating veins, or thrill and bruit the diagnosis is easy. Demonstration of the arteriovenous fistula connections by arteriography confirms the diagnosis. There may be indirect evidence of the arteriovenous fistula seen by arteriography (early venous fill, dilated tortuous proximal arteries, etc.) which suggests the diagnosis. However, in the absence of these physical signs and arteriographic findings, it seems hazardous to render a diagnosis of an arteriovenous fistula soley on the basis of hypertrophy, cutaneous hemangioma, or varicose veins. The problem is more difficult in fistulas that are small or hypoactive. Such fistulas are more often in the lower extremity, rarely have a thrill or bruit or Branham's sign and are frequently associated with a cutaneous hemangioma of the "port-wine" variety. In such hypoactive fistulas, one must rely on careful arteriography to demonstrate evidence of the fistula. In such patients there may be dilated superficial veins, however, the deep venous system is usually normal. It appears feasible to separate the patients with arteriovenous fistulas, including hypoactive fistulas, from a distinct group of patients with the triad of varicose veins, cutaneous hemangioma of the "port-wine" variety and bony and soft tissue hypertrophy who have no evidence of an arteriovenous fistula either clinically or by arteriography.[5] In such patients the basic underlying vascular defect appears to be absence or obstruction of the deep venous system rather than arteriovenous fistula. Both of these lesions cause venous hypertension which probably accounts for the dilated superficial veins and for the bony and soft tissue hypertrophy.

During the past 15 years, 14 patients

TABLE 1a *Diffuse Congenital*

Case	Sex	Age at Onset	Location	Limb Enlargement	Dilated Veins	Thrill and/or Bruit	Cutaneous Hemangioma
1. J. R.	F	20 Yrs.	Arm	Yes	Yes	Yes	Yes
2. C. B.	F	11 Yrs.	Arm	Yes	Yes	Yes	No
3. E. B.	F	Birth	Arm	No	Yes	Yes	No
4. R. S.	M	Birth	Fingers	Yes	Yes	Yes	No
5. P. W.	F	20 Yrs.	Thigh	Yes	Yes	Yes	No
6. B. A.	M	20 Yrs.	Thigh	Yes	Yes	No	No
7. P. E.	F	15 Yrs.	Arm	No	No	Yes	No
8. L. C.	M	18 Yrs.	Arm	Yes	Yes	Yes	No
9. J. B.	F	11 Yrs.	Leg	Yes	Yes	Yes	No

with congenital arteriovenous fistulas and 25 patients with the Klippel-Trenaunay Syndrome have been seen and treated at the University of Michigan Medical Center. They represent two distinct groups of patients and form the basis of this report.

Congenital Arteriovenous Fistula

The 14 patients with congenital arteriovenous fistulas comprised two groups. There were nine patients with diffuse active macrofistulous connections and five patients with localized, hypoactive, microfistulous connections (Table 1a and 1b). Among the nine with diffuse fistulas there were six females and three males. The upper extremity was involved in six and the lower in three. Limb enlargement, dilated veins, either a thrill or bruit or both, and increased warmth was present in almost every case. In five there were masses and in two the masses were pulsatile. A cutaneous hemangioma was present in only one patient. Branham's sign was positive in two patients. All nine patients underwent multiple excision of the lesions and in six there was evidence of residual fistulous connections. In three patients there was no evidence of residual fistula. One patient required amputation of two fingers.

TABLE 1b. *Localized Congenital*

Case	Sex	Age at Onset	Location	Limb Enlargement	Dilated Veins	Thrill and/or Bruit	Cutaneous Hemangioma
1. R. W.	M	13 Yrs.	Finger	No	No	No	No
2. L. M.	M	3 Yrs.	Thigh	No	No	No	No
3. M. E.	M	1 Yr.	Foot	No	No	No	No
4. V. W.	F	?	Ankle	No	Yes	No	No
5. C. P.	M	1½ Yrs.	Thigh	No	No	No	No

Arteriovenous Fistula

Branham's Sign	Mass	Increased Warmth	Arteriogram	Treatment	Result
Yes	No	Yes	Multiple AV fistulas	Partial excision	Residual fistula—asymptomatic
?	No	Yes	Multiple AV fistulas	Ligation and excision × 5	Residual fistula—asymptomatic
Yes	Yes pulsating	Yes	—	Ligation × 1	Residual fistula—minimal discomfort
?	Yes pulsating	Yes	Multiple AV fistulas	Ligation × 2 amputation × 2	Residual fistula—asymptomatic
?	Yes	Yes	Multiple AV fistulas	Excision	No residual
No	Yes	Yes	Multiple AV fistulas	Excision × 2	No residual
No	Yes pulsating	Yes	Multiple AV fistulas	Excision × 3	No residual
No	No	Yes	Multiple AV fistulas	Ligation and excision × 2	Residual fistula
?	No	Yes	Multiple AV fistulas	Ligation and excision × 6	Residual fistula—painful cutaneous ulcer

In only one patient was the residual fistula associated with significant symptoms. Arteriography was utilized in eight of the nine patients and demonstrated multiple fistulous connections in all.

In the five patients with localized hypoactive microfistulous connections, there were four males and one female. The lesion was located in the lower extremity in four and the upper extremity in one. In almost all of these patients there was no limb enlargement, thrill or bruit, dilated veins, cutaneous hemangioma or Branham's sign. The lesion was a painless mass usually with increased warmth of the part. Arteriography was not utilized and the true nature of the mass was not often suspected preoperatively. The vascular abnormality was well appreciated at operation and the diagnosis confirmed by histologic examination. The lesion often required multiple excisions, however, in all patients it finally was completely excised with no recurrence. The similarities and differences between localized and diffuse congenital arteriovenous fistulas are summarized in Table 2. The following case reports illustrate typical patients with congenital arteriovenous fistulas.

