

THE PSYCHOSOCIAL IMPACT OF PRIMARY LYMPHOEDEMA

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This article reviews the literature on the psychosocial impact of non-cancer related lymphoedema. Relevant articles were obtained through database searches, although only a limited number of articles investigated psychological or social factors. Overall, the following psychological factors appeared to be relevant for patients with lymphoedema: body image, mobility, relationship problems, anger, anxiety and depression, although the prevalence and extent of problems pertaining to these factors is not known. Psychosocial factors appear to be under-represented in research to date, suggesting that this needs to be the focus of further work to ensure that these key issues are addressed appropriately in practice.

Key words

Psychosocial impact
Management
Treatment
Quality of life

Lymphoedema is a condition characterised by swelling, pain, heaviness in affected limbs, skin problems and other complications, and has a range of causes, including infection, trauma or immobility (Williams, 2006). More specifically, lymphoedema is described as the abnormal accumulation of protein-rich fluid in the interstitial space (Finegold et al, 2008). The disorder is generally classified into 'primary' and 'secondary' lymphoedema for the purposes of distinguishing between varying presentations of the disease by aetiology. Primary or inherited lymphoedema is a chronic oedema caused by malformation of the lymphatic system. Associations have been found between mutations in

genes, such as vascular endothelial growth factor receptor 3 (VEGFR-3) and congenital lymphoedema (Ferrell et al, 1998; Connell et al, 2008). Secondary lymphoedema is a result of damage to the lymphatic system from, for example, trauma or cancer. A predisposition to secondary lymphoedema has been identified in HGF/MET gene mutations, posing the question 'is secondary lymphoedema a primary disease?' (Rockson, 2008).

The exact prevalence of lymphoedema is not known, however it is estimated that lymphoedema of various causes affects around 0.13% of the population (Franks et al, 2003; Moffatt et al, 2003). Although the condition is widespread, the aetiology is only just beginning to be unravelled. According to Pereira de Godoy et al (2002), some lymphoedemas can be distinguished from lymphoedema resulting from breast and other cancers, although it is difficult to distinguish between the impact on quality of life on those with and without cancer because of studies using mixed groups of patients. It has also been suggested that those with and without the cancer-related condition form two groups, and that patients without cancer delay presenting, which leads to complex problems with management (Sitzia et al, 1998). This distinction is further supported by research suggesting

that patients with lymphoedema not associated with cancer experience a range of difficulties which are not always identified and recognised by healthcare professionals, including incorrect diagnosis and inappropriate treatment (Williams et al, 2004). Delays in obtaining a diagnosis mean that patients often present with lymphoedema that is both severe and complicated (Moffatt et al, 2003), raising important questions about how to enhance management of this group.

According to Morgan and Moffatt (2006), there is a dearth of studies looking at the impact of lymphoedema unrelated to breast cancer. Furthermore, because of funding constraints, provision of care and support for non-cancer lymphoedema is limited (Sitzia et al, 1998) and the needs of patients might not be met (Williams, 2006). Much of the literature has focused on the biomedical aspects of the condition; that is, the pathophysiological and biological mechanisms underlying diagnosis and treatment of lymphoedema, including genetic factors (Ferrell et al, 1998; Evans et al, 1999). For example, current National Institute for Health and Clinical Excellence (NICE) guidelines only include guidance on the use of liposuction for chronic lymphoedema (NICE IPG251, 2008), rather than offering any guidance on psychosocial

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issues or support required. Considering that lymphoedema is a chronic condition resulting in limited function, which can also be visible, understanding the psychosocial consequences is an important goal for successful management of the condition. Psychosocial consequences include any outcomes pertaining to cognitive, affective or behavioural factors, and any issues relating to interpersonal relationships, including personal and professional relationships.

The aim of this article is to review the literature on the psychosocial impact of primary lymphoedema, with a focus on the patient's perspective and offer directions for future research. Lymphoedema can also be present in children and adolescents and the needs of this younger age group may be specific and different to those in adults (Rogge, 1993; Todd, et al 2002). Consequently, this review also covers the issues for this group, because experiences at a younger age can have lifelong psychological implications which may impact on the future management of the condition and the extent to which patients are able to lead full and active lives as adults. Despite the burgeoning literature on the psychosocial aspects of lymphoedema in cancer patients (Radina and Armer, 2001; Radina et al, 2004), this review excludes this patient group in order to uncover the salient psychosocial issues that are not related to cancer.

