Aims and objectives

The purpose of this study is to show our experience and results and also to describe the technique of contrast enhanced MR-lymphography for the management and diagnosis of lower and upper limbs lymphedema and lymphorrea.
INTRODUCTION

LYMPHEDEMA

Lymphedema is a pathologic condition of the lymphatic system where there is interstitial accumulation of lympha (a protein rich fluid) secondary to a compromised lymphatic system that impedests and diminishes lymphatic return. Subsequently there is inflammation, hypertrophy of adipose tissue and finally fibrosis.

It causes significant physical and psychological morbidity in patients and can be difficult to treat.

Furthermore, it is a frequently underdiagnosed and undertreated pathology.

Lymphedema can be classified as primary or secondary.

Primary limphedema is caused by defects in vascular morphogenesis during the embryophetal period and develope in lymphedema syndroms. These diseases involve almost exclusively the lower limbs. They are caused by congenital abnormalities but can turn clinically evident later in life.

Secondary lymphedema is caused by an acquired defect in the lymphatic system, frequently associated with malignancy, infection, obesity and trauma.

The most common cause of secondary lymphedema worldwide is filariasis, causing permanent lymphedema.

In industrialized countries, the most common causes of secondary lymphedema are neoplasms and their treatment. In this cases, the disease can be secondary to obstruction from primary lymphoma or cancer metastases or develop after lymph node dissection and excision.

TREATMENT OF LYMPHEDEMA:

A conservative management is the linchpin in the treatment of lymphedema. Most patients have an appropiate evolution without surgery, with over 80% of patients with lower limb lymphedema treated succesfully with physiotherapy. For this reason surgery is performed in selected patients after physiotherapy fails, such as in patients who have already developed adipose tissue hypertrophy.
Surgical therapy can be performed with three different ways:

- Debulking or resection approach: A surgical excision of subcutaneous tissue is made with or without excision of overlying skin. It provides comfort to the patients but the underlying disease persists.

- Microsurgery: The aim is to correct the underlying pathology, by creating anastomoses between lymphatic vessels and veins, between lymph nodes and veins, or between distal and proximal lymphatics. These techniques have proved good long-term results for peripheral lymphedema, with reduction in limb volume as well as in the incidence of cellulitis.

- Removal of subcutaneous fatty tissue: It is a recently developed technique in which a circumferential liposuction of the involved limb is made, with early results showing improvement in symptoms and appearance.

IMAGING STUDIES OF THE LYMPHATICS

Conventional lymphography has been the reference for decades, but nowadays is no longer performed routinely due to potential lifethreatening complications (Fig. 1 on page 7).

Lymphoscintigraphy (LS) with colloid-bound technetium-99m is the first imaging study performed these days. However the temporal resolution of the technique is low and requires ionization exposure.

MRI, in comparison with LS, has higher spatial and temporal resolution, capacity of proportionating three-dimensional (3D) images and no ionization exposure is needed. The aim of MR-lymphography (MRL) is the assessment and planning of the surgical treatment of the lymphedema with a lymphovenous anastomosis.

HOW WE PERFORM A RM-LYMPHOGRAPHY

1. Establishing a reference system

It is a fundamental step to perform a MRL, which allows to create an arbitrary reference system for both radiologist and surgeon. We apply a cartesian coordinate system so any
lymphatic and venous structure can be located. As limbs are anatomical zones where natural land-marks do not exist, we have to create them.

We trace a line that connects the acromioclavicular joint and the second metacarpal-phalangeal joint in upper limb and a line that connects the anterosuperior iliac spine and the first metatarsal-phalangeal joint in the lower limb. Then we place MR-compatible skin marks each 10 cm in cranecaudal sense. (Fig. 2 on page 7, Fig. 3 on page 7, Fig. 4 on page 8 and Fig. 5 on page 9)

2. Contrast application

After local asepsia we inject intracutaneously a mixture of a gadolinium based contrast media/local anesthetic mixture (gadodiamide 0.5mmol/ml and mepivacaine 1%) in each interdigital space (1ml in each space) and at the lateral border of the medial thigh in the foot or hand respectively, and additionally in the lateral border of the foot. The gadolinium based contrast media in the interstitial space is collected by the lymphatic system an will circulate in the lymphatics. (Fig. 6 on page 9)

3. MRL acquisition

A 3T equipment is necessary for MRL. In our institution the examinations are performed on a 3T MR system (Magnetom VERIO, Siemens Medical Solutions), using a coronal T1-weighted 3DGRE sequence with fat saturation.

Three anatomical levels are subsequently examined ascending from the lower leg, knee, and upper leg as well as hand and distal forearm, elbow and armshoulder in the upper limb. Usually, exploration of the entire limb is performed at 15, 30 and 45 minutes after contrast injection (Fig. 7 on page 9).