Arteriovenous Fistula

Branham's Sign	Mass	Increased Warmth	Arteriogram	Treatment	Result
No	Yes	No	—	Excised × 2	No recurrence
No	Yes	Yes	—	Excised × 1	No recurrence
No	Yes	No	—	Excised × 1	No recurrence
No	Yes	Yes	—	Excised × 4	No recurrence
No	Yes	Yes	—	Excised × 4	No recurrence

TABLE 2. *Comparison of Diffuse and Localized Arteriovenous Fistulas*

	Sex	Site	Limb Enlargement	Dilated Veins	Thrill and/or Bruit	Branham's Sign	Mass	Increased Warmth	Results of Treatment
Diffuse AV Fistula	F	Upper Extremity	Yes	Yes	Yes	Sometimes	Yes Sometimes pulsating	Yes	Usually incomplete
Localized AV Fistula	M	Lower Extremity	No	No	No	No	Yes	Sometimes	Can be completely excised

Case Reports

Case 1. R. W. *Localized Arteriovenous Fistula.*

A 13-year-old boy was admitted to the University of Michigan Medical Center on Januatry 3, 1957. For the past several years he had noted a slowly enlarging mass along the lateral aspect of the proximal phalanx of the fourth finger of the left hand which had caused some pain and tenderness. Examination revealed a 2 × 1 cm. non-pulsatile, slightly tender, non-compressible mass along the lateral aspect of the proximal phalanx of the fourth finger. There were no dilated veins, thrill or bruit or increased warmth associated with the mass. The lesion was slightly adherent to the underlying tissues. The preoperative impression was that this was a fibroma. On January 4, 1957 the lesion was excised. Pathologic examination revealed it to be a small localized arteriovenous fistula. He returned in April 1958 with a similar, but slightly larger mass underneath the operative scar. On April 19, 1958 the lesion was more widely excised. Pathologic examination again revealed an arteriovenous fistula. He was followed through November 1960 with no evidence of recurrence and remains asymptomatic.

Case 2. J. R. *Diffuse Arteriovenous Fistula.*

A 20-year-old woman was admitted to the University of Michigan Medical Center on 12–2–63 with a history of a swollen and red right forearm and wrist for the past several years. Examination revealed the right distal forearm and wrist to be warmer and red in color and to contain many engorged veins. A prominent thrill and bruit were present at the wrist. The right arm was longer than the left and the forearm and hand were greater in girth. Compression of the brachial artery caused the pulse to slow from 100/min. to 82/min. and the measured cardiac output to fall by 20%. The oxygen saturation in the brachial artery was 83% and in the brachial vein 80%. An arteriogram (Fig. 1) revealed a diffuse arteriovenous fistula with many large fistulous connections arising from the ulnar artery which was dilated. The chest x-ray revealed no cardiomegaly or pulmonary congestion. The patient had successfully completed two pregnancies.

On December 9, 1963, the mass of fistulous connections in the hand and wrist were subtotally excised preserving the ulnar artery and the tendons and nerves in the hand. At the time of her last follow-up in 1966 she had a residual bruit and the hand was still slightly warmer. Branham's sign could not be elicited and the venous engorgement was much less. She had no symptoms relative to her hand and had normal function.

Case 3. P. W. *Diffuse Arteriovenous Fistula.*

A 20-year-old woman was admitted to the University of Michigan Medical Center with a 2-year history of an asymptomatic mass in the left thigh which had slowly increased in size. Examination revealed a 15 × 20 cm., non-pulsatile mass in the medial aspect of the left thigh. The left leg was increased in girth, but the length of

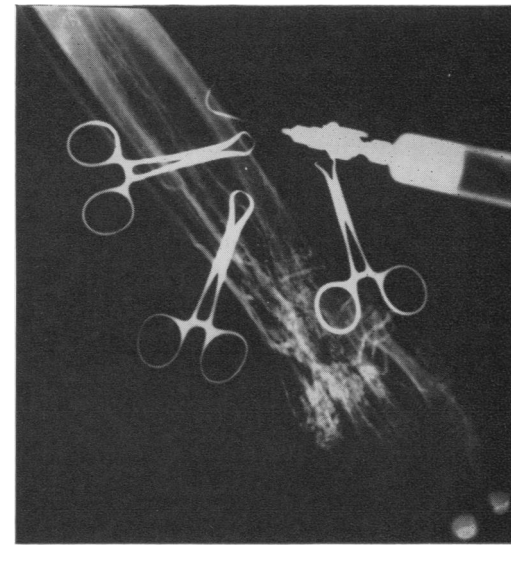

FIG. 1. Case 2. J. R. Photograph of arteriogram demonstrating multiple large fistulous connections in the hand and wrist arising from the ulnar artery which is dilated.

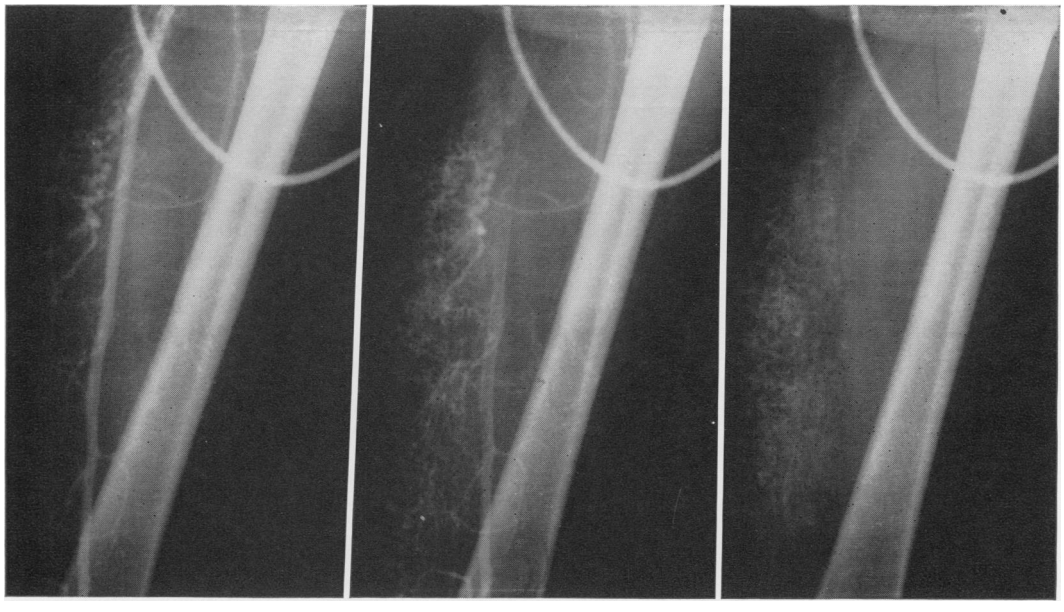

FIG. 2a. (left) Case 3. P. W. Photograph of femoral arteriogram demonstrating abnormal tortuous branches arising from the superficial femoral artery.

FIG. 2b. (center) Case 3. P. W. Photograph of latter stage in femoral arteriogram demonstrating multiple diffuse arteriovenous connections.

FIG. 2c. (right) Case 3. P. W. Photograph of still later phase of femoral arteriogram showing diffuse venous filling.

both extremities was equal. The left thigh was increased in warmth and normal in color. There were many small dilated subcutaneous veins present. A bruit was present over the mass but no thrill could be felt. A femoral arteriogram revealed many large arteriovenous connections in the medial aspect of the thigh arising from branches of the superficial femoral artery (Fig. 2a, 2b, and 2c). A chest x-ray was normal. There was no tachycardia.

