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LYMPHOSCINTIGRAPHY PATTERNS IN NEWBORNS AND CHILDREN WITH CONGENITAL LYMPHATIC DYSPLASIA

C. Bellini, G. Villa, G. Sambuceti, C. Traggiai, C. Campisi, T. Bellini, G. Morcaldi, D. Massocco, E. Bonioli, F. Boccardo

Patologia e Terapia Intensiva Neonatale (CB,CT), Dipartimento Alta Intensità di Cure, Istituto Gaslini; IRCCS Azienda Ospedaliera Universitaria San Martino - IST (GV,GS)- Istituto Nazionale per la Ricerca sul Cancro, Medicina Nucleare, Università di Genova; Sezione di Chirurgia e Microchirurgia dei Linfatici (CC,FB), Dipartimento di Chirurgia, IRCCS Azienda Ospedaliera Universitaria San Martino - IST - Istituto Nazionale per la Ricerca sul Cancro, Università di Genova; and Dipartimento di Neuroscienze, Riabilitazione, Oftalmologia (TB,GM,DM,EB), Genetica e Scienze Materno-Infantili Università di Genova, Genova, Italy

ABSTRACT

We performed lymphoscintigraphy on 31 patients (newborns and children) affected by congenital lymphatic dysplasia according to our previously published protocol. Congenital lymphatic dysplasia may present with various degrees of clinical severity, ranging from nonimmune hydrops fetalis with visceral effusions to lymphedema alone. We recommend that lymphoscintigraphy should be strongly considered in all patients with signs of lymphatic dysplasia, including those with minimal and initial signs of lymphatic impairment, in order to obtain a very early diagnosis and to start treatment. Lymphoscintigraphy is safe and useful in the diagnosis of lymphatic dysplasia in the newborn and children. Moreover, it is well tolerated by patients and well accepted by their parents.

Keywords: lymphoscintigraphy, congenital lymphatic dysplasia, lymphedema, chylous reflux, newborn, children

In 2005, *Lymphology* published an article describing our efforts to recommend a protocol for the use of lymphoscintigraphy in newborns and children who were suspected of having congenital lymphatic dysplasia (1). Congenital lymphatic dysplasias are a group of congenital diseases which are characterized by a congenital maldevelopment of the lymphatic system causing early onset of peripheral lymphedema and/or effusion of chyle or lymph into the pleural, pericardial, or peritoneal cavities (2-7).

Although lymphoscintigraphy is employed widely, it is rarely reported as a technique utilized in pediatrics (8-12). In an effort to evaluate the effectiveness of our protocol for lymphoscintigraphy in the newborn and children, we retrospectively evaluated the methodology previously described (1) and examined the lymphoscintigraphic patterns that were obtained in a wide series of neonatal and pediatric patients presenting with signs and symptoms of congenital lymphatic dysplasia.

PATIENTS AND METHODS

Patients

Newborns who presented at least one of the following signs at birth were considered

eligible for our study: idiopathic hydrops fetalis, hydrothorax, hydropericardium, ascites, edema of the limbs, or edema of the genitalia. Newborns with a diagnosis of immune hydrops fetalis or non-immune hydrops fetalis caused by infectious, cardiac, hematological, renal, or gastrointestinal disorders were excluded from the study (7). Children who presented at least one of the following signs were taken into consideration for the study: any degree of limb edema, or abdominal, thoracic or pericardial effusion. Children with a diagnosis of cardiac, hematological, renal, or gastrointestinal disorders related to edema were excluded from the study. Cavity effusion, when present, was evaluated in all enrolled patients on the basis of the previously published criteria (13,14). Lymphedema was staged on the basis of the Consensus Document of the International Society of Lymphology (15-17). The diagnosis of chylous effusion in enteral fed patients was based on the following conditions: milk-like appearance, triglyceride levels > 110 mL/dL, and the presence of chylomicrons (13,18). Fecal -1-antitrypsin excretion was evaluated in all patients in an effort to identify intestinal protein-losing syndrome, possibly suggesting intestinal lymphangiectasia. All patients eligible for the study underwent lymphoscintigraphic study as soon as their clinical conditions were suitable. All patients or parents received appropriate information and gave written informed consent approving the procedure. A flowchart of the consecutive methodological steps for enrolling patients is presented in *Fig. 1.* Six excluded patients were lost at follow-up, thus lymphoscintigraphy was not performed.

Lymphoscintigraphic method

Lymphoscintigraphy was performed according to a previously described protocol (1), with the key features outlined herein.