4. Post-proccesing

Three types of reconstructions are useful in MRL. 5 and 15-mm-thick maximum-intensity projections (MIPs) (Fig. 8 on page 10) show with detail lymph vessels. In normal conditions, lymph vessels appear beaded (while veins have lineal contours) with a stronger enhancement than veins.

Multiple projection reconstructions (MPR) are useful for a better anatomical understanding (Fig. 9 on page 11).

Volume Rendering (VR) reconstructions provide a global imaging of the limb and the lymphatics in it (Fig. 10 on page 12).
OUR STUDY

From July 2012 to November 2014, 42 female patients (age range 17-74 years) (Table 1 on page 13) were admitted in our unit under the diagnosis lymphedema.

11 patients suffered primary lymphedema (all of them in lower limbs), 28 patients suffered secondary lymphedema (21 in upper limbs, 7 in lower limbs) (Table 2 on page 14) and 3 of them with lymphorrea.

MR-lymphography was performed in all patients with the technique explained before.
Fig. 1: Conventional lymphography of the inguinal region (left) and the leg (right).

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Fig. 2: Reference system in the upper limb: line from the acromioclavicular joint to the second metacarpal-phalangeal joint, with marks arranged every 10cms in cranio-caudal sense.

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**Fig. 3:** Reference system in the lower limb: line from the anterosuperior iliac spine to first metatarsal-phalangeal joint, with marks arranged every 10cms in cranio-caudal sense. Cartesian system is used to locate any point at the images: - X: external or internal location (negative or positive values of X) - Y: cranial or caudal dispanses (positive or negative values of Y) - Z: depth

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Fig. 4: Reference system in the upper limb.

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Fig. 5: Reference system in the lower limbs.

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Fig. 6: Intracutaneous contrast application under asepsia.

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Fig. 7: Images are acquired 15, 30 and 45 minutes after the contrast injection.

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Fig. 8: MIP reconstruction. Note the enlarged and enhancing lymphatic vessel in the lateral region of the left leg.

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Fig. 9: MPR reconstruction.

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Fig. 10: VR reconstruction. Note de skin marks easily seen.

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Table 1: Patient ages.

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Table 2: Origin of the lymphedema in our group of patients.
Results

The imaging findings in lymphedema were the following:

- Delayed flow pattern due to diminished speed of lymph flow.
- Numerous tiny lymphatic vessels located in the skin and dermal backflow in the lower part of the leg (Fig. 12 on page 18) followed by one or two dilated lymphatic vessels in the upper part of the limb.
- Groups of very dilated and significantly enhanced lymphatic vessels located mainly in the medial and the lateral portion of the proximal limb (Fig. 13 on page 18).
- Radiating enhanced vessels in lower part of the limb that run up to the medial portion and finally go up to (Fig. 14 on page 19).
- Discontinuous and lightly enhanced but dilated vessels in the medial portion of the limb.

MR lymphography allowed the study of the lymphatic system in all patients but in two, in whom the MR was not carried out due to claustrophobia.

In one patient, the study was performed twice, after the first study was non diagnostic as a result of movement of the patient during the acquisition of images. The rest of the studies were diagnostic.

Over the 90% of the studies had a good or excellent quality (Table 3 on page 20). We can observe that lower limb studies have in general a better quality, widely due to the better positioning of lower extremities in the gantry (Table 4 on page 20).

Evaluation and planning of lymphaticovenous anastomosis as a surgical definitive treatment of the lymphedema (both primary or secondary) was carried out. (Fig. 15 on page 21).
In those patients with lymphorrea, the injured lymphatic vessel was determined. An exact intraoperative correlation with imaging findings was observed. (Fig. 16 on page 21).
Fig. 12: MRL in lower limbs. Sigs of lymphedema in the left limb, with asymmetry in volume. We can see small-sized lymphatic vessels located in the skin of the forefoot. A dilated lymphatic vessel is seen in the lateral part of the leg.

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Fig. 13: MRL in upper (Left) and lower (right) limb. Dilated and enhanced lymphatic vessels in the lateral portion of the limbs.

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**Fig. 14:** Enhanced vessels in lower part of the limb that go up to.

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**Table 3:** Quality in our studies.

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Table 4: Lower limb studies have better quality than upper limb studies.

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**Fig. 16:** Patient with lymphorrea after several knee prosthesis surgeries (left). In the images in the middle (VR reconstruction) and right (MIP reconstruction), the enlarged and enhanced lymphatic vessel causing the lymphorrea can be located.

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Conclusion

MR-Lymphography allows high-resolution isotropic imaging of the peripheral lymphatic system with excellent anatomical detail and morphological information. In our experience, it is a reliable, accurate, reproductible and secure technique for the diagnosis, management and surgical planning of lymphedema and lymphorrea.
References