On August 5, 1959, the arteriovenous fistula, which involved the sartorius and adductor muscle mass, was excised. All of the sartorius and a portion of the adductor muscle mass was excised en-bloc. The superficial femoral artery was preserved. The patient had an uneventful postoperative course and was last seen in 1963 with no evidence of recurrent fistula.

Klippel-Trenaunay Syndrome

Twenty-five patients appeared to correspond to the vascular anomaly described by Klippel and Trenaunay in 1900 (Table 3). Most of these have been seen and followed at the University of Michigan Medical Center. There were 13 males and 12 females. The average age was 22.3 years, the youngest 2 years and the oldest 51 years. The average age at which medical attention was sought was 11.6 years. In all patients one or more of the manifestations was noted at birth or shortly thereafter. Often they appeared when the child first started to walk. In most, the first manifestation was the cutaneous hemangioma or the varicosities. The varicosities often became more prominent when the child spent more time upright. The presenting symptoms were: varicose veins and hemangioma—8; varicose veins, hypertrophy and hemangioma—5; hypertrophy and hemangioma—4; hemangioma alone—4; varicose veins and hypertrophy—3; varicose veins alone—1.

In all instances the lesion was confined to one extremity. In 23 the lower extremity was involved; the left lower extremity in 10 and the right lower extremity in 13. In one patient the right upper extremity was involved and in a second the left upper extremity.

TABLE 3. *Summary of Patients*

Case	Age	Sex	Extremity	Onset	Hemangioma	Length	Girth
1. F. N.	23	M	Lt. lower	Birth	Extensive	Greater	Greater
2. L. B.	11	F	Rt. lower	Birth	Extensive	Greater	Greater
3. P. B.	10	M	Rt. lower	Birth	Moderate	Greater	Less
4. M. C. H.	34	F	Lt. lower	Birth	Extensive	Shorter	Less
5. E. S.	17	M	Rt. upper	Birth	Extensive	Greater	Greater
6. D. D.	26	F	Lt. lower	Birth	Extensive	Equal	Greater
7. R. W.	26	M	Rt. lower	Birth	Extensive	Equal	Greater
8. E. G.	41	F	Lt. lower	Birth	None	Greater	Greater
9. M. F.	30	F	Rt. lower	Birth	None	Greater	Greater
10. F. F.	38	M	Lt. lower	Birth	Present	Greater	Greater
11. W. W.	21	M	Rt. lower	Birth	Extensive	Equal	Greater
12. F. M.	34	M	Lt. lower	Birth	Extensive	Greater	Equal
13. G. P. B.	27	F	Rt. lower	Birth	Extensive	Equal	Greater
14. V. P.	29	M	Rt. lower	Birth	Present	Equal	Equal
15. D. B.	6	F	Lt. lower	Birth	None	Shorter	Greater
16. G. J.	52	F	Lt. lower	Birth	Extensive	Greater	Greater
17. P. S.	9	M	Rt. lower	Birth	Extensive	Greater	Greater
18. R. S.	51	M	Rt. lower	Birth	Extensive	Greater	Greater
19. P. D. P.	20	F	Rt. lower	Birth	Extensive	Greater	Greater
20. J. C.	18	M	Rt. lower	Birth	Extensive	Shorter	Equal
21. M. C.	8	M	Lt. upper	Birth	Moderate	Equal	Greater
22. L. P.	2	F	Rt. lower	Birth	Extensive	Greater	Greater
23. P. M.	17	F	Lt. lower	Birth	Extensive	Shorter	Greater
24. L. P.	24	M	Rt. lower	Birth	Extensive	Equal	Less
25. D. M. B.	5	F	Lt. lower	Birth	Extensive	Shorter	Less

In one patient a faint hemangioma was present in the contralateral leg, and in another, ipsilateral arm and trunk hypertrophy was present. In two cases minimal varicosities of far less magnitude were present in the contralateral lower extremity. In one, the patient was extremely obese, and in another the lesions appeared during pregnancy.

The classic triad of cutaneous hemangioma, varicose veins and soft tissue and bony hypertrophy was present in 16 patients. The hemangioma was present in 22 and absent in three. When present, it was of the so-called "port-wine" variety and confined to the skin. The hemangioma was patchy in distribution and generally extended over the ipsilateral buttock and trunk to the thoracic region. It frequently stopped abruptly at the midline with a sharp linear border. The lesions varied in hue from a fairly bright reddish-orange to a faint erythematous blush. In one, on the contralateral leg, the color was much fainter and much less extensive. Varicosities were present in all cases. The varicosities were usually extensive and not the commonly seen patterns of varicose veins. The veins usually were present over extensive areas of the extremities and one could not discern a greater or lesser saphenous system. Bony hypertrophy and concomitant increase in length was present in 13 cases. In five the affected extremity was shorter and in seven the leg length was equal. Soft tissue hypertrophy with increase in girth of the affected extremity was present in 18 cases. In four, the leg girth was less than the opposite limb and in three the girth was equal. In 11 paients pedal edema was present. In no instance did a thrill, murmur, hum, pulsating

Varicosities	Edema	Venogram	Arteriogram	Treatment	Results
Marked	3+	Absent deep calf veins	Normal	Ligation and stripping	Worse
Marked	0	Absent deep veins	—	Epiphysodesis	Worse
Very marked	0	Absent deep veins	Normal	None	No change
Marked	3+	Absent deep veins	Normal	Ligation and injection	Worse
Moderate	0	Absent deep veins	—	Exploration, no basilic or brachial vein	No change
Marked	3+	Absent deep veins	Normal	Ligation and stripping	Worse
Marked	0	Absent deep veins	Normal	Ligation and stripping	Worse
Marked	1+	—	—	Ligation	Worse
Marked	0	Absent superficial femoral vein	Normal	Excision, x-ray to calf	Worse
Marked	2+	—	—	Ligation and stripping	Worse
Marked	0	—	Normal	None	Worse
Marked	2+	—	—	Ligation and stripping	No change
Marked	2+	—	—	Ligation and stripping	Worse
Moderate	2+	—	—	Ligation and stripping	Worse
Marked	0	? constricting band	—	Exploration, no superficial femoral vein	Worse
Marked	3+	—	—	Ligation and stripping	Worse
Moderate	0	Absent deep veins	—	None	No change
Marked	1+	Absent deep veins	Normal	Ligation and stripping	Worse
Mild	0	—	—	Epiphysodesis	No change
Marked	0	Absent deep veins	—	Ligation and excision	Worse
Mild	0	—	Normal	None	No change
Marked	0	—	—	None	No change
Marked	2+	Stenosis, left iliac vein	Normal	Relieve stenosis	Improved
Marked	2+	Absent deep calf veins	Normal	Ligation and excision	Worse
Mild	0	—	—	None	No change

veins, cardiomegaly or heart failure suggest the presence of an arteriovenous fistula. Branham's sign was not elicited in any. Venous oxygen saturation was measured in three patients. In two it was decreased on the affected side and in one it was equal in both extremities.