Method

Databases including Academic Search Premier, the British Nursing Index, CINAHL, (Cumulative Index to Nursing and Allied Health Literature), ISI Web of Knowledge, PsycINFO, Pubmed and Science Direct, were searched for relevant articles using the search terms 'lymphoedema', 'lymphoedema, primary', 'lymphoedema, psychological impact', 'lymphoedema, psychosocial impact', 'lymphoedema, psychological outcome'. Inclusion of articles in the study was based on their coverage of psychosocial factors. Exclusion criteria included articles relating specifically to breast cancer or

cancer survivors, case reports or case studies, genetic studies, those articles only focusing on aetiology rather than psychosocial impact, and studies only including a small number of patients with lymphoedema. Further articles were found through the reference lists of the included articles. Overall, few articles reported on psychosocial factors specifically, although some mentioned other subjective patient-reported outcomes, such as quality of

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life (QoL). Of the 48 articles specifically relating to lymphoedema identified through the searches and reference lists of articles, 10 were included in this review. The reasons for exclusion of the remaining 38 articles were as follows: cancer (18); psychological impact not investigated (9); case study/small number of participants in adults (5) or in children (4); not peer-reviewed/unpublished (2).

Review of the literature

A search of the ISI Web of Knowledge suggests that the number of published articles per year relating to primary lymphoedema has varied between 2–14 over the last four decades, and that since 2005, the rate of publication has been above 13 per year, reflecting a greater interest in the field in recent years. Of the articles identified through the database searches, methodology type and quality of reported studies varied. Considering the relatively small number of studies derived from this search, all relevant peer-reviewed articles were included, regardless of methodological quality. The database search revealed a number of case reports or case studies which illuminate the complexity of individual cases, but

do not provide robust evidence of the key psychosocial consequences. Furthermore, empirical studies focused almost exclusively on the biomedical aspects of the condition (Wolfe and Kinmonth, 1981; Browse, 1986). The exclusion of psychosocial factors by most studies is surprising, given that these have a key role in the management of other conditions, such as chronic pain (Richardson et al, 2006).

Some of the key findings from the reviewed articles are summarised in *Table 1*, including the psychosocial consequences of living with lymphoedema (excluding cancer-related lymphoedema). In general, very little detail is provided regarding these psychosocial factors, including the consequences of a changed body image, whether depression is concomitant with lymphoedema or the causal relationship between these factors. This review is structured by method type and begins with a discussion of review articles, followed by cross-sectional, longitudinal and outcome studies, and finally, qualitative studies.

Reviews

Although assessing patient QoL is an important aim, the psychosocial impact is only one dimension of this multidimensional outcome of which little is known. In a comprehensive review of the literature on health-related quality of life (HRQoL), Morgan et al (2005) identified a number of studies using a variety of methods and assessing a range of outcomes. Reviewed papers included qualitative studies (e.g. semi-structured interviews using a phenomenological approach or grounded theory), cross-sectional surveys assessing a range of outcomes including functional impairment, functional status, psychological morbidity, pain and so on, and longitudinal studies investigating the impact of treatment or rehabilitation on health and functioning. A number of key factors were identified from this review, including the impact of the emergence of symptoms, the limited resources available, the lengthy period of adjustment to the condition, impact on body image and the general

