

# Journal of the University of Malaya Medical Centre

#### Volume 9 Number 2

2006

#### **Editor**

Rosmawati Mohamed, MBBS, MRCP, M.Med, MD

#### **Board Members**

Low Wah Yun, Ph.D. CPsychol, AFBPSS Atiya Abdul Sallam, MBBS, MPH, Msc Azad Hassan Abdul Razack, MBBS, FRCS Ikram Shah Ismail, MBBS, Ph.D, FRCP, FAMM Lim Chin Theam, MBBS, FRCPCH, FRCP, FAMM Maude Elvira Phipps, Ph.D. Peh Suat Cheng, MBBS, MPATH, MD, Ph.D., FRCPath, FRCPA Saw Aik, MBBS, M.Med, FRCS Sazaly Abu Bakar, Ph.D. Debra Sim Si Mui, Ph.D. Suzita Mohd. Noor, MMedSc.

#### **Production**

Tam Lye Suan Doris Ng

#### Secretary

Rajeswari K

#### Correspondence

All manuscripts, general correspondence and enquiries should be addressed to:

The Editor c/o Dean's Office, University of Malaya Medical Centre 50603 Kuala Lumpur, Malaysia Tel: (03) 7949 2077 Fax: (03) 7956 8841 E-mail: zakiiah@ummc.edu.my

#### Publisher

JUMMEC is published twice a year by the University of Malaya Medical Centre, 50603 Kuala Lumpur, Malaysia

#### Cover

Grade IV hypertensive retinopathy and choroidopathy (Courtesy of Dr Tengku Ain Fathlun Tengku Kamalden, Department of Ophthalmotology, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia)

#### **Printed by**

Millennia Comms, Petaling Jaya, Selangor Darul Ehsan, Malaysia

## Journal of the University of Malaya Medical Centre

## **Instructions to Authors**

JUMMEC publishes both basic and applied science and clinical research studies on any area of medicine. JUMMEC welcomes manuscripts on all aspects of medicine in the form of original articles, case reports, review articles, short communications, clinicopathological conference abstracts and letters to the Editor. Manuscripts should be submitted to:

The Editor JUMMEC c/o The Dean's Office University of Malaya Medical Centre 50603 Kuala Lumpur, Malaysia Tel: (03) 7949 2106 Fax: (03) 7956 8841 E-mail: rajeswari@ummc.edu.my

**Manuscripts:** Manuscripts must be in English and should not exceed 3,000 words. It should be submitted in duplicate, typed on one side of A4 size paper and double-spaced with at least 2.5 cm margin. A computer diskette (3.5 in) or compact disc (CD) containing the manuscript in Microsoft Word and a covering letter, stating that the work has not been published nor under consideration for publication elsewhere, should be submitted to the Editor. Presentations at meetings are not classed as prior publication. The text of the manuscript should be in the following form:

**Title page:** The title page should contain a concise title of the article. It should identify all the authors, the name(s) of the institution(s) and their full addresses where the work was carried out. Contact information of the corresponding author including name, address, telephone, fax number and e-mail should also be indicated.

**Abstract and Keywords:** The second page should contain an abstract of about 150-200 words. It should state the purpose of the study, a brief description of the procedures employed, main findings and principal conclusions. Three to five keywords should also be listed below the Abstract.

*Text:* Wherever possible, the text should consist of an introduction, materials and method, results, discussion, conclusions, references and acknowledgements.

**References:** Number references consecutively in the order in which they are first mentioned in the text. References in the text should be indicated by a figure within parenthesis (). The titles of journals in the list should be abbreviated according to the style used in the Index Medicus. Authors are responsible for the accuracy of all references. Examples of correct forms of references are given as follows:

#### i) Journal articles:

Roberts CW, Alexander J, Bossi L, *et al.* Studies on a murine model of congenital toxoplasmosis. Parasitol 1992; 104:19-23.

#### ii) Personal author(s) of book:

Osler AG. Complement: mechanisms and functions. Englewood Cliffs: Prentice-Hall, 1976.

#### iii) Chapter in book:

Weinstein L., Swartz MM. Pathogenic properties of invading microorganisms. In: Sodeman WA Jr, Sodeman WA, Eds. Pathologic physiology: mechanisms of disease. Philadelphia: WB Saunders; 1974; 457-72.

#### *iv)* Agency publication:

World Bank. Intensifying action against HIV/AIDS in Africa: responding to a development crisis. 2000, 89p.

#### v) Journal article on the Internet:

Foley KM, Gelband H, editors. Improving palliative care for cancer. Washington National Heading Press; 2001 *www.nap.edu/books/0309074089/html* (accessed 14 Apr 2006). **Abbreviations, Symbols and Nomenclature:** A list of acceptable abbreviations is published in the Uniform Requirements for Manuscripts submitted to Biomedical Journals (also known as the Declaration of Vancouver). For more information, refer to:

International Committee of Medical Journal Editors. Uniform requirements for manuscripts submitted to Biomedical Journals. BMJ 1991; 302: 338-41.

Only generic names of drugs may be used. Quantitative data must be reported in SI units.

**Tables:** Type each table on a separate sheet and number in arabic numerals. The tables should be as few and as simple as possible, with the title above and any notes or description below. Explain all abbreviations. If a table or figure has been published before, written permission must be given by the owner for its reproduction.

**Figures:** Graphs, drawings and photographs should be submitted as clear, glossy prints measuring 12 cm by 17 cm. Figures should be identified on the back with the title of the article and figure number (in light pencil) and an arrow to indicate the top. Legends to the figures should be submitted on a separate sheet. Explain all abbreviations and symbols used.

**Letter of Consent:** Submissions must be accompanied by a letter of consent, signed by **all** authors, containing the following text:

"The manuscript represents original, exclusive and unpublished material. It is not under consideration for publication elsewhere. Further, it will not be submitted for publication elsewhere, until a decision is conveyed regarding its acceptability for publication in the JUMMEC. If accepted for publication, I agree that it will not be published elsewhere, in whole or in part without the consent of the Journal of the University of Malaya Medical Centre. The undersigned author(s) hereby transfer/assign or otherwise convey all copyright ownership of the manuscript entitled (*the title of article*) to the Journal of the University of Malaya Medical Centre."

**Reprints:** Author will receive 20 reprints free of charge. Additional reprints can be purchased by writing to the Editorial Office.

## JUMMEC: TAKING STOCK, CONSOLIDATING AND GAINING IN STRENGTH

The year 2005 marked the rebirth of JUMMEC when the Editorial Board took over from its previous Editor with modest but realistic expectations. One year on, we have successfully achieved our initial expectation, that is, to encourage greater participation from our junior academics to write and publish in our very own journal, JUMMEC. As we head towards the end of 2006 and prepare to usher in 2007, we see JUMMEC consolidating and gaining in strength. Let us reflect on its past achievements and our expectations for the future.

In the bumper 2003-2005 issue, many articles written by our Faculty members were published. Among some of these articles include an article pertaining to health issues and challenges in the 21st century, where changes in disease pattern, environmental health, demographic impacts on health, migrant issues and health, effects of globalization on health, mental health and wellness, and equity in health care were reviewed (1). Other reviews on cardiovascular exercise testing as a valuable physiological assessment tool in research and clinical practice (2), and an overview on peripartum cardiomyopathy, where challenges that lie in diagnosing and managing this rare yet lethal disease were highlighted (3). Apart from clinical studies, epidemiological studies such as, lifestyle practices in relation to overweight and obesity were also reported in a community-based study, where risk factors, such as physical exercise, cigarette smoking, dietary habits, eating pattern, chronic diseases and family history were studied (4).

In conjunction with the centennial celebration of the University of Malaya in 2005, the Faculty of Medicine had organized the International Conference on Health Sciences: 'Partnership in Healthcare – Lessons Learnt and Future Challenges'. Some international speakers spoke at the conference and their extended abstracts are published in Supplement I, 2006. Darryl M from UNESCO wrote on biomedical ethics; Khoo SK from Brisbane, Australia touched on the clinical implications on ageing (or longevity); Booker PD from Liverpool, UK wrote on the recent advances in paediatric intensive care; Ng CH from Melbourne, Australia described the burden of depression in the Asia Pacific region; David Hui SC from Hong Kong on obstructive sleep apnoea and cardiovascular complications; Peter Eu from Melbourne, Australia on PET radiopharmaceutical production; Kevin Tse KM from Hong Kong who gave the Hong Kong experience in the pursuit of an ideal imaging protocol in PET and PET-CT; Herman S from Indonesia who gave a current perspective on cervical cancer in Indonesia; Partha G from Singapore wrote on PET-CT clinical image interpretation of various carcinomas; David JG from Glasgow, UK dealt with the screening for colorectal cancer; Mok QQ from London, UK who deliberated on the new strategies in ventilation; Kenneth MC from Scotland, UK shared their UK solutions to the challenges of postgraduate medical education; John R from Toronto, Canada reviewed the recent advances in neurotology; and Jeremy DMR and Bill F from Coventry, UK who described the basic principles of the Values-based Practice: A new approach to understanding clinical interactions. And also our very own experts in their respective areas of specialty, such as, Goh KL, Lam SK, Tan CT, Azila AMN, Liam CK, Shyamala DS, Vijayan R, Chan LL, Yeoh PH, Ednin H, and so forth, whose abstracts were highlighted in this issue. The Editorial Board would like to thank Professor Liam Chong Kin, the Scientific Chair for compiling these abstracts.

The 2006 Issue 9 (1) bears witness to the strengthening of JUMMEC in line with the research activities of the Faculty. In that issue, you will find Chia YC's Editorial on more rational prescribing (5). Among others, that issue also carried articles on the resistance to antimicrobials and its problems, steroid withdrawal in transplant renal recipients, body-fat comparison of sport players, complementary medicine, usage of a simple neural network architecture to diagnose angina and a review on the trends of maternal mortality.

In this current issue, apart from the two papers on interdisciplinary research on impaired mobility disability and rehabilitation and the breast cancer review, many case reports – Grade IV hypertensive retionopathy, lumphangioleiomyomatosis, congenital anomaly of the urinary tract and neurogenic bladder, and squamous cell carcinoma of the tongue were presented.

Before we move on to the new year, we wish to record our thanks and appreciation to the Editorial Board members for their endearing support, energy and hard work in making the rebirth of JUMMEC a success. We know that the journey to JUMMEC had been long and challenging. We also wish to thank our panel of reviewers for assisting us with the review of articles. We look forward to an exciting year ahead and we invite you, the authors to continue contributing your articles for publication in JUMMEC.

#### References

 Wong YL, Mohd Amin J. Malaysian society and health: Issues and challenges in the 21st century. JUMMEC 2003-2005; 8: 2-8.

- (2) Elina RA, Husain R, Lang CC. Cardiopulmonary exercise testing: Utility in research and patient care. JUMMEC 2003-2005; 8: 9-17.
- (3) Wan Azman WA, Haizal HK. Peripartum cardiomyopathy: An overview. JUMMEC 2003-2005; 8: 18-22.
- (4) Moy FM, Atiya AS. Lifestyle practices and prevalence of obesity in a community within a university campus. JUMMEC 2003-2005; 8: 33-38.
- (5) Chia YC. Editorial. Towards more rational prescribing. JUMMEC 2006; 9(1): 1.

#### Rosmawati Mohamed, MD Low Wah Yun, Ph.D

**Low Wan Tun, Ph.D** Faculty of Medicine University of Malaya Kuala Lumpur

## BREAST CANCER AS A CHRONIC ILLNESS: IMPLICATIONS FOR REHABILITATION AND MEDICAL EDUCATION

#### Loh SY<sup>1</sup> and Yip CH<sup>2</sup>

Department of Allied Health Sciences, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia

<sup>2</sup> Department of Surgery, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia

ABSTRACT: Evidence suggests that breast cancer is taking the form of a chronic illness. This will add on to the present burden of managing chronic diseases in the healthcare delivery system. The burden of breast cancer being a chronic illness, calls for greater efforts to address the many neglected, physical-psycho-social and occupational functioning consequences. Timely efforts are needed to identify and implement interventions that are aim at improving the quality of life of women with breast cancer. At present, research evidence is highlighting that chronic diseases may best be managed using a self-management approach, and best treated by a balance of traditional medical care and the day-to-day practice of self-management skills. This paper presents the perspective of breast cancer as a chronic illness and its implication for rehabilitation and medical education. It is imperative that health professionals be made aware of these survivorship issues through medical education. The goals are to reduce the many disability risks, encourage patient-health provider communication and enhance partnership in care, within a timely, holistic therapeutic program to improve the quality of life of women with breast cancer. (*JUMMEC 2006; 9(2): 3-11*)

KEYWORD: Breast cancer, rehabilitation, medical education, self-management, chronic disease

#### Introduction

Worldwide, breast cancer is the second most commonly diagnosed cancer (1.15 million) after lung (1.35 million) (1). Incidence and mortality rates in breast cancer vary widely from one region of the world to another. Incidence rates are high in most of the developed areas (except for Japan, where it is third after colorectal and stomach cancers), and is modest in Eastern Europe, South America, Southern Africa, and Western Asia, but it is still the most common cancer of women in these geographic regions (1). The variations are attributed to the environmental differences in diet and lifestyle. These incidence rates can be up to five-fold higher in western countries than those reported from the Asian region (2). Age standardized incidence rates in developed countries are around 100/100,000 women with mortality rates of about 25/100,000 (3). The unfavourable trend of an increasing incidence of breast cancer is global, due in part to the increase in risk factors (decreased childbearing and breast-feeding, increased exogenous hormone exposure; and detrimental dietary and lifestyle changes, including obesity and less physical activity) (4). With its high incidence rates and relatively good prognosis (and a reducing mortality rate), breast cancer is the most prevalent cancer in the world today; with an estimated 4.4 million survivors up to five years following diagnosis (compared with just 1.4 million survivors – male or female – from lung cancer) (1,5).

In Malaysia, the International Agency for Research in Cancers estimated a crude incidence of about 34.86 per 100,000 population in 2000 (6). Breast cancer in Malaysian women occurs more commonly in younger

Correspondence: Loh Siew Yim Department of Allied Health Sciences Faculty of Medicine University of Malaya 50603 Kuala Lumpur, Malaysia Fax: 603-7967 4766 Email: syloh@um.edu.my women, aged between 40-49 years, with many presenting in the advanced stages similar to developing countries like Thailand and Pakistan (7). Despite its significant rise in incidence and a significant rise in long term survivorship, not much importance is being considered for the rehabilitation of women with breast cancer. There are many issues for women to manage, following the diagnosis and treatment for breast cancer. In the curative period, intervention are often too medically-focused, without due consideration to other aspects of functioning. In the survivorship period, follow-up medical appointments are often, narrowly and primarily confined to detection of recurrences (1), neglecting any needs for physical and psychosocial rehabilitation. Clinical manifestations of breast cancer can range from a localised to a metastatic neoplasm, and the treatment options are broad ranged, reflecting the heterogeneity of the disease. Despite many years of interests in evidence-based practice, it was recently highlighted in health services research that one of the most unfortunate, consistent finding is the gap between best practice (as determined by scientific evidence), and that of the actual clinical care. Even developed countries such as the United States had at least 30-40% of patients who do not receive care according to current scientific evidence (8). Adjustment to breast cancer implies coping and dealing with self-care issues as a result of the more obvious physical complications as well as the less obvious ones such as an altered body image, changes in relationship with partner and children, living with any ongoing side effects, and living with the intrusive thoughts of tumour recurrence, and the uncertainties of the futures. In Australia, only a minority of oncology patients with depressive illnesses are identified by staff and referred appropriately (9), although a study found that as high as 45% of women with early-stage breast cancer reported clinically significant levels of depression or anxiety (10). Health professionals need to explore their roles on how best to support these social adjustments through the illness trajectory. The traditional medical model of healthcare tends not to acknowledge that the mind and body are connected, prolonging a healthcare system that focus predominantly on curative and on the obvious physical impairments.