Arteriograms were obtained in 11 patients and were all normal with no evidence of anomalous arteriovenous connections. Venograms, performed in 14 cases, showed absent deep veins in the popliteal region or superficial femoral region in 13. Occasionally, absent deep veins were observed in the calf. In no instance was a completely normal deep venous system visualized. In one patient obstruction of the common iliac vein was noted.

Fourteen patients were treated with multiple venous ligations and stripping. Thirteen were made worse with the onset of edema or an increase in edema already present. An almost universal observation was the rapid appearance of recurrent varicosities, frequently with associated leg discomfort; in one instance there was no change. Two patients underwent surgical exploration which confirmed the absence of deep veins. Two patients underwent successful epiphysodesis with correction of limb length inequality. Ten patients received no surgical treatment directed towards the varicose veins. In seven it was felt that there was no change in the varicosities during the period of observation. Three showed increased prominence of their varicosities. However, the increase in the prominence of the varicose veins in these instances was much less than that seen in the patients treated by multiple ligations and stripping. In no instance was pedal edema seen in those patients who

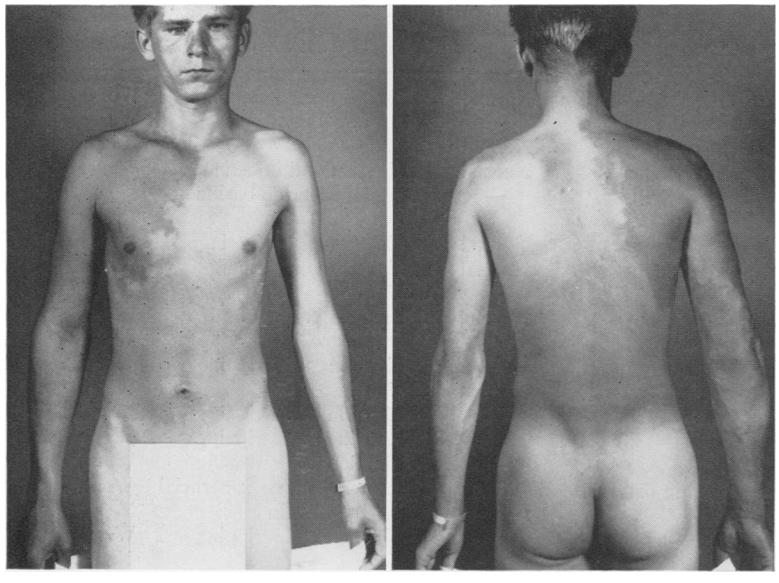

FIG. 3a, 3b. Case 4. E. S. Seventeen-year-old boy with cutaneous hemangioma, varicose veins and increased length and girth of right upper extremity since birth. No evidence of arteriovenous fistula. Exploration of upper arm revealed normal arterial system and absence of basilic and brachial veins with an intact axillary-subclavian system.

became worse without treatment. In one patient stenosis of the common iliac vein was relieved with improvement.

The following case reports illustrate typical patients with the Klippel-Trenaunay Syndrome:

Case 4. E. S. *Klippel-Trenaunay Syndrome* (Fig. 3a, 3b).

A 17-year-old boy was seen at the University of Michigan Medical Center in August, 1962 with a history of a birth mark present in the right upper extremity since birth. For as long as he could remember, his right arm and hand had been larger and stronger and dilated veins had been present. He was a healthy appearing adolescent. Pulse was 60 and regular. Positive findings were confined to the right upper extremity where a "port-wine" type cutaneous hemangioma extended over the entire arm, shoulder, right lower neck and the upper half of the right chest. Peripheral pulses were equal with no bruit or thrill. There were varicosities along the lateral and medial aspects of the forearm and arm, posterior axillary fold and along the lateral aspect of the chest overlying the area of the latissimus dorsi muscle. The right arm appeared grossly larger in length and circumference, measuring 1¼ inches longer and 2¼ inches greater in circumference at the mid-arm and 2 inches greater at the mid-forearm. The right wrist was ¾ inch greater in girth. Venograms (Fig. 3c) revealed

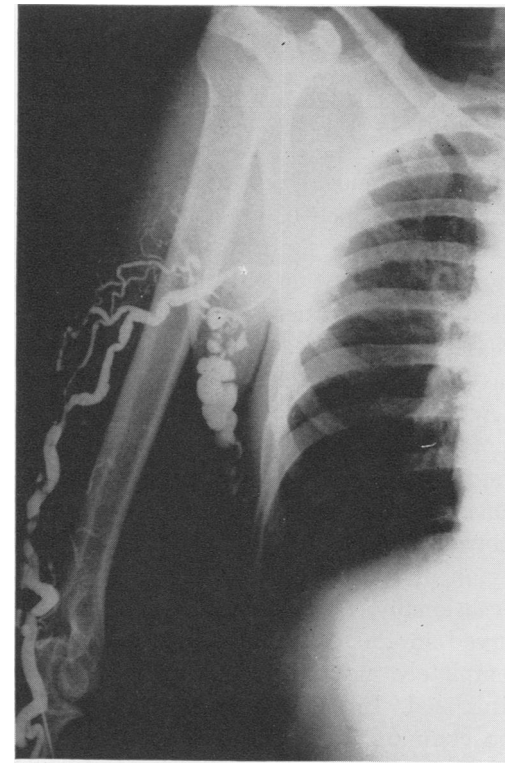

FIG. 3c. Case 4. E. S. Photograph of venogram of right upper extremity demonstrating absence of deep veins in the arm and superficial varicosities.

a large superficial vein, measuring 1 cm. in diameter, draining over the lateral aspect of the arm and across the posterior axillary fold and ending in a large mass of varicosities over the lateral chest wall. The upper arm was explored and no basilic or brachial veins could be found. Operating venogram showed a myriad of tiny veins confluescing in a patent axillary subclavian system. No further treatment was given.

Case 5. D. B. *Klippel-Trenaunay Syndrome.*

A 6-year-old girl was first seen at the University of Michigan Medical Center in October 1960 with a history of dilated veins of the left lower extremity and left buttock noted 2 weeks after birth. They had progressed until 1 year of age and had remained stable thereafter. The mother had likewise noted an increased girth of the affected extremity. Examination revealed numerous dilated subcutaneous veins and varicosities over the entire left lower extremity and buttock (Fig. 4a, 5a). No hemangioma was present. Peripheral pulses were normal with no bruit or thrill. The left leg was ¼ inch shorter in length and 1 inch greater in circumference at the thigh and ⅛th inch greater at the calf. The right lower extremity was entirely normal. There was no cardiomegaly or evidence of congestive heart failure. Teleoroentgenograms revealed the left lower extremity to be shorter by 2.1 cm. Venograms (Fig. 6a, 6b) revealed a patent ileo-femoral system but suggested the presence of a constricting band over the left femoral vein.