Table 1

Study findings and commonly reported psychosocial sequelae

Authors	Method	Findings
Augustin et al, 2005	Longitudinal study, questionnaire development and testing	QoL assessed with a disease-specific QoL questionnaire. QoL was most compromised for everyday living, satisfaction and emotional well-being
Bogan et al, 2007	Qualitative study	Three themes were identified; nowhere to turn, turning point and making room. These relate to issues around diagnosis and treatment, the experience of being an inpatient and self-management
Cho, 2004	Qualitative study	Healthcare-seeking behaviour of female patients from Korea in three systems; professional, folk and popular
King, 2006	Brief report	Body image due to limb size and shape, reduced mobility and activities due to weight gain, affected posture, difficulty buying clothes, relationship problems
Lam et al, 2006	Cross-sectional study	Dissatisfaction with consultations reported. Lymphoedema swelling impacted on daily living, social life, work and close relationships. Problems with pain, confidence and body image reported
Morgan et al, 2005	Review	Review of studies investigating health-related QoL. Qualitative studies: patients did not receive adequate information, healthcare professionals were not informed and specialist resources were not always available. Psychological factors included shock, disappointment, anger; fear; problems with self-image, life disruption, difficulty adjusting, anxiety and stress. Cross-sectional and longitudinal studies: poorer psychological adjustment, reduced physical and social functioning, greater anxiety and depression than those without lymphoedema. Lack of social support and maladaptive coping associated with psychological and social distress and pain
Sitzia and Sobrido, 1997	Longitudinal study	Overall improvement in QoL assessed with the NHP-I four weeks following conservative treatment. Largest change occurred in the physical dimension. No change detected in psychological and emotional dimensions
Smeltzer et al, 1985	Longitudinal study and review	This follow-up study addressed use of and results of treatment of primary lymphoedema in children and adolescents. The review identified a number of psychological issues for adolescents with the condition, including: self-consciousness over appearance, an altered self-image, being different from others and an impact on self-esteem
Todd et al, 2002	Qualitative study	Six themes relating to the experience of parents with children with lymphoedema were identified. These related to difficulty receiving a diagnosis and obtaining treatment; failure to refer children due to inadequate knowledge of healthcare professionals; receiving inaccurate information; nature and type of support available; challenges of day-to-day management, including the psychological impact on the child; concerns about and uncertainty of the future
Williams et al, 2004	Qualitative study	Issues relating to three broad areas were identified; experience of diagnosis, experiencing and dealing with lymphoedema and treatment

disruption to the lives of patients. Specific psychological concomitants included anger, fear, anxiety and depression, as well as the effect of social support and coping styles on mediating distress. The review also highlighted that patients frequently report pain, which the authors suggested might be underestimated by healthcare professionals and thus needs to be addressed more robustly. Although a number of key implications and avenues for further research are identified, it is difficult to distinguish those factors pertaining to cancer and non-cancer related lymphoedema. More recently King (2006), reporting on the assessment and management of the condition, has devoted only a single paragraph to psychosocial factors, such as body image relating to limited mobility, weight gain and posture, difficulty purchasing clothing, limitations on leisure activities and relationship problems. Such issues clearly impact on the QoL of patients, however, how these issues would be tackled in practice was not discussed.

Few studies examine lymphoedema in children and adolescents, and it has been suggested that psychological aspects have attracted little attention both in research and in the clinic (Smeltzer et al, 1985). Although it is possible that this is due to the low prevalence of this condition in children and adolescents (affecting 1.15 of 100,000 under the age of 20), low prevalence does not justify this inattention. The review by Smeltzer et al (1985) discusses how lymphoedema has been defined in this group, and the range and results of treatments in use at the time. The authors briefly discuss the psychological consequences, such as self-consciousness over appearance, an altered self-image, discomfort as a result of being different from others and the impact on self-esteem. They also suggest that it is important for these issues to be addressed, particularly in adolescents, so that they are able to cope with their condition more effectively. Although they do not propose ways in which this could be done, reference is made to the importance of counselling. It is

clear that more up-to-date data on this important issue in children and adolescents is warranted, particularly by studies in the UK, to enable us to apply the findings more readily.

Cross-sectional, longitudinal and outcome studies

The development and testing of reliable, valid and appropriate outcome measures is essential to the assessment of the impact of lymphoedema. Presently, a number of generic health-related quality of life (HRQoL) instruments have been applied, such as the short-form 36 (SF-36) questionnaire, which has