Indeed, studies on breast cancer and its rising health burden should be conducted alongside studies on current treatment, with careful consideration of its potential impact on functioning and quality of life, by both clinicians and medical researchers. However, despite an increasingly obvious significant survivorship, clinical research and therapy is slow and not forthcoming. The aim of this paper is to discuss the implication of breast cancer taking a form of chronic illness, for medical education and rehabilitation.

#### Breast Cancer – An Emerging Chronic Illness

Despite years of research to find the cause of breast cancer, the aetiology remains largely undetermined till today. What researchers know are merely the risks factors, and what clinicians can inform their patients are merely that they are at a certain level of risk. Scientist have been postulating on the many risk factors. Obesity, a lifestyle factor, is said to be specifically linked to lower risk in pre-menopausal women but the risk increases in post-menopausal women (11). The highest risk is linked to the BRCA genes and geographical location (12). In short, with an aetiology that remains idiopathic with numerous risk factors to blame, coupled with better treatment contributing to longer survivorship, breast cancer is slowly taking the form of a chronic illness (13).

Scientific evidence, from the highest level of randomized controlled trials down to expert's opinions, at the lowest, is therefore important. In January 2005, an International Consensus Panel of experts met during the 9th Conference on Primary Therapy of Early Breast Cancer in St. Gallen, Switzerland, to develop a series of guidelines and recommendations for adjuvant systemic treatments in specific patient populations (14). Use of tamoxifen, an antiestrogenic drug, has been the gold standard of treatment with its excellent results on women with oestrogen receptor (ER) positive tumours (15). However, new evidence suggests that aromatase inhibitors, which hinders the aromatase enzyme catalyzing the final step in oestrogen biosynthesis, have shown equally successful result in the prevention and treatment of breast cancer (16). Table I presents an overview of the survival rate in breast cancer, according to staging. With earlier detection and better treatment, these cancer survival rates are improving and have been reported to improve up to above 50% a decade ago (17) and the rates are still increasing. Better treatment leads to better prognosis and patients need to manage beyond adjuvant treatment periods, since longer survivorship often bring a myriad of survivorship issues.

Although some believe little can be done to reduce breast cancer risk through primary prevention (12), chemoprevention has been reported as an effective primary strategy (18). There must be further secondary prevention through screening (self and clinical breast examination and mammography), with rehabilitation continuing even after the curative phase to enable patients to adjust to the numerous survival issues. Thus, self-management skill at managing modifiable risks should now be taught to patients, besides the self-management education to equip them to be a

Stage	Disease-free survival (%)	Breast cancer specific survival (%)	Frequency of follow-up (monthly)			
			Year I	Years 2–3	Years 4–5	
DCIS†	90–95	98–100	3–4	6	6-12	
I‡	80–95	80–98	3–4	6	6-12	
11†	50–75	50–70	3	4–6	6-12	
III	50–66 (I5–35 <sup>‡</sup> )	30-60 (15-40‡)	3	3–6	3–6	
IV – metastatic	0–5	5–20	3	3	3–6	

Table 1.	Disease-free and 'BC specific sur	vival' by stage of breast cancer, a	at five years following optimal
	management, and suggested frequ	iency of follow-up	

\* Data from Stuart et al, 2006

<sup>†</sup> Treated with breast conserving technique (Stuart et al, 2006)



Figure I.	Yearly	risk	of	recurrence	in	breast	cancer
-----------	--------	------	----	------------	----	--------	--------

Source: http://www.webmd.com

partner in managing their breast cancers, in collaboration with their health providers.

Meta-analysis of the risk of breast cancer recurrence in each year during the ten-year period after a diagnosis of early breast cancer in patients who received no adjuvant treatment (treatment following surgery with or without radiation), indicates that risk of recurrence occurs throughout the ten-year period, but is highest in the first three years after a diagnosis, and in the absence of adjuvant follow-up treatment. Risk of recurrence is low after ten years survivorship (Figure 1). However, issues of recurrence can affect survivors' quality of life. Quality of life, is an increasingly accepted important outcome criteria in healthcare, and even amongst the early stages, many breast cancer patients often spend the remainder of their life in uncertainties, under the shadow of the possibility of a recurrence in the future.

As an emerging long-term (chronic) illness, breast cancer must be differentiated from acute and terminal illnesses. Acute illnesses are typically short-lived, have no permanent disruption of functioning and cure being almost always certain with predictable outcome. In contrast, terminal illnesses are life threatening with the mostly 'end-of-life' expected outcomes. The difference between acute and chronic illness are substantially different. Chronic illnesses, however, are those with an insidious onset and over a longer course duration. One definition of chronic illness is; "a disease which has one or more of the following characteristics: (a) is permanent, leaves residual disability; (b) is caused by non-reversible pathological alternation; (c) requires special training of the patient for rehabilitation, or may be expected to require a long period of supervision, observation, or care (19). With cure being uncertain, and its challenges over a longer time span, chronic illnesses can have an enduring effect

on quality of life. Therefore, any factors that affect quality of life can be critical intervention points.

## Burden of Breast Cancer as a Chronic Illness

As the health status of women with breast cancer can be impaired by the side effects of treatment and/or reactions to the cancer diagnosis and experiences, breast cancer like many other chronic illnesses, has an impact on many performance areas in a patient's daily life. Firstly, impairment and disability arising from breast cancer under the various functioning aspects can be wide ranging, and have wide ranging impact on a person's functional status and quality of life. Secondly, the disability in breast cancer to some extent is rather invisible, unlike the visually crippling disability in more physical-form chronic conditions like arthritis and stroke. Thirdly, in terms of duration, although impairment may not be permanent, the impact from living with uncertainty for a long duration is a real burden, and can be taxing on the person's quality of life.

Physical, psychosocial and psychosexual functioning are important sequelae that affects quality of life in women with breast cancer. Evidence suggested that primary treatment have resulted in impairment in emotional functioning, body image, sexual functioning, social-work role (20,21); anxiety, depression, fatigue and insomnia (22); impaired body image and self concept (23); pain, limited range and lymphoedema from surgical treatment (24-26); with an incidence rate of 10% axillary node dissection, to 24% dissection with radiation (27). Occupationally, a diagnosis of breast cancer can also hinder women from working for a minimum of a few months to years, and some women may never return to work. The reasons may be wide ranging from fears, myths, physical-psychological disabilities, as well as, to more positive reasons like revaluing life and life priorities. Some women see the diagnosis of breast cancer as a wake-up call and can afford to leave a paid job because, "I realized money is not everything, I want to spend time with my family and do things I enjoyed doing". However, not all women can afford not to work, as another survivor; "the treatment is so costly ... and with two small children to feed, I cannot afford not to work" (28). The implication of being occupationally 'unable' and/or 'disabled' can lead to other complications like lowered income, self-esteem, and reduced social contact with impact on the survivor's quality of life. Another important issue facing women with breast cancer is co-morbidity. Breast cancer is higher in advancing age, with risk of other illnesses (i.e., osteoporosis, heart disease and hypertension) setting in. Factors like complicated

#### Impact on Physical Functioning Issues

Evidence suggests breast cancer patients suffered impairments of the upper limb, even at one year after operation and many experiencing limited activities of daily living with negative impact on quality of life. Lymphoedema of the upper limb can occur even 20-30 years after axillary treatment (29), and it causes much mental distress. Estimates on the risk of severe lymphoedema can range from 6% with one treatment modality up to 30% with the combination of axillary clearance and radiotherapy. The persistence of a distorted body image, made worse with visible lymphoedema limbs can aggravate the difficulty to resolve the initial shock and denial, and/or the later adjustment phases. It is expected that with better and earlier screening, stages are detected earlier with less aggressive treatment and thus, lower complications, but more research is due in this area. One study found that two strong predictors of survivors' quality of life are arm-dysfunction and doctor-patient communication (30). These two aspects have been observed to be grossly lacking in our healthcare. Both deficits need to be highlighted in the medical education of the undergraduate as well as the postgraduate. Table 2 highlights many other common side-effects of breast cancer treatment can have an impact on the quality of life. An awareness of these consequences can help the health professionals in supporting their patients (29).

#### Impact on Psycho-sexual Functioning Issues

Breast cancer can have a debilitating impact on a woman's feelings of well-being, wholeness, sexuality and attractiveness to varying degree, since the breasts has been regarded as a symbol of femininity and beauty. Thus, a woman's choice of treatment can be influenced by her age, her belief and cultural background, religion and faith, as well as her hopes and fears. A single women who had a mastectomy shared, "society sees women as having two breasts ... and I felt I have lost my femininity" (28). Younger ones appear to have more difficulties, especially suitor-marriagechildbearing issues or societal-expected issues. However, guidelines stipulating age, as a criteria for convenience or based on popular beliefs, rather than per medical evidence is an issue for some women. A survivor in her 60s was angry that she was never told she could opt for breast reconstruction, and was 
 Table 2. Common physical side-effects from cancer treatment

#### Surgery

- 'Pulling' over the scar/pain/discomfort
- Scar contracture
- Paraesthesia in the axilla and medial upper arm (due to an axillary clearance)
- Lymphoedema
- Physical imbalance and difficulties with neck pain due to breast tissue loss (for those with big breasts)
- Intermittent jabbing neurological breast pains
- Intermittent pain in the upper arm on the side of an axillary clearance (may settles over 3-6 months)

#### Chemotherapy

- Tiredness/fatigue
- Hair loss
- Bitter taste in mouth (may clear few weeks after chemotherapy)
- · Peripheral neuropathy may persist

#### Radiotherapy

- Breast oedema and tenderness
- Hyperpigmentation in the first year
- Later, an increased density of the breast tissue
- Small decrease in the size of the residual breast

#### Hormonal therapy (tamoxifen)

- Hot flushes
- Tender breasts
- Gastrointestinal upset
- Vaginal dryness and discharge
- Decreased libido
- · Abnormal vaginal bleeding

#### Hormonal therapy

#### (Aromatase inhibitors or AI)

- Hot flushes
- Musculoskeletal pain (may have to stop AI)
- Vaginal dryness
- Osteoporosis

Adapted from Stuart et al, 2006

furious when told that the offer was only for women below 40 years old (28). Lack of plastic surgeons to perform the job should be addressed and immediate action taken to overcome the shortage, rather than addressing the issue at the expense of the patients.

In a multicultural society, ethnicity as a barrier to treatment adherence must also be considered. A

woman shared about her Muslim-convert sister who had vehemently refused mastectomy because of the fears that her husband may look for another wife. The decision has cost her life, and her survivor sister justifies, "I would rather have one breast and live, than to have two breasts and die" (28). This apt message has been adopted as a key message for women attending the four week SAMA (staying abreast, moving ahead) self-management program scheduled for Malaysian women. [The clinical trial program is currently conducted at the University of Malaya Medical Centre Kuala Lumpur]. Although she felt that she was luckier and more rational to choose life over a breast, it cannot be denied that losing a breast is a loss, there is a process of grieving over any lost and the impact would vary from one to another. Unfounded beliefs and myths (highlighted in several focus group with women with breast cancer) (28), and the fear of recurrence also interfere with intimacy. One example is the myth that toxicity of chemotherapy and cancer cells can spread through intimacy, and that sexual excitement can cause recurrences. These concerns on sexuality, attractiveness and beliefs aggravating a woman's sexual problems, are part and parcel of the psycho-sexual issues with implications on their occupational life roles. Thus, women need extra support and time to develop openness, and acceptance in overcoming the changes due to body image, fear of rejection, doubt of attractiveness, low self-esteem and mental distress.

#### Impact of Breast Cancer on Psychosocial Functioning

The nature of the breast cancer illness, its severity, the treatment and side-effects; coupled with many other personal-environmental factors like personality, finances, lifestyle prior to the illness and the access and level of social support, impact the ability to 'journey' and adjust to the realities of this long-term condition. Mental distress can occur in response to the uncertainty of recurrence, leading to depression and complicated grief. All these can be further aggravated by any prevailing cultural-religious myths, such as breast cancer being caused by bad 'karma' or a punishment for a past misdeed. Like any major illness, an expected search for the 'why' questions, or the need to find solace and hope occurs at a time when nothing else makes sense, which makes support so crucial.

A cultural stigma of a cancer diagnosis may also complicate patients' social functioning and roles. The consequences of the patients' own reactions, as well as others' reactions, can lead to increase in withdrawal and isolation, sometimes made worst by family who are profoundly affected, and unable to cope and/or unsure of how to react; and may need professional support. Women and their family may also need assurance that breast cancer is not a punishment from God for their bad deeds. One survivor shared on how she was ostracised by her in-laws who continuously injected the idea that she was receiving her dues for her past misdeeds. A belief that they are not alone in the challenges that they face can certainly add quality to life. They need the message that breast cancer is highly prevalent and can affect anyone, good and noble women, as well as even men.

Further losses come in the form of having to relinquish roles and responsibilities, because of myths like stress causes breast cancer. One survivor shared that her overly protective family barred her from doing any housework and she was told to just sit around and not to work to avoid stress. However, staying away from work causes one to lose the support system, as well as the financial security and become a potential burden to loved ones. These also may take a toll on one's self-esteem, self-worth and self-image; leading to more shame and possibly resentment and anger. Such myths must be nipped in the bud so that whatever decision women take are choices based on informed decisions. This is because the consequences of abandoning functional roles, leads to a feeling of dependency, and the emotions around the loss of status and control can threaten the once - healthy, capable and functioning individual.

The traditional medical model of healthcare tends not to acknowledge that the mind and body are connected, prolonging a healthcare system that focus predominantly on curative and on the obvious physical impairments. This care delivery is rather limiting and not in line with the progression of the development of the breast cancer disease. The neglects on the nonphysical impairment need to be redressed, with an acknowledgement that a lowered emotional-distress, and social participation can greatly impact the body in a positive way. Intervention must target at both the seen and on the 'unseen' impairments in broader life scope. For example, cognitions or thoughts must be deliberately considered, as potent determinants of how patients feel and they influence the choices patients make. A diagnosis of cancer, often trigger people's inner struggle around their faith, and at times, bring out self-imposed blame and punishment for having the condition. When a belief or thought is based on a premise that is distorted and self-defeating, an increase in distress may be experienced unnecessarily. Therefore, cognitive-restructuring coping skills must be taught to refute negative automatic thoughts or myths that underline patients' anxiety or depression. Such cognitive reappraisal enables them to focus on the positives, identify personal strengths and enhance their self-efficacy in coping and managing the condition. During this crisis period, the empathetic ability and/or training of the health professionals to render support can enable patients to reclaim the meaning of life behind the illness and/or assert their gratitude for their life. Health professional must be alerted on these extensive, at times, tremendous impact, and despair brought about by a cancer diagnosis. The capability of health professionals to respond sensitively to patients and their families is increasingly being seen as fundamental in this partnership in cancer care. Self-management of emotions, in collaboration with relevant health professionals, is crucial and it can improve the goal setting and problem solving skills for self-managing the condition more effectively.