On November 15, 1960, operative exploration of the left femoral fossa was undertaken. The arterial system was normal with no evidence of an arteriovenous fistula. The common femoral iliac veins appeared normal. No deep venous channel distal to the common femoral vein could be identified. No constricting band was present.

The patient has been followed closely through July 1969. There has been essentially no change in the varicosities (Fig. 4b, 5b). She remained asymptomatic until 1969 when she had slight fatigue and discomfort at the end of the day following athletic participation. There has been no pedal edema. Leg length inequality has gradually decreased over the intervening years. Teleoroentgenograms revealed a 1.1 cm. difference (left shorter) in 1962 and since 1968 the leg length has been equal. Thigh girth on left has remained increased. The child has otherwise grown and developed normally and participates actively in athletics in school. In July 1969 elastic hose were prescribed for athletic participation.

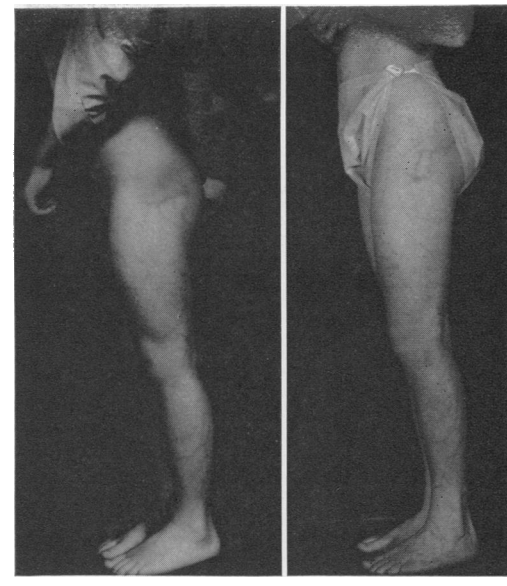

FIG. 4a, 4b. Case 5. D. B. Photograph of patient D. B. in 1960 (4a) and 1967 (4b) demonstrating minimal varicosities with essentially no progression. No hemangioma present.

Case 6. L. B. *Klippel-Trenaunay Syndrome* (Fig. 7a, 7b, 8).

An 11-year-old girl was first seen at the University of Michigan Medical Center in September, 1951 at 17 hours of age with a history of a birth mark and enlargement of the right lower extremity noted since birth. Her mother's pregnancy and delivery had been uncomplicated. On Examination the infant was healthy. In the right lower extremity there was a cutaneous hemangioma over the entire leg, right buttock, right labia and anterior abdomen to the midline. The right thigh was ½ inch greater, and right calf ⅜ inch greater in circumference, then the left. Leg length was equal. No treatment was given. The child was followed yearly and varicosities of the right lower extremity slowly appeared and became more prominent. At 1 year of age the right lower extremity was ¼ inch longer. In 1959 at age 8 years the varicosities were marked and elastic hose were prescribed. At this time the thigh was 3¼ inches greater and the calf 2 inches greater in circumference. Teleoroentgenograms revealed an increase of 3 cm. in bone length on the right. The hemangioma was unchanged. Leg length on the right was greater by 1¼ inches with a pelvic tilt and compensatory scoliosis. A ½ inch sole and ¾ inch heel lift was prescribed. In 1960 the leg length inequality increased to 1½ inches and teleoroentgenograms revealed a 3.3 cm. discrepancy. Be-

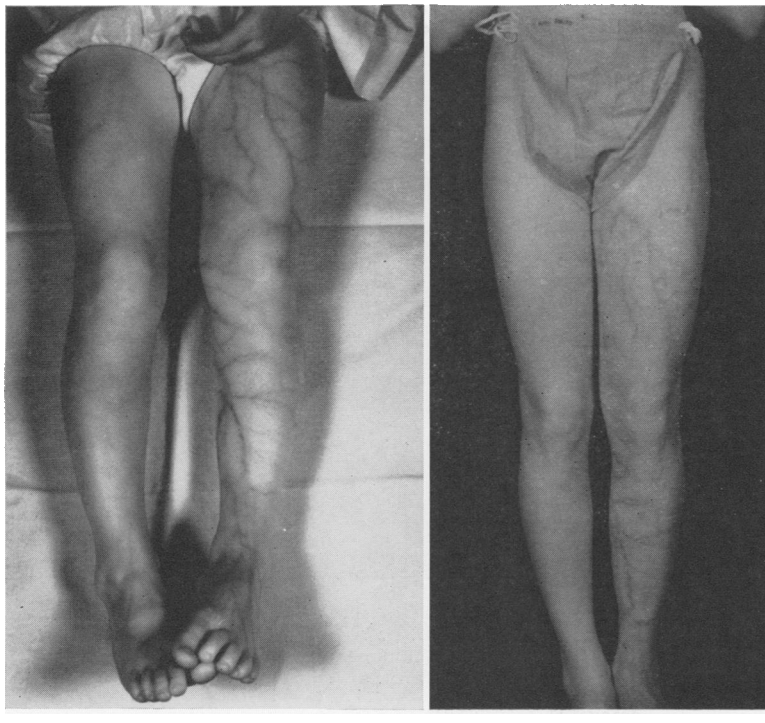

FIG. 5a, 5b. Case 5. D. B. Photographs of patient D. B. in 1960 (5a) and 1967 (5b) utilizing infrared technic. No significant progression of varicosities.

cause of this an epiphysodesis of the proximal tibial and fibular epiphysis was performed on the right. In September, 1962 the leg length inequality was 2 inches and the bone length inequality was 3.1 cm. The right thigh girth was greater by 3 inches and the calf greater by 2 inches. The right foot required a shoe 3 sizes wider and 1 size longer in addition to a ¾ inch heel and ½ inch