Radiotracer. We used microcolloidal sulfide particles labeled with Technetium-99m- (99mTc) (Nanocol®; GE Healthcare, UK), 8 MBq in 0.15 ml each, with a particle diameter ranging from 20 to 80 nm. To evaluate the deep lymphatic circulation, two aliquots of tracer (0.1 ml each) were injected into the II and III metacarpal and metatarsal-phalangeal spaces of the hands and feet. To evaluate superficial lymphatic circulation, two doses of tracer were administered intradermally (epifascial) in the hands and feet. All injections were done at approximately the same time for newborns and planar acquisitions were obtained in newborns by planar imaging. In children, we preferred to inject lower and upper limbs at different times because the collimator can scan the whole body of the newborn at once while body size in older children and in adults requires movement of the camera. Both subfascial injection of radiotracers to investigate the deep lymphatic system of the limbs followed by epifascial injection for the superficial system are optimal to obtain better differentiation of the various types of edema.

Parameters for obtaining images

Images were recorded using a high-resolution parallel-hole collimator. Images were acquired using a great field, double-head gamma camera (GE Millenium, Knoxville, USA) equipped with a high resolution parallel-hole collimator with a 10% window centered on the 140-KeV photopeak of 99mTc; 300 second planar images were acquired while the scan speed for total body acquisition was 10 cm/min. Data are shown with the upper level set to display the small fraction of tracer that emigrates from the injection site to the nodes. The gamma-camera is positioned as close as possible and anteriorly to the patient, who is in a supine position. Early dynamic acquisition (20 sec/frame, 128 x 128 matrix) allows visualization of the main lymphatic channels and the first lymph node drainage as well as calculation of the quantitative parameters. At the end of the

