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## CLINICAL STUDIES IN PRIMARY LYMPHEDEMA

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Results of clinical observations of 120 subjects suffering from primary lymph edema of lower extremities, were presented. Particular attention was paid to the relationship between the degree of edema and lymphographic picture, to the lymphangitis complicating edema and to the coexistence of morbid venous lesions. No parallelism between radiologic picture and clinical signs was found. The highest percent of lymphangitis was found in a group of patients with the so-called aplasia of lymphatic vessels. Morbid venous lesions were found only in a negligible percent of patients being under observation. The latter lesions do not seem to cause the edema.

In this presentation results of further studies of patients with primary lymphedema will be described (3), and the following questions will be answered:

1. Is there any relationship between the intensity of lymphedema and lymphangiographic findings?
2. How often does lymphangitis occur in different groups of lymphedema?
3. What is the incidence rate of thrombophlebitis in primary lymphedema?

### METHODS

Examinations were carried out in 120 patients with the clinical diagnosis of primary lymphedema divided into 4 groups according to the lymphangiographic findings. Group 1 (so-called aplasia) consisted of 46 patients (37.4%), group 2 (so-called hypoplasia) of 16 patients (13%), group 2b (so-called obstructive hypoplasia) of 24 patients (19.5%), group 3 (hyperplasia) of 9 patients (7.3%), and group 4 of 25 patients (25%) with normal lymphangiogram.

Clinically lymphedema was classified according to the intensity and stability of swelling into 5 groups. Lymphangiograms in each of these groups were thoroughly evaluated.

The incidence rate of lymphangitis was assessed in each group basing on the data from the past history.

In 50 patients phlebography of the affected extremity, pelvis and inferior vena cava was performed. The rate of postthrombophlebitis changes in different groups of lymphedema was assessed.

RESULTS

1. Relationship between the intensity of lymphedema and lymphangiographic findings.

The intensity of edema in different lymphangiographic groups was evaluated and results are presented in Fig. 1. Clinically edema was classified into 5 groups as in Table I.

Table I

| Group | Part of limb involved | Type of edema   |
|-------|-----------------------|---|
| I     | foot                  | soft, subsiding after night rest                              |
| II    | foot, calf            | soft, diminishing after night rest                            |
| III   | foot, calf            | hard in the foot, diminishing slightly in the calf after rest |
| IV    | whole limb            | hard, stable  |
| V     | only calf             | stable  |

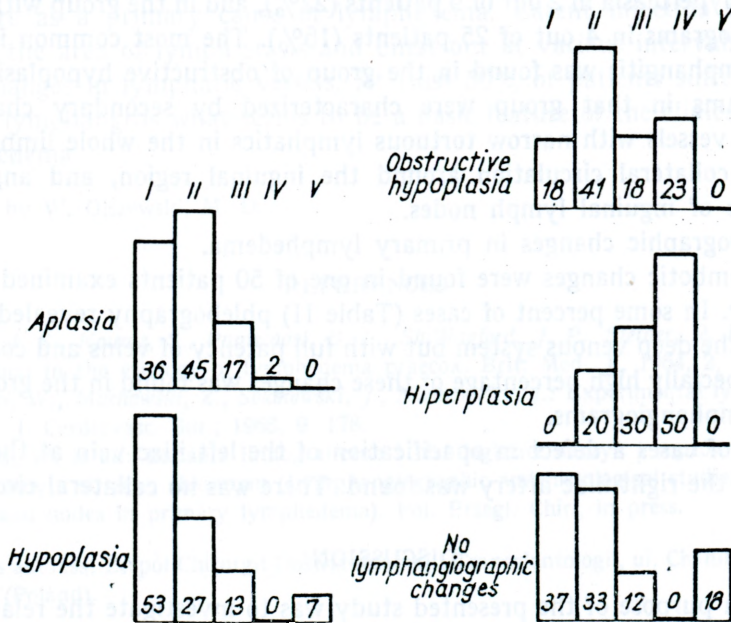


Fig. 1. Extent and stability of edema in different groups of lymphedema. Numbers indicate the percentage of patients in various groups classified as on Table I. I — edema limited to the foot, II — foot and calf (pitting), III — foot and calf (hard, stable), IV — involving the whole limb, V — involving only calf.