#### The Crux of Managing Breast Cancer as a Long-term Illness

This review raises two important questions: (a) are there unique issues for women with a longer term survivorship, and (b) should women with breast cancer, as an emerging chronic illness, be treated differently from those yesteryears? The answer to the first question would depend on more research findings. The answer to question two is a strong definite 'yes'. In fact, breast cancer, once a short term potentially fatal illness, has now added to the burden of chronic illnesses as rising incidence rates and better treatments have contributed to more cases and longer survivorship. As the burden of chronic illness continues to rise and takes over as the number one source of healthcare expenditure worldwide, it is imperative to identify efforts and implement timely, culturally sensitive, and long-term, cost-efficient interventions for women with breast cancer.

Rehabilitation interventions for long-term conditions must now focus proactively on integration of medicalemotional-role management with a primary prevention emphasis on promoting healthy lifestyle; as well as, on secondary prevention defining risk reduction habits to keep cancer at bay, and to enhance participation and quality of life. Patients must be alerted on the need to make changes in behaviour and lifestyle in order to promote risk reduction habits. This is so pertinent in breast cancer as getting patients to re-focus on health (rather than looking for cure) makes much more sense as survivorship increase dramatically with better treatment. Our rehabilitation aim must now extend beyond reducing present disabilities (visible and invisible ones) towards more concerted efforts on facilitating im-



Silicone prosthesis



Attachable nipple

Figure 2. Some basic accessory aids for enhancing body image, posture and managing lymphoedema

Pressure garment

provement in participation and quality of life. These may require aids and adaptations, and prosthesis to adjust to impairments such as the commonly available aids in Figure 2 which is making an entry, albeit a slow one, in our therapy settings.

## The Essence of Self-management Interventions

Along the concept of self-care, is a current interest (in the ongoing conceptualization) of 'self-management', which is not about patients going alone managing by themselves. The essence of self-management is about collaborative care with health professionals. Von Korff and colleagues (31) assert that chronic disease is best treated by a balance of traditional medical care and the day-to-day practice of self-management skills; both can be enhanced by effective collaboration from the patients, their families and healthcare providers. Collaborative management is said to occur when patients and care providers have shared goals, a sustained working relationship, mutual understanding of role and responsibilities, and requisite skills for carrying out their roles (31). This is important since the impact of breast cancer is extensive and drawing from both experts (patient as expert in day-to-day management, and the healthcare providers as experts in the medical care) is the best management approach to reduce hospitalization cost and improve healthcare delivery. Self-management program supports patients to live the best possible quality of life with their chronic condition. Under this broad category of interventions, some may include informational and educational strategies, cognitive and behavioural skills, vicarious learning and peer support methods that focus on changing specific thoughts and healthy habits. Training to prepare patients to be informed, and activated may include problem solving skills, resource utilization, relaxation training and stress-coping training. The idea is rooted on the chronic care model where optimal chronic care is achieved when a prepared, proactive practice team work collaboratively with an informed, activated patient (31). A partnership approach to enable patients to self-manage is an integral collaboration for quality primary care.

## Implications for Medical Education and Rehabilitation

The recognition that breast cancer is gradually taking a form of a long-term illness requires a cognitive shift in understanding its unique implications. It differs from that of an acute illness or a terminal illness and this requires a paradigm shift in our management of breast cancer. As a long-term illness, breast cancer is comprehensive in its effects, as well as being tremendously varied from one person to another. The implication is more far ranging than merely treating the illness and symptoms as studied in many chronic illnesses. Thus, medical care for chronic illness is rarely effective in the absence of adequate self-care. Medical care with selfmanagement should be actively encouraged and promoted, and health professionals must be willing to adopt a less authoritarian approach towards more collaborative care. The attitude of 'I know best' may more likely interfere rather than complement the management of an increasing chronic condition and an increasingly affluent society. Self-management and partnership is the best way to manage a condition that can lasts for years or decades and its success requires a dire change, not just in the roles of our patients but in the knowledge, skills and attitudes of our health professionals.

Adjustment to breast cancer implies coping and dealing with self-care issues as a result of the more obvious physical complications as well as the less

obvious ones such as an altered body image, changes in relationship with partner and children, living with any ongoing side-effects, and the fear of tumour recurrence, and the uncertainties of the futures. Occupational therapists working with people with chronic illnesses spend considerable time rehabilitating them to improve their independent functioning in self-care and the overall activities of daily living. Each person's illness is very unique and can range from mild to severe limitation in participation and quality of life. Instead of resuming or returning to a previous lifestyle and functioning (as in an acute illness), or struggling with life (terminal illness), people with breast cancer are now faced with having to adjust to long term, and very likely, lifelong symptoms and limitations. The constant adjustment needed along the way requires ongoing support. Recently in Malaysia, two cases of death were reported; (a) the late first lady who was battling with a recurrence and (b) a 55-year old survivor of ten years from Alor Star, Kedah (32). Both were highlighted in the media. The contrast was the former fought till the end; whilst the latter, a mother of two boys, after being ten years of being disease-free, opted to terminate her life. This case should trigger a revisit on the clinical practice guidelines for management of breast cancer. Appropriate therapeutic intervention can reduce the high levels of emotional distress experienced by many women with breast cancer. Having an awareness of these medical and survivorship issues allows the health professional to provide important support to both the patient and her family. The role of rehabilitation after the active curative treatment must be initiated for breast cancer, to optimize quality of life. Optimizing quality of life is not only an important outcome in itself, but it is also associated with longer survival and lower levels of depression in breast cancer (33).

#### Conclusion

Ideas improve the healthcare delivery, but not till they are transformed into feelings (and sufficient enough to trigger action). Quality of life is often linked to what patients can do (the visible), less on how they feel and even less on what they think. Yet the reverse may be equally, if not more significant, in breast cancer. Thus, the recognition that healthy living is also important in such long-term conditions, must also be balanced with an awareness that it depends on positive attitude and an awareness on how to self-manage living with breast cancer successfully. Attempting to self-manage the actual illness on a daily basis, and the profound nature of the changing emotional consequences can lead to feelings of helplessness and hopelessness. Living with hope, when cure is so fleeting, is an important element that must be emphasized in rehabilitation. It brings hope and a belief that they can live through another day, in spite of experiencing the disease and the uncertainties of a recurrence. The aim of rehabilitation being to promote global physical and psychosocial adjustment by targeting at all modifiable levels, i.e., strengthening individual coping resources through psychotherapy, improving quantity and quality of familial and social support, and restoring optimal physical functioning (34). Managing health, emotions and roles as well as managing risk-reduction, thus, play a crucial part in the overall management of breast cancer, as in many chronic illnesses. Medical care with self-management should be actively encouraged and promoted, and health professionals must be willing to adopt a less authoritarian approach towards more collaborative care. Self-management and partnership is the best way to manage a condition that can lasts for years or decades and its success requires a dire change in the roles of patients as well as in the health professionals.

#### Acknowledgement

The authors wished to thank Prof Low Wah Yun for co-facilitating, and Ms Tan Wah Chah, Ms Peggy Lim and Dr Selina Khoo for taking turns to help out with the focus groups interviews held between June-August 2005. A report of the focus group qualitative study will appear in APJPH (2007: Vol. 19, No. 3). The SAMA clinical trial (4-week program) is currently recruiting patients for the clinical trial.

#### References

- Rojas M, Telaro E, Russo A, et al. Follow-up strategies for women treated for early breast cancer. Cochrane Database Syst Rev. 2000; 4 (CD001768).
- Lacey J, Devesa S, Brinton L. Recent trends in breast cancer incidence and mortality. Environ Mol Mutagen 2002; 39: 82-88.
- 3. Greenlee R, Murray T, Bolden S, *et al.* Cancer Statistics. CA Cancer J Clin 2000; 50: 7-33.
- Parkin DM, Fernandez LMG. Use of statistics to assess the global burden of breast cancer. Breast J. 2006; 12 (Suppl 1): S70-S80.
- 5. Parkin DM, Bray F, Ferlay J, et al. Global Cancer Statistics, 2002. CA Cancer J Clin 2005; 55(2): 74-108.
- Ferlay J, Bray F, Pisani P, et al. Globacan 2000: Cancer Incidence, Mortality and Prevalence Worldwide. Lyon; 2001. Report No.: Version 1.0 No. 5.

- Hisham AN, Yip CH. Overview of breast cancer in Malaysian women: a problem with late diagnosis. Asian J Surg. 2004; 27, 2: 130-3(2): 130-133.
- Grol R, Wensing M. What drives change? Barriers to and incentives for achieving evidence-based practice. Med J Aust 2004; 180 (6 Suppl): S57-S60.
- Ford S, Fallowfield L, Lewis S. Can oncologists detect distress in their out-patients and how satisfied are they with their performance during bad news consultations? Br J Cancer. 1994; 70: 767-770.
- Kissane D, Clarke D, Ikin J, et al. Psychological morbidity and quality of life in Australian women with early-stage breast cancer: A cross-sectional survey. Med J Aust 1998; 169(4): 192-6.
- Kuller L.The etiology of breast cancer from epidemiology to prevention. Public Health Rev 1995; 23: 157-213.
- Kelsey J, Bernstein L. Epidemiology and prevention of breast cancer. Annu Rev Public Health 1996; 17: 47-67.
- Fallowfield L. Evolution of breast cancer treatments: current options and quality-of-life considerations. Eur J Oncol Nurs 2004; 8S, S75-S82.
- Goldhirsch A, Glick JH, Gelber RD, et al. Meeting Highlights: International Expert Consensus on the Primary Therapy of Early Breast Cancer 2005. Ann Oncol 2005;16: 1569-1583.
- Jordan V. Estrogens and antiestrogens. In: Holland JF FE, Bast RC, Kufe DW, Morton DL, Weichselbaum RR Eds. Cancer Medicine. Philadelphia: Lee and Febiger; 1993. pp 857-865.
- Campos S. Aromatase inhibitors for breast cancer in post-menopausal women. Oncologist 2004; 9: 126-136.
- American Cancer Society. Breast cancer facts and figures, 2005-2006. Atlanta, Georgia: ACS; 2005.
- Kinsinger L, Harris R, Lewis C, et al. Chemoprevention of breast cancer – systematic review. Rockville, Maryland: RTI/University of North Carolina; 2002. http://www.ncbi. nlm.nih.gov/books/bv.fcgi?rid=hstat3.chapter.2527. (accessed 20 Jan 2006).
- DHA. Delaware Healthcare Association Glossary of Health Care Terms and Acronyms. In: Dover D, Ed. Health Care Professional Resources. Dover, Delaware: Delaware Healthcare Association; 1998.
- Irvine D, Brown B, Crooks D, et al. Psychosocial adjustment in women with breast cancer. Cancer 1991; 67(4): 1097-117.

- Irvine D. A critical review and meta-analysis of the literature investigating psychosocial adjustment in breast cancer. Ottawa: Health Canada: 1996.
- 22. Shimozuma K, Ganz P, Petersen L, *et al.* Quality of life in the first year after breast cancer surgery: rehabilitation needs and patterns of recovery. Breast Cancer Res Treat 1999; 56: 45-57.
- Rowland JH, Desmond KA, Meyerowitz BE, et al. Role of breast reconstructive surgery in physical and emotional outcomes among breast cancer survivors. J Natl Cancer Inst 2000; 92(17): 1422-1429.
- Tasmuth T, Von Smitten K, Kalso E. Pain and other symptoms during the first year after radical and conservative surgery for breast cancer. Br J Cancer 1996; 74: 2024-2031.
- Knobf M. Symptoms and rehabilitation needs of patients with early stage breast cancer during primary therapy. Cancer 1990; 15(66): 1392-1401.
- Knobf MT, Sun Y. A longitudinal study of symptoms and self-care activities in women treated with primary radiotherapy for breast cancer. Cancer Nurs. 2005; 28(3): 210.
- Johansson K, Ingvar C, Albertsson M, et al. Arm lymphoedema, shoulder mobility and muscle strength after breast cancer treatment. A prospective 2-year study. Adv Physiotherapy 2001; 3: 55-66.
- Loh SY, Packer T, Yip CH, et al. Perceived barriers to self-management in Malaysian women. Asia Pac J Public Health 2007; 19(3). (In press)
- Stuart K, Brennan B, French J, et al. Life after breast cancer. Aust Fam Physician 35(4), April 2006; 35: 177-258.
- Engel J, Kerr J, Schlesinger-raab A, et al. Predictors of quality of life of breast cancer patients. Acta Oncologica 2003; 42(7): 710-718.
- Von Korff M, Gruman J, Schaefer J, et al. Collaborative management of chronic illness. Ann Inter Med. 1997; 127: 1097-1102.
- 32. Embun M. "Cancer Mum I cut my wrist". The Star Newspaper (Nation) 2005 20th Oct; Sect. 23.
- Coates A, Hurny C, Peterson H, et al. Quality-of-life scores predict outcomes in metastatic but not early breast cancer. J Clin Oncol 2000; 18(22): 3768-74.
- Ronson A, Body J. Psychosocial rehabilitation of cancer patients after curative therapy. Support Care Cancer 2002; 10(4): 281-291.

## INTERDISCIPLINARY RESEARCH ON IMPAIRED MOBILITY, DISABILITY AND REHABILITATION IN SELANGOR

Manderson L<sup>1</sup>, Zaliha O<sup>2</sup>, Rameezan BAR<sup>2</sup>, Nooreini AH<sup>2</sup>, Soh SB<sup>2</sup> and Disler P<sup>3</sup>

<sup>1</sup> School of Psychology, Psychiatry and Psychological Medicine, Monash University, 3800 Australia

<sup>2</sup> Department of Allied Health Sciences, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia

<sup>3</sup> School of Rural Health, Monash University, 3800 Australia

ABSTRACT: Demographic, economic and social changes have had major impact on health and illness globally, including in Malaysia, and present significant challenges to the structure and delivery of health services. While these changes have influenced the epidemiology of disease, the diagnosis, experience and response to changes in health status for individuals and their families are influenced by additional environmental and personal factors. We describe these factors in relation to our ongoing research program on personal and social aspects of impairment and disability. The Resilience study aims to understand how people with impairments and their families live with chronic health conditions, how these conditions impact on self-esteem, social relationships and societal participation, and how structure, context and environment affect individual functioning, disability and well-being. We described our methodology and summarize the baseline data that will inform our future enquiries. (JUMMEC 2006; 9(2): 12-17)

KEYWORDS: Disabled persons, locomotor activity, Malaysia, quality of life, social impact

#### Introduction

The global increase in the prevalence of chronic diseases in middle and low as well as high income countries reflects both the success of public health interventions and improvements in the treatment of disease, and social changes related to industrialization, urbanization and globalization. In Malaysia, the demographic and epidemiological transition is such that patterns of morbidity and mortality are now similar to those of advanced economies (Western Europe, USA). Cardiovascular disease, cancer and diabetes, mental health problems and accidents and injury are the major causes of burden of disease. These conditions require ongoing medical treatment and appropriate health and welfare programs. The ability of individuals to maximize health and wellbeing following the development of chronic illness and impairment varies as a result of differential access to and the affordability of secondary prevention and treatments. In many cases, individual and community needs have outpaced developments in healthcare provision and expenditure (1).

