Lymphology 30 (1997) 98-104

*LYMPHSPIRATION*

# LYMPHATIC DYSFUNCTION IN CONJUNCTION WITH DYSREGULATED HYPERDYNAMIC BLOOD FLOW (THE HYPERSTOMY SYNDROME)

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Forty years ago Pratesi (1) and Malan (2) called attention to a clinically unusual condition of dysregulated blood flow in the extremities characterized by premature venous filling often in conjunction with segmental venous obstruction. Pratesi termed this phenomenon "hyperstomy syndrome," but a better designation is probably "angiodyskinesia." This phenomenon was subsequently studied and reviewed in considerable detail by Amir-Jahed (3), Dramez et al (4), and Haimovici (5). In general, this vascular entity was invoked to explain distal extremity ischemia in the absence of arterial occlusion and was examined in some detail experimentally by DeBakey et al (6), who referred to the syndrome as "borrowing-lending" or "hemometakinesis."

During the course of treating lymphatic circulatory disorders, we encountered several patients who angiographically displayed this hyperdynamic dyskinetic blood flow picture and clinically had marked peripheral lymphedema. Indeed some of these patients had undergone previous unsuccessful operative procedures for control of lymphedema (Thompson, Charles/Kondoleon). Once the dysregulated arteriovenous flow was pinpointed, however, operative ligation of the offending arteries not only resulted in long-term control of edema but restored these severely handicapped individuals to a productive livelihood.

Although I (RCM) devoted a monograph (written in Portuguese) to this entity in 1976 (7), we now highlight the clinical background and long-term outcome in four such patients.

## *CASE REPORTS*

1. A 20-year old man with primary lymphedema presented with persistent right lower leg lymphedema after a failed Thompson operation *(Fig. 1)*. Posterior tibial arteriography using hand injection showed abnormal muscular arterial branches in the calf with premature filling of varicose soleus veins with valvular incompetence *(Fig. 2)*. At operation, the varicose veins were resected along with three large gastrocnemius arterial feeders. Although he wore an elastic stockinette thereafter, follow up after several years showed he is free of edema *(Fig. 1)*.

2. A 46-year old woman developed chronic left leg lymphedema from multiple episodes of cellulitis/lymphangitis. Initially, debulking was done along with split thickness skin grafts (Charles/Kondoleon operation) but repeated infections thereafter led to marked incapacitation for 15 years *(Fig. 3)*.

