

Diagnosis & Treatment

The Journey of a Patient with
Malignant Pleural Mesothelioma

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Diagnosis and treatment: the journey of a patient with malignant pleural mesothelioma





OFFICE OF THE GOVERNOR

SYDNEY 2000

Message from Her Excellency, Professor Marie Bashir AC CVO

Foreword

Diagnosis and Treatment – The Journey of a Patient with Malignant Pleural Mesothelioma

Mesothelioma is a deadly cancer caused primarily by asbestos.

For many years, Australia has been one of the biggest users of asbestos in the world and there continues to be a large amount of asbestos in Australian buildings and other infrastructure. As a result, Australia has one of the world's highest rates of mesothelioma and it has yet to reach its peak.

Those diagnosed with mesothelioma are innocent victims - sick as a result of being exposed to a substance that has been used widely around the world.

Once inhaled, asbestos can lie dormant for decades and then suddenly strike without warning. Mesothelioma is a painful, devastating disease that can affect anyone.

However –with research, there is hope.

Diagnosis and Treatment – The Journey of a Patient with Malignant Pleural Mesothelioma is a much needed contribution to the resources available for people looking for help and guidance.

This book is designed to bring information, support and hope to patients diagnosed with mesothelioma. It outlines the patient journey – the diagnostic steps, possible treatment paths and illustrates the importance of knowing that as a mesothelioma patient you are not alone.

This book is an extremely valuable resource for both patients and caregivers. It is a unique blend of both research and evidence translated into a practical and useful guide for all those affected by this deadly disease.

Diagnosis and Treatment – The Journey of a Patient with Malignant Pleural Mesothelioma is an essential resource for mesothelioma patients and carers.

It is a much needed and valuable contribution to the treatment and support of all those facing the fear, uncertainty and challenge of mesothelioma.

I congratulate and thank Jocelyn McLean and Assoc. Professor Brian McCaughan for identifying the need in our community for a publication such as this. Their work is exceptional and their commitment to their patients and carers is outstanding.

Professor Marie R Bashir AC CVO
Governor of New South Wales



Dedication

This book is dedicated to the hundreds of heroic patients with mesothelioma that we have treated. They have taught us very important lessons in managing them and their mesothelioma. One lesson has been the importance of supporting and informing patients, carers and caring families during this stressful experience. We hope that the information provided in this book will assist future patients on their journey so that they can be better informed and able to negotiate what often appears to be an impossible path.

This is our 'thank you' to all those inspirational patients who have allowed us to assist them in their cancer journey. We feel honoured to have been part of their care.

When things were not going so well for one brave lady she gave us this reminder:

*DON'T WAIT FOR THE STORM TO PASS, LEARN TO DANCE IN THE RAIN.
Thank you JP.*

Jocelyn & Bryan

Thank You

Thank you to Dr MoMo Tin, Radiation Oncology and Dr Steven Kao, Medical Oncology for reviewing the scripts related to their specialty.

Thank you to The Baird Institute for their unfailing support of our mesothelioma patients, and in particular to Michelle Sloane and Sue Moore for their vital role in coordinating the communication related to running the support group.

Thank you also to the Comcare Asbestos Innovation Fund for their financial grant to run the Wellness Living Programme.

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PART 1

Background

Asbestos History

Asbestos mining and use has been part of Australian industry and manufacturing for many years. The following are some of the facts about our Australian experience.

Asbestos was mined in NSW at Baryulgil on the north coast, between 1940–1979, at Barraba on the northern tablelands, between 1918–1923 and at Wittenoom in WA from 1940–1966. Australia imported white asbestos from Canada, brown asbestos from South Africa and manufactured asbestos products from the UK, USA, Germany and Japan.

Asbestos was used widely in the manufacturing, transport and building industries. More than 60 per cent of production and 90 per cent of asbestos consumption was in cement manufacturing. In 1954, Australia was fourth in the Western world in consumption of cement products, after USA, UK, and France. Between WWII – 1954, 70,000 asbestos cement houses were built in NSW (52% of all houses).

Consumption peaked around 1975, at 70,000 tons per year, although regulation of the use of asbestos products began in the 1970s, with removal of production during the 1980s. Asbestos was removed from building boards between 1981 and 1983, from corrugated products around 1985, from cement pipes in 1987 and it was removed from use in brake pads and linings in 2003. In 2003, there was an Australia-wide ban on asbestos being sold, but some was still imported. Mining of asbestos in north America for export to Third World countries has just been banned in 2013.

Asbestos was a chosen product because of its durability and insulation properties. Interestingly, the dangers were noted 4,500 years ago, when a Greek geographer and Roman naturalist observed the “sickness of the lungs” in the slaves who wove asbestos into cloth. However, it was not until 1974, that the dangers of asbestos were first raised in the Australian media.

Today, there is an increasing concern about what is called the “third wave” of this disease. This relates to past and current do-it-yourself home renovators, who may have been unknowingly exposed to asbestos dust during the renovation processes. Education campaigns are required, to raise awareness of the potential dangers of renovating and to prevent exposure to this lethal dust.

Information obtained from:

The Asbestos Diseases Research Institute (ADRI). Website: www.adri.org.au

The Safe Work Australia 2011 report, titled "Mesothelioma In Australia Incidence 1982 to 2008, Deaths 1997 to 2007". Website: www.safeworkaustralia.gov.au