In recognizing the prevalence of chronic illness and impairment, and the discrimination and stigma that often flow from them, the UN Economic and Social Commission for Asia and the Pacific declared a Decade for Disability in 1992-2001. The second decade (2002-2011) aims to continue the work to promote inclusive national and state government policies. This initiative has proceeded without a substantial evidence base, and there is still little research data on how people in the region live with chronic diseases and physical and mental impairments. In this paper, we introduce the research we are conducting in Malaysia on this subject, and highlight the importance of an interdisciplinary approach to

Correspondence: Lenore Manderson, PhD FASSA ARC Federation Fellow and Professor School of Psychology, Psychiatry and Psychological Medicine Faculty of Medicine, Nursing and Health Sciences Monash University, 3800 Australia Tel: 613-9903 4047 Fax: 613-9903 2501 Email: lenore.manderson@med.monash.edu.au address the social, economic and medical challenges that derive from this epidemiological transition.

#### Impairment and Disability in Malaysia

Malaysia's population is 25.6 million; life expectancy is around 70 for males and 75 for females (2). Most Malaysians can expect to remain independent and productive for a longer period than their parents, but increased longevity brings with it an increased incidence of chronic disease, with attendant problems for individuals, families and communities. Some 50% of Malaysian men smoke, an important risk factor of numerous cancers, cardiovascular disease and chronic obstructive pulmonary disease; traffic accidents are still common (2,3). Malaysia has escalating rates of hypertension and diabetes mellitus, in part associated with changes in diet and activity: nearly 30% of the population at the time of the National Health and Morbidity Survey 1997 (the most recent community data available) were obese or overweight and less than 12% of the population aged 18 years or older had "adequate" physical exercise (2). In addition to impairments following these conditions, approximately 7% of the Malaysian population are affected by congenital disability or disabilities acquired from accident, injury and disease in childhood and throughout adulthood (3,4,5). An additional 10-15% of the population are kin or care-givers to this population.

#### **Classifying Function and Disability**

Until recently, people with physical or intellectual impairments were classified using the International Classification of Impairment, Disability and Handicap (ICIDH) (1980). This system of classification emphasised clinical impairments, and while addressing appropriate management strategies for medical rehabilitation teams, it overlooked the social and environmental context and the influence of individual and structural factors on individual outcomes. Consequently, in May 2001, the 54th World Health Assembly adopted the International Classification of Functioning, Disability and Health (ICF) (6).

The revised classification system and its published guidelines provide a framework of disability that acknowledges the different perspectives of health from biological, individual and social perspectives, and provides a tool to describe and compare the health of populations in an international context. The overall aim of the classification system is to provide a unified and standard language and framework to describe health and health-related states. While it describes body functions and structures, it also incorporates broader health-related domains affecting activities and participation such as family structure, economic status, workforce participation and education. The ICF consequently has the potential to identify various disabling and beneficial psychosocial and environmental factors that are relevant in ensuring the full inclusion of persons with impairments. The ICF identifies meaningful sets of related physiological functions, anatomical structures, actions, tasks, and areas of life. The term 'functioning' encompasses all body functions, activities and participation. With the development of the ICF, WHO has moved away from classifying impairment and disability as a 'consequence of disease' classification (i.e., based on the impact of disease) to become a 'components of health' classification (7).

While criticism of this instrumentation continues (8,9), the approach in the ICF is consistent with the contemporary broad-based rehabilitation perspective, with emphasis on the links between health, functioning, and social context. In many respects, the ICF captures the work that has always been a component of medical rehabilitation, including as understood and practised in Malaysia. Rehabilitation medicine is the interdisciplinary management of a person's functioning and health. It aims to restore and maintain body structure and function, minimize activity limitation (impairment/ disability) and restriction in participation (handicap) and prevent further disablement. Depending upon the condition, rehabilitation involves physicians and surgeons, occupational, speech and physiotherapists, nurses, prosthesis and orthotics specialists, psychologists and social workers. Rehabilitation interventions may not change the underlying pathology of a given condition, but can improve a person's symptom control and minimize disability, so enhancing quality of life, improving participation in the community, and decreasing health care costs (7).

Associated personal and environmental factors are critical from a rehabilitation perspective. These have policy and community-wide consequences. For example, providing a wheelchair to someone unable to walk, and training the person to use the wheelchair, is of limited value if curbing and guttering in streets are inadequate, if there are no ramps to allow access to buildings and if public transport is not designed for access by people using wheelchairs. The use of a wheelchair alone also will not change social attitudes or discriminatory and stigmatizing practices that exclude people with physical disabilities from full participation in society and prevent them from maximizing their capabilities in employment, and family and community life. Our understanding of the complex factors that result in disability, however, is limited. In particular, the personal factors that facilitate outcomes remain to be identified, and further research is required to enhance our understanding of how different people respond to impairment and disability. Research is also still required to elucidate the social, cultural and economic factors that shape these personal aspects and contribute to the environment of rehabilitation and adaptation, in order to provide better clinical care and to contribute to appropriate policies and programs at a national and local level. Our ongoing research on social aspects of disability and disablement responds to this need.

## Resilience: Studying Social Aspects of Impairment and Disability

As noted, in a clinical context, rehabilitation involves an interdisciplinary team which acknowledges the social and physical environmental factors involved in meeting the needs of individuals who are born with or loose various functions. In research that addresses the broader context, this interdisciplinary approach expands to include social scientists, in order to examine individual and cultural understandings and social and economic consequences of impairment disability. The Resilience Study, the first stages of which we summarize here, was developed in this context. RESILIENCE is an acronym for REsearch into Social Inclusion, Locomotor Impairment and Empowerment through Networking, Collaboration and Education, and is a multi-country, interdisciplinary and collaborative study focusing on living with acquired disability in Australia, Thailand and Malaysia. The study brings together social scientists, public health researchers, clinical medical researchers and practitioners, and includes researcher-consumers, with the aim of generating information of relevance to social policy, rehabilitation medicine and community health.

The Resilience Study is designed to explore the experiences of disability at household and community level. The research focuses on the social context of disability and explores how personal, social, demographic and structural factors moderate or compound the impact of impairment. Drawing on both quantitative and qualitative data using approaches common in medical anthropology (10), the study explores lay understandings and life experiences of people who live with physical difficulties. Their health conditions are often complicated by psychological problems and cognitive difficulties, but also by social isolation and low economic status. Continued disability results in high costs at personal and societal levels in terms of medical and allied healthcare, and in everyday living. There is considerable variation within and between

societies and nation states in terms of how care is provided and how the costs of care are met.

The first phase of research focused on local patterns, understandings and life experiences of people who have locomotor impairments for any number of reasons, and the consequent marginalization of individuals and their families. In order to identify the experience of impaired mobility within the community, we adopted a broad definition of impairment, and so included any condition that was considered to affect mobility. These included conditions such as limb amputation from disease, impaired mobility from accident (resulting in spinal cord injury, for example), and loss of function from stroke, but also conditions such as morbid obesity, fractures, arthritis, and asthma which restrict the involvement of the individual in social and economic life.

## Quantitative and Qualitative Approaches

The baseline study was conducted in Selangor in 2003-2004. The quantitative component of the project recruited 210 participants aged 18 years or older. Following stratification into urban and rural areas, a two-stage modified cluster sampling method was employed. Thirty communities were selected randomly, and from health centres in each of these communities, seven individuals were identified as having impaired mobility and were recruited to participate in the study. While some of these participants had access to rehabilitation medicine or community-based rehabilitation services, others had no access to routine services, and no access to physiotherapy or occupational therapy. The quantitative survey used closed questions and scales to gather demographic, socioeconomic and psychometric data, with a structured schedule to collect basic demographic information and socio-economic data. General health and well-being was characterized using the EQ-5D (European Quality of Life Short Form) (11), which includes questions on mobility, self-care, pain and discomfort, ability to undertake usual activities, anxiety and depression, and general health status. In addition, a purpose-specific instrument - Perceived Impact of Problems Profile (PIPP) - was designed and used in the survey to identify self-perceived impact and distress from impaired mobility across a series of domains, including self, relationships, mobility, participation, employment, independence and future. Additional questions were open-ended.

The qualitative component was conducted in a subset of six communities, with a purposively selected population including people who had participated in the survey, who elaborated on their experiences of impairment. Other qualitative methods, including interviews with carers and key informants (e.g., health workers, community leaders), group discussions, community mapping and observation provided further contextual information. The qualitative interviews provided participants with an opportunity to discuss at length their individual circumstances, perceptions and experiences, to describe in their own ways and to reflect on their own understandings of the accident or disease, treatment and care, and the changes they experienced personally and socially. The interviews were open-ended; each began with an explanatory statement by the researcher, with each question building on the response of the participant to the prior question. Interviews were taped, checked and reviewed, and tapes were then transcribed for data analysis.

The quantitative data provided us with information on trends and patterns of behaviour, activities and ideas of people with disabilities and their carers. These were explained in depth from the qualitative data. We were not interested primarily in incidence, prevalence or causal links, but rather in identifying common patterns that emerge among people with disabilities. The complementary approach of quantitative and qualitative methods allows us to ensure internal validity, coherence and quality. The approach is inductive, concerned with identifying meaning and patterns rather than designed to test a strict set of hypotheses, and so, gathering "meaning rich" data takes precedence over aggregation and prevalence.

#### **Preliminary Results**

The study sample included 94 men (44.8%) and 116 women (55.2%), ranging in age from 18-90 (60.2 ± 16.6); men were on average younger than women (57.1 ± 17.5 for men, 62.7 ± 15.5 for women). Almost three guarters (72.4%) resided in rural areas, and partly reflecting this, mean years of education was low  $(5.1 \pm 4.2)$  and the majority (72.4%) had primary level education only. Sixty per cent of respondents were married and living with their spouses at the time of the study; 10% had never married and 29.5% were separated, divorced or widowed, the latter reflecting the age range of this population. Most (70.5%) were Malay and Muslim (71.9%); 13.8% were Chinese and 15.2% Indian with Buddhist, Confucian, and Hindu religious affiliation. A summary of the health status of the respondents in set out in Table 1, and illustrates that most (83%) experiencing some problems in

Table 1.	Health	profile of	study	participants	by a	age $(N = 210)$	
----------	--------	------------	-------	--------------	------	-----------------	--

	Age Range (Years)						
	Total	<u>&lt;</u> 55	56 – 70	71+			
	N = 210	N – 71	N = 79	N = 59			
		(33.8%)	(37.6%)	(28.6%)			
Duration of mobility problems							
– Range (years)	0 - 60	0 - 50	0 - 60	I – 25			
– Mean years & SD	6.8 ± 10.9	10.4 ± 14.5	5.4 ± 9.8	4.5 ± 4.3			
Cause of mobility problems (%)							
– Sickness/Illness	61.0	45.1	65.8	73.3			
– Accident	23.8	43.7	19.0	6.7			
– Since birth	2.4	4.2	2.5	0.0			
– Don't know	11.9	8.5	13.9	13.3			
– Others	9.0	8.5	5.1	15.0			
Other health problems (%) <sup>1</sup>							
– Hypertension	43.8	23.9	59.0	47.5			
– Diabetes	35.4	26.8	44.3	33.9			
– Arthritis	29.0	16.8	36.7	33.3			
– Stroke	18.2	7.0	25.3	22.0			
– Heart disease	14.8	7.0	12.7	26.7			
<ul> <li>Lung disease (including asthma)</li> </ul>	8.7	2.8	11.5	11.8			
- Tuberculosis	2.9	1.4	5.1	1.7			
– Others	12.1	15.5	9.1	11.9			

<sup>1</sup>Participants reported more than one health problem

walking, with 9% confined to bed. Only a small proportion of the sample (8%) was able to walk about without problems, for example, because they felt confident using a gait aid. More than half (62%) of the sample perceived that they had no problems with self care, and one third (31%) felt that they had some problems in this respect. The remaining 7% of the sample were not able to wash or dress themselves. More than half of the sample (53%) felt that they could perform their "usual" activities, including working or doing housework or family activities, but with some problems. While 26% of the respondents did not have problems with performing their usual activities, 21% reported that such activities were impossible to perform.

More than half of the respondents (63%) reported experiencing moderate pain or discomfort at the time of interview, and more than one quarter (27%) experienced extreme pain or discomfort. Only a small proportion (10%) was free of pain or discomfort. While nearly one quarter of the respondents (23%) reported no feelings of anxiety or depression, 46% and 31% respectively were moderately or extremely anxious or depressed. When asked about their feelings regarding general health (using the scale of 0-100), the single most frequent response was the mid-point of 50 (mean = 60).

As described, the PIPP instrument collected data across a number of domains. The major differences were that there was a greater impact on relational items (confidence, mood, satisfaction) and on the ability to have a close relationship for younger people than older respondents. There was also a trend towards greater impact in the emotion items for urban participants compared with rural participants in terms of overall satisfaction with life, moods and feelings, and sense of confidence, and greater distress on the relational items for younger people in terms of ability to relate to people in authority, ability to relate to neighbours and friends and ability to relate to relatives. Interview data with health workers emphasized that these people often lacked social support to allow them to interact with others and participate in society, and in consequence the social exclusion is one reason for people's reported distress and depression.

The qualitative data allows for some interpretation of these findings. People drew attention to the impact of loss of mobility on their mental health and on their independence and autonomy. One man, for example, recalled that he was very depressed: "I used to walk and live an independent life, then suddenly I can't do everything. I realized it's fate but what can I do? I just

have to be really patient and leave it up to God." Many participants drew on faith-based ideas – that their health condition was in God's hands, for example, or that their health condition was the result of fate. Older participants reflected on the inevitability of aging, and drew solace from the fact that they had already lived a full life. Participants also reflected on the difficulty of being as independent as possible, rejecting family support that they felt was inappropriate and demeaning. Hence the comment, "I feel annoyed most of the time when people come to me and try to do things for me. Let me do it myself. If I can't, then I'll ask for help."

While people aspired to be as independent as possible, data from health workers and community members indicate that there is considerable misunderstanding about the capability of individuals with limited mobility. One health worker, for example, reflected that "if they (people with limited mobility) do have support, it is only partial ... and they have low self-esteem and low self-confidence." The assumption that such people have "low self-confidence", in turn influences how others in the community interact with them. Thus a village headman explained that "we invite all residents including the disabled, whenever we conduct any program for the village, but whether they want to join in or not is their own decision. We never force them to be involved. Maybe because some of them have low self-esteem, although we never treat them differently." Yet at the same time, this participant and others acknowledged other practical barriers to social participation and inclusion, such as lack of an attendant carer, transportation problems, financial difficulties and environmental barriers.

