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The role of physical and rehabilitation medicine specialist in lymphoedema☆

Le rôle du médecin de médecine physique et de réadaptation dans la prise en charge du lymphœdème

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1. English version

1.1. Introduction

This paper discusses lymphology and the role of the Physical and Rehabilitation Medicine (PRM) specialist in the management of people with lymphoedema. PRM specialists have a defined role on managing these people and are particularly interested in those with primary and secondary lymphoedema [41]. The aim of the paper is to assist PRM specialists inform their colleagues and payers of services as to their role and to the range of patients, who will gain most advantage from their clinical care.

Lymphoedema is a chronic and progressive condition resulting from an abnormality of, or damage to the lymphatic system. It contrasts from simple oedema due to conditions, such as limb immobilisation or hypoalbuminaemia and is the most readily recognizable attribute of lymphatic vascular incompetence in the presence of the characteristic swelling of tissues, which arises as a consequence of insufficient lymph transport. The accumulation of interstitial fluid in interstitial space leads to swelling, most often in the arms, legs, and other parts of the body.

Lymphoedema is a problem more and more current because its incidence is increasing, linked to the improvement of the survival after breast neoplasm, which represents the first etiology. The early physical treatment is recommended as first-line treatment (HAS, Actualités et Pratiques 2011, n° 28). The PRM allows a global approach of this problem, by integrating the physical problems (the oedema, its functional repercussions, its complications) but also the psychological dimension (always present), the problem of the social and professional integration and finally the quality of life.

1.1.1. Pathophysiology

The main function of the lymphatic system is to drain protein-rich fluid, macromolecules and cells from the interstitium to lymph collectors, then to larger collecting lymph vessels, to the lymph nodes and into the venous circulation. Chronic lymph stasis typically stimulates an increase in the number of fibroblasts, adipocytes, and keratinocytes in the skin as well as a large infiltration of neutrophils. The profound stimulus to collagen deposition in

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the integument seems to be unique to chronic lymphatic oedema, although this biology remains largely unexplained. Mononuclear cells (chiefly macrophages) often demarcate the chronic inflammatory response.

1.1.2. Primary lymphoedema

Poorly developed or missing lymph nodes or lymphatic channels may be the cause of primary lymphoedema. Primary lymphoedema can occur at birth, at the onset of puberty or in the adulthood.

1.1.3. Secondary lymphoedema

Secondary lymphoedema may remain a significant, long-term problem for many cancer survivors secondary lymphoedema [24]. The incidence of lymphoedema after breast-cancer surgery varies across studies from 6 to 70% depending on the criteria used for diagnosis and the follow-up interval [3]. Classically, it develops due to filariasis.

Lymphoedema is deeply disturbing for patients' physical and mental health, involving loss of function and psychological distress, resulting in diminished quality of life. Loss of function can be caused by decreased range of motion and heaviness of the affected limb, impaired wound healing predisposing the patient to infection, and pain. Psychological morbidity includes anxiety, fear, depression, loss of body-image and self-esteem, and decreased sexual drive. There is a current deficiency of the clinical research in the domain and there was thus no recent progress concerning the care of lymphoedema. There are only 11 randomized and controlled studies on this topic. The recommendations are based thus on experts' advice.

1.1.4. Treatment

Treatment is tailored to the individual patient's presentation of the condition and the underlying disease. Early diagnosis and treatment are essential to prevent worsening of the condition and to prevent the psychologic consequences of the disease. Specific type, amount and combination of these treatments continue to be debated. Psychological distress can impact on compliance with physical treatments.

The broad training enables the PRM specialist to diagnose, assess and manage the severity of health problems in lymphoedema [31,33,36].

1.2. Management of Lymphoedema by Physical and Rehabilitation Medicine specialists

1.2.1. Diagnosis

The diagnosis of lymphoedema in PRM predominantly is a clinical diagnosis. The PRM doctor takes the patient's history and performs the clinical investigation [17]. In some cases, imaging techniques have to be applied [60,67,68]. Additionally, the impact of lymphoedema on functioning (referring ICF-domains of body functions, activities and participation) and quality of life needs to be assessed by PRM specialists.

1.2.1.1. History taking. History taking of patients with lymphoedema has to include the dynamic (slowly, fast), the localization and size and texture of oedema. The history of causally connected diseases (trauma, inflammatory diseases, radiotherapy and others) as well as other conditions inducing oedema also are of major importance (cardiac insufficiency, renal diseases, venous disease, and others) [21]. Symptoms possibly related to malignant disease have to be investigated very carefully. Rapid development, pain and nerve lesions as well as the known general signs (weight loss, fever, and others) are red flags to diagnose malignant disease.

The evaluation of pain by visual analog scale is particularly important. The psychological context must be addressed: anxiety and especially depression, fear of cancer recurrence, sleep disorders, sexuality problems, asthenia and even sometimes cognitive abnormalities [22]. Moreover, the social and professional consequences have to be evaluated.

1.2.1.2. Clinical Investigation. Clinical investigation includes inspection of localization and size of lymphoedema as well as changes in skin and palpation of lymph nodes. Palpation also looks for mechanical properties of skin and subcutaneous connective tissue e.g. to diagnose fibrosis. A special tool is the so called Stemmer sign that proves if a skin fold in fingers or toes can be lifted or not (the latter being a sign of lymphostasis) [16] Systematic search for a truncal neurological suffering (median and ulnar nerve), as well as for skin complications: fibrosis, lymphangitis, erysipelas, intertrigo, ulcer with lymphorrhoea, papillomatosis, rare malignant transformation are part of clinical investigation [82].

For the purpose of prognosis and defining a treatment strategy, it is important to stage the lymphoedema [14,27,59]:

- stage 1: reversible, no fibrosis, oedema reduction in putting the extremity up, connective tissue is still plastic (e.g. by pressing with finger);
- stage 2: still reversible, but no significant oedema reduction by putting the extremity up, connective tissue is hard and not plastic any more, skin cannot be moved from subcutaneous connective tissue (Stemmer sign);
- stage 3: tissue characteristics as stage 2, but very big oedema (elephantiasis).

Still very useful is the measurement of circumference in extremities with lymphoedema, in comparison to the contralateral side and to look for changes during therapy. It has to be done on standardized sites (e.g. distance from finger tip or sole of foot) and avoiding compression to the tissue. A minimum of two measurements is recommended [15].

The methodology to measure the volume of lymphoedema has been refined by several approaches [15,65]:

- measurement of circumference of the whole leg or arm in 4 cm intervals (method according to Kuhnke). This method is easy to apply, very sensitive to changes in volume but time consuming;
- calculating oedema volume from at least two measurements of circumferences on both sides using a calculator (method according to Herpertz);