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been found to be appropriate for use with patients with lower limb lymphoedema (Franks et al, 2006), although many of the reported studies are of cancer-related lymphoedema. A German study reported on the development of a 92-item, condition-specific questionnaire: the FLQA-I (German acronym for lymphoedema specific standardised QoL questionnaire) (Augustin et al, 2005), which assesses several dimensions of QoL pertaining to lymphoedema, including physical complaints, everyday life, social life, emotional well-being, treatment, satisfaction, and issues relating to the patient's profession or household. Low scores (1) represent good QoL and high scores (5) poor QoL. The instrument demonstrated good internal consistency with Cronbach's alpha for the seven scales ranging from 0.85–0.94 (Cronbach's alpha should be between 0.70 and 0.90 to demonstrate acceptable internal consistency reliability [Streiner and Norman, 1995]). For test-retest reliability, correlations between baseline and 12–14 days

later ranged from 0.59–0.87. With the exception of the scale addressing household work, sensitivity to change was demonstrated with significant changes occurring four weeks after lymphatic therapy. Both before and after treatment, QoL was most compromised for everyday living (pre 2.92, post 2.26), satisfaction (pre 3.08, post 2.73) and emotional well-being (pre 2.68, post 2.28). The authors concluded that this was a valid measure for use with patients with lymphoedema. However, it does not appear to have been applied to UK populations which would be a useful addition to understanding the psychosocial impact of the condition.

Although patients with primary lymphoedema were few (n=10 of 34), Sitzia and Sobrido (1997) investigated the HRQoL of patients undergoing conservative treatment consisting of the wearing of multilayered bandaging, with either manual lymph drainage or simple massage using the Nottingham Health Profile (NHP-I). The NHP-I is a self-completion questionnaire addressing the following dimensions of disease: pain, social isolation, emotional reactions, physical mobility, sleep and energy. Given the nature of the treatment, it is perhaps not surprising that the significant improvements in QoL were only found for the physical dimensions. Although the authors conclude that the NHP-I is less useful in assessing the psychosocial domains, applying a more holistic intervention aimed at improving these dimensions might prove more successful, given that improvements in physical aspects of QoL are not necessarily associated with improvements in psychosocial dimensions. Improving psychosocial well-being in patients may require a more targeted and tailored approach than merely addressing a reduction in symptoms, such as swelling or pain. This study provides a useful indication of the impact of conservative treatment on QoL. However, monitoring QoL over time in this way does not tell us which aspects of treatment, if any, brought about change or specifically addressed the impact of the condition on psychosocial well-being.

It appears that much of the impact of lymphoedema has been assessed in terms of its effect on QoL, rather than focusing on specific psychosocial dimensions. Although these factors are accounted for by QoL instruments, there is a lack of clear evidence of impact on more specific psychosocial dimensions, such as social support, and possible psychological sequelae, such as depression and anxiety. However, future work should consider the work that has been done to date in developing and testing appropriate holistic outcomes (such as QoL) in order to ensure that the dimensions that they purport to measure are being assessed.

Qualitative studies

In an attempt to uncover a more in-depth account of the experience of patients with primary or non-cancer related lymphoedema, a number of studies have adopted qualitative methodology. Williams et al (2004) used a phenomenological approach to investigate the experience of patients with both primary and secondary lymphoedema; although their focus was on those with primary lymphoedema due to their recognition of the limited literature relating to this group. Three themes were identified from in-depth interviews.

The first related to the uncertainty and anxiety around the experience of diagnosis, where there was often a long interval between first experiencing symptoms and receiving a diagnosis. Associated with this was an apparent lack of knowledge among healthcare professionals, which appeared to be related to being poorly managed and limited or lack of information.

The second theme accounts for the sense of isolation and impact on personal relationships. For example, the stigma associated with the visibility of the condition was described, and issues relating to self-image, communicating to others about the condition, making sense of it, and devising coping strategies to manage it.

Substantive issues around treatment were identified in the third theme, and

these included the multidimensionality of the range of outcomes that would reflect successful treatment. Despite the illuminating nature of this study, the authors acknowledged its methodological limitations. Moreover, the distinction within the themes to those relevant to patients with the primary or secondary condition is not always clear, further highlighting the lack of understanding of the consequences of the non-cancer related condition.

Overall, the qualitative studies suggest major themes relating to the impact of and issues surrounding diagnosis, relationships, body image and the psychological factors relating to acceptance of and adjustment to the condition.