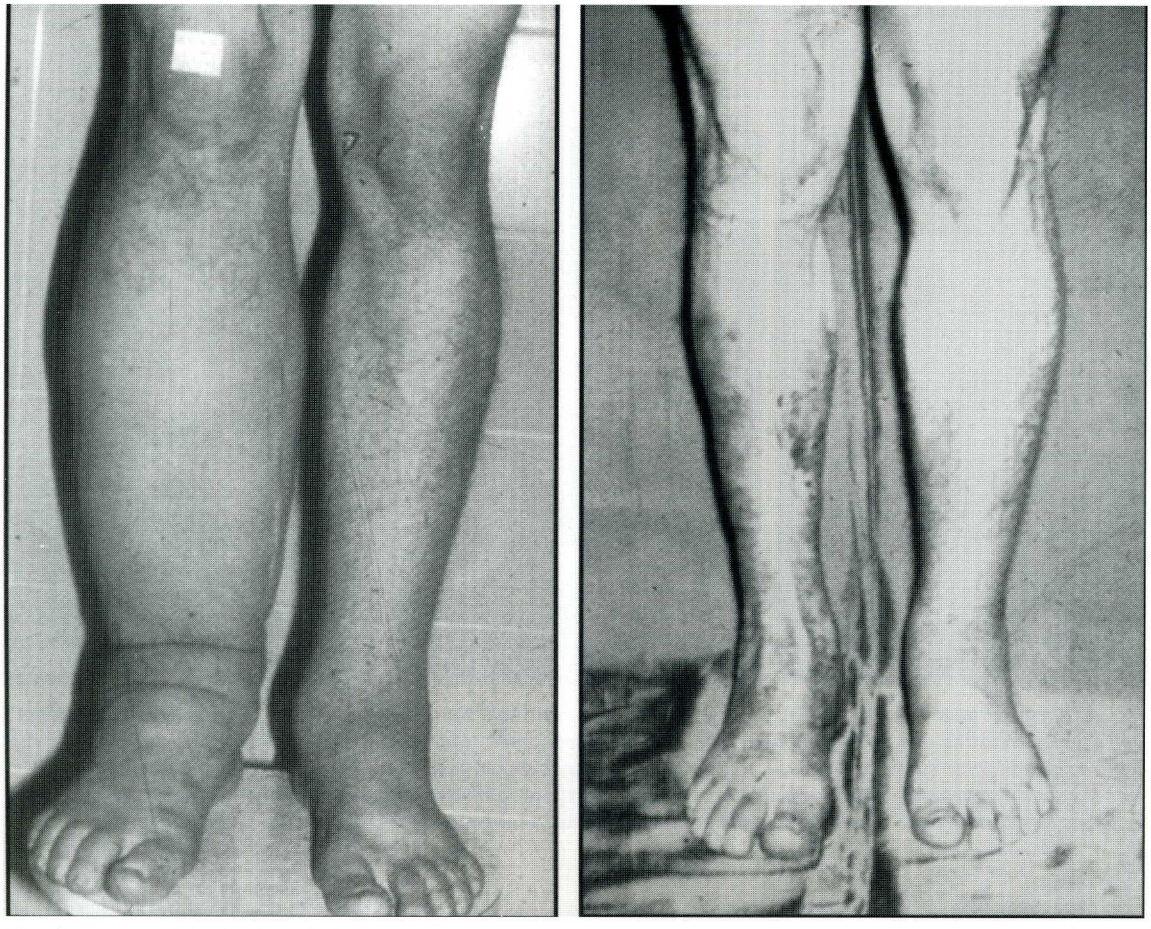

*Fig. 1. LEFT—right leg lymphedema after a failed Thompson procedure (notice the vertical scar on the medial portion of the lower leg). RIGHT—appearance several months after ligation of gastrocnemius arterial feeders and removal of varicose soleus veins.*

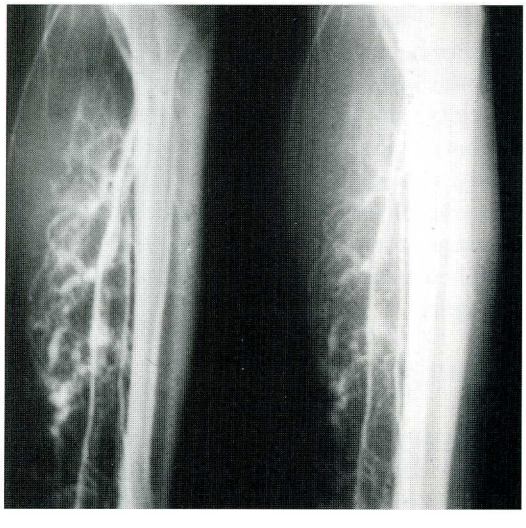

*Fig. 2. Popliteal arteriogram of patient 1 (Fig. 1) demonstrating abnormal arterio-venous communications with premature filling of soleus veins with "lakes." After ligation of these abnormal arterial branches, lymphedema regressed (Fig. 1R).*

Phlebography of the left leg showed virtual occlusion (compression) by the bulky thigh mass *(Fig. 4)*. Femoral arteriography depicted six anomalous large arterial branches within the adductor (Hunter) canal *(Fig. 5)*. After ligation of these abnormal arterial branches, the thigh mass softened/shrunk considerably, and after debulking and resurfacing, the leg was markedly improved *(Fig. 3)*. Repeat phlebography showed the femoral vein now

