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AN IMAGING EVALUATION OF ANGIODYSPLASIA SYNDROMES*

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ABSTRACT

Current imaging techniques such as magnetic resonance, magnetic resonance angiography, computer tomography, ultrasound, plain x-rays, and lymphangioscintigraphy have enhanced the ability to define blood and lymph vascular malformations in more precise pathophysiologic terms. Not only can these imaging modalities distinguish arterial anomalies from lymphatic and venous angiodysplasia, but they also readily differentiate edema in the epifascial as opposed to the subfascial peripheral compartments. Moreover, visceral lymphangiectasia (e.g., chylous and non-chylous reflux), bone and muscle overgrowth, agenesis, and fat deposits can also be delineated. Clinical examples are provided including an algorithm for approaching these conditions.

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Imaging Algorithm for Evaluation of Lymphedema-angiodysplasia Syndromes

Modality	A-V	Venous	Lymph-edema	Lymph-angioma	Chylous	Lymphangio-myomatosis	KT-S	Lipedema
MRI		++++	++	++++	++++		++++	++++
MRA		+++		+++			++++	
LAS			++++	+++	++++		++++	++++
CA	++++							
CL					++			
CT			++			++++		
PLAIN		+					++	
US	+++	+++	++ (filariasis)					
CV		+++						

MRI = magnetic resonance images; MRA = magnetic resonance arteriography; LAS = whole body lymphangioscintigraphy; CA = conventional arteriography; CL = conventional lymphography; CT = computed tomography; PLAIN = radiographs; US = ultrasonography; CV = conventional venography; A-V = arteriovenous malformation; KT(-S) = Klippel-Trenaunay (-Serville) syndrome