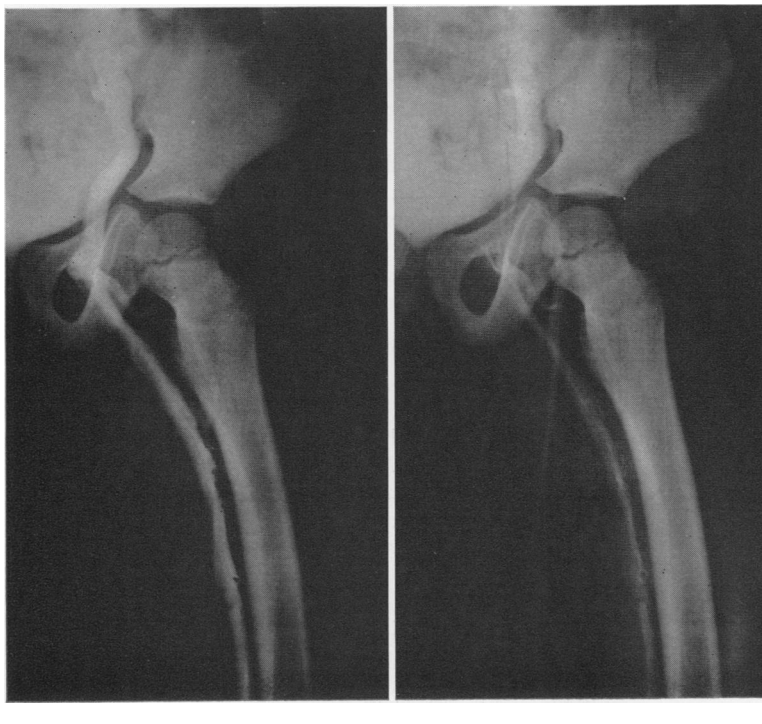

FIG. 6a, 6b. Case 5. D. B. Venograms suggesting presence of constricting band at common femoral-superficial femoral vein junction. Exploration revealed absence of superficial femoral and profunda femoris veins with only superficial veins joining the common femoral.

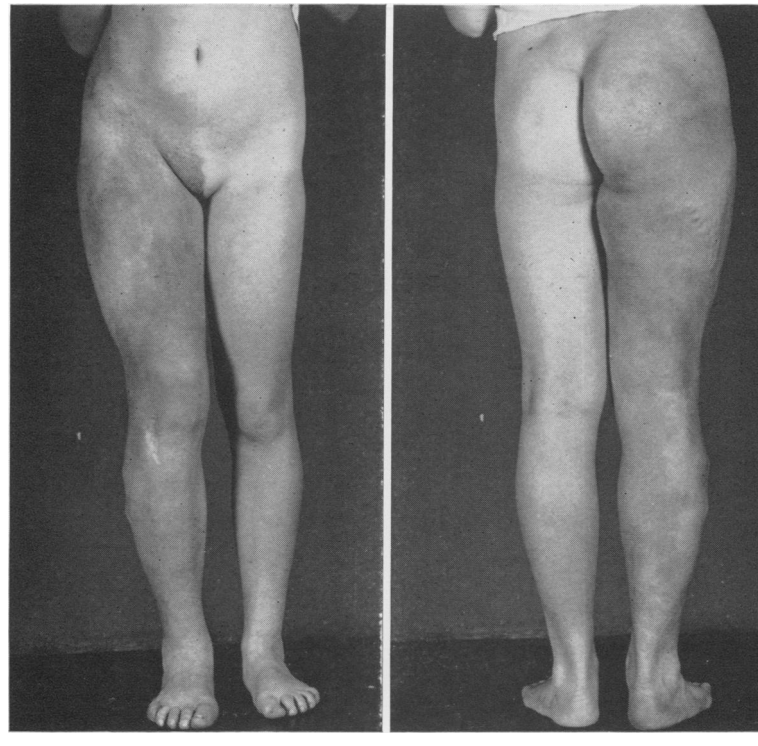

FIG. 7a, 7b. Case 6. L. B. Eleven-year-old girl with varicosities, cutaneous hemangioma and bony and soft tissue hypertrophy in right lower extremity noted at birth. Minimal progressive enlargement of varicosities with no treatment.

sole lift. Varicosities had become more prominent. No edema was present. At no time was a thrill or bruit evident nor was there clinical evidence of arteriovenous fistula. Venograms (Fig. 9a, 9b) showed absent deep venous channels in the popliteal and thigh region.

By December 8, 1965 the limb length was equal. When last seen in March 1967 limb length remained equal and teleoroentgenograms revealed the uninvolved leg (left) to be greater in length by 1.1 cm. The pelvic tilt and scoliosis were no longer present and the sole and heel lift was no longer required. The right thigh and calf has remained greater in girth. The varicosities remain stable and the patient is asymptomatic.

Case 7. P. M. *Klippel-Trenaunay Syndrome* (Fig. 10a, 10b).

A 17-year-old girl was admitted to the University of Michigan Medical Center for the first time in August 1968 with a complaint of aching in the left calf and pedal edema of one years' duration. A cutaneous hemangioma and varicose veins had been noted in the left lower extremity since birth. The varicose veins had slowly progressed in severity in recent years. Examination revealed a healthy white female. There were marked varicosities over the entire left lower ex-

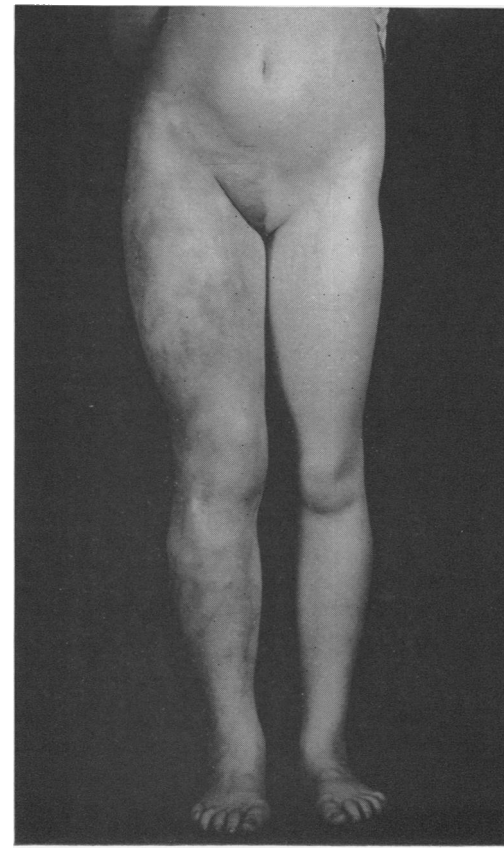

FIG. 8. Case 6. L. B. Infrared photograph demonstrating varicosities to better advantage

