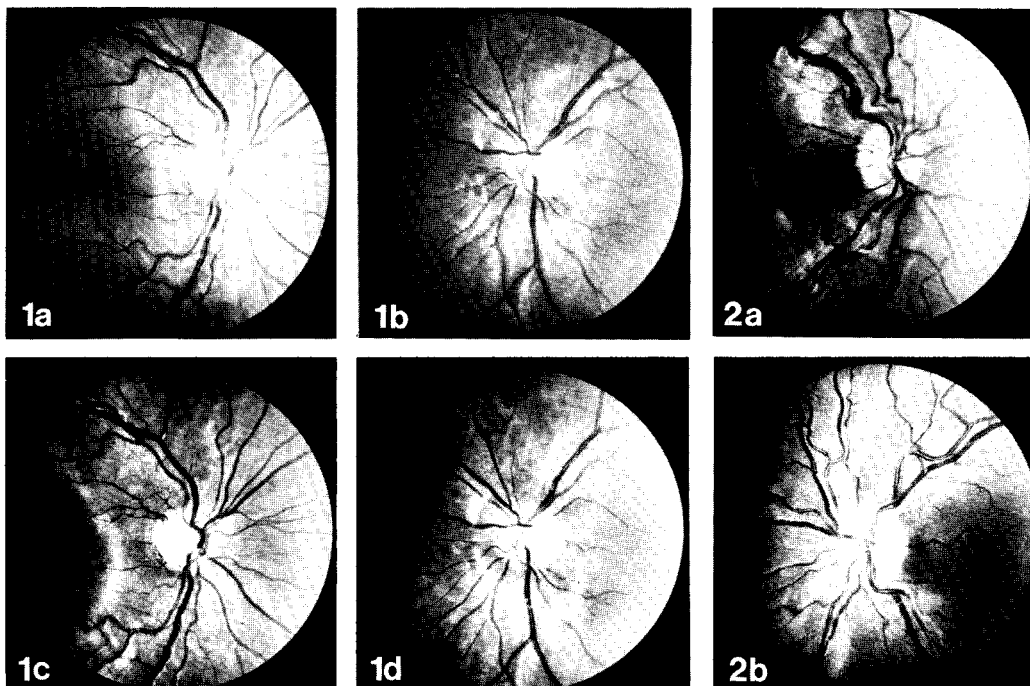


## LYMPHOGRAPHIA

### RETINAL EDEMA IN PATIENTS WITH GENERALIZED PRIMARY LYMPHEDEMA

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Fundoscopic pictures with Figures 1a-d.

Bilateral retina edema (Fig. 1a, b) in a seventeen year-old girl with generalized congenital primary lymphedema, pleural effusions and lymphostatic protein-losing enteropathy. Treatment by "decongestive physiotherapy" for three weeks not only decreased lymphedema of the soft tissues but also slightly improved retinal edema (Fig. 1c, d).

Figure 2a, 2b. Optic fundus of a 25-year-old woman with edema of the retina bilaterally associated with congenital lymphedema of all four extremities, the face and conjunctivae (Fig. 2a, 2b). Improvement followed physiotherapy (massage and compression).

## COMMENT:

Acute edema of the optic nerve, the retina and its blood vessels ("lymphostatic retinal hemangiopathy" develops after experimental blockade of cervical lymphatics (1). In the more chronic stage, deposition of collagen fibers is observed in dilated and edematous perivascular spaces of retinal blood vessels (2). Although there are no lymphatics in the brain, optic nerve and retina, these organs are dependent nonetheless on lymphatic drainage. Thus, numerous connections between the subarachnoid space via leptomenigeal sheets of brain nerves and adventitial "perivascular spaces" with lymphatics of the neck are well described (3). Direct communication between the subarachnoid space and the "perivascular space" of central blood vessels of the retina have also been documented (4).

One of us (MF) has emphasized (1) that with edema, one of three forms of lymphatic insufficiency exists: 1) lymphostatic edema or low output failure of the lymphatic system; 2) high lymph flow failure where lymphatics are intact and lymph flow is maximally accelerated but nonetheless overwhelmed by an excessive tissue fluid and protein lymph load; and 3) "safety valve insufficiency" where lymphangiopathy

predominates in combination with an increased lymphatic fluid load. Because prolonged retinal lymphostatic edema is likely to generate progressive fibrosis in the "perivascular spaces" early treatment is warranted. The results of these two patients suggest that decongestive physiotherapy is a useful therapeutic adjunct in this regard. Despite absence of lymphatics in the central nervous system, the retina of the eye is linked to the lymphatic network via the subarachnoid space and its various communications to cervical lymphatic trunks.

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