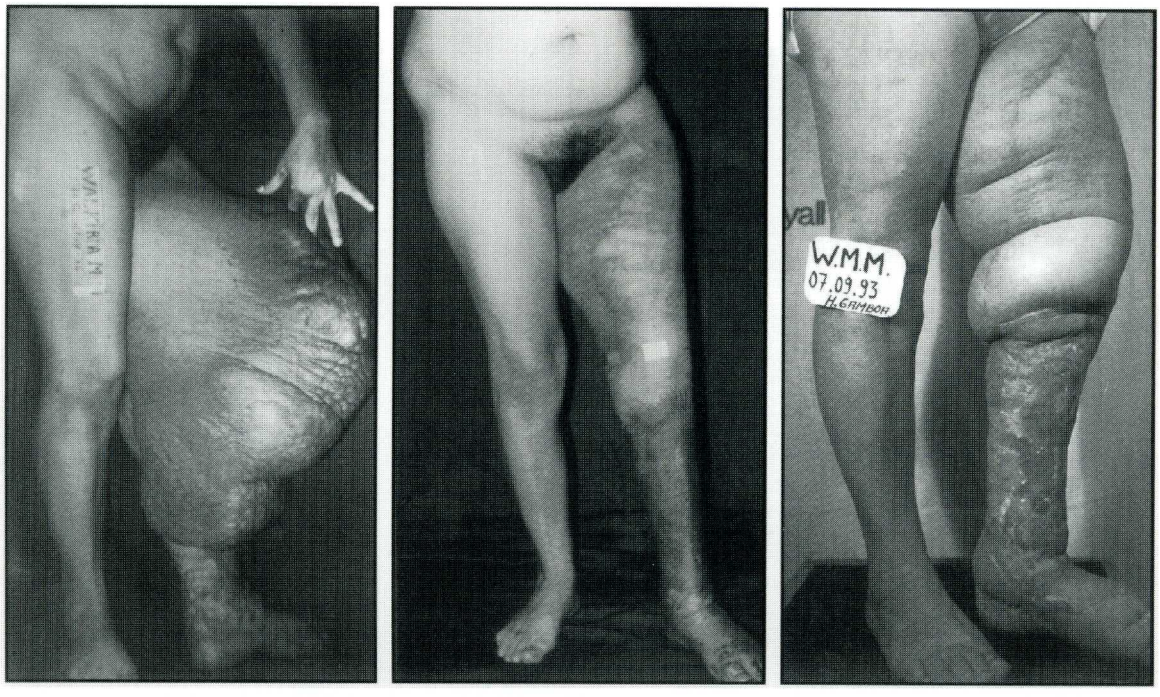


*Fig. 3. Forty-six year old woman with massive thigh mass and scaling skin changes of the lower leg after multiple episodes of cellulitis and debulking by a Charles/Kondoleon operation (left). After ligation of anomalous arteries within the adductor canal (see Fig. 5), and excision of the softened mass with resurfacing of the skin contour she was markedly improved (middle). Nineteen years later, despite some worsening in foot and thigh edema (right) she remains functionally active without elastic stockinette, drug therapy or other non-operative treatment.*

to be widely patent *(Fig. 4)*. Nineteen years later without elastic stockinette support or other treatment, she showed some progression of lymphedema *(Fig. 3)* but is nonetheless ambulating and fully active.

3. A 43-year old man had intractable lymphedema with skin ulceration for 41 years after traumatic injury to the foot and ankle at 2 years of age *(Fig. 6)*. Conventional oil contrast lymphography depicted mild hypoplasia. Phlebography was unremarkable but popliteal arteriography showed several anomalous large muscular branches *(Fig. 7)*. Histopathology of a biopsy of the skin ulcer showed squamous cell carcinoma (Marjolin ulcer). After perfusion for 7 days with antineoplastic agents into the anterior tibial artery, the edematous soft tissue with residual tumor was excised and the anomalous arterial branches including the feeder to the malignant ulcer was interrupted. Nine years later, the patient remains well *(Fig. 6)*.

4. An adult man developed severe right leg lymphedema secondary to repeated episodes of cellulitis *(Fig. 8—left)*. Distal arteriography *(Fig. 9)* showed three anomalous branches of the posterior tibial artery with premature, intense filling of varicose soleus veins. After ligation of these abnormal arterial branches (Dr. Josias), lymphedema was permanently corrected *(Fig. 8—right)*.