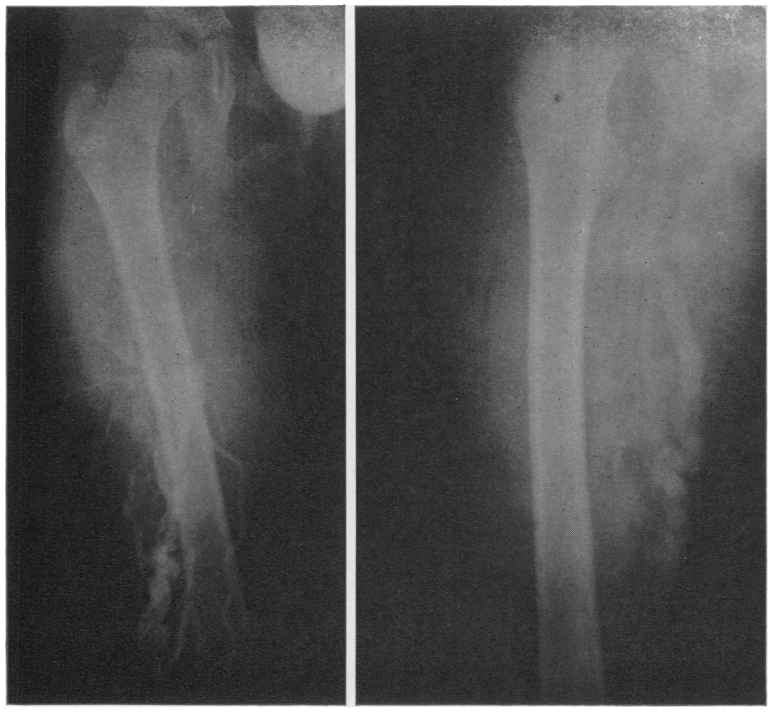

FIG. 9a, 9b. Case 6. L. B. Photographs of venogram demonstrating absent deep thigh veins and superficial varicosities.

tremity with an extensive "port-wine" type cutaneous hemangioma. The left leg was ½ inch shorter in length and 1½ inches greater in girth at the calf and thigh. Peripheral pulses were normal with no thrill or bruit present. The right leg was normal.

A chest x-ray showed no abnormalities. An arteriogram showed no evidence of an arteriovenous fistula. A venogram revealed superficial varicosities and patent deep veins in the calf and thigh (Fig. 11). In the left common iliac vein there was a high grade stenosis past which a catheter could not be advanced (Fig. 12a). The venous press at this point measured 17 cm. of water. Teleoroentgengrams revealed the bone length to be greater on the right by 1.8 cm. Venous oxygen saturation at the ankle on the left was 71.5% and on the right 76%.

On August 27, 1968 the left iliac was explored by operation. It was dilated and 3 times greater in size than the right. The left iliac vein was compressed by the right iliac artery. The aorta was more to the left of the inferior vena cava than normal accentuating the compression of the left iliac vein. The right iliac artery and the left iliac vein were freed of adherent fibrous tissue resulting in a noticeable decrease in the dilatation of the left iliac vein. A venotomy revealed no internal stenosis or septum. A portion of peri-

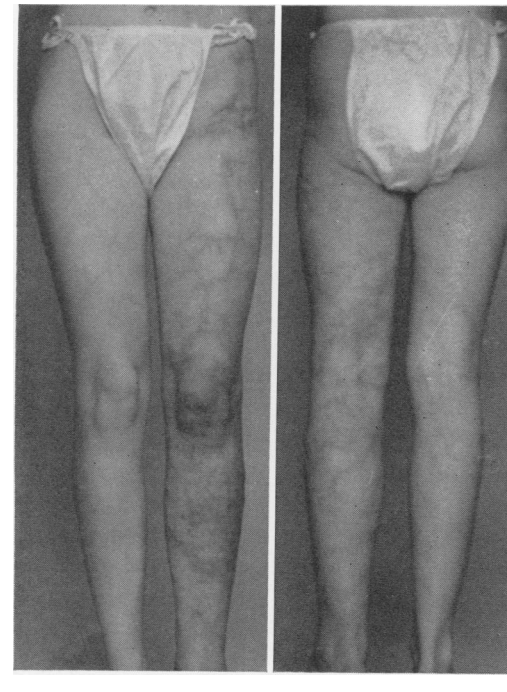

FIG. 10a, 10b. Case 7. P. M. Photographs of 17-year-old girl with cutaneous hemangioma and varicosities in left lower extremity since birth. The leg is greater in girth and slightly shorter in length.

toneum and adjacent soft tissue was interposed between the iliac artery and vein.

A postoperative venogram (Fig. 12b) revealed the stenosis to be gone. The venous press was equal in both common iliac veins and in the inferior vena cava (11 cm. water). The patient was seen 3 months postoperatively and was much improved.

Discussion

In a patient with an enlarged limb, varicosities, increased warmth, and a cutaneous hemangioma the presence of a congenital arteriovenous fistula or the Klippel-Trenaunay Syndrome should be considered. The presence of a thrill or bruit, Branham's sign, a pulsating mass or pulsating veins strongly suggests a congenital arteriovenous fistula. This can almost always be confirmed by arteriography. The absence of these additional signs suggests the Kippel-Trenaunay Syndrome. In such patients, angiography will show a normal arterial system and absence or obstruction of a portion or all of the deep venous system. In no patient with the Klippel-Trenaunay Syndrome in this series was there clinical or arteriographic evidence of an arteriovenous fistula.

Localized hypoactive congenital arteriovenous fistulas usually appear as a mass in a male and are most often in the lower extremity. They can be readily dealt with by local excision. Though they may recur, repeated excision eventually will result in cure.

Diffuse active congenital arteriovenous fistulas most commonly occur in the upper extremity in females. This is similar to the experience of Malan and Puglionisi.[7] The treatment of active diffuse fistulas is diffcult and seldom satisfactory. Most often the fistulous connections arise from branches of main arteries. This lends itself to excision of the branches with preservation of the main channel. Ligation of the main trunk has proven to be unsatisfactory. If sufficient collateral exists, the fistula persists. If collateral around the site of ligation is inadequate distal ischemic symptoms occur or amputation will be required. Surgical excision should consist of an en-bloc excision of the mass of tissue containing the fistulous connections with preservation of the main arterial channels. Because of the extensiveness of the fistulous connections, excision can seldom be complete and probably should be done in stages. Moreover, small fistulous connections may be overlooked at the time of initial excision and will later enlarge and become clinically apparent. This repeatedly occurred in patients in this series. Such fistulous connections can then be excised at a later date.