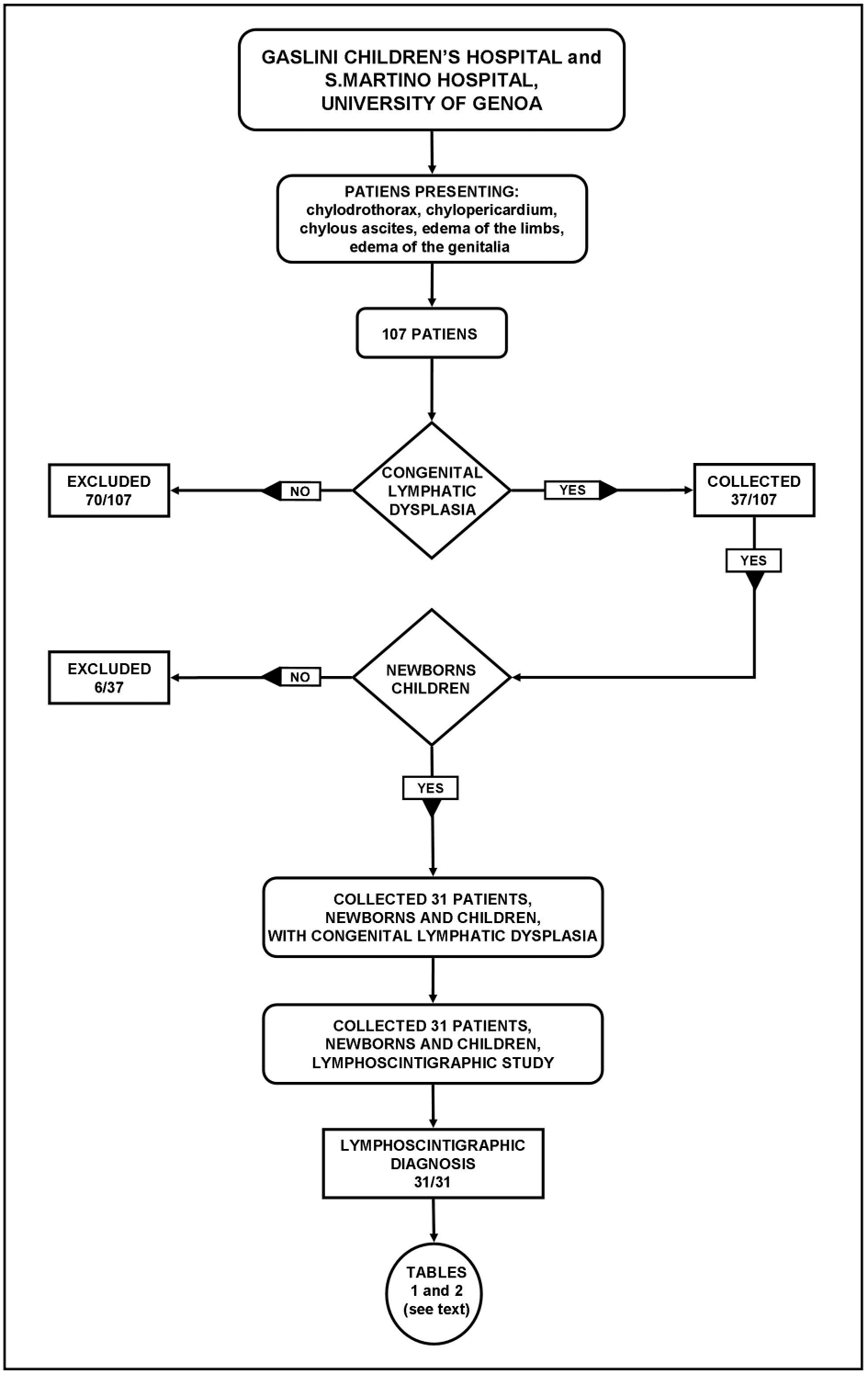

Fig. 1. Flowchart of consecutive methodological steps for subject accrual used in this study.

dynamic acquisition, the study continues with static images using the following parameters: matrix 256 x 256; zoom 1.

Clearance rate and regional lymph node measurements were performed. Semi-quantitative evaluation of lymphatic drainage was established by a numeric index (transport index: TI) (19) calculated by the formula TI = K + D + 0.04T + N + V, where K is lymphatic transport kinetics (no delay, low grade delay, extreme delay, and lack of transport are 0, 3, 5, and 9, respectively), D is the distribution pattern of the tracer (normal, partial diffuse, diffuse, and transport stop are 0, 3, 5, and 9, respectively), T stands for how long it takes (in minutes) for the tracer to appear before it can be observed in the lymph nodes (no appearance is 9) and scans are carried out until these data have been obtained, but for no longer than one hour, N is assessment of lymph node visualization (no appearance is 9), and V is assessment of lymph vessel visualization (clearly demonstrated, faint visualization, hardly recognizable, or no visualization are 0, 3, 5, and 9, respectively). Each parameter ranges from 0 to 9, so total evaluation ranges from 0 (normal) to 45 (pathological). Transport index in healthy extremities is less than 10 (19).

Sedation is usually not required for a technically satisfactory examination, but in some patients who cannot or do not want to co-operate, mild sedation may be necessary. On the other hand, movement must be very limited in order to obtain good quality images. We decided not to apply rigid containment and to allow limited movement if necessary. Following injection into the upper and lower extremities, a massage was carried out and lukewarm packages were positioned on the inoculation sites. The newborn was then allowed to carry out spontaneous motor activity for 20 minutes. Lastly, the newborn was contained so as to obtain images. Older children were allowed to have 15 minutes of spontaneous motor activity after injection.

RESULTS

In each case, the established protocol for performing lymphoscintigraphy was applied, and all patients underwent lymphoscintigraphic study without difficulty. A summary of the data reporting clinical and lymphoscintigraphic patterns, treatment, and follow-up of each patient who was included in the study, and a summary of the clinical lymphodysplastic features of the patients are shown in *Tables 1 and 2*, respectively. A highly detailed lymphoscintigraphic pattern was obtained for each patient (*Table 1*). Data reporting the clinical pattern, treatment, and follow-up of each patient in the study are reported in *Table 1*. Altogether, 31 patients were eligible for the study. Five of the 31 were newborns and were first seen at birth, while the remaining 26 patients ranged from 1 to 8 years of age when first seen (median age 4 years and 9 months). Lymphoscintigraphy was performed within the first two months of age in the 5 patients who were seen at birth, while in the remaining 26 patients, lymphoscintigraphy was performed during the first hospital admission. The initial clinical conditions of the newborns were quite variable, ranging from severely ill newborns who underwent mechanical ventilation, pleural drainage for non-immune hydrops fetalis complicated by chylothorax or chylopericardium, and the need for total parenteral nutrition, to clinically stable patients who presented only with swelling of the genitalia and/or limbs. Thoracocenteses were performed in all patients with chylothorax.

In the presence of respiratory distress, pleural drainage was performed in association with mechanical ventilation. Paracentesis was performed in all cases of chylous ascites. Lymphatic dysfunction was demonstrated in all patients. Lymphoscintigraphic studies showed delay, asymmetric or absent visualization of regional lymph nodes, “dermal back-flow,” asymmetric visualization of lymphatic channels, collateral lymphatic

TABLE 1
Summary of Data Relating Clinical and Lymphoscintigraphic Patterns, Treatment, and Follow-up of Each Patient in the Study