*COMMENT*

Dysregulated arteriovenular flow was initially described to explain peripheral ischemia in the absence of arterial occlusion

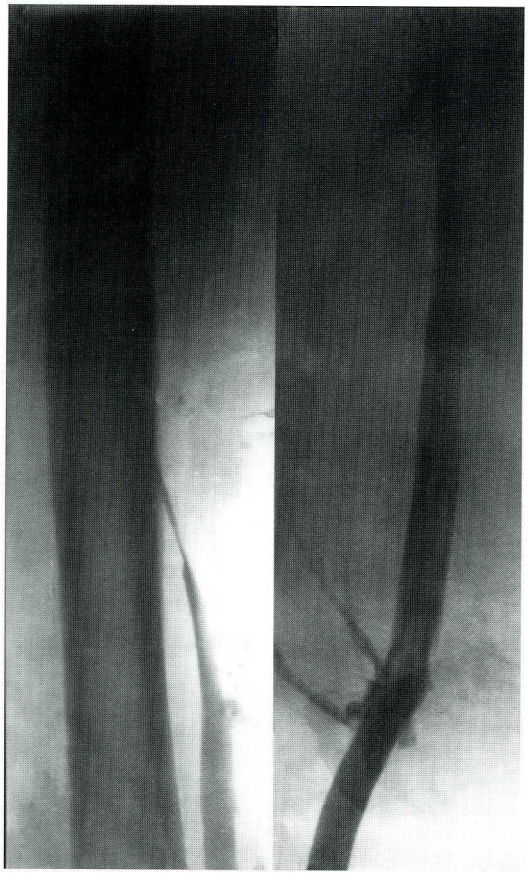

*Fig. 4. Phlebogram of the left leg (Patient 2) showing virtual occlusion of the femoral vein (left) and restoration of patency (right) after physically displacing the thigh mass laterally (see Fig. 3).*

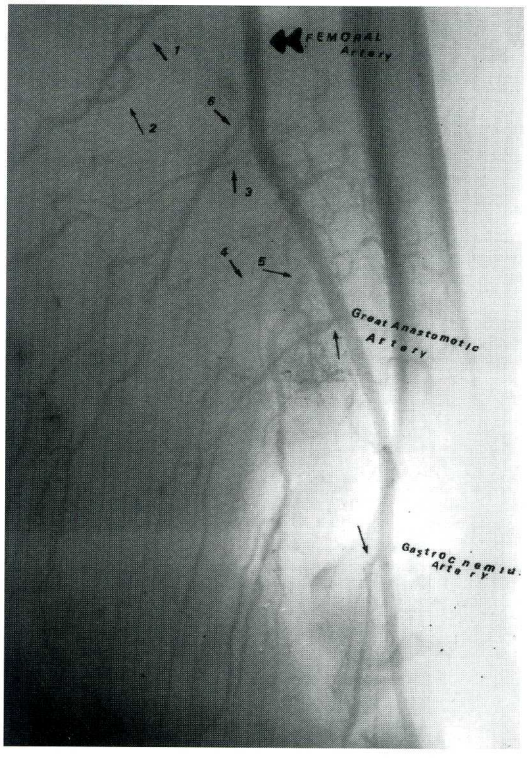


*Fig.5. Femoral arteriogram showing several anomalous muscular arterial branches derived from the superficial femoral artery (Patient 2).*

(1,2). Later the phenomenon was expanded to encompass certain venous and lymphatic disorders. Two patterns emerged—primary, that is, unknown cause, and secondary, that is, in conjunction with documented vascular perturbation such as postphlebitic syndrome, lymphostasis, and obliterative atherosclerosis. Trauma, fractures, or other causes of venous occlusion also are associated with the development of anomalous arterial branches and dysregulated flow.

In the patients presented here, lymphatic dysfunction was aggravated by arterial circulatory dyskinesia, and treatment of lymphedema was successful only in conjunction with ligation of the anomalous feeding arteries. Whereas the true significance of these vascular findings needs further investigation, it may be appropriate to reexamine the morphologic and functional status of the peripheral blood circulation along with lymphatic dynamics in primary and secondary lymphedema syndromes.

*ACKNOWLEDGMENT*

The authors thank Drs. Marlys and Charles Witte for their invaluable assistance in the writing of this text.