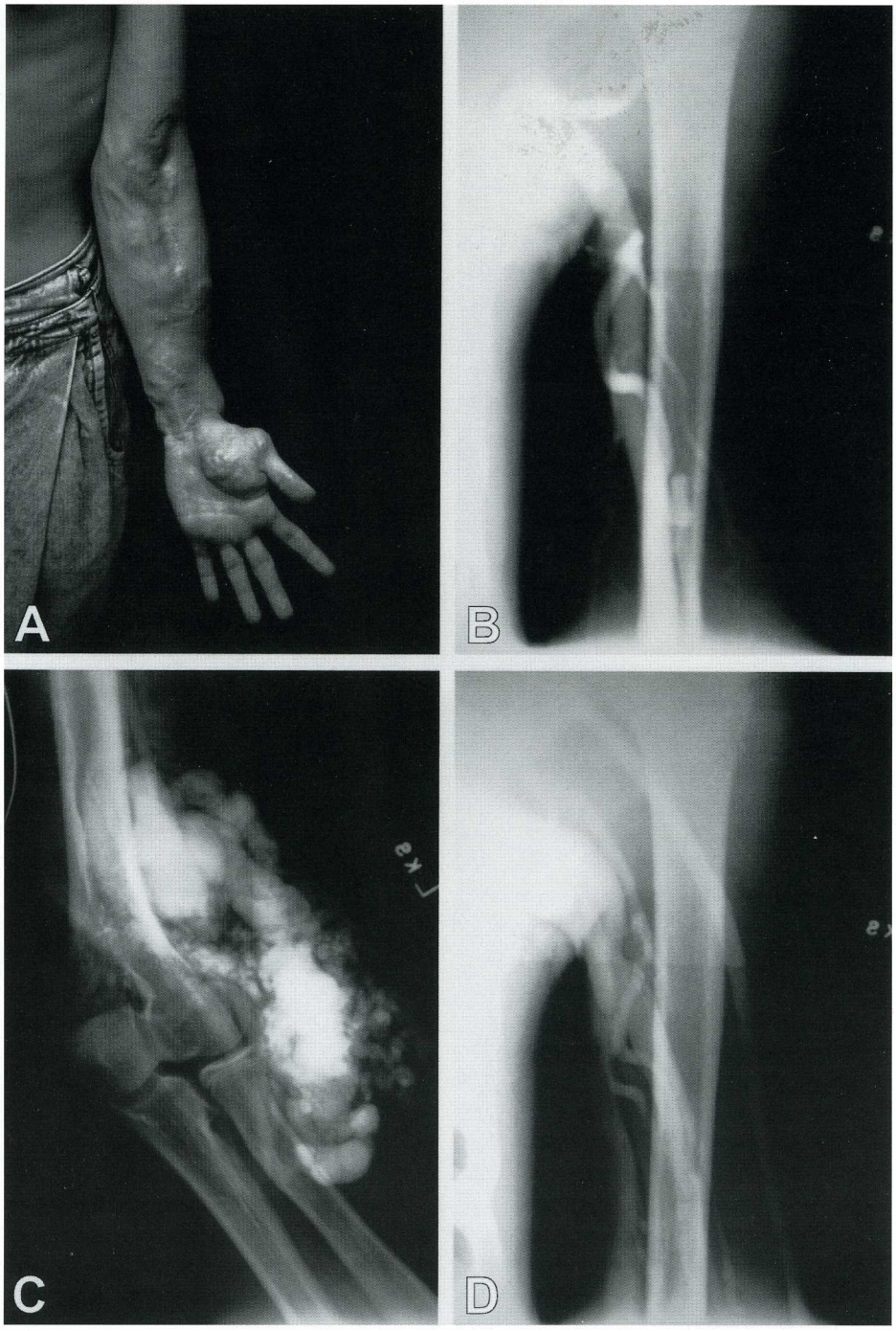

Fig. 1. 24 year-old man with major arteriovenous fistulae of the left arm. Note (A) diffuse venous collaterals with bulbous thenar eminence; (B) angiography shows a large axillary-brachial artery with (C) extensive arteriovenous malformation in the antecubital fossa and (D) huge axillary-brachial vein. Despite multiple operations/ embolizations, the patient ultimately developed acute congestive heart failure and forequarter amputation of the arm was done as a life-saving maneuver.