Case	Clinical Pattern	Treatment	Outcome	Lymphoscintigraphic evaluation	
				Pattern	T1 Transport Index
1	non-immune hydrops fetalis, chylothorax, chylopericardium, limb lymphedema, genital lymphedema, intestinal lymphangiectasia	mechanical ventilation, continuous positive airway pressure, intensive care at birth, total parenteral nutrition, medium-chain triglyceride diet	stable lymphedema, medium-chain triglyceride diet	delay, dermal back flow, interrupted lymphatic structures	32
2	chylothorax, limb lymphedema, genital lymphedema, intestinal lymphangiectasia, pulmonary lymphangiectasia	mechanical ventilation, continuous positive airway pressure, intensive care at birth, total parenteral nutrition, medium-chain triglyceride diet	stable lymphedema, medium-chain triglyceride diet	delay, dermal back flow, nearly complete absence of visualization of right inguinal lymph nodes	32
3	non-immune hydrops fetalis, chylothorax, limb lymphedema	mechanical ventilation, continuous positive airway pressure, intensive care at birth, total parenteral nutrition, medium-chain triglyceride diet	minimal residual lymphedema	delay, absent tracer uptake by right inguinal lymph nodes	30
4	non-immune hydrops fetalis, chylothorax, chylous ascites, limb lymphedema, genital lymphedema, pulmonary lymphangiectasia	mechanical ventilation, continuous positive airway pressure, intensive care at birth, total parenteral nutrition, medium-chain triglyceride diet	progressive lymphedema, medium-chain triglyceride diet	delay, dermal and thoracic back flow, interrupted lymphatic structures	32
5	non-immune hydrops fetalis, chylothorax, chylous ascites, genital lymphedema, pulmonary lymphangiectasia	mechanical ventilation, continuous positive airway pressure, intensive care at birth, total parenteral nutrition, medium-chain triglyceride diet	initial lymphedema, medium-chain triglyceride diet	delay, dermal-thoracic and iliac lumbar back flow, interrupted lymphatic structures	30
6	chylothorax, chylous ascites, limb lymphedema, intestinal lymphangiectasia	pleurodesis, medium-chain triglyceride diet	stable lymphedema, medium-chain triglyceride diet, minimal residual chylous ascites	delay, dermal back flow, interrupted lymphatic structures	29
7	limb lymphedema, genital lymphedema	medical treatment	stable lymphedema	dermal back flow, interrupted lymphatic structures	18
8	limb lymphedema, intestinal lymphangiectasia	medical treatment, medium-chain triglyceride diet	stable lymphedema, medium-chain triglyceride diet	reduced tracer uptake by left inguinal lymph nodes, lack of visualization of bilateral iliac lymph nodes	22
9	genital lymphedema	medical treatment	stable genitalia lymphedema	interrupted lymphatic structures	16

Table 1 (continued)

10	genital lymphedema	medical treatment	stable genitalia lymphedema	delay, interrupted lymph structures	18
11	chylous ascites, limb lymphedema, intestinal lymphangiectasia, pulmonary lymphangiectasia	laparoscopic–microsurgical treatment, medium-chain triglyceride diet	recovered chylous ascites, stable lymphedema, medium-chain triglyceride diet	dermal and iliac lumbar back flow, interrupted lymphatic structures	32
12	chylous ascites, genital lymphedema, intestinal lymphangiectasia	laparoscopic–microsurgical treatment, medium-chain triglyceride diet	recovered chylous ascites, stable lymphedema, medium-chain triglyceride diet	delay, dermal and iliac lumbar back flow, interrupted lymphatic structures	34
13	chylous ascites, limb lymphedema, intestinal lymphangiectasia	laparoscopic–microsurgical treatment, medium-chain triglyceride diet	recovered chylous ascites, stable lymphedema, medium-chain triglyceride diet	delay, dermal and iliac lumbar back flow	26
14	limb lymphedema	medical treatment, medium-chain triglyceride diet	stable lymphedema	absent visualization of iliac-inguinal lymphnodes, minimal visualization at vescical level	22
15	non-immune hydrops fetalis, chylothorax, chylous ascites	mechanical ventilation, continuous positive airway pressure, intensive care at birth, total parenteral nutrition, medical treatment medium-chain triglyceride diet.	recovered congenital chylothorax, medium-chain triglyceride diet	normal visualization lower and upper limbs; minimal tracer uptake at thoracic level	32
16	limb lymphedema	medical treatment	stable lymphedema	delay, little tracer uptake by iliac and lumbar-aortic lymphatic pathways; delay in tracer uptake by upper limbs	18
17	limb lymphedema	medical treatment	stable lymphedema	absent tracer uptake right upper limb	22
18	chylothorax	medical treatment, medium-chain triglyceride diet	recovered congenital chylothorax	normal visualization lower and upper limbs	16
19	chylopericardium, chylous ascites, intestinal lymphangiectasia	laparoscopic–microsurgical treatment, medium-chain triglyceride T diet	recovered chylous ascites, medium-chain triglyceride diet	absent tracer uptake by iliac and lumbar-aortic lymphatic pathways	19
20	limb lymphedema	medical treatment	stable lymphedema	delay, absent tracer uptake by iliac and lumbar-aortic lymphatic pathways on the right, reduced on the left	22
21	limb lymphedema, genital lymphedema	medical treatment, medium-chain triglyceride diet	stable lymphedema	delayed visualization of iliac-inguinal lymph nodes, reduced visualization at vescical level	16
22	limb lymphedema, intestinal lymphangiectasia	medical treatment medium-chain triglyceride diet	progressive lymphedema, medium-chain triglyceride diet	dermal and iliac lumbar back flow, interrupted lymphatic structures	20
23	chylous ascites	laparoscopic–microsurgical treatment, medium-chain triglyceride diet	recovered chylous ascites, medium-chain triglyceride diet	delay, absent visualization of iliac-inguinal lymph nodes, dermal back flow	30
24	limb lymphedema	medical treatment, medium-chain triglyceride diet	stable lymphedema	delay, dermal and iliac lumbar back flow, interrupted lymphatic structures	28