Bogan et al (2007) echoed some of the findings from the Williams study. By contrast, however, they adopted a more temporal perspective in interpreting their findings. Participants experienced difficulties obtaining a diagnosis, which was subsequently associated with feelings of fear and depression, and a lack of adequate treatment and a sense of isolation through the embarrassment of being noticed, all of which were also found by the Williams study. By contrast, the second and final theme relates to the importance of finally obtaining a diagnosis, and the consequences of this through the need to adopt appropriate self-management strategies. Similar psychological issues emerged within this theme, including problems with body image.

With regard to children and adolescents, case studies or case reports appear to be a common research method, particularly the reporting of the rare condition of primary lymphoedema of the genitalia (Ross et al, 1998), but studies using this methodology were excluded from this review. In a qualitative

study however, Todd et al (2002) examined the experience of six parents of children with primary lymphoedema and identified six different themes. In the first, parents reported difficulty receiving a diagnosis and obtaining treatment, including delays and misinterpretation of symptoms. A failure to refer children to an appropriate specialist was also reported, which parents believed was due to the inadequate knowledge of healthcare professionals. Related to this was the fact that parents often received inadequate and sometimes inaccurate information, for example, regarding the prognosis of the lymphoedema. The nature and type of support available was discussed by parents from the clinic, family and school. The theme which addressed the psychosocial impact on the child in the most detail related to the challenges of day-to-day management, including altered body image and the self-consciousness associated with it, being different from other children, being unable to wear the clothes that they wanted, difficulty in adhering to wearing compression garments and being frustrated and upset. The final theme addressed parental concerns about their child's future, including worries about the progression of the condition, and anxiety about school and how the child would cope. What has not been investigated in full is the role that psychological factors play in the aetiology of the condition.

Overall, these qualitative studies reveal some of the pertinent issues for patients living with lymphoedema, although there is a need for further studies that focus more on the psychological factors that affect the experience and consequences of living with the condition. There may also be important cultural issues that remain unexplored. One study has contributed to our understanding of this, but focuses not on the psychological impact per se, but the way people seek help for their condition in other cultural settings (Cho, 2004). Overall, the qualitative studies suggest major themes relating to the impact of and issues

surrounding diagnosis, relationships, body image and the psychological factors relating to acceptance of and adjustment to the condition.

Discussion

This review has focused on some of the key studies exploring the consequences of living with non-cancer related lymphoedema. What has not been investigated in full is the role of psychological factors in the aetiology of the condition. For example, an early study showed that lymphoedema of the hand could present following unresolved grief (Dopson, 1979). Psychosocial factors may be important in whether and when people present to an appropriate healthcare professional, and thus may have a key role in diagnosis and decisions relating to appropriate treatment. As with many other chronic conditions, key dimensions include factors relating to aetiology, maintaining factors and treatment or management issues. Rather than focusing on the biomedical aspects of each of these, a broader biopsychosocial perspective is more encompassing of the range of relevant factors affecting the patient as an individual.

Considering the lack of literature examining the patient's perspective and the range and extent of the psychosocial impact of the condition, this review has identified some important avenues for further research that will enable healthcare professionals to manage the care of patients more effectively and help facilitate self-management. Although it is worth noting that there is some emerging work investigating the psychosocial impact of lymphoedema using a biographical approach to explore the experience of living with lymphoedema (Waters, 2007), this has not yet been published in peer-reviewed journals. However, preliminary findings suggest that patients with primary lymphoedema had difficulty accessing services and information relating to diagnosis and treatment and that they believed that the knowledge of healthcare

professionals was inadequate. The challenge of managing cellulitis and pain was also highlighted and the fear and the restrictions associated with this. The psychosocial impact varied, although it included feelings of stress, embarrassment, problems with body image and self-esteem, uncertainty about the future, relationship difficulties and more serious mental health problems such as depression. The final theme identified by Waters related to the perceived importance of self-help groups in providing support. There is currently limited evidence for the best practice for psychosocial care of this group, and there appear to be no randomised controlled trials investigating the less biomedical aspects of care. Developing an effective evidence base to assist patients in managing the psychological sequelae of living with lymphoedema and establishing the most appropriate patient-reported outcomes that move beyond swelling and pain is required.