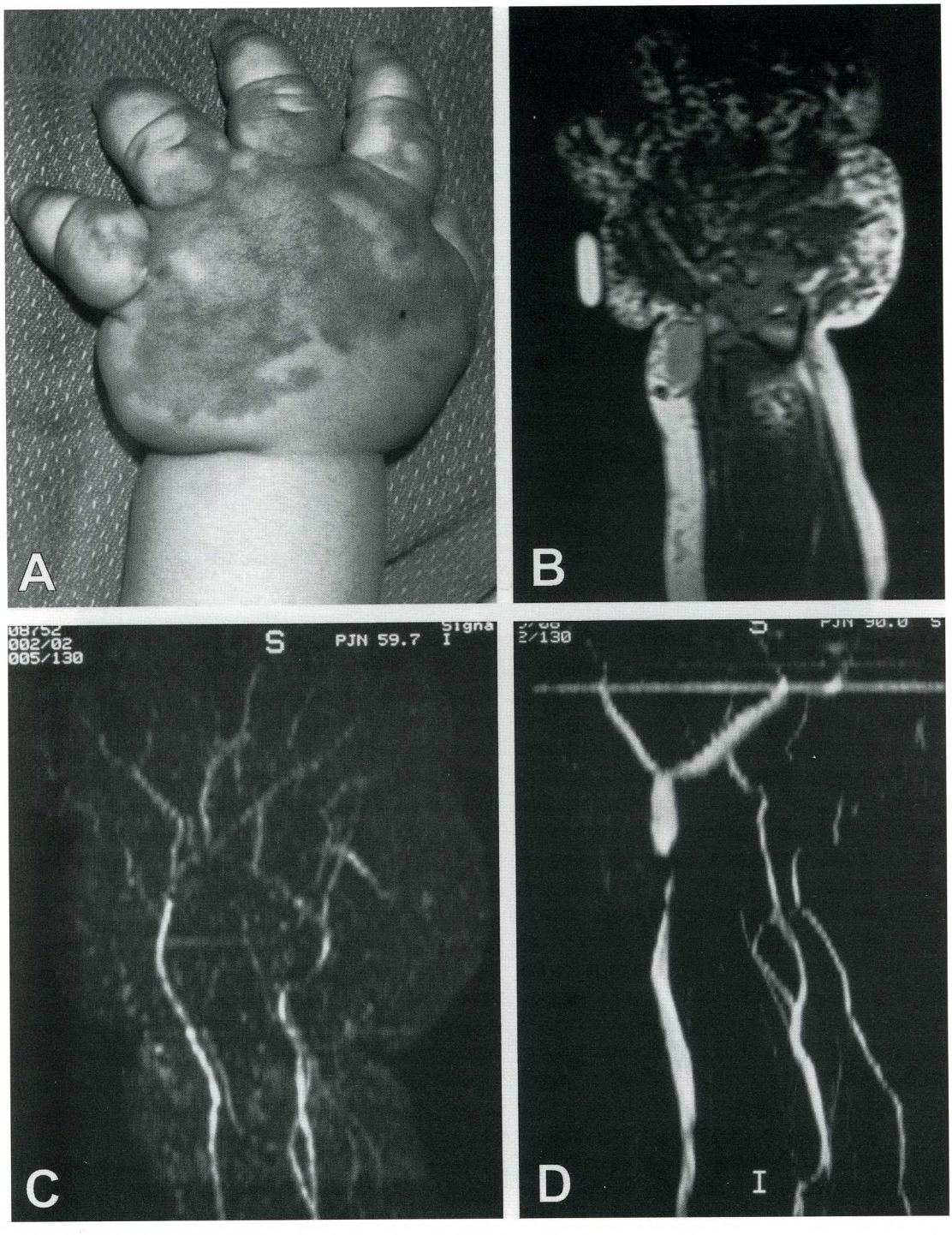

Fig. 2. 4 month old girl with Klippel-Trenaunay malformation of the left hand (A). Magnetic resonance (MR) imaging (B) shows extensive blood vascular anomaly of the soft tissues of the hand. MR angiovenography (C and D) shows intact arterial inflow without arteriovenous fistula (C) whereas numerous strictures of corresponding draining veins are evident (D). The child had a similar malformation of the right pectoral girdle.