Table 1 (continued)

Case	Clinical Pattern	Treatment	Outcome	Lymphoscintigraphic evaluation	
				Pattern	TI Transport Index
25	limb lymphedema, intestinal lymphangiectasia	medical treatment, medium-chain triglyceride diet	stable lymphedema, medium-chain triglyceride diet	dermal and iliac lumbar back flow, interrupted lymphatic structures	22
26	non-immune hydrops fetalis, chylothorax, limb lymphedema	medical treatment, medium-chain triglyceride diet	stable lymphedema	dermal and iliac lumbar back flow, interrupted lymphatic structures	26
27	limb lymphedema, intestinal lymphangiectasia	medical treatment, medium-chain triglyceride diet	stable lymphedema, medium-chain triglyceride diet	delayed visualization lower and upper limbs; minimal tracer uptake at thoracic level	30
28	chylothorax, limb lymphedema	medical treatment, medium-chain triglyceride diet	stable lymphedema, recovered congenital chylothorax	delayed visualization lower and upper limbs; tracer uptake at thoracic level	22
29	limb lymphedema	medical treatment, medium-chain triglyceride diet	stable lymphedema	delay, interrupted lymphatic structures	18
30	limb lymphedema	medical treatment, medium-chain triglyceride diet	stable lymphedema	reduced tracer uptake by iliac and lumbar-aortic lymphatic	16
31	limb lymphedema	medical treatment, medium-chain triglyceride diet	stable lymphedema	delay, dermal and iliac lumbar back flow, interrupted lymphatic structures	26

TABLE 2
Summary of Clinical Lymphodysplastic Features of Study Patients
(many patients presented multiple features)

Total number of cases	31
Non-immune hydrops fetalis	6/31
Lymphedema	23/31
Genital lymphedema	9/31
Chylous ascites	9/31
Chylothorax	10/31
Chylopericardium	2/31
Intestinal lymphangiectasia	11/31
Pulmonary lymphangiectasia	4/31

channels, and interrupted lymphatic structures and lymph nodes of the deep lymphatic system *(Fig. 2 and Table 1*). Transport index results are shown in *Table 1.* No adverse effects to the procedure were observed.

Medical and surgical treatment and follow-up

Total parenteral nutrition using a percutaneous central line was initiated in each case of cavity effusion and was maintained until resolution. When cavity effusion recovery was obtained, we waited at least 1 week before the first attempt at enteral feeding, which then consisted of a medium-chain triglyceride formula for newborns and a hypolipidic diet made up of medium-chain triglycerides in children (lipid concentration at 22-25%, including 18% medium-chain triglycerides). If this resulted in the recurrence of effusion, enteral feeding was discontinued and total parenteral nutrition was resumed. Pleurodesis was performed in case # 6 and recovery from pleural effusion was obtained.

All patients were discharged and sent home. Chylothorax recovery appeared to be complete in all cases. Varying degrees of lymphedema of the limbs or genitalia were present at discharge in all previously affected subjects.