#### Conclusion

The Malaysian Government is faced with numerous challenges to current health policies and systems that derive from changes in standards of living, changing disease patterns (from communicable to noncommunicable diseases) and demographic changes. The increased incidence in chronic and degenerative diseases related to changes in lifestyle and behaviour, for example, highlights the importance for changes to occur in the mix, integration and delivery of health services, and changes in public health and prevention. The need for review and, as necessary, reform of medical and health services has arisen concurrently with increased commitment to ensuring equity and maximizing the social participation and capabilities of all citizens. The former Ministry of National Unity and Social Development (now the Ministry of Women, Family and Community Development) presented a draft Persons with Disabilities Act for discussion in parliament in 2002. This Act provides the legislative means for strategies and programs to ensure such participation and inclusion. Improved and appropriate social and medical strategies are required to ensure that Malaysians with chronic illness or disability enjoy productive lives and maximize their capabilities. The consequent changes in policies, programs and strategies to ensure universal access to appropriate health care and services, and to ensure the inclusion of people with impairments, require a strong evidence base. This points to the need for continued research regarding social as well as medical aspects of chronic disease, impairment and disability. Our research on social aspects of impairment is a first step towards this end.

#### Acknowledgement

The baseline research was facilitated by the Australian Research Council (ARC) Federation Fellowship Grant (Manderson, No. FF0241243), an ARC Discovery Grant (Manderson, Bennett and Disler DP0449614, The University of Melbourne), and an Intensified Research Priority Area (IRPA) Grant No. 06-02-03-0534. The research teams from all three countries met as a consortium in June 2003 in Khon Kaen (northeast Thailand) and in May 2004 in Vientiane, Lao PDR to share data methods, instruments and analysis. Ethics approval for the research in Malaysia was provided by the University of Malaya Medical Centre, Medical Ethics Committee (Ethics No. 344.12). Team members involved in various ways in the development of instruments, data collection and data management and analysis are Akmal Hafizah, Kumaran s/o Ramakrishnan, Yi Lin, Noor Ezawanee Badaruddin, Kavitha Uma, Maslina Yazid, Shaari Mohd Yatim, Elizabeth Bennett, Julie Pallant, Sari Andajani-Sutjahjo and Cate Bailey.

#### References

- Mathers CD, Sadana R, Salomon JA, et al. Healthy life expectancy in 191 countries, 1999. Lancet 2000; 357: 1685-1691.
- Hussein RHT, Anuar H. Report of the Second National Health and Morbidity Survey Conference, Hospital Kuala Lumpur, 20-22 November 1997. Kuala Lumpur: Public Health Institute, Ministry of Health, Malaysia 1997.
- Karim HA. The elderly in Malaysia: demographic trends. Med J Malaysia 1997; 52, 3: 206-212.
- Mustaffa BE. Diabetes epidemic in Malaysia. Med J Malaysia 2004; 59, 3: 295-296.
- Sherina MS, Rampal L, Mustaqim A. The prevalence of depression among the elderly in Sepang, Selangor. Med J Malaysia 2004; 59, 3: 45-49.
- WHO (World Health Organization) International Classification of Functioning, Disability and Health: ICF. Geneva: WHO, 2001.
- Stucki G, Ewert T, Cieza A. Value and application of the ICF in rehabilitation medicine. Disabil Rehabil 2002; 24, 17: 932-938.
- Imrie R. Demystifying disability: a review of the International Classification of Functioning, Disability and Health. Sociol Health IIIn 2004; 26, 3: 287-305.
- Schuntermann MF. The implementation of the international classification of functioning, disability and health in Germany: experiences and problems. Int J Rehabil Res 2005; 28, 2: 93-102.
- Manderson L. Gender, normality and the post-surgical body. Anthropology and Medicine 1999; 6(3): 381-394.
- Brooks R. EuroQoL: the current state of play. Health Policy 1996; 37, 1: 53-72.

## MALIGNANT SERTOLI CELL TUMOUR: AN UNCOMMON TESTICULAR TUMOUR

#### Sia SF<sup>1</sup>, Dublin N<sup>1</sup>, Nurul B<sup>2</sup> and Wong KT<sup>2</sup>

Department of Surgery, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia

<sup>2</sup> Department of Pathology, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia

ABSTRACT: We report a case of an 86 year old Chinese man who presented with a painless right testicular swelling that had persisted for one year. There was no history of maldescend or cryptorchid testes. Clinical and ultrasound examination revealed testicular tumour with two round masses within the right scrotal sac, with minimal fluid seen within the sac. Tumour markers were normal. He subsequently underwent a right inguinal orchidectomy under local anaesthesia as he had an underlying cardiac insufficiency. Histopathological examination revealed malignant Sertoli cell tumour. True Sertoli cell mesenchyme tumours constitute less than 1% of all testicular cancers. Current literature on histopathological and clinical features and treatment options are reviewed. (JUMMEC 2006; 9(2): 18-21)

**KEYWORDS: Malignant Sertoli cell tumours** 

#### Introduction

Sertoli cell tumour is a rare testicular stromal neoplasm in adults and constitutes less than 1% of all testicular tumours. It is usually benign (1), and only approximately 10% are malignant. However, it is among the most common testicular neoplasms in canines (2). In humans, more than 130 cases of malignant Sertoli cell tumours have been reported worldwide (2). We report the first case of malignant Sertoli cell tumour of the testis in Malaysia.

#### **Case Report**

An 86 yearold Chinese man with underlying ischaemic heart disease presented with painless right testicular swelling. The swelling had been present for one year. There was no history of maldescend or cryptorchid testes. He denied testicular or groin pain, fever, chills, weakness, weight loss or other constitutional symptoms. Physical examination revealed an enlarged right testicle. There was no gynaecomastia. Ultrasound examination revealed a testicular tumour with two round hyperechoic masses within the right scrotal sac with minimal hydrocele. Serum levels of  $\alpha$ -fetoprotein,  $\beta$ -human chorionic gonadotropin and lactate dehydrogenase were normal. After taking his underlying cardiac insufficiency into consideration, he underwent right inguinal orchidectomy under local anaesthesia.

#### Histopathology

Grossly, the testis measured 6 cm x 3.5 cm x 3 cm, and the haemorrhagic epididymis was approximately 3.5 cm in length. Cut section of the testis showed a multiloculated cystic structure containing serous fluid, measuring 2 cm x 1.8 cm x 1.5 cm and an adjacent well circumscribed firm yellowish lesion measuring 3 cm x 3 cm x 2.5 cm. Both the lesions were within the tunica albuginea (Figure 1).



Figure 1. Multiloculated cystic with firm mass (arrow) in orchidectomy specimen

Correspondence: Dr Sia SF Department of Surgery Faculty of Medicine University of Malaya 50603 Kuala Lumpur, Malaysia Tel: 603-7949 3852 Email: sfsia02@um.edu.my Histological sections showed that the yellowish mass consisted of sheets of tumour cells exhibiting mainly tubule formation and an alveolar like pattern. The tumour cells were large, polyhedral, with small hyperchromatic nuclei and clear cytoplasms which contained fat (as demonstrated by oil red O stain) (Figure 2). There was little glycogen within the cytoplasm. Mitosis and necrosis were present (Figure 3). In addition some papillary areas were noted. There was infiltration by neutrophils. No hyaline globules were identified and immunohistochemistry for  $\alpha$ -



**Figure 2.** Shows large, polyhedral cells with small hyperchromatic nuclei and clear cytoplasm which contain fat (oil red O stain: original magnification x 40)

fetoprotein was negative. Although this tumour was largely encapsulated, there was one area of capsular and vascular invasion (Figure 4).

The spermatic cord (surgical margin) was free of tumour. A section from the cyst wall showed thick mature fibrous tissue mainly devoid of lining epithelium and some areas were lined with single layer cuboidal cells. Inactive seminiferous and hyalinized/ fibrosed tubules were present outside the cyst wall. No *in situ* malignant change was noted.



Figure 3. Shows area of necrosis (arrow) (Haematoxylin and eosin stain: original magnification x 10)



**Figure 4.** Shows area of capsular and vascular invasion (*arrow*) (Haematoxylin and eosin stain: original magnification x 10)



Figure 5. CT scan shows ill-defined mass at right kidney (white arrow)

Computerised tomography of the chest, abdomen and pelvis did not reveal any presence of mediastinal, paraortic or pelvic lymphadenopathy. The liver, spleen, pancreas and left kidney were normal. However, the right kidney appeared irregular in shape with an ill-defined mass seen at the right mid and upper pole measuring 2.9 cm  $\times$  2.2 cm  $\times$  2.5 cm (Figure 5).

As the general condition of the patient was poor, the family declined further treatment. No further investigations were performed, although it was speculated that the right kidney was involved by the Sertoli cell tumour.

#### Discussion

Sertoli cell tumours also known as androblastoma or gonadal stromal tumours, are rare testicular stromal neoplasm and comprise 0.4-1.5% of all testicular tumours (3). Patients with malignant Sertoli cell tumours show a wide age range, similar to that observed in patients with benign Sertoli cell tumours. Sertoli cell tumours have been reported in newborns (2,3) and in patients as old as the octogenarian in the present case. Mean age of these patients was reported as 39 years old (4). Most patients present with painless testicular swelling. These tumours occur with equal frequency in either testes, and are occasionally associated with cryptochid or maldescend testes. Almost 30% of reported cases have been associated with gynaecomastia (5). It has also been reported that the majority of cases were hormonal inactive and laboratory hormonal assays were inconsistent (3). Eble *et al* (4) reported that tumour size varies, and that size is not a reliable indicator of tumour behaviour or prognosis.

Diagnosis by fine needle aspirate has been proposed (6). However, this is not normal practice and the authors do not favour this method as there is a risk of cutaneous dissemination. The only absolute criterion for the diagnosis of malignancy is made by histopathology of the orchidectomy specimen along with the demonstration of metastasis. The diagnosis of this tumour can be difficult, frequently misinterpreted as seminoma (7).

The basic pathological pattern is of cells resembling normal Sertoli cells arranged in the nests and tubules, with or without lumina. Mitotic figures are occasionally present, and are associated with poorer prognosis. Three distinct patterns have been reported, (a) well demarcated nodules (b) replacement of the entire testis and (c) infiltration (4).

In the present case, malignancy was diagnosed on the basis of microscopic features, namely the combination of mitosis, necrosis and evidence of local vascular invasion.

Sertoli cell tumours commonly metastasize via the lymphatic system. Haematogenous spread to the lung, bone, pleura, thigh and adrenal gland has been reported (5). In a large series of Sertoli cell tumours, no synchronous renal mass or renal metastasize were reported. In this case the renal pathology could have been a metastatic deposit or a synchronous renal cell carcinoma. A renal biopsy would have been useful.

Radical inguinal orchidectomy remains the mainstay of treatment. Retroperitoneal lymph node dissection and adjuvant radiation therapy have been advocated to offer the best result (4), however, results of chemotherapy have been disappointing. These tumours may recur years after the ochidectomy, therefore long term follow-up is recommended (8).

#### References

- Peterson RO. Testis in Urologic Pathology. Philadelphia: JB Lippincott 1992; 487-491.
- 2. Talerman A. Malignant sertoli cell tumour of the testes. Cancer 1971; 28(2): 446-455.
- Kolon TF, Hochman HI. Malignant sertoli tumour in a prepubescent boy. J Urol 1997; 158(2): 608-609.
- 4. Eble JN, Hull MT, Warfel KA. Malignant sex cord stromal tumour of testes. J Urol 1984; 131(3): 546-550.
- 5. Hopkins GB, Parry HD. Metastasizing sertoli cell tumour (Androblastoma). Cancer 1969; 23(2): 463-467.
- Nijhawan R, Dey P, Savitri K, et al. Fine needle aspiration cytology in malignant sertoli cell tumours of the testes. A case report. Acta Cytol. 1992; 36(4): 563-565.
- Henley JD, Young RH, Ulbright TM. Malignant sertoli cell tumours of the testes: a study of 13 examples of a neoplasm frequently misinterpreted as seminoma. Am J Surg Pathol. 2002; 26(5): 541-550.
- Herrera LO, Wilk H, Wills JS, et al. Malignant (Androblastoma) sertoli cell tumours of testes. Urology 1981; 18(3): 287-290.

## A CASE OF CONGENITAL ANOMALY OF THE URINARY TRACT AND NEUROGENIC BLADDER

#### Sia SF and Dublin N

Department of Surgery, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia

#### Introduction

Anomalies of the genitourinary tract occur in approximately 10% of the population and account for about one third of all congenital malformations in children (1). Congenital malformations involving the genitourinary system are major therapeutic challenges faced by the practising urologist.

#### **Case Report**

A 31 year old Chinese man with spina bifida presented with right loin pain and fever. Ultrasonography of the abdomen showed right hydronephrosis and absent left kidney. He had a long history of lower urinary tract symptoms mainly, frequency with occasionally difficulty in passing urine since childhood. There was, however, no history of urinary incontinence. A nephrostomy tube was inserted and antibiotic administered which successfully treated the urinary tract infection. Antergrade pyelography showed ureteric obstruction in the distal ureter at its ectopic insertion. The patient complained of loin pain when attempts were made at clamping the nephrostomy tube. Cystoscopy and ureteroscopy showed an ectopic ureteric insertion at the prostatic urethra (Figure 1) and a huge dilated tortuous right ureter with gross hydronephrosis. The bladder was also trabeculated with saccules (Figure 2). The patient had mildly impaired renal function.

Urodynamics done showed a hypocompliant bladder (10.7 ml/cm H20) with a maximum cystometric capacity (MCC) of 250 cc. (Maximum Pdet at MCC is 55 cm H20). Micturating cystourethrogram (MCU) showed grade 5 refluxing megaureter. The DPTA nuclear scintigraphy showed a grossly hydronephrotic right kidney with a glomerular filtration rate (GFR) of 52 ml/min with a prolonged drainage (> 60 mins). Plain radiograph showed spina bifida at L4, L5 and S1 levels with scoliosis of the lumbosacral spine. Computerised tomography of the abdomen revealed absence of the left kidney.



**Figure I.** An ectopic ureteric insertion at the prostatic urethra (*arrow*)



Figure 2. Endoscopic appearance of the trabeculated bladder with saccules

Correspondence: Dr Sia SF Department of Surgery Faculty of Medicine University of Malaya 50603 Kuala Lumpur Malaysia Tel: 603-7949 3852 Email: sfsia02@um.edu.my The patient subsequently underwent reimplantation of his right ureter (Figure 3) and an augmentation ileocystoplasty was done for his hypocompliant bladder. Post-operatively, he had to do clean intermittent catheterization (CIC). In the first year after surgery, he had several episodes of UTI which were successfully treated with antibiotic. There was no evidence of deterioration in renal function.



Figure 3. Tapered ureter implanted to bladder wall

#### Discussion

This paper presents a unique combination of malformations. Solitary kidneys are relatively rare with an incidence of approximately I in 1000 autopsies. In clinical practice, a solitary kidney is discovered approximately I in 1500 urological patients, and are twice as common in males as in females (2).