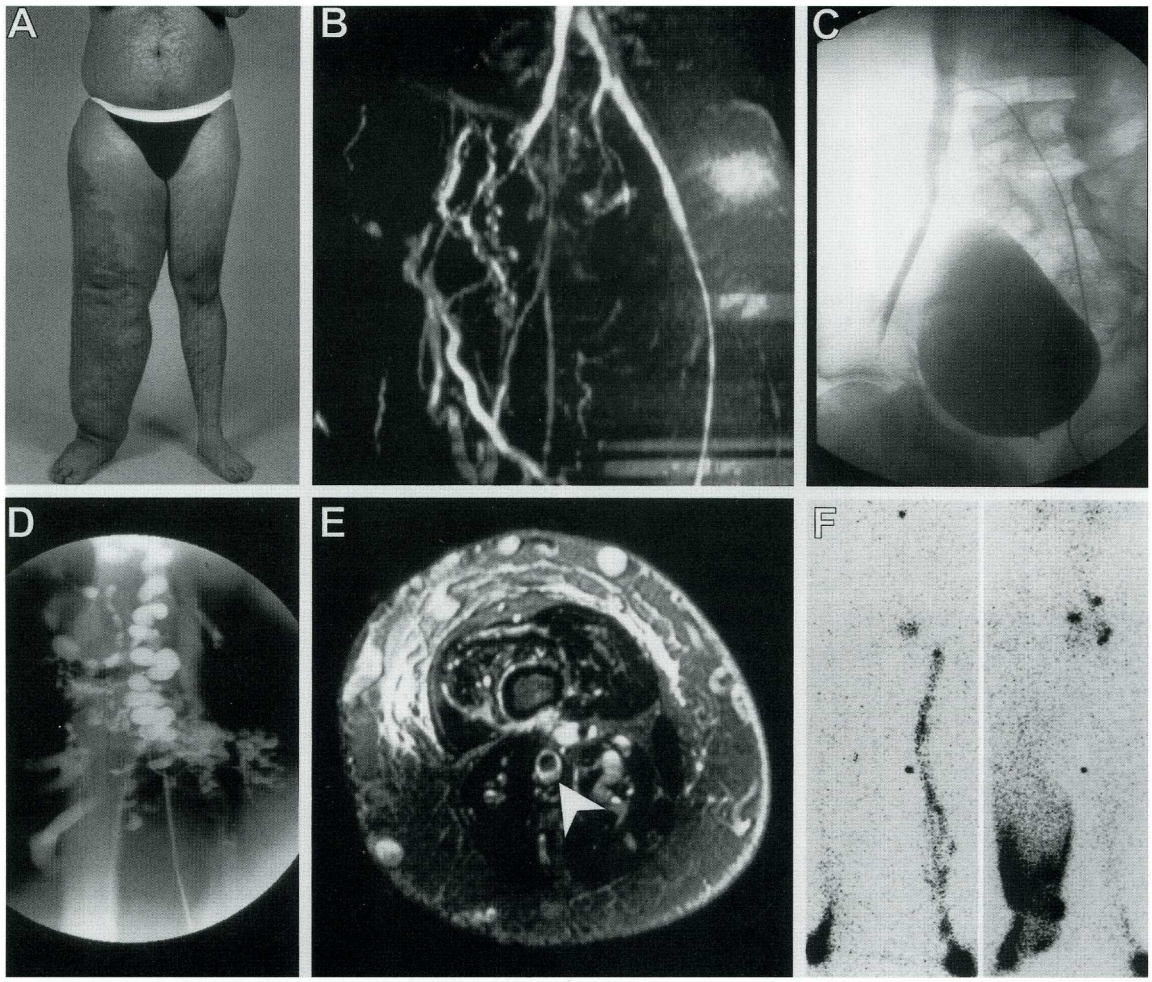

Fig. 3. 26 year-old man with Klippel-Trenaunay-Servelle malformation of the right leg and pelvic girdle (A). Magnetic resonance (MR) angiography shows intact iliac veins but atresia of the right common, superficial and profunda femoral veins (B). These findings were confirmed by direct venography via the left leg (C and D). Transaxial MR at the knee shows a patent popliteal vein (arrowhead) (E). Lymphangioscintigraphy (F) shows severe lymphatic hypoplasia of the right leg with prominent dermal backflow and absence of regional nodes. Radiotracer transport in the left leg is unremarkable. A venous bypass graft from the popliteal vein to the external iliac vein with a small posterior tibial arteriovenous fistula was constructed but the bypass failed to remain patent. Because of repeated phlebitis and life-threatening infectious episodes, the right leg ultimately was amputated by disarticulation at the hip.

Whereas most angiodysplastic birthmarks are minor cosmetic imperfections, major angiodysplasias are often accompanied by bulky soft tissue overgrowth and edema, represent challenges in characterization, and carry disabling and even life-threatening consequences. Some such as Klippel-Trenaunay (K-T) and Proteus syndrome are well recognized, but others represent forme-frustes or contain mixed elements of these entities. Some have a prominent lymphatic component (e.g. K-T-Servelle variant), a few display arterial anomalies (e.g. Parkes-Weber variant) while still others are "pure" venous dysplasias. These conditions all too often are lumped together as "hemangiomas" and