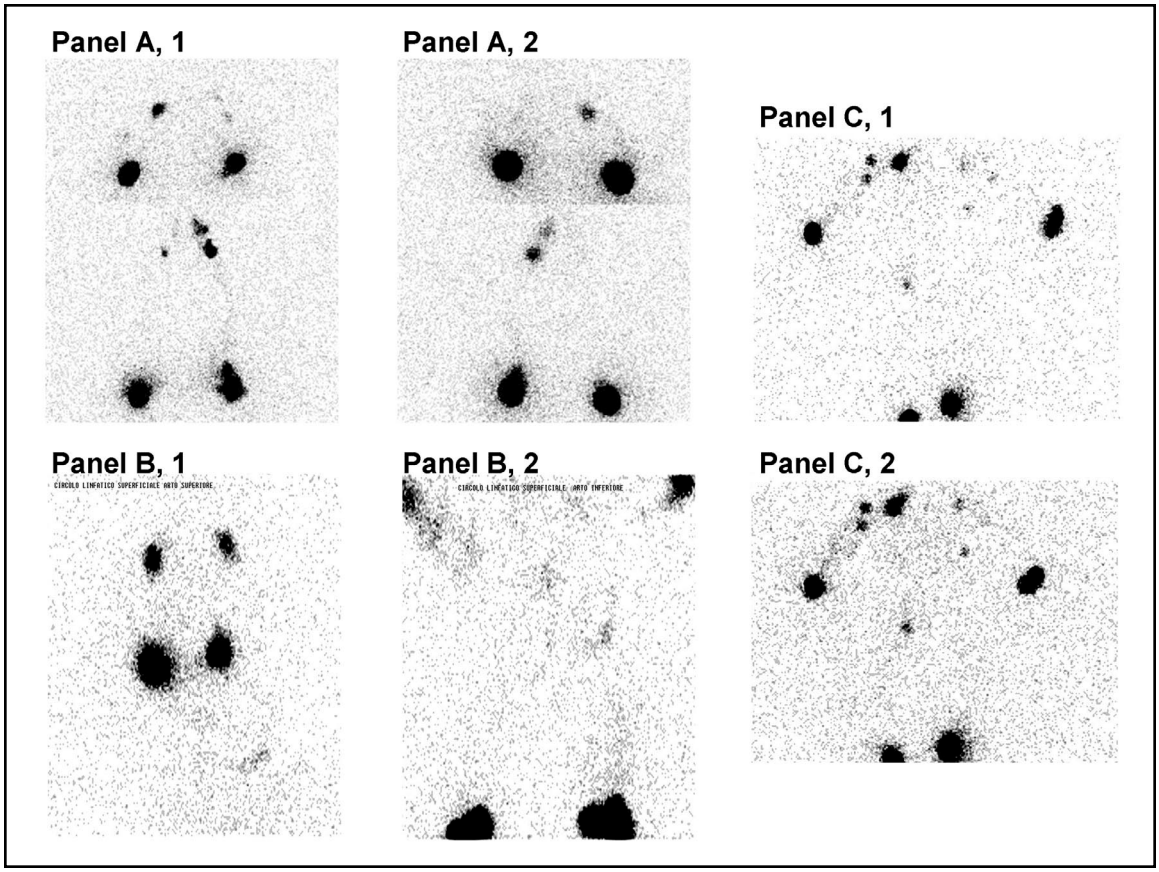

Fig. 2. Stable lymphedema of the lower right limb (panels A) displays delayed and reduced tracer uptake by right inguinal and left axillary lymph nodes (1=anterior; 2=posterior views). Congenital chylothorax (panels B) displays normal visualization of the upper limbs, absent visualization of right iliac-inguinal lymph nodes, reduced tracer uptake by left inguinal lymph nodes, and minimal tracer uptake at the thoracic level. Dermal back flow is seen in the left lower limb. (1=upper limbs; 2=lower limbs). Congenital lymphedema of the lower limbs (panels c) presents with absent visualization of left iliac-inguinal lymph nodes, reduced and delayed visualization of right inguinal and left axillary lymph nodes. (1=early; 2=delayed views).

DISCUSSION

We present our experience regarding lymphoscintigraphic evaluation of a total of 31 pediatric patients (newborns and children) affected by congenital lymphatic dysplasia. Lymphoscintigraphic studies of all patients showed various pathological features, including delay, asymmetric or absent visualization of regional lymph nodes, "dermal back-flow," asymmetric visualization of lymphatic channels, collateral lymphatic channels, interrupted lymphatic structures and lymph nodes of the deep lymphatic system, thoracic duct retrograde reflux, and chylous ascites, i.e., back-flow in the iliac-lumbar aortic area (*Fig. 2*). These features are all consistent with congenital lymphatic dysplasia. In fact, these results allowed us to identify a) congenital aplasia or hypoplasia of the peripheral lymphatics, b) congenital abnormalities of the abdominal or thoracic lymphatic trunks, and c) congenital lymphatic valvular incompetence, usually associated with megalymphatics. Lympho-scintigraphic patterns depicting normal