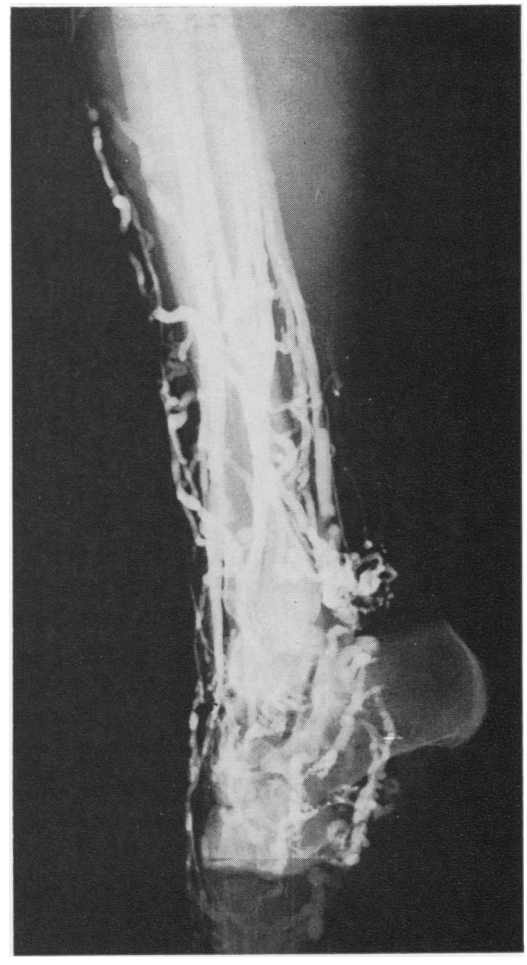

FIG. 11. Case 7. P. M. Photograph of peripheral venogram demonstrating superficial varicosities and patent deep venous system.

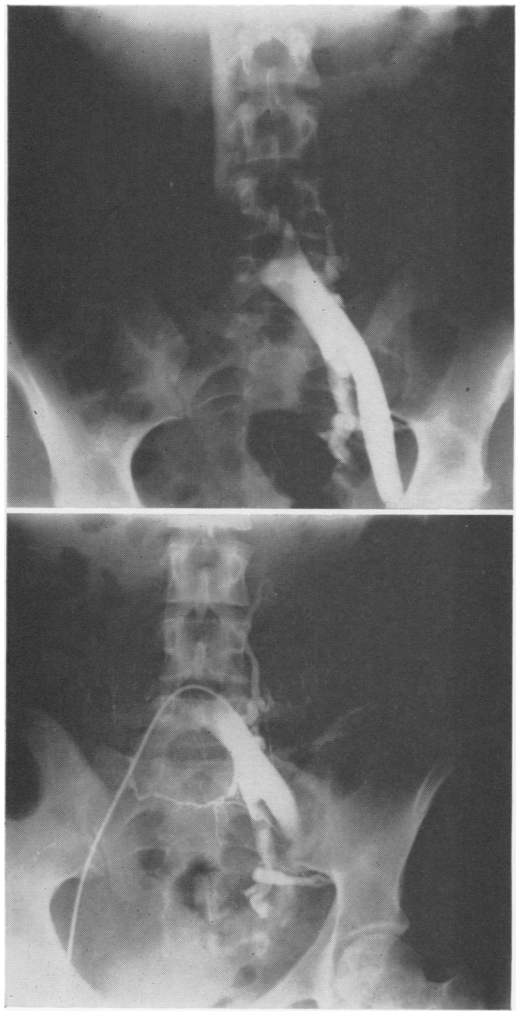

FIG. 12a. Case 7. P. M. Photograph of preoperative iliac venogram demonstrating high grade stenosis of the proximal left common iliac vein.

FIG. 12b. Case 7. P. M. Photograph of postoperative iliac venogram demonstrating relief of stenosis of left common iliac vein.

Only one patient had significant symptoms, and the mere presence of a congenital arteriovenous fistula should not be reason enough to recommend surgical removal. In contrast to patients with traumatic arteriovenous fistulas, patients with congenital arteriovenous fistulas seldom develop cardiac difficulties, degenerative arterial changes, or aneurysmal dilatation of the proximal arterial system. Therefore, surgical intervention should be undertaken only for compelling symptoms. When undertaken, excision should be conservative and the presence of recurrent or residual fistulous connections should not be considered a poor result. These patients tolerate residual fistulas with very few symptoms. Often times complete eradication of the fistulous connections can only be accomplished by amputation. When the fistulous connections involve bone this is always the case. Conservative sub-total excision should be attempted before resorting to amputation.

In patients with varicose veins, cutaneous hemangioma, and bony and soft tissue hypertrophy confined to one extremity, in whom there is no clinical or arteriographic evidence of an arteriovenous fistula, venograms should be obtained before any surgical therapy is contemplated. In such patients agenesis or obstruction of a portion of the deep venous system will likely be found. This was true in the 14 patients in this series who underwent venography. Similar observations have been made by Servelle,[15, 16] Poulet, and Ruff,[14] Malan and Puglionisi[6] and Olivier.[10] Commonly the superficial femoral, popliteal, or tibial veins are absent or obstructed. In Servelle's[15] experience obstruction is far more common than agenesis. In this series, agenesis of a portion of the deep venous system was encountered in 13 patients and obstruction in only one. It is important that the entire deep venous system be visualized including the iliac vessels. In the one patient with a surgically remediable lesion due to deep venous obstruction, the site of obstruction was the proximal common iliac vein. Iliac vein involvement has also been reported by Martorell,[8] Foster and Kirtley,[8] Peck[13] and Oda *et al.*[9]

In patients with obstruction of the deep venous system an attempt should be made to relieve the obstruction. This was done in one patient with gratifying results. Similar improvement following relief of an obstructing lesion has been described by Servelle.[16]

The patients with agenesis of the deep venous system who underwent ligation and stripping almost all were made worse by the procedure with increase in leg discomfort, recurrence of varicosities, and worsening of pedal edema. This has also been observed by Foster and Kirtley [3] and Malan and Puglionisi.[6]

Such patients are best treated by elastic hose for external support. In 10 patients so treated, there was no change in seven and minimal progression in three. Only two patients had limb lengthening that required surgical intervention and in both of these epiphysodesis was successful in reducing the leg length inequality.

Summary

During the past 15 years, 14 patients with congenital arteriovenous fistulas and 25 patients with the Klippel-Trenaunay Syndrome without evidence of arteriovenous fistula were seen and treated at the University of Michigan Medical Center.

By means of careful clinical evaluation and angiography, these two groups of patients can be defined.

Five patients with localized hypoactive arteriovenous fistulas were treated successfully by local excision.

Nine patients with diffuse active congenital arteriovenous fistulas were treated by subtotal excision with good results in eight. Surgical management of these patients should be conservative because of the minimal symptoms and lack of long term morbidity due to residual fistulous connections.

In 25 patients with unilateral varicose veins, cutaneous hemangioma of the "port-wine" variety and bony and soft tissue hypertrophy present from birth, no evidence of congenital arteriovenous fistula could be demonstrated. In 14 patients undergoing venography, agenesis or obstruction of the deep venous system was demonstrated. Operative intervention in these patients should only be undertaken to relieve deep venous obstruction if present or to correct leg length inequality. Removal of superficial varicosities is contraindicated and will result in worsening of existing symptoms.

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