Ectopic ureters are relatively uncommon but their true incidence is uncertain because many are asymptomatic. Embryologically, the distal insertion of an ectopic ureter is the result of the ureteric bud arising more cephalad than usual on the mesonephric duct. Consequently, the ureter remains attached to the mesonephric duct longer, migrates more caudally than normal and is absorbed or inserted into either the urinary tract distal to the bladder or the reproductive tract. The most common sites of the ectopic ureteric insertion in male are posterior urethra (47%), seminal vesicle (33%), prostatic utricle (10%) (3,4). Ectopic ureters can also drain into other wolffian duct structures, like the epididymis and the vas deferens. As all these locations are proximal to the external sphincter, males with ectopic ureters do not usually suffer from urinary tract infection but occasionally from constipation, pelvic pain, discomfort during ejaculation, and infertility (5).

The combination of a solitary kidney with an ectopic ureter is rare but the combination of an ectopic ureter of a solitary kidney with prostatic insertion, associated with an underlying neurogenic bladder and vertebral anomalies have not been reported previously to the best of our knowledge.

In conclusion, this unique case report stresses the importance of:

- investigating a male patient with urinary tract infection;
- congenital malformations of the genitourinary system could be associated with other complexed medical or developmental problems, as seen in this case of neurogenic bladder.

#### References

- Rubenstein M, Meyer R, Bernstein J. Congenital abnormalities of the urinary system: a post-mortem survey of developmental anomalies and acquired congenital lesion in a children's hospital. J Pediatric 1961; 58, 356.
- 2. Brown, M. Congenital unilateral renal agenesis. Thesis. The University of California, Los Angeles, 1966.
- 3. Ellerker AG. The extravesical ectopic ureter. Br J Surg 1958: 45: 344-53.
- Schlussel RN, Retik AB. Anomalies of the ureter. In Walsh PC, Retik AB, Vaughen ED Jr, Wein AJ, Eds. Campbell's Urology (7th edition), Vol 2, Philadelphia: WB Saunders, 1998; 1820-1829.
- Squadrito J Jr, Rifkin MD, Mulholand SG, et al. Ureteral ectopia presenting as epididymitis and infertility. Urology 1987; 30:67.

### MODIFIED VACUUM-ASSISTED CLOSURE

#### Sia SF and Fong EP

Division of Plastic Surgery, Department of Surgery, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia

ABSTRACT: The treatment of large ulcers is a problem. Closure with flaps and even skin grafts in compromised patients is often fraught with risks. Dressing these large wounds is also a problem, as they are often heavily exudative. We report our experience using the vacuum-assisted closure technique in managing difficult ulcers in four patients. A modification of the technique using cheap, easily available materials is described. The vacuum-assisted suction is an effective treatment for large ulcers in compromised patients. It facilitates considerable contraction of the wound, and in smaller wounds; the vacuum-assisted closure allows sufficient contracture to enable delayed primary closure. We recommend the use of this method of wound dressing and closure in large wounds particularly, in high-risk patients where extensive flap coverage or even split skin graft would incur considerable risk. (*JUMMEC 2006; 9(2): 24-27*)

KEYWORDS: Modified vacuum-assisted closure, difficult ulcers

#### Introduction

The vacuum-assisted closure (VAC) system, which uses sub-atmospheric pressure and is non-invasive, has been widely used in the management of ulcers.

In this technique, the negative pressure is applied uniformly to the wound. This allows the surrounding arterioles to dilate, increasing the local circulation. Negative pressure also removes excess fluid from the wound and reduces oedema. In addition, it also enhances wound healing by promoting the growth of granulation tissue (1,2). In many instances, it can be replaced by the use of simple, cheap, easily available material. Modification of this technique using the conventional closed disposable vacuum suction in the ward has produced equally good results.

We report our experience using a modified vacuumassisted closure technique in the management of difficult ulcers.

#### **Case Reports**

#### Case 1

A 60 year old lady presented to us with a large, sloughy diabetic ulcer over the left inner thigh area, which had existed for a month. Following debridement, the resulting cavity measured  $160 \times 120$  mm in

diameter (Figure 1). Following irrigation of the wound, a 14F-size nasogastric tube was placed in the depths of the wound. The proximal end was then bought out at the edge of the cavity. The cavity was loosely packed



Figure 1. Diabetic ulcer over the left thigh following debridement

Correspondence: Dr Sia SF Department of Surgery Faculty of Medicine University of Malaya 50603 Kuala Lumpur, Malaysia Tel: 603-7949 3852 Email: sfsia02@um.edu.my



Figure 2. Diabetic ulcer over the left thigh following application of vacuum-assisted closure dressing



Figure 3. Dehiscence of the midline laparotomy wound



Figure 4. Wound almost closed following vacuum-assisted suction dressing

with iodine-soaked gauzes. An occlusive dressing (Opsite, Smith & Nephew, Hull, UK) was placed over the defect to achieve airtight closure, with the edges of the dressing extending 3-5 cm over the adjacent intact skin, converting the open wound into a controlled closed wound. The drainage catheter was connected to a wall suction vacuum unit continuously to maintain a tightly collapsed state and crinkled appearance of the occlusive dressing. The vacuum pressure was maintained at 150mmHg (-200mBar). The wound was inspected and the dressing changed daily. Analgesia in the form of pethidine injection was given prior to each dressing change. The management of the patient's wound in this manner did not hinder her mobility, as she was instructed to disconnect the tubing from the wall unit if she wished to mobilize or use the toilet or shower. The tubing was simply plugged with a stopper during mobilization.

Within one week of treatment, the wound had contracted to one third of its original size, allowing split skin graft (Figure 2).

#### Case 2

A 61 year old man underwent elective right hemicolectomy for carcinoma of the caecum one year ago. He was admitted for a recurrent tumour at the right iliac region of the abdominal wall as well as the underlying small bowel. Excision of the recurrent tumour was done and the resultant abdominal wall defect closed using a loco-regional tensor fascia lata flap with underlying prolene mesh. Two days post-operatively, however, an area of wound dehiscence was noted at the midline laparotomy incision as well as the edge of the tensor fascia lata flap (Figure 3). Both wounds were connected with extensive undermining.

The wound was dressed with vacuum-assisted suction with total closure of the deep wound and the reepithelization of the surface within three weeks (Figure 4).

#### Case 3

A 70 year old lady had bilateral total knee replacement done for severe osteoarthritis. There was left knee surgical wound dehiscence associated with significant undermining, making the cavity double the size of the skin defect (Figure 5). Modified vacuum-assisted suction dressing was applied enabling wound contracture and split skin graft was done one week later (Figure 6).

#### Case 4

A 91 year old diabetic lady developed nercotizing fasciitis in the ward following a coronary artery bypass graft procedure. She had suffered a myocardial infarct



Figure 5. Wound dehiscence following total knee replacement



**Figure 8.** Vacuum-assisted suction was done with two nasogastric tubes placed in the wound bed. Both tubes were joined with a Y-connector before connecting to the main suction tubing.



Figure 6. The wound following application of split skin graft



Figure 9. The wound at one month just prior to split skin graft



**Figure 7.** Wound at posterior thigh following debridement prior to the application of suction dressing

six months previously. The extensive necrotic area on the posterior aspect of the right thigh and gluteal region was debrided twice and daily dressing of the wound with modified vacuum-assisted suction was done (Figures 7 and 8). After one month, the wound had contracted considerably with a healthy granulating bed allowing a simple split skin graft procedure (Figure 9).

#### Discussion

In 1911 the application of mechanical stress onto wounds was described (3), which accelerated the healing process by promoting angiogenesis and tissue growth. Mechanical deformation of the cells resulted in an increase in protein and matrix molecule synthesis.

The use of topical negative pressure onto wounds has been well published and its value as a treatment modality well documented. Application of controlled sub-atmospheric pressure to a wound remove the excess interstitial fluid, reduces tissue turgor, decreases the oedema and enhances local blood flow. This in turn will improve the local tissue oxygenation and nutrient supply, increase the outflow of toxin and inhibitory factors and decrease tissue bacterial levels (1,2,4,5). Thus, healthy granulating tissue appears much more rapidly. The original conventional sub-atmospheric pressure system which employs a medical grade reticulated polyethane ether foam dressing with embedded non-collapsible drainage tube is costly and not easily available especially in small district hospitals. Modifying the technique by using absorbent gauze, occlusive dressing, nasogastric tubing and continuous controlled wall suction or a portable vacuum suction pressure machine, we have found the technique simple and effective both in terms of cost and result. It is easily taught to nursing staff. There is immediate improved local hygiene with rapid formation of granulation tissue.

Some series have reported that exposed bone and tendon in orthopaedic patients may be covered with VAC technique (6). In our patient (Case 3), the skin and soft tissue cover over the knee was thin with significant oedema making retraction of tissue off the underlying bone and bony exposure a real risk. Modified vacuum-assisted dressing provided rapid wound contracture.

Although in our series, the negative pressure is connected to wall suction apparatus, the therapy did not require undue immobilization of any of these patients. The patients were allowed to disconnect the drainage tube from the wall suction entry and ambulate as advised.

Bacterial counts were reported to drop from approximately  $10^7$  per gram of tissue to  $10^2$  to  $10^3$  per gram tissue in animal studies and in human samples by day 5 (1,2). Bacterial levels of less than  $10^5$  per gram of tissue are associated with progressive successful wound healing.

The four patients reported here were staying in the ward throughout the treatment. No difficulties were encountered during the dressing procedure. Successful healing without surgical intervention was obtained in Case 2. In the other three patients, a simple split skin graft procedure could be performed on the healthy granulating contracted wound.

Some patients may require analgesia with dressing change. Some degree of pain may be experienced when the tubing is first connected to the suction apparatus and sub-atmospheric pressure is transmitted to wound. Gradual negative pressure increment resulted in better tolerance as opposed to a sudden increment. Occasionally, movement caused displacement of the occlusive dressing leading to loss of vacuum. To remedy this situation, the patients were advised to check the seal (i.e., constant airtight closure with crinkling of the occlusive dressing). Should an air leak be present, this could simply be resolved by reinforcing with additional layer of occlusive dressing, overlapping the edge of the first layer. However, if excess egress of exudate or soiling of the wound with faecal material accompanies loss of seal, the dressing should be changed completely.

Bleeding from the wound is another possible complication of using the vacuum-assisted suction. This may occur during dressing change. Hemostasis can be easily secured with pressure.

#### Conclusion

The modified vacuum-assisted suction technique is an effective treatment for large or difficult ulcers in compromised patients. It enhances and facilitates considerable contraction of the wound and in smaller wounds; the vacuum-assisted suction allows sufficient contracture to enable delayed primary closure. We recommend this method of wound dressing and closure in large wounds particularly, in high risk and compromised patients. Cost efficiency can be optimized in the care of many patients.

#### References

- Argenta LC, Morykwas MJ. Vacuum-assisted closure: a new method for wound control and treatment. Clinical experience. Ann Plast Surg 1997; 38(6): 563-567.
- Morykwas MJ, Argenta LJ, Shelton-Brown El, et al. Vacuum-assisted closure: a new method for wound control and treatment. Animal studies and basic foundation. Ann Plast Surg 1997; 38(6): 553-562.
- 3. Thoma R. Ueber die Histomechanik des Gesfasssystems und die Pathogenase der Angioskleroose. Virchows Arch. F. Pathol. Anat 1911; 204:1.
- Shaer WD. Inexpensive vacuum-assisted closure employing a conventional disposable closed suction drainage system. Plast Reconstr Surg 2001(1); 107: 292-293.
- Collier M. Know how: vacuum-assisted closure (VAC). Nurs Times 1997; 93: 32-33.
- DeFranco AJ, Argenta LC, Marks MW. The use of vacuum-assisted closure theraphy for the treatment of lower extremity wound with exposed bone. Plast Reconstr Surg 2001; 108(5): 1184-1191.

## FREE RADIAL FOREARM FLAP AFTER PARTIAL GLOSSECTOMY FOR SQUAMOUS CELL CARCINOMA OF THE TONGUE

#### Chuah UC, Kamarul T, Sara T

Department of Orthopaedic Surgery, University of Malaya Medical Centre, 50603 Kuala Lumpur, Malaysia

ABSTRACT: Squamous cell carcinoma of the tongue is a highly malignant condition and results in high mortality and morbidity in patients despite its early detection (1). Early surgical interventions have been found to reduce mortality but in many reports, tongue reconstructions using live grafts have been found to reduce normal tongue function of speech, swallow and taste. In contrast, our report using free radial forearm flap (FRFF) to reconstruct the defect left over after a radical tongue resection in a 38-year-old gentleman with oral cancer has shown promising results. This type of reconstruction has left the patient with a functional and cosmetically acceptable tongue with minimal alteration in recognizable speech. (JUMMEC 2006; 9(2): 28-31)

KEYWORDS: Radial forearm flap, tongue reconstruction, free flap

#### Introduction

Oral cancer is a debilitating condition and although the condition is malignant, early detection has resulted in good results with prolonged survivability. However, due to the aggressive nature of the disease, despite early detection, the morbidity following extensive resection is undesirable. Our technique of using free radial forearm flap (FRFF) has proven to reduce the morbidity of this condition and in addition, to help replace the function of a tongue loss in the resection.

#### **Case Report**

A 38 year old Chinese man was referred from Selayang Hospital in May 2005 to University of Malaya Medical Centre with a 4–5 months history of pain and progressive ulcer over the right side of his tongue. He smokes occasionally but has no history of betel nut chewing habit. No family history of cancer is noted.

He was initially seen at our Maxillofacial Department and a biopsy was taken in April 2005. The HPE result confirmed the diagnosis of squamous cell carcinoma (SCC) which was moderately differentiated. Subsequently, he was referred to the Hand and Microvascular Surgical Unit for pre-operative assessment and assistance. On examination, there was a hard mass noted on the right side of his tongue with an overlying ulcer (Figure I). The tongue was also oedematous. There were palpable firm lymph nodes on the right side of the neck.



Figure 1. Cancerous lesion found on the tongue which was very similar to one seen in the patient in our report (6)

Correspondence: Dr Chuah Uei Chyi Attn: Dr. Tunku Kamarul Zaman Tunku Zainol Abidin Department of Orthopaedic Surgery Faculty of Medicine, University of Malaya Tel: 603-7949 2061 Email: ucchuah@perdana.um.edu.my Email: tkzrea@yahoo.com Surgery was performed involving the joint efforts of the Maxillofacial Department, ENT Department and the Hand and Microsurgery Unit. This involved tumour resection and tongue reconstruction. The mandible was split using a surgical saw. This approach offered an excellent exposure to the excision and reconstruction of the tongue. A near total glossectomy was done over the right side. Careful resection and clearance of the tumour margin was performed. While the Maxillofacial team began their resection, radical neck dissection was performed by the ENT team.

Simultaneously, a FRFF was harvested from the left forearm. The defect over the donor site was then closed partially, leaving a minimal area of space devoid of skin without much tension over the surrounding skin. This area was then covered using a split-skin-graft from the left thigh. During this surgery, the radial forearm flap was harvested with a cuff of brachioradialis muscle to facilitate contouring of the reconstructed right tongue. Anastamosis of the radial artery of the flap was performed to the nearby mandibular artery. Successful anastamosis was confirmed with bleeding observed at the edges of the free flap.