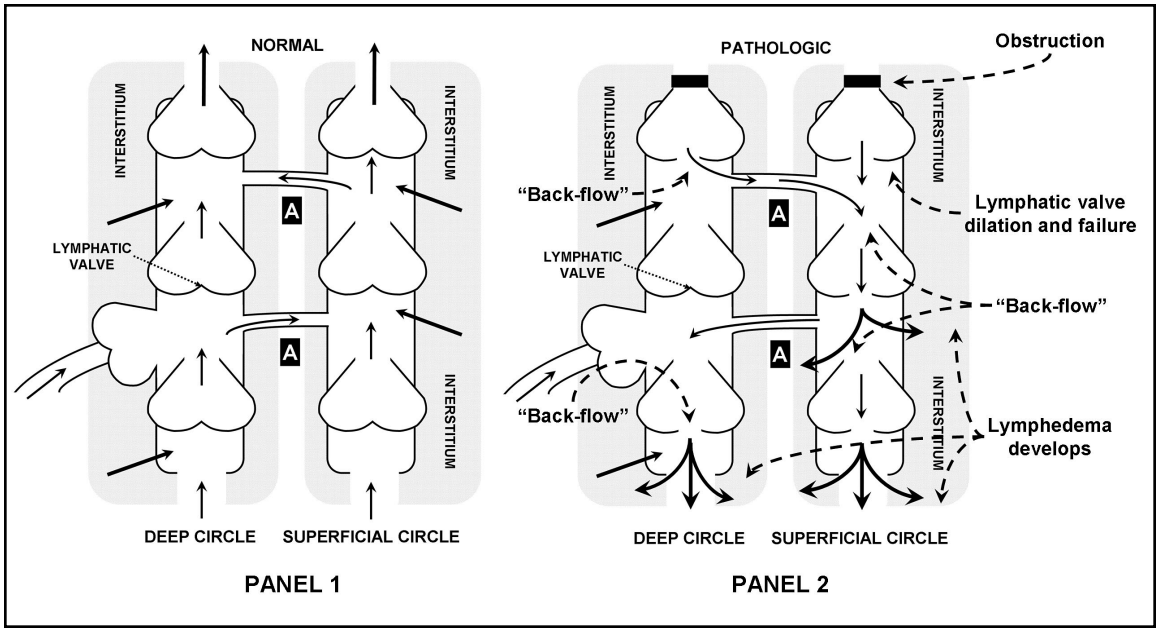

Fig. 3. Schematic drawings depicting basic lymphoscintigraphic patterns. Note the small collateral vessels interconnecting the 2 systems (letters A). Normal lymphatic flow (panel 1) in deep and superficial systems is contrasted with flow alterations seen in lymphedema (panel 2) developing from obstruction, dilation of valves, valvular insufficiency which can lead to subsequent reversal of lymphatic flow (back-flow). Arrows: direction of lymph flow. Dotted arrows: functional indications. Partially modified from http://emedicine.medscape.com /article/1087313-overview.

patients and those with conditions of lymphatic vessel aplasia, hypoplasia, or hyperplasia are shown in *Fig. 3*.

Lymphoscintigraphy is widely considered the main investigative tool for establishing the diagnosis of lymphedema and visualizing peripheral lymphatics (10, 15-17).

Although lymphoscintigraphy is widely used to assess lymphatic function in adults, few reports are currently available describing this technique as a diagnostic tool in newborns and children (20,21, and included refs). Lymphoscintigraphy relies on the function of the lymphatic system to transport large molecules from the interstitial space back to the vascular compartment. Based on this important function, injection of a large molecule such as a protein or colloid with a radioactive label [technetium (99mTc)] into the interstitial space allows the highlighting of lymphatic function and pathways. The use of lymphoscintigraphy is essential in order to obtain a "functional" evaluation of congenital lymphedema. The fine anatomical details can only currently be demonstrated by direct X-ray lymphography (22), which is very difficult to perform in the pediatric age and nearly impossible in the neonatal age. Furthermore, use of X-ray lymphography is contraindicated in pediatrics if congenital lymphatic dysplasia is suspected since the use of this technique may further damage lymphatic vessels. A newer possibility is the use of indocyanine green (ICG) lymphography to evaluate lymphatic dysplasia (23-26). Lymphoscintigraphy cannot be performed at the bedside and a high level of proficiency is required to obtain clear lymphoscintigraphic images. Thus, ICG lymphography can be considered a valid diagnostic option, though keeping in mind that ICG lymphography does not provide comparable diagnostic

accuracy as lymphoscintigraphy. Lymphoscintigraphy can study both superficial and deep lymphatic vessels, while ICG lymphography can only study the superficial lymphatic circulation. ICG lymphography may be a valid approach in severely ill newborns who cannot be transported, but this technique cannot replace lymphoscintigraphy.

