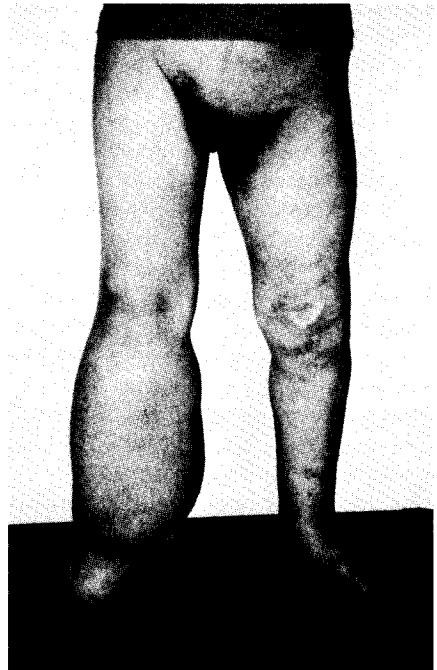
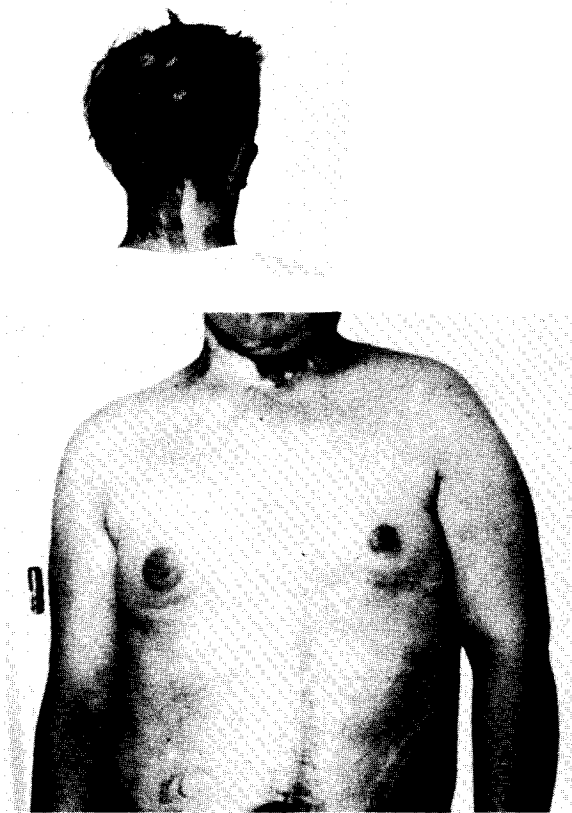


## LYMPHOGRAPHIA UNUSUAL CHYLOLYMPHEDEMA OF CONGENITAL ORIGIN

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*Fig. 1: Unusual appearance of a 12-year-old boy with congenital peripheral lymphedema. Note the broad skull, left-sided hyperpigmentation (neck, abdomen, and groin), bilateral gynecomastia, and hypoplastic genitalia.*

### CASE HISTORY

A 12-year-old boy developed progressive, intractable peripheral lymphedema (Fig. 1). Two years earlier he had undergone repair of a cardiac abnormality

and coarctation of the aorta with a decrease in systemic arterial pressure from 280/150 to 180/100mmHg. Thereafter peripheral edema rapidly increased. His skull was especially prominent, the hair thin with multiple sites of alopecia, the

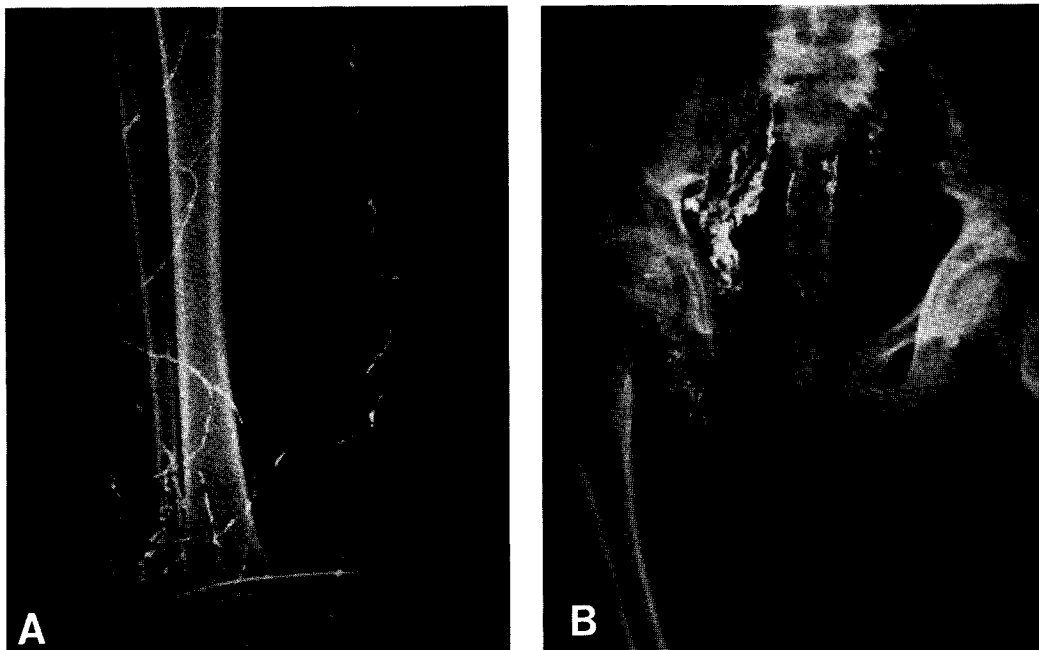


Fig. 2: Lymphangiography demonstrates marked lymphatic ectasia with reflux (A) and failure of contrast to pass beyond the true pelvis (B).

facies brachymorphic with an aged appearance and the abdomen pudgy. There was notable gynecomastia bilaterally as well as left sided hemipigmentation on the neck, left hemiabdomen, and left groin. The genitalia were also hypoplastic including unilateral cryptorchidism (Fig. 1). Bilateral lens dislocation with corneal thickening (MacCune-Albright Syndrome) was present. Overall intelligence seemed unimpaired.

Lymphangiograms (Fig. 2A) demonstrated abnormally ectatic, tortuous collectors and 15 hours later failed to visualize beyond the aortic-iliac region (Fig. 2B). A reduction Kondoleon-type operation was unsuccessful. Subsequently, 10 subinguinal lymphatic-venous anastomoses were attempted (Degni technique) but these thrombosed followed by chylous drainage from the wound. One year later, via an extraperitoneal approach, large retroperitoneal lymphatic lakes and matted channels (6-7mm in diameter) were en-

countered. Despite construction of numerous lymphatic-venous shunts and six months later radical excision of these lymphatic varices, severe peripheral edema persisted.

#### EDITORIAL COMMENT

*This patient demonstrates the common occurrence of peripheral lymphedema in association with a variety of other rare congenital anomalies (some based on well-worked-out chromosomal abnormalities) as well as the continued frustration of attempts to alleviate intractable lymphedema by both older and newer operative procedures. (CLW)*

#### REFERENCE

1. Degni, M: New technique of lympho-venous anastomoses for the treatment of lymphedema. *Vasa* 3 (1974), 479-483.