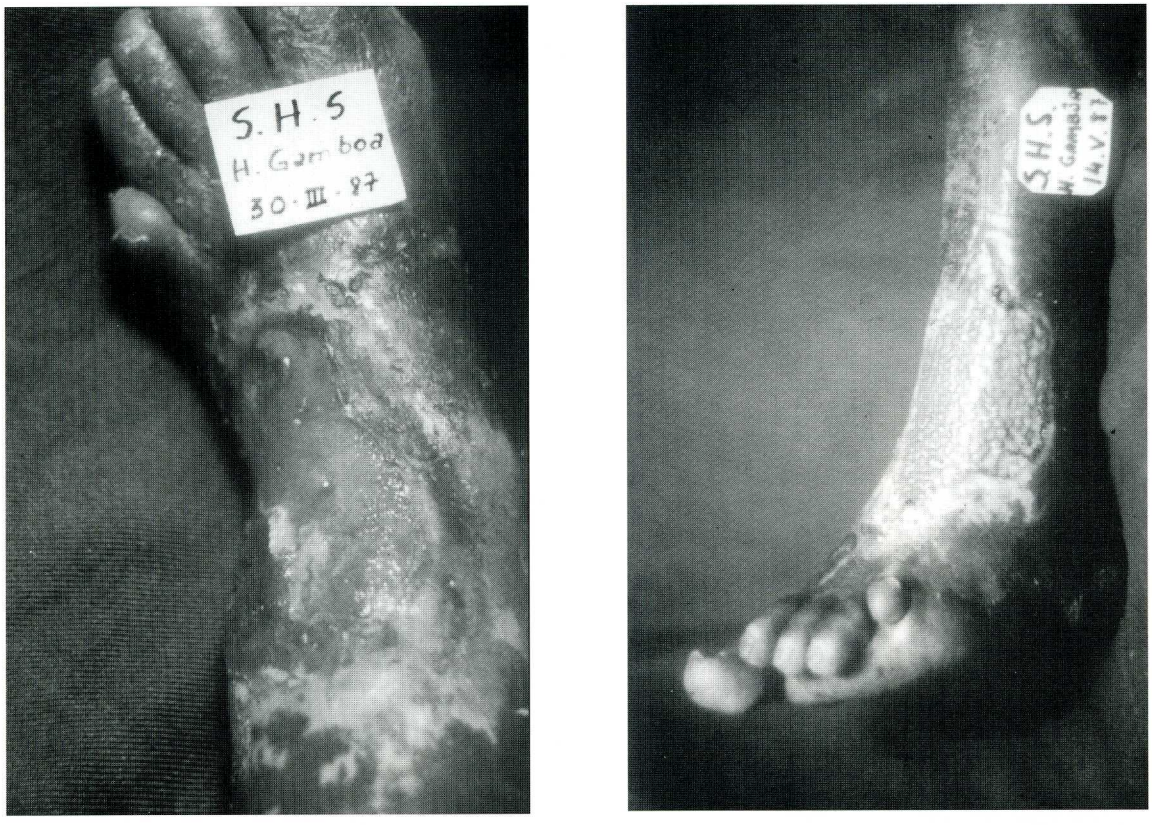


*Fig. 6. Left—intractable lymphedema with Marjolin ulcer (squamous cell carcinoma). Right—appearance after arterial perfusion with chemotherapy, ligation of the anomalous arterial branches (see Fig. 7), excision of the ulcerated tumor and placement of a split thickness skin graft.*

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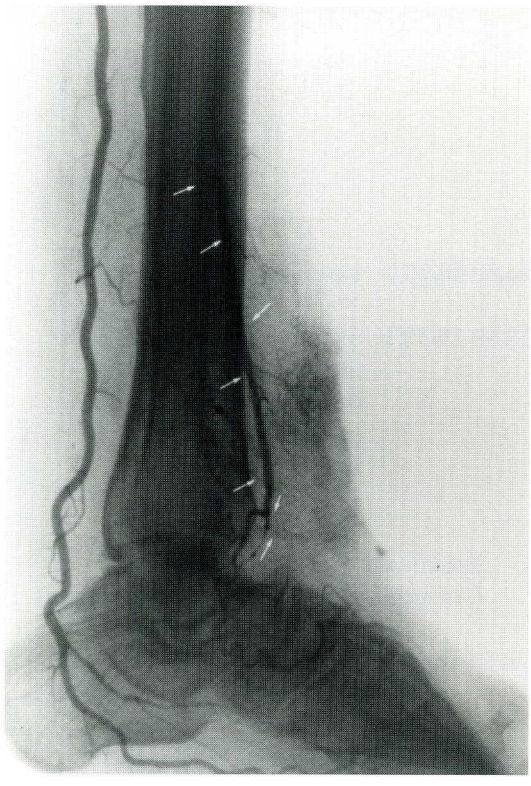