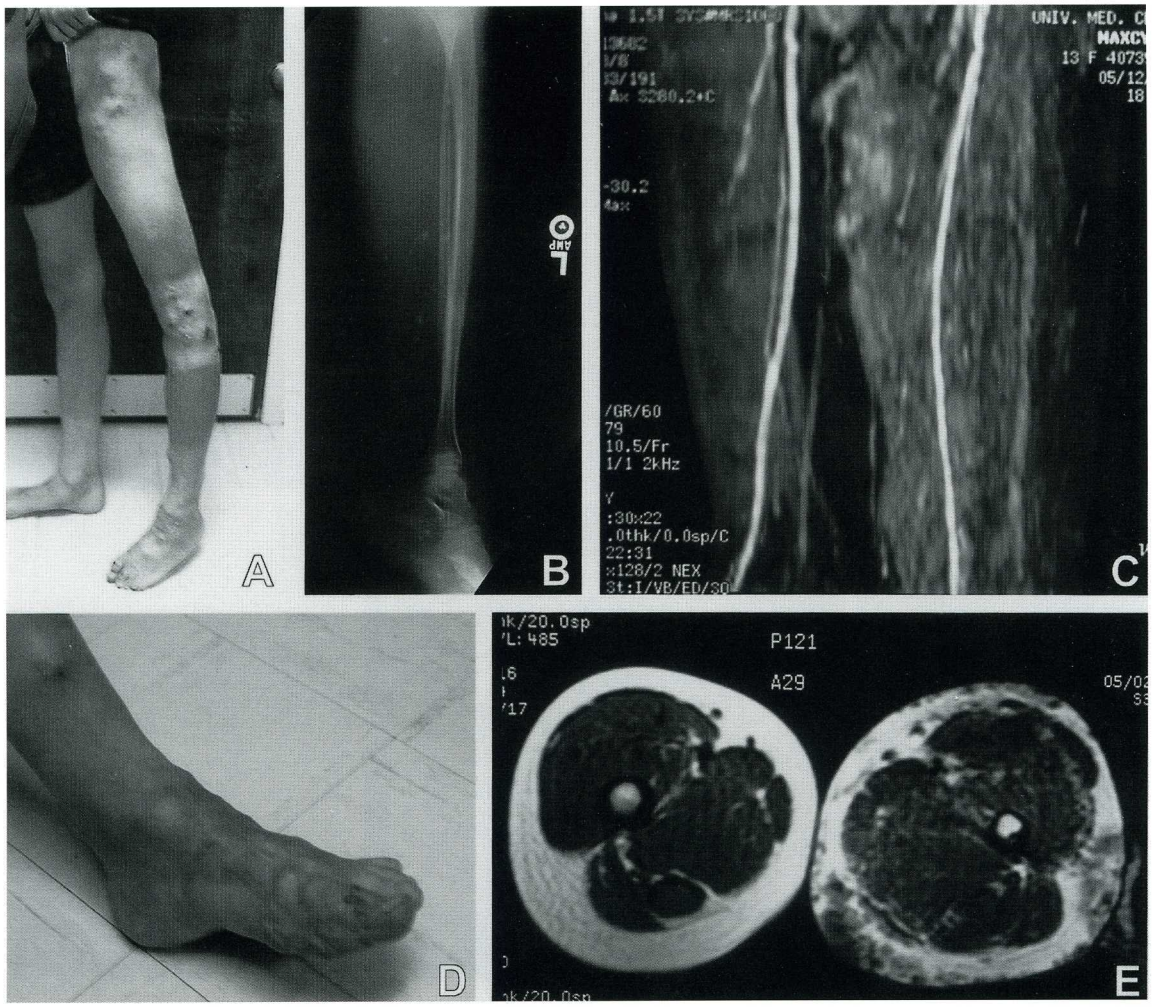

Fig. 4. 14 year-old girl with abnormal lengthening of the left leg with pelvic tilt accompanied by extensive venous malformation of the pelvic girdle, leg and foot (A and D). Plain x-rays (B) show several phleboliths in the calf (faintly seen as white circular densities). Coronal MR arterial venography (C) shows intact arteries with widespread venous malformation involving both the superficial and deep (muscular) compartments. Transaxial views (D) also show extensive radiolucent masses consistent with diffuse venoangiodysplasia.

therapy commonly recommended without appreciation of their full complexity.

Current availability of imaging techniques such as magnetic resonance (MR) magnetic resonance angiography (MRA) with and without contrast (gadolinium) to complement computer tomography (CT), ultrasound (US) and plain x-rays along with lymphangioscintigraphy (LAS) have sharply enhanced the ability to define these abnormalities in physiologic/pathologic terms rather than simply applying eponyms. These imaging techniques, can delineate and depict arterial anomalies, lymphangiomas, lymphatic dysplasia, phleboliths (typical of venous angiodysplasia) and edema in the subfascial in contradistinction to the epi-fascial compartment. Moreover, malformed blood vessels can be distinguished from maldeveloped lymphatics and patterns of