There is some confusion in the literature between what is meant by primary and secondary lymphoedema, although this review has focused on non-cancer related lymphoedema, which may include lymphoedema related to trauma rather than to cancer and, thus, may also be defined as secondary. However, as Rockson (2008) highlights, given that some individuals may be predisposed to the condition, this could also be defined as a primary disease. Consequently, a further goal for future research should be to focus on any salient issues that discriminate between patients presenting with primary and secondary lymphoedema. Despite the importance of recognising each patient as an individual, identifying common psychosocial consequences would undoubtedly show an understanding that currently appears to be lacking in patient interactions with healthcare providers (Williams et al, 2004).

In addition to the gaps in the literature described, little seems to be known about gender differences in psychological impact, coping in patients with lymphoedema, or the

Key points

- ▶▶ Few studies have investigated the psychosocial impact of non-cancer related lymphoedema in adults.
- ▶▶ Research to date has been very biomedical in its focus.
- ▶▶ Notable qualitative studies have highlighted the patient's perspective, although many relate to lymphoedema associated with breast cancer.
- ▶▶ More research is needed to assess the psychosocial impact on both patient and carer, and to adopt a broader biopsychosocial perspective.
- ▶▶ Such research should form part of the management of patients with lymphoedema, including the training of healthcare professionals and the development of effective self-management strategies as an adjuvant to conventional treatment.

role of socio-demographic factors in aetiology and maintenance of the condition. Furthermore, it is not just the patients with lymphoedema that suffer, but the families, caregivers and significant others, the impact on whom has not fully been explored with the exception of a study examining the parents of children with primary lymphoedema (Todd et al, 2002). Despite the relatively low prevalence of lymphoedema in children, further research should also explore the experience of children in greater detail.

Conclusion

Much of the literature discusses primary lymphoedema as if it were synonymous with non-cancer related

lymphoedema, despite the fact that the secondary condition may come about through trauma, in addition to being a consequence of cancer. Furthermore, genetic research has highlighted the potential vulnerability to secondary lymphoedema in some patients, which raises the question of whether the secondary condition is actually a primary disease (Rockson, 2008).

Despite the issues surrounding definition and distinction between the primary and secondary condition, this review has focused on non-cancer related lymphoedema in its broadest sense. In relation to this, the following factors appeared to be most salient for patients with lymphoedema: the delay in receiving a diagnosis; lack of information; uncertainty; problems with body image; communication and interpersonal relationships. Identifying comorbid anxiety, depression and other common mental disorders, regardless of their relationship to the diagnosis is clearly an important goal. Psychosocial factors are under-represented in studies of primary lymphoedema, which tend to focus primarily on biomedical factors of the disease.

It is time for a refocus on the broader biopsychosocial issues of lymphoedema, and future research needs to address this to ensure that there is a sound evidence base for such factors to be managed appropriately in practice. Robust quantitative studies and systematic reviews should be undertaken to fill this current knowledge gap for both adults and younger patients with the disease. JL