The patient recovered well post-operatively. The flap survived and the skin healed. During the follow-up, the patient was able to talk in a recognizable voice and he has no regurgitation or swallowing difficulties. On the third month follow-up, the patient maintains a good life with minimal functional disability.

#### Discussion

Squamous cell carcinoma of the oral tongue typically affects men from the sixth through the eighth decades of life, usually after many years of alcohol or tobacco abuse. Less than 4% of these lesions occur in patients younger than 40 years of age. Recent reports suggest that there is an increased incidence of oral SCC in the under-40 age group. Retrospective analyses performed suggested that the disease follows a more aggressive course in young patients but there are no significant differences in the outcome exist between the different age groups (1).

There is a 10% mortality noted in the first two years of follow-up in the older age group and 45% in the younger group. Therefore, effort should be made on identifying young patients whose disease behaves aggressively and require radical treatment (1). Metastatic spread of SCC of the tongue is facilitated by its rich lymphatic network and tends to increase with the size of the primary tumour. Approximately 50% of tongue cancers present with lymph node involvement. Early and recognizable cancer of the tongue often allows surgical intervention as part of the treatment management. However, in order to strike a good therapeutic control of the disease, a wide primary excision is often needed. This is followed by an optional radiotherapy treatment. This procedure usually leaves the patient with a large defect in the tongue and oral cavity. The absence of the tongue will impair the articulation, swallowing and vocalization processes. It is therefore prudent that reconstruction is needed for a good functional substitution for the defect. As for this patient, a non-innervated FRFF was used. As with any surgical reconstruction procedures, four main issues arise:

- 1. Survivability of patient following surgery;
- 2. Functional status of the reconstructed tongue;
- 3. Cosmetic appearance following reconstruction for both donor and recipient sites; and
- 4. Morbidity as a result of the surgery.

The radial forearm flap is commonly used for reconstruction of tongue defects following tumour excision. This flap is easy to harvest and offers thin tissue with large-caliber vessels. However, its use leaves behind a conspicuous aesthetic deformity in the forearm and requires the sacrifice of a major artery of that limb, the radial artery. The survival rate of FRFF is estimated at 92%. In one study, the disadvantages of the radial forearm flap included donor site morbidity which included partial loss of skin graft (11%), abnormal sensations (26%), poor appearance (8%), and reduced grip strength (11%) (2). In some patients, the donor-site scar of the forearm acted as a social stigma, preventing these patients from leading a normal life (3).

Some surgeons advocate an innervated FRFF in an attempt to produce a more favourable outcome (4). It is their belief that while the flap effectively fills the defect, it serves as an insensate reservoir in which food and saliva can collect. However, a number of studies have shown that very little and insignificant benefit is achieved when compared to an insensate flap (4,5). It has also been suggested that sensory innervated flap other than the FRFF is much more superior as compared to non-innervated flap (4). In their study, the lateral antebrachial cutaneous nerve to the (divided) lingual nerve (4) was used. On the other hand, some authors argued otherwise by saying that although the trend in this study is towards improved function with the innervated flaps, these flaps do not appear to offer major intra-oral functional advantage over the non-innervated flaps, which attain reasonably effective sensory recovery from neural ingrowth, if the lingual nerve is intact (5).



Figure 2. Post-operative picture of reconstruction of the tongue

Other methods of tongue reconstruction following a partial glossectomy were also mentioned in literature. More recently, the anterolateral thigh flap in a thinned form, has been used to reconstruct defects of the tongue with functional results equivalent to that of the radial forearm flap. The advantage of this flap is that it reduces the donor site morbidity as wounds can be closed primarily. Another neurovascular island flap used for reconstruction can be derived from the infrahyoid. With the neurovascular infrahyoid flap, defects of the tongue base can be reconstructed successfully after partial resections or total glossectomies. The main advantage is the voluntary innervations of this flap by means of the ansa cervicalis and the prevention of scarring and atrophy of the reconstructed tongue. This flap, however, is only suitable for small tumours of the tongue and offers very little to the more common situations where wider resections are required. Less commonly, lateral upper arm free flap, pectoralis major flap and a combined brachiaradialisradial forearm flap can be used instead of the FRFF. However, having compared these flaps to FRFF, we have yet to find further advantage other than the fact that the anterior thigh flap offers slightly improved cosmetics.

Functional outcome of a reconstructed tongue is seen by its ability to mimic the original function of a normal tongue prior to the diseased state. In this case, it is to review its function to assist deglutition, speech and taste. It is also important to factor the patient's satisfaction following the surgery. Because the free flap does not have the taste sensory, the return of taste cannot be expected in our procedure. However, swallowing and speech in this patient has returned to near normal with satisfaction rated very high at three months following surgery. Compared with primary closure of the tongue defect, some authors suggest it is better to reconstruct it with a FRFF when more than 50% of the tongue is resected (1,6). Although previous reports indicated that articulation intelligibility was better in patients who did not receive grafts as compared to those who received grafts, our patient showed otherwise (6). This is because reconstructions with flaps have been accused of interfering with the flexibility and mobility of the tongue which may contribute to articulatory impairment.

On the overall, patient satisfaction for function and cosmesis of the operated sites following any surgery with regards to tongue reconstruction rates high when using flaps as a closure following wide resection of cancer of the tongue (1,2,4,6,7). Although the surgery involved is lengthy and requires the joint effort of various expertise, the outcome for patients with this deadly disease is rewarding with high recovery rates being reported in cases where detection is early. Furthermore, by improving our techniques of using functional flaps as closures as opposed to mere resection, not only can the cancer be treated successfully, the morbidity following surgery can be greatly reduced whilst maintaining the function of resected part.

#### References

- Popovtzer A. Squamous cell carcinoma of the oral tongue in young patients. Laryngoscope 2004; 114(5): 915-917.
- Chen C-M. Complications of free radial forearm flap transfers for head and neck reconstruction. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2005; 99(6): 671-676.
- Boyd B, Mulholland S, Gullane P, et al. Reinnervated lateral antebrachial cutaneous neurosome flaps in oral reconstruction: are we making sense? Plast Reconstr Surg 1994; 93(7): 1350-1359; discussion 1360-1352.

- Kuriakose MA, Loree TR, Spies A, et al. Sensate radial forearm free flaps in tongue reconstruction. Arch Otolaryngol Head Neck Surg 2001; 127(12): 1463-1466.
- Netscher D, Armenta AH, Meade RA, et al. Sensory recovery of innervated and non-innervated radial forearm free flaps: functional implications. J Reconstr Microsurg 2000; 16(3): 179-185.
- 6. http://www.who.int/oral\_health/publications/fact\_sheet\_ tobacco/en/index1.html. (accessed 7 Dec 2006).
- Chuanjun C, Zhiyuan Z, Shaopu G, et al. Speech after partial glossectomy: a comparison between reconstruction and non-reconstruction patients. J Oral Maxillofac Surg 2002; 60(4): 404-407.

## GRADE IV HYPERTENSIVE RETINOPATHY AS A RARE FIRST PRESENTATION OF HYPERTENSION: A CASE REPORT

#### Tengku Kamalden TA, Choo MM and Fauzi A

Department of Ophthalmology, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia

ABSTRACT: Malignant hypertension affects less than 1% of people with high blood pressure, and is a hypertensive emergency. It is rare for patients to present initially with this form of elevated blood pressure, which is almost always associated with acute target organ damage, which can manifest in many forms including ocular, neurological, cardiac and renal. This treatable condition is associated with a high rate of morbidity and mortality therefore, early detection and immediate management is of paramount importance. (JUMMEC 2006; 9(2): 32-34)

KEYWORDS: Malignant hypertension, hypertensive emergency, mortality

#### Introduction

Hypertension is a major risk factor for the development of serious complications affecting the heart, brain, kidneys, eyes and the vascular system, leading to death and disability on a huge scale globally. As more and more Malaysians are diagnosed with hypertension every year, it is important for clinicians to be aware of the acute and chronic changes seen in a fundoscopic examination.

#### **Case Report**

A 45 year old previously well Malay man presented with gradual blurring of vision in both eyes over a period of two weeks. This was preceded by six weeks of intermittent occipital headaches which were aggravated by eye straining to focus when reading. There was no nausea or vomiting. He denied any limb weakness or numbness.

On examination, visual acuity was OD 6/12, ph 6/12 and OS 2/60. The anterior segment examination was normal for both eyes, with no relative afferent papillary defect. Dilated fundus examination showed bilateral papilloedema with splinter haemorrhages temporal to the disc in the left eye and multiple cotton-wool spots at the posterior pole (Figure 1). A macular star and mild macular oedema was also noted, which explained the poor left vision. There was narrowing of the arterioles with arteriovenous nipping. His blood pressure was recorded as 230/130 mmHg with a pulse rate of 84 beats per minute. Neurological and cardiovascular assessments were normal. The rest of the systemic examination was unremarkable.

He was diagnosed as having malignant hypertension with Grade IV hypertensive retinopathy and choroidopathy. He was referred to the physicians for urgent management of hypertensive crisis. His blood pressure was reduced by titration of isosorbide dinitrate infusion to maintain a reading of lower than 160/90 mmHg. Renal function tests showed renal failure with elevated serum creatinine and urea levels of 16.6mmol/l and 732µmol/l respectively. Urine examination showed gross proteinuria at 5g/l, and 24-hour urine protein was markedly elevated with a reading of 1.8g. An ultrasonography of the kidneys showed bilateral small kidneys which measured 7.60cm and 8.05cm in bipolar length in the right and left kidneys, respectively. This was suggestive of bilateral renal parenchymal disease. All these evidence confirmed the presence of chronic renal failure.

Correspondence: Dr Tengku Ain Fathlun Tengku Kamalden Department of Ophthalmotology Faculty of Medicine University of Malaya 50603 Kuala Lumpur, Malaysia Email: ain@ummc.edu.my



**Figure 1.** The right fundus showed multiple cotton-wool spots with papilloedema. There was an early macular star in the left eye as shown by the black arrow. (OD: right eye; OS: left eye)



Figure 2. Fundus photos taken two months later showed almost complete resolution of the hypertensive retinopathy and choroidopathy changes with minimal residual hard exudates at both macula.

His blood pressure normalized to 130/80 mmHg by the third day of admission, and his left vision improved to OS 6/18. He was counseled regarding renal failure and the future need for dialysis. He was subsequently discharged with oral anti-hypertensive agents and iron supplements.

Three months following his discharge, his vision improved to a score of 6/9 in both eyes, and fundus examination showed complete resolution of the hypertensive changes and papilloedema (Figure 2). By the eighth month following the first presentation to the eye clinic, his best corrected vision recovered fully to 6/6 in both eyes. There was no evidence of optic atrophy in either eye.

#### Discussion

Malignant hypertension, or accelerated hypertension, is a rare syndrome consisting of rapid and severe elevation of blood pressure with the systolic component above 200 mmHg or the diastolic blood pressure greater than 140 mmHg. The presence of systemic findings defines malignant hypertension. These include ocular, cardiac, renal and cerebral injury. Persistent malignant hypertension can lead to a rapidly fatal course with heart failure, myocardial infarction, stroke or renal failure.

Besides retinopathy, hypertensive choroidopathy may also occur, causing serious detachment, Elshnig's spots

and Siegrist's streaks. These changes are caused by late leakage of choroidal vessels and focal necrosis of the retinal pigment epithelium. Cotton-wool spots usually resolve by 3-6 weeks, but arteriolar narrowing and AV nipping changes are permanent (1).

Immediate management of malignant hypertension is aimed at lowering the blood pressure in a controlled fashion to a level that minimises end-organ damage. Too rapid a decline can lead to ischaemia of the optic nerve head, brain and other vital organs, resulting in permanent damage. Treatment in this patient was initiated in a controlled, monitored setting under the close supervision of a physician.

With appropriate clinical diagnosis, hypertensive emergencies can be successfully treated and the complications can be prevented with timely intervention (2). From a systemic viewpoint, the diagnosis of a malignant hypertensive crisis represents a medical emergency. Untreated, it has a high mortality rate of 50% at two months and 90% at one year (1). Thus early recognition of signs and symptoms, and early diagnosis is imperative to prevent fatalities.

The visual prognosis in malignant hypertension is good, with most patients resuming normal vision. However, on the rare occasion of visual loss, this may result from retinal pigment changes secondary to retinal detachment or from optic atrophy due to prolonged papilloedema. The visual prognosis in this patient was good, with full recovery of vision over time, along with the control of his blood pressure.

#### References

- Yanoff M, Duker J, Eds. Ophthalmology, 2nd ed. St Louis: Mosby, 1999; 849-53.
- Tuncel M, Ram VC. Hypertensive emergencies: etiology and management. Am J Cardiovasc Drugs 2003; 3(1): 21-31.

## A CASE OF LYMPHANGIOLEIOMYOMATOSIS WITH MULTIPLE ANGIOMYOLIPOMAS IN THE KIDNEYS: A CASE REPORT

#### How SH<sup>1</sup>, Azlin S<sup>2</sup>, Pang YK<sup>3</sup> and Liam CK<sup>3</sup>

<sup>1</sup> Department of Internal Medicine, Kulliyyah of Medicine, International Islamic University Malaysia, P.O. Box 141, 25710 Kuantan, Pahang

<sup>2</sup> Department of Radiology, Kulliyyah of Medicine, International Islamic University Malaysia

<sup>3</sup> Department of Medicine, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia

ABSTRACT: Lymphangioleiomyomatosis (LAM) is a rare disease. The reported prevalence is around one per million in the West but the exact prevalence in Asia is unknown. It affects primarily young women of reproductive age, with a mean age of onset at 34 years. The disease is characterised by an abnormal proliferation of smooth muscle-like cells that grow over a course of time to obstruct airways, lymphatic and blood vessels. We report a case of LAM in a young woman who presented with a spontaneous pneumothorax. Computed tomography scan of the thorax and abdomen showed homogenous thin-walled cystic lesions with normal intervening lung parenchyma and multiple lesions in her kidneys with Hounsfield units similar to that of fat. (JUMMEC 2006; 9(2): 35-38)

KEYWORDS: Lymphangioleiomyomatosis, renal angiomyolipoma, spontaneous pneumothorax, cystic lesions of lungs

#### Introduction

Lymphangioleiomyomatosis (LAM) is a rare disease. The reported prevalence is around one per million population in the United Kingdom, France and the United States of America (1). The disease has been well documented in many Asian countries. However, the prevalence data of this disease in Asia is not available (1). It affects primarily young women of reproductive age, with a mean age of onset at 34 years. The disease is characterized by an abnormal proliferation of smooth muscle-like cells that grow over a course of time to obstruct airways, lymphatic and blood vessels. It usually occurs sporadically, but occasionally may occur in association with tuberous sclerosis complex. A case of LAM associated with renal angiomyolipoma is described with a particular focus on the diagnostic difficulty encountered.