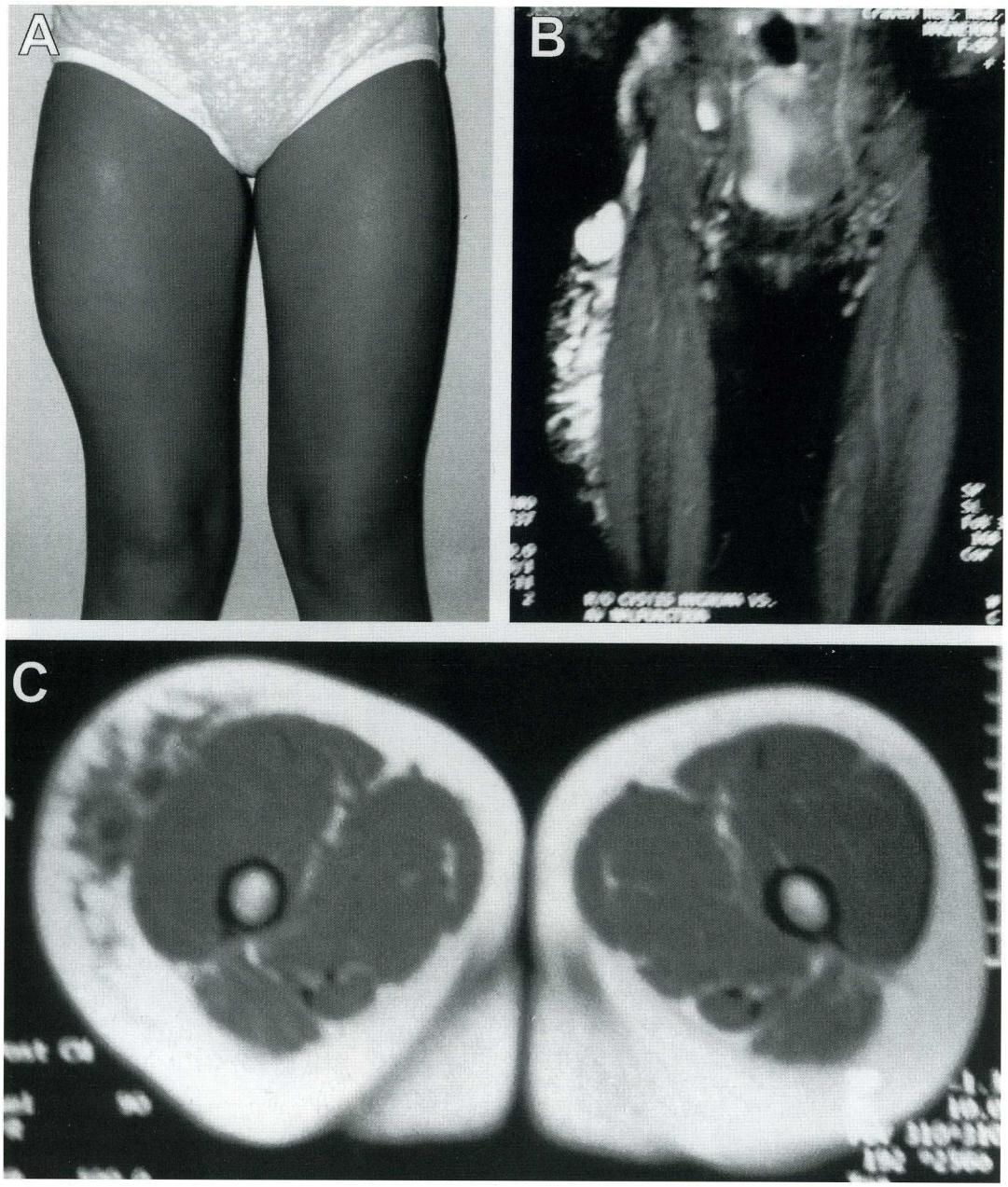

Fig. 5. 16 year-old girl with large right thigh mass (A). Coronal (B) and transaxial (C) magnetic resonance imaging views demonstrate a fluid filled mass superficial to the fascia and muscle consistent with a cavernous lymphangioma.

visceral lymphangiectasia (chylous and non-chylous reflux syndromes) as well as bone and muscle overgrowth or agenesis and fat deposition) can be delineated. Representative imaging examples of hematic and lymphatic angiodysplasia and the potentially confusing entity of morbid obesity are shown.

From these illustrative, albeit select clinical examples, the following imaging option algorithm is suggested for evaluation of