*Fig. 7. Arteriogram showing occlusion of the distal anterior tibial artery with multiple anomalous arterial branches (white arrows) (Patient 3).*

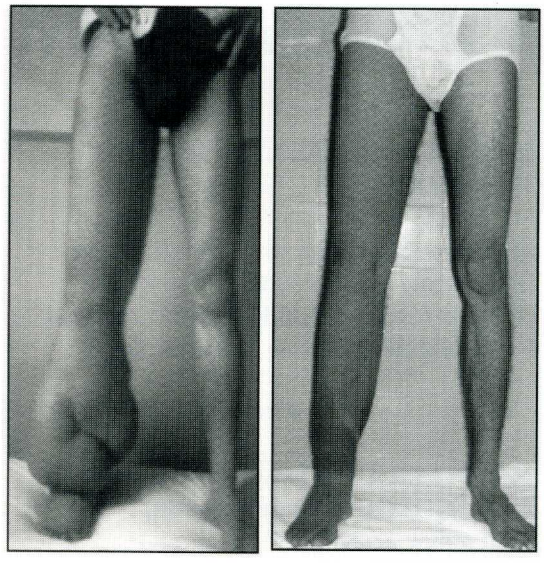

*Fig. 8. Young man with secondary lymphedema before (left) and after (right) ligation of anomalous branches of the posterior tibial artery (see Fig. 9).*

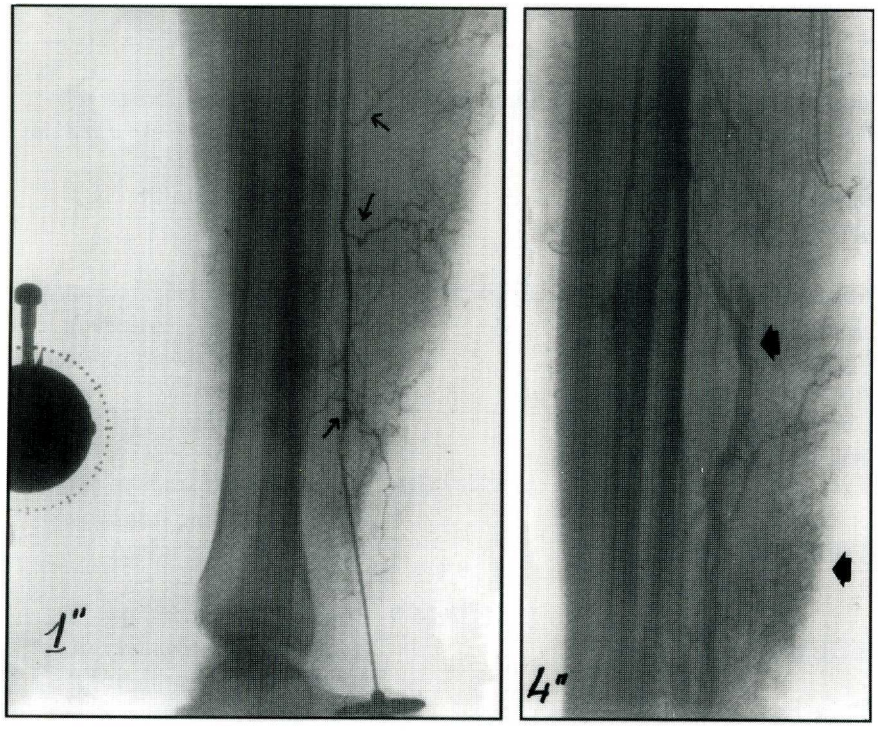


*Fig. 9. Distal arteriogram of patient shown in Fig. 9 demonstrating anomalous branches of posterior tibial artery (small arrows—left) with premature and intense filling of varicose soleus veins (large arrows—right).*