References

- Augustin M, Bross F, Földi E, Vanscheidt W, Zschocke I (2005) Development, validation and clinical use of the FLQA-I, a disease-specific quality of life questionnaire for patients with lymphedema. *Vasa*. 34(1): 31–5
- Bogan LK, Powell JM, Dudgeon BJ (2007) Experiences of living with non-cancer-related lymphedema: implications for clinical practice. *Qual Health Res* 17(2): 213–24
- Browse NL (1986) The diagnosis and management of primary lymphedema. *J Vasc Surg* 3(1): 181–4
- Cho MO (2004) Health care seeking behavior of Korean women with lymphedema. *Nurs Health Sci* 6(2): 149–59
- Connell F, Brice G, Mortimer P (2008) Phenotypic characterisation of primary lymphedema. *Ann N Y Acad Sci* 1131: 140–6
- Dopson C (1979) Unresolved grief presenting as chronic lymphedema of the hand. *Am J Psychiatry* 136(10): 1333–4
- Evans AL, Brice G, Sotirova V, Mortimer P, Beninson J, Burnand K, Rosbotham J, Child A, Sarfarazi M (1999) Mapping of primary congenital lymphedema to the 5q35.3 region. *Am J Hum Genet* 64(2): 547–55
- Ferrell RE, Levinson KL, Esmen JH, Kimak MA, Lawrence EC, Barmada MM, Finegold DN (1998) Hereditary lymphedema: evidence for linkage and genetic heterogeneity. *Hum Mol Genet* 7(13): 2073–78
- Finegold DN, Schacht V, Kimak MA, Lawrence EC, Foeldi E, Karlsson JM, Baty CJ, Ferrell RE (2008) HGF and MET mutations in primary and secondary lymphedema. *Lymphat Res Biol* 6(2): 65–8
- Franks PJ, Moffatt CJ, Doherty DC, Williams AF, Jeffs E, Mortimer PS (2006) Assessment of health-related quality of life in patients with lymphedema of the lower limb. *Wound Repair Regen* 14(2): 110–18
- King B (2006) Oedema, lymphovenous oedema and lymphoedema. *Nursing Standard Supplement* 1(1): 31–8
- Lam R, Wallace A, Burbidge B, Franks P, Moffatt C (2006) Experiences of patients with lymphoedema. *J Lymphoedema* 1(1): 16–21
- Moffatt CJ, Franks PJ, Doherty DC, Williams AF, Badger C, Jeffs E, Bosanquet N, Mortimer PS (2003) Lymphoedema: an underestimated health problem. *QJM* 96(10): 731–8
- Morgan PA, Franks PJ, Moffatt CJ (2005) Health-related quality of life with lymphoedema: a review of the literature. *Int Wound J* 2(1): 47–62
- Morgan P, Moffatt C (2006) The National Lymphoedema Framework project. *Br J Community Nurs* 11(4): S19–22
- NICE (2008) Treating chronic lymphoedema by liposuction. Information about NICE interventional procedure guidance 251. IPG251
- Pereira de Godoy JM, Braile DM, de Fatima Godoy M, Longo O (2002) Quality of life and peripheral lymphedema. *Lymphology* 35(2): 72–5
- Radina ME, Armer JM (2001) Post-breast cancer lymphedema and the family: a qualitative investigation of families coping with chronic illness. *J Family Nurs* 7(3): 281
- Radina ME, Armer JM, Culbertson SD, Dusold JM (2004) Post-breast cancer lymphedema: understanding women's knowledge of their condition. *Oncology nursing forum* 31(1): 97–104
- Richardson C, Adams N, Poole H (2006) Psychological approaches for the nursing management of chronic pain: part 2. *J Clin Nurs* 15(9): 1196–202
- Rockson SG (2008) Secondary lymphedema: Is it a primary disease? *Lymphatic Res Biol* 6(2): 63–4
- Rogge H (1993) Psychosocial problems of lymphedema patients in adolescence. Two case reports. *Z Lymphol* 17(1): 12–4
- Ross JH, Kay R, Yetman RJ, Angermeier K (1998) Primary lymphedema of the genitalia in children and adolescents. *J Urol* 160(4): 1485–9
- Sitzia J, Sobrido L (1997) Measurement of health-related quality of life of patients receiving conservative treatment for limb lymphoedema using the Nottingham Health Profile. *Qual Life Res* 6(5): 373–84
- Sitzia J, Woods M, Hine P, Williams A, Eaton K, Green G (1998) Characteristics of new referrals to twenty-seven lymphoedema treatment units. *Eur J Cancer Care* 7(4): 255–62
- Smeltzer DM, Stickler GG, Schirger A (1985) Primary lymphedema in children and adolescents: A follow-up study and review. *Pediatrics* 76(2): 206–18
- Streiner DL, Norman GR (1995) *Health Measurement Scales: a practical guide to their development and use*. 2nd edn. Oxford University Press, Oxford
- Todd M, Welsh J, Moriarty D (2002) The experience of parents of children with primary lymphoedema. *Int J Palliat Nurs* 8(9): 444
- Waters B (2007) Unknown Stories: Biographies of Adults with Primary Lymphoedema – update on research activity. *LymphLine: Newsletter of the Lymphoedema Support Network*. 4–5
- Williams A (2006) Lymphoedema: care in the community. *Primary Health Care* 16(6): 25–9
- Williams AF, Moffatt CJ, Franks PJ (2004) A phenomenological study of the lived experiences of people with lymphoedema. *Int J Palliat Nurs* 10(6): 279–86
- Wolfe JH, Kinmonth JB (1981) The prognosis of primary lymphedema of the lower limbs. *Arch Surg* 116(9): 1157–60