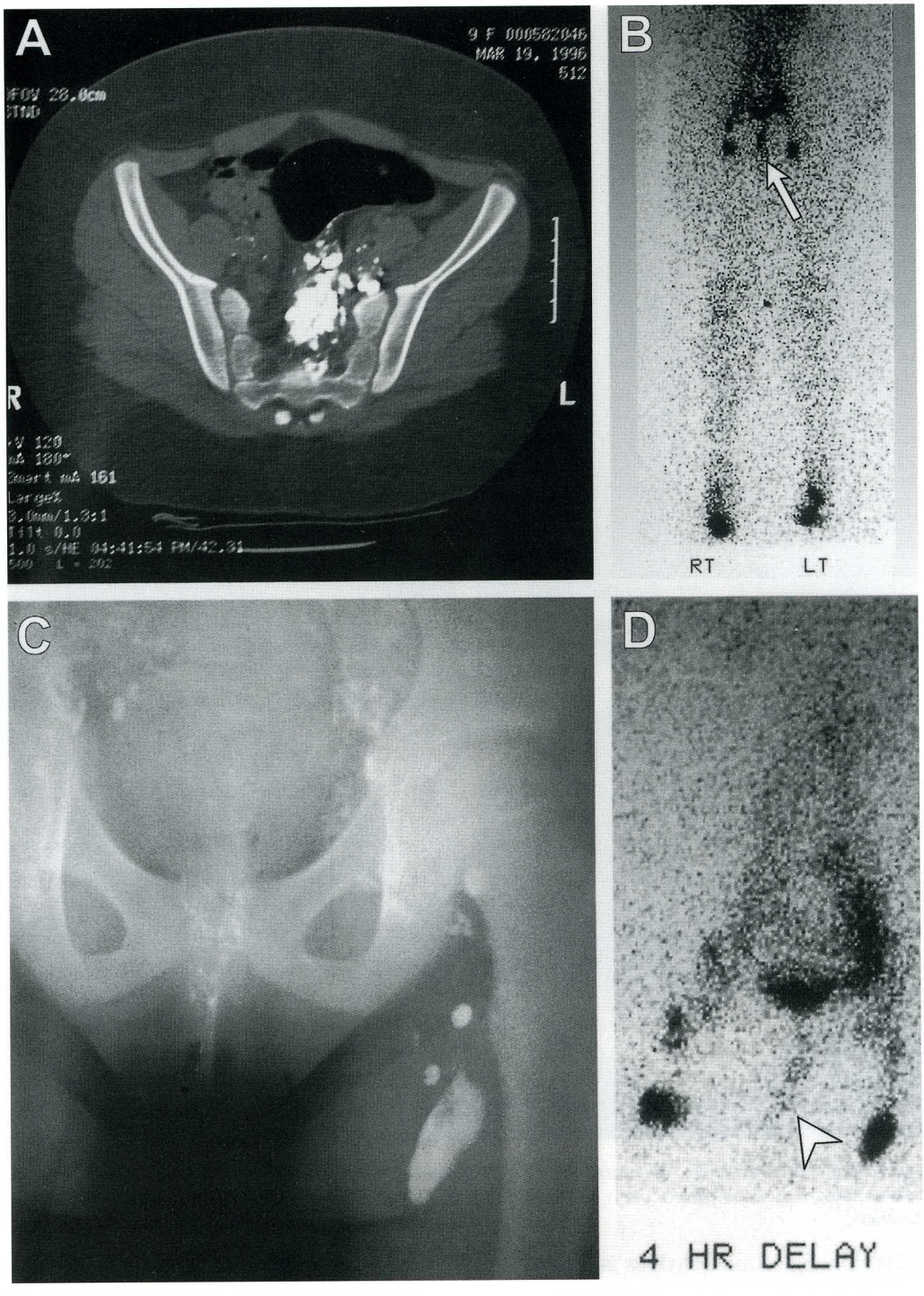

Fig. 6. 14 year-old girl with chylocolporrhea from intestinal lymphangiectasia. (A) Transaxial CT scan after direct conventional lymphography shows lymphatic "lakes" in the true pelvis while plain X-rays demonstrates oil-contrast (ethiodol) leaking into the vagina (arrow) (C). Lymphangioscintigraphy demonstrates that lymphatic transport in the legs is intact but radiotracer is escaping into the vagina (arrow) (B) with close up (arrowhead) (D).

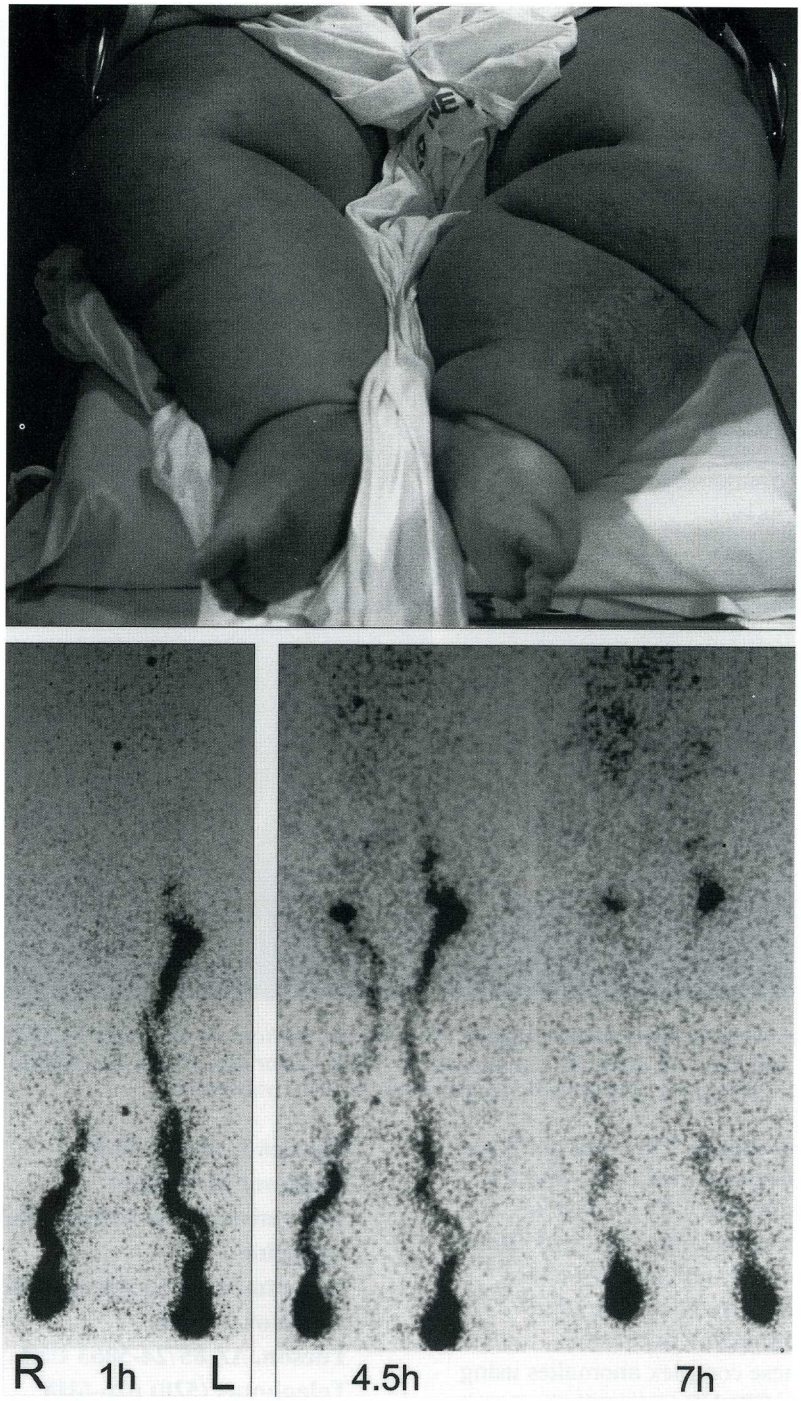

Fig. 7. 56 year-old morbidly obese woman (>200kg) (above). Lymphangioscintigraphy (lower panels) shows markedly delayed radiotracer transport but otherwise an intact lymphatic system. Lower leg swelling, composed of fat and edema fluid, derive from chronic leg dependency, lack of skeletal muscle contraction and forward venous flow in conjunction with sluggish lymphangion contractions (functional lymph stasis). This condition can be confused with true lymphangiodysplasia (lymphedema).