#### **Case Report**

A 36 year old Chinese woman first presented in October 2001 to the University of Malaya Medical Centre with a complaint of cough with episodes of haemoptysis for six months. She did not have reduced effort tolerance, fever, weight loss or reduced appetite. She denied taking any oral contraceptive pill. She used to smoke about ten cigarettes per day for three years during her teens. There was no history of tuberculosis infection or tuberculosis contact. She worked as an office clerk after completing her secondary school education.

Physical examination during the first hospital admission showed that she was comfortable at rest with no signs of respiratory distress. She did not have digital clubbing or fever. Findings on respiratory examination were unremarkable. Her chest radiograph was reported as "abnormalities consistent with bilateral generalised bronchiectasis". Her full blood and differential white cell counts, serum immunoglobulin and alpha-I-antitrypsin levels were normal.

Correspondence: Dr Pang Yong Kek Lecturer Department of Medicine Faculty of Medicine, University of Malaya 50603 Kuala Lumpur, Malaysia Email: ykpang@ummc.edu.my

#### JUMMEC 2006: 9(2)

Conventional spiral computed tomography (CT) scan of her thorax and abdomen performed after hospital discharge revealed "multiple cysts of varying sizes in the lungs, as well as multiple bilateral hypodense lesions in the kidneys with a Hounsfield unit of fat. Mild ascites and abdominal lymphadenopathy were also noted (Figure I and 2). These CT scan findings were thought to be consistent with LAM and multiple renal angiomyolipoma, probably secondary to underlying tuberous sclerosis. However, the patient was treated as having bronchiectasis despite the CT scan findings.



Figure 1. White arrows show multiple thin-wall cystic lesions, which are quite homogenous in size scattered throughout both lungs. The intervening lung parenchyma appears normal.



**Figure 2.** Contrasted axial-CT at the level of both kidneys revealed a lobulated mass in the right kidney (shown with black arrow) with density similar to that of fat (margins shown by white arrows). This finding is typical of renal angiomyolipoma.

Sixteen months later in February 2003, she presented with a right-sided hydro-pneumothorax. Her chest radiograph also showed reticulonodular infiltrates and small cystic lesions in both lungs with blurring of the cardiac margins. The left lung volume was preserved. The right hydro-pneumothorax was drained by an intercostal chest tube under water-seal. Minimal straw-coloured fluid was drained. She was discharged two weeks later when the right lung was fully reexpanded.

On clinic follow-up, she remained quite well except for occasional episodes of haemoptysis. A review of the patient's thoracic CT scan which had been taken in 2001, revealed that she had had diffuse cystic lesions in the lungs without a predilection for central or peripheral involvement. These cystic lesions were thinwalled, quite homogenous in size and not adjacent to the vasculature of lung. The intervening lung parenchyma was normal. The clinical diagnosis was revised in February 2004 from bronchiectasis to that of LAM. A respiratory function test performed revealed a forced expiratory volume in the first second (FEV<sub>1</sub>) of 58% of predicted; forced vital capacity (FVC), 78% of predicted; total lung capacity (TLC), 96% of predicted; residual volume (RV), 130% of predicted and an FEV<sub>1</sub>/ FVC ratio of 62%. Her lung diffusing capacity (DLCO) was only 54% of predicted. She underwent an open lung biopsy of her right upper, middle and lower lobes which revealed emphysematous changes without typical changes of LAM on histopathological examination. As she only had minimal symptoms, she was not keen to undergo another lung biopsy and treatment. A repeat CT examination in January 2005 did not show any change in the lung lesions nor the size of the renal angiomyolipoma.

#### Discussion

Despite being described in scientific literatures since 1937, the aetiology of LAM remains elusive. However, hormonal factors particularly, oestrogen and progestorone seem to play a major role in the initiation, progression and retardation of the disease. This is based on the observations that the disease occurs exclusively in women, worsens during pregnancy and exacerbates after hormonal therapy in post-menopausal women (1). Treatment with progesterone and/or oophorectomy in certain reported series seems to afford retardation of disease progression or even improvement of the condition (1). Furthermore, oestrogen and progesterone receptors have been found in LAM tissue. In a study in the United Kingdom (UK) by Johnson (2), the commonest presentations of LAM are pneumothorax (38.7%), dyspnoea (36.7%) followed by chylous pleural effusion (6.1%). Renal angiomyolipoma was found in 12.0% of the patients in this series. If the incidence in Malaysia with a population of about 25 million is comparable to that of UK, we would expect to see around 25 cases of LAM in the country and we expect only two or three patients to have renal angiomyolipoma. Pulmonary function test may show an obstructive (commonest), restrictive or mixed pattern with reduced DLCO. As in this patient, the residual volume is usually increased (3).

The diagnosis of LAM can be difficult. In one series (3), the exact diagnosis was made after an average delay of 44 months (ranging from 1 to 219 months after the first symptoms). In our patient, the definite diagnosis was made after 36 months from the time of first presentation even though the diagnosis was suspected by a radiologist on the first presentation. This was probably due to the lack of awareness among physicians on this rare condition. Plain chest radiograph during the early stages of the disease is often normal in appearance. Later, when the disease advances, mild reticulonodular shadows or small cystic lesions would appear (1). These changes, however, are often misinterpreted as some other more common diseases, e.g., bronchiectasis, as seen in this patient. Hence, when clinical features of the disease are not compatible with radiological diagnosis based on chest radiograph, further investigation with high resolution CT scan is advisable.

The classical findings of LAM described on chest radiograph are bilateral reticulonodular interstitial infiltrates, with normal or increased lung volumes (3). However, these findings also occur in other diseases, such as chronic hypersensitivity pneumonitis, pulmonary histiocytosis X, sarcoidosis and emphysema coexisting with idiopathic pulmonary fibrosis (3). High resolution thorax CT scan findings (as described above for this patient) are characteristic but not pathognomonic of LAM because patients with chronic histiocytosis X may have similar CT appearances (2). Open lung biopsy has been the gold standard to diagnose this disease. However, in the presence of typical clinical features and characteristic CT findings of LAM, open lung biopsy may be omitted (1). Furthermore, abdominal CT scan in this patient also revealed features consistent with renal angiomyolipoma, ascites (which could have been chylous) and abdominal lymphadenopathy, which are well known association with LAM.

#### Conclusion

In conclusion, the diagnosis of LAM should be considered when a young female presents with spontaneous pneumothorax and CT scan findings of homogenous thin-wall cystic lesions and normal intervening lung parenchyma.

#### References

- Johnson S. Lymphangioleiomyomatosis: clinical features, management and basic mechanisms. Thorax 1999; 54: 254-264.
- Johnson R, Tattersfield AE. Clinical experience of lymphangioleiomyomatosis in the UK. Thorax 2000; 55: 1052-1057.
- 3. Sullivan EJ. Lymphangioleiomyomatosis: a review. Chest 1998; 114: 1689-1703.

## LIST OF REVIEWERS

The Editorial Board of JUMMEC acknowledge with gratitude the following individuals for having completed reviews.

Associate Professor Saimy Ismail University of Malaya Kuala Lumpur

Associate Professor Dato' S Noor Abdul Ghani University of Malaya Kuala Lumpur

Professor Wan Azman Wan Ahmad University of Malaya Kuala lumpur

Professor Mohd Afandi Muhamad University of Malaya Kuala Lumpur

Professor Suresh Kumar University of Malaya Kuala Lumpur

Professor Wang Chee Woon University of Malaya Kuala Lumpur

Professor Mohd Rais Mohd University of Malaya Kuala Lumpur

Dr Azizudin Hj Kamaruddin University of Malaya Kuala Lumpur

Professor Dato Khairudin Yusof Royal Perak College of Medicine Ipoh, Perak

Professor Chan Yoo Kuen University of Malaya Kuala Lumpur

Professor Patrick Tan University of Malaya Kuala Lumpur Associate Professor David Choon Siew Kit University of Malaya Kuala Lumpur

Professor Raymond Azman Ali University Kebangsaan Malaysia Kuala Lumpur

Professor Dato Tg Sara Tg Ahmad Yahya University of Malaya Kuala Lumpur

Professor S Sengupta University of Malaya Kuala Lumpur

Professor Datin Norella Kong University Kebangsaan Malaysia Kuala Lumpur

Dr Morris Woo Universiti Kebangsaan Malaysia Kuala Lumpur

Professor Kamaruddin Mohd Yusof University of Malaya Kuala Lumpur

Professor Zahurin Mohamed University of Malaya Kuala Lumpur

Professor Ikram Shah Ismail University of Malaya Kuala Lumpur

Professor Chan Siew Pheng University of Malaya Kuala Lumpur

Professor Maniam Thambu University Kebangsaan Malaysia Kuala Lumpur Professor Hatta Shaharom University Kebangsaan Malaysia Kuala Lumpur

Associate Professor Jamiyah Hassan University of Malaya Kuala Lumpur

Dr Hoh Hon Bing Pantai Medical Centre Kuala Lumpur

Professor R Krishnan University of Malaya Kuala Lumpur

Associate Professor Haizal Dato Haron Kamar University of Malaya Kuala Lumpur

Professor Md. Idris Mohd. Nor Hospital Universiti Kebangsaan Malaysia Kuala Lumpur

Professor Tan Si Yen University of Malaya Kuala Lumpur

Dr Clarence Lei Chang Moh Normah Medical Specialist Centre Kuching, Sarawak

Professor Liam Chong Kin University of Malaya Kuala Lumpur

Professor Raman Rajagopalan University of Malaya Kuala Lumpur

Professor Zainal Ariff bin Abdul Rahman University of Malaya Kuala Lumpur

#### JUMMEC 2006: 9(2)

Dr Margaret Leow University of Malaya Kuala Lumpur

Associate Professor Azad Hassan Abdul Razack University of Malaya Kuala Lumpur

Professor Peh Suat Cheng University of Malaya Kuala Lumpur

Professor Tan Chong Tin University of Malaya Kuala Lumpur

Dr Loh Chit Sin Gleaneagles Intan Medical Center Kuala Lumpur

Professor Dato' Tan Hui Meng Adjunct Professor University of Malaya Subang Jaya Medical Center Subang, Selangor

Dr Evelyn Ho Megah Medical Specialist Centre Kuala Lumpur

Dr Harjit Kaur Putra Jaya Hospital Putrajaya, Selangor

Associate Professor Jamal Sam I Ching University of Malaya Kuala Lumpur

Dr Noran Naqiah Mohd Hairi University of Malaya Kuala Lumpur

Associate Professor Hakimi Zainal Abidin University Technology Mara (UiTM) Shah Alam, Selangor

Dr R Jayamalar Subang Jaya Medical Center Subang Jaya Selangor

Professor Atiya Ab Salam University of Malaya Kuala Lumpur Professor Saroja Krishnaswamy Penang Medical Center Penang

Dr Ng Chirk Jenn University of Malaya Kuala Lumpur

Associate Professor Saw Aik University of Malaya Kuala Lumpur

Associate Professor Vickneswaran CM University of Malaya Kuala Lumpur

Professor Adeeba Kamarulzaman University of Malaya Kuala Lumpur

Associate Professor Christina Tan Phoay Lay University of Malaya Kuala Lumpur

Dr Sylvia McCartney Hospice Malaysia, Cheras Kuala Lumpur

Professor Mustafa Ali Mohd University of Malaya Kuala Lumpur

Professor K Subramaniam University of Malaya Kuala Lumpur

Professor Looi Lai Meng University of Malaya Kuala Lumpur

Professor Gita Jayaram University of Malaya Kuala Lumpur

Professor Rohela Mahmud University of Malaya Kuala Lumpur

Professor Ramani Vijayan Sannasi University of Malaya Kuala Lumpur Associate Professor Quek Kia Fatt Monash University Malaysia Sunway Selangor

Professor Hussain Habil University of Malaya Kuala Lumpur

Associate Professor Khoo Ee Meng University of Malaya Kuala Lumpur

Professor Ng Kee Peng University of Malaya Kuala Lumpur

Dr Maimunah A Hamid Institute of Public Health Kuala Lumpur

Professor V Sivanesaratnam University of Malaya Kuala Lumpur

Dr Lee Sing Hong Gleneagles Intan Medical Center Kuala Lumpur

Professor Hematram Yadav International Medical University Bukit Jalil Kuala Lumpur

Professor Normadiah M Kassim Uinversity of Malaya Kuala Lumpur

Dr Hamidah Hj Abd Karim University of Malaya (Retired) Kuala Lumpur

Professor Nafisah Adeeb University Kebangsaan Malaysia Kuala Lumpur

Associate Professor Taufik Teng Cheong Lieng International Medical University Bukit Jalil Kuala Lumpur

## CORRECTIONS

The paragraphs in the article, *Pseudomonas Aeruginosa*: Epidemiology of bacteremia and antimicrobial susceptibility pattern in a teaching hospital in Kuala Lumpur by Nadeem SR, Rina K, Hamimah H and Savithri DP should read as follows:

#### Discussion

#### Paragraph 1, Line 5:

"The patients were mainly distributed among the intensive care units, surgical and hematological wards, whereas another study (6) had found a predominance of patients from the general surgical and transplant services.

#### Paragraph 2 Line 4:

In the study by Aliaga et al (1), only 12% of cases were community-acquired whereas Weinstein et al reported a rate of 40% (6).

#### Paragraph 4, Line 1:

Polymicrobial bacteremia in this study was seen in 33% of the bacteremic cases, which was much higher than the 17% previously reported (12).

#### Paragraph 4, Line 6:

In Aliaga et *al*'s study, patients with polymicrobial bacteremia were clinically worse and had a higher mortality rate compared to patients with monomicrobial *P. aeruginosa* bacteremia (12).

#### Paragraph 4, Line 11:

The most common organism isolated along with *P. aeruginosa* in Aliaga et *al*'s study was *Enterococcus* spp. (12), whereas in our study, *Staphylococcus aureus* was the commonest organism isolated, followed by *Enterococcus* spp.

#### Paragraph 5, Line 6:

A previous study found that the respiratory tract was the commonest source for *P. aeruginosa* septicemia (6).

The Editorial Board regrets any inconvenience caused.

# Journal of the University of Malaya Medical Centre

## Volume 9 Number 2 CONTENTS 2006

Editorial: JUMMEC: Taking stock, consolidating and gaining in strength	I
Breast cancer as a chronic illness: Implications for rehabilitation and medical education	3
Interdisciplinary research on impaired mobility, disability and rehabilitation in Selangor I Manderson L, Zaliha O, Rameezan BAR, Nooreini AH, Soh SB and Disler P	2
Malignant sertoli cell tumour: An uncommon testicular tumour Is Sia SF, Dublin N, Nurul B and Wong KT	8
A case of congenital anomaly of the urinary tract and neurogenic bladder	2
Modified vacuum-assisted closure	4
Free radial forearm flap after partial glossectomy for squamous cell carcinoma of the tongue 24 Chuah UC, Kamarul T, Sara T	8
Grade IV hypertensive retinopathy as a rare first presentation of hypertension: A case report	2
A case of lymphangioleiomyomatosis with multiple angiomyolipomas in the kidneys: A case report	5
List of reviewers	9

## JUMMEC



## Volume 9 Number 2 2006



Journal of the University of Malaya Medical Centre