In group 1 (aplasia) swelling of the extremity was limited to the foot and lower part of the calf in 81% of cases. There was no case with edema affecting the whole limb. In group 2 (hypoplasia) edema was also limited to the foot and lower part of the calf. Still on lymphangiography a patent, single lymphatic vessel could be seen in the calf. In group 2b (obstructive hypoplasia) 18% of patients suffered from edema of the foot, 59% of foot and calf, and 23% of the whole limb. In group 4 all patients had advanced edema involving the whole limb in 50% of cases, never limited to the foot. In group 4 with normal lymphangiograms usually foot or foot and part of the calf were affected.

Table II

|                                     | Aplasia | Hypoplasia | Obstructive hypoplasia | Hyperplasia | Normal |
|-------------------------------------|---------|------------|------------------------|-------------|--------|
| Veins of the limb                   | 12.5    | 22         | 33                     | 0           | 56     |
| Pelvic veins and inferior vena cava | 0       | 10         | 17                     | 0           | 0      |

### 2. Incidence rate of lymphangitis in various groups of lymphedema.

Recurrent lymphangitis was found in the aplastic group in 21 patients (45%), in the hypoplastic group in 6 (37.5%), in obstructive hypoplasia in 16 patients (66%), in hyperplasia in 2 out of 9 patients (22%), and in the group with normal lymphangiograms in 4 out of 25 patients (16%). The most common incidence rate of lymphangitis was found in the group of obstructive hypoplasia. Lymphangiograms in that group were characterized by secondary changes in lymphatic vessels with narrow tortuous lymphatics in the whole limb, dermal backflow, collateral circulation around the inguinal region, and angora-like appearance of inguinal lymph nodes.

### 3. Phlebographic changes in primary lymphedema.

Postthrombotic changes were found in one of 50 patients examined phlebographically. In some percent of cases (Table II) phlebography revealed uneven outline of the deep venous system but with full patency of veins and competent valves. Especially high percentage of these changes was found in the group with normal lymphangiograms.

In 27% of cases a defect in opacification of the left iliac vein at the site of crossing of the right iliac artery was found. There was no collateral circulation.

## DISCUSSION

The main purpose of the presented study was to investigate the relationship between the intensity of swelling of the limb and the character of lymphangiographic changes. According to our findings there was no direct relationship

between the two. In the group of aplasia, where the pathological changes in lymphatics seemed to be most advanced, edema was limited to the foot and lower part of the calf. On the other hand in the group of obstructive hypoplasia, with multiple, narrow, tortuous lymph vessels and often a network of lymphatics in the foot edema affected most parts of the extremity. The largest swellings were observed in the group of hyperplasia, with multiple dilated vessels. In 20% of patients there was evident edema of foot with normal lymphangiographic appearance. In that last group major changes could probably be found more distally from the lymphatic collectors, namely in the lymphatic capillaries. Lack of direct relationship between the clinical state of edema and radiological findings was also found in our experimental studies (2).

Phlebographies performed in a large group of patients with lymphedema helped to exclude the venous factor in the mechanism of development of edema. Minor changes were found on phlebograms, never encountered in cases with the postthrombophlebitic syndrome.

A defect in opacification of the left iliac vein compressed by the right iliac artery was found in 27% of cases, but it seems to be a physiological finding. No correlation between the rate of incidence of compression of the left iliac vein and edema on the same side was found (1).

Recurrent lymphangitis of the edematous limb was typical of cases with a- and hypoplasia. Every recurrency brought about enhancement of edema. Basing on our data it has been impossible to establish infection of the lymphatic vasculature as a primary cause of lymphedema. Latent infection may still persist in the area of lymph stasis and burst out at various intervals causing further changes in lymphatic vessels. At least 50% of patients suffered from recurrent lymphangitis what seems to be a basic feature of the clinical picture of lymphedema.

Translated by W. Olszewski, M. D.

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