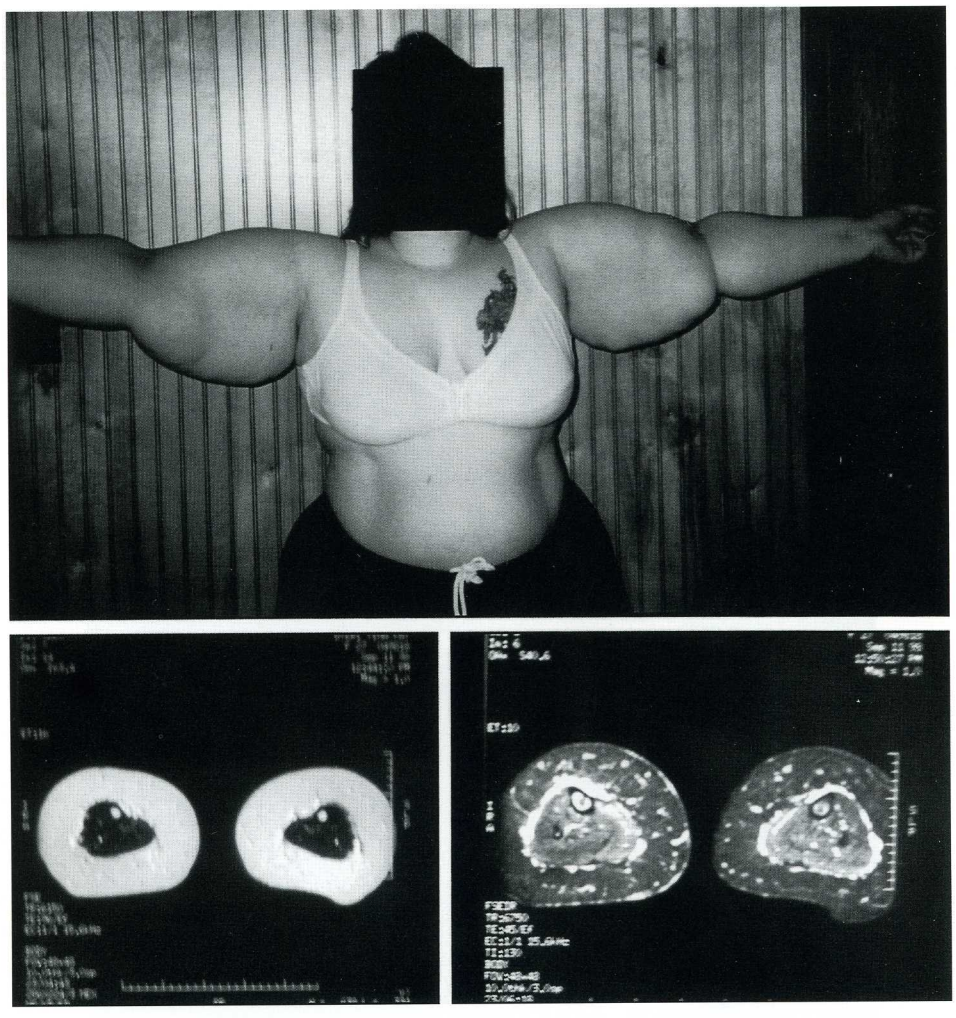

Fig. 8. 38 year-old morbidly obese woman (upper). MR with and without fat subtraction (STIRS) (lower panels) confirms that the soft tissue excess of the arms is entirely fat. A similar appearance was demonstrated by MR and STIRS in the lower extremities. This condition maybe confused with lymphedema.

(Lymph) Angiodysplasia. The most preferred imaging study is represented by 4+ and the lesser helpful studies by 1-3+ *(see Table)*.

Although these recommended procedures are expensive and technologically require advanced radiologic and "cutting edge" computerized equipment, accurate description of these complex anomalies using LAS, MR, MRA, CT, US (primarily in filariasis) provides a blueprint for diagnosis and thereby a rational basis for operative, physical and drug intervention as well as therapeutic innovations for these conditions.

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