Lymphoscintigraphy is useful for detecting both the superficial level of the lymphatic system (epifascial), which primarily drains the skin and subcutaneous tissues, and the deep level (subfascial), which mainly drains the musculature. In order to demonstrate both superficial and deep lymphatic drainage, various injection sites are used, as reported in detail in the Methods section. It has been suggested, although there is no general agreement in the literature (21), that the two systems should be examined separately in an effort to avoid difficulties in interpreting the scan images, since different lymphatic pathways may be taken by the tracer. Thus, the tracer which was, for example, injected subfacially could be drained via the superficial circulation, or on the contrary, routing of the epifascially injected tracer through the deep system might reveal an abnormality in the superficial system. We usually perform simultaneous epifascial and subfascial injections (1). We did not observe any scan images that were difficult to interpret, and both superficial and deep levels of the lymphatic system of our patients were shown in detail.

The onset of primary lymphedema may occur from birth to beyond 25 years of age, though it usually appears very early in life. Congenital maldevelopment of lymphatic vessels causes accumulations/effusion of lymph or chyle in the limbs (lymphedema), and into the pleural (chylothorax), pericardial (chylopericardium), or peritoneal (chylous ascites) cavities. Syndromic occurrence of lymphedema has been extensively reviewed (2,6,27,28). Most patients with primary lymphedema have either unilateral or bilateral swelling, involving the legs more often than the arms, and the occurrence of edema may be an isolated phenomenon with no familial pattern.

Congenital lymphatic dysplasia may present with various degrees of clinical severity. At birth, if congenital lymphatic dysplasia presents as idiopathic hydrops fetalis, it may remain unrecognized due to severe newborn conditions usually leading to premature death (7). Limb lymphedema is the most immediate symptom pointing to lymph vessel maldevelopment, although it may not be evident at birth or during the early years of life, despite the presence of severe visceral impairment (i.e., chylothorax, chylous ascites, etc). In our experience, this fact is underlined by patients # 14, # 16, and # 17 of our study, in whom lymphoscintigraphy was extremely useful in demonstrating pre-clinical lymph vessel impairment, despite the lack of clinical evidence of limb or genital lymphedema. It might be hypothesized that the younger the patient, the better lymphedema may be tolerated owing to the high degree of elastic fibers in the subcutaneous tissue and the wide interstitial space.

Present data suggest that congenital chylothorax, chylous pericardial effusion, and chylous ascites, as well as the presence of lymphangiectasia (intestinal, pulmonary, or affecting other organs), either alone or in association, are indications for performing lymphoscintigraphy during the neonatal age, even in cases in which peripheral lymphedema is not evident.

Although the diagnosis of lymphedema remains mainly clinical and relies primarily on family history and physical examination, early diagnosis during the neonatal age or childhood is very important to prevent the rapid evolution of lymphedema itself (4,29,30).

Conditions characterized by functional lymphatic insufficiency currently lack cures, and reducing the progression of lymphatic impairment would appear to be the sole concrete option (4,29-31).

On the basis of our recent experience, we confirm that lymphoscintigraphy is a mildly invasive technique that is easy to perform, safe, and reliable (4,18,19). There is little discomfort, and it offers objective evidence to help distinguish lymphatic pathology from non lymphatic causes of peripheral edema. It should be considered a mandatory and very valuable diagnostic tool in newborns and children affected by or suspected of congenital lymphatic dysplasia in whom conventional contrast lymphography is potentially dangerous and difficult to perform.

Lymphoscintigraphy is very useful in the diagnosis of lymphedema, and the quantitative analysis that is obtained by performing transport index improves the diagnostic and prognostic power of the technique in the very early diagnosis of lymphatic disorders.

Early diagnosis of lymphedema is important in any period of life and during the neonatal age it is very important in order to prevent the rapid evolution of lymphedema and of visceral chylous effusions in an effort to reduce disease progression and prevent a range of complications that may eventually lead to severe disability or even death.

In conclusion, we suggest carrying out lymphoscintigraphy as soon as possible in all patients who are at risk of developing lymphedema and possibly lymphedematous tissue fibrosis, sclerosis, and fat accumulation. Lymphoscintigraphy is safe and useful in the diagnosis of lymphatic dysplasia in newborns and children and semiquantitative evaluation of the lymphatic transport with lymphoscintigraphy reliably depicts abnormalities in the lymphatic circulation. Lymphoscintigraphy can establish if leg swelling is due to lymphatic system malfunction or to other causes. Finally, lymphoscintigraphy is well tolerated by patients and well accepted by their parents, who can fully understand the usefulness and benefits of the procedure.

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Dr. Carlo Bellini, MD, PhD
Patologia e Terapia Intensiva Neonatale
Istituto G. Gaslini,
Largo G. Gaslini, 5
16147 Genova, Italy
Tel: +39 10 5636762
Fax: +39 10 3770675
Email: carlobellini@ospedale-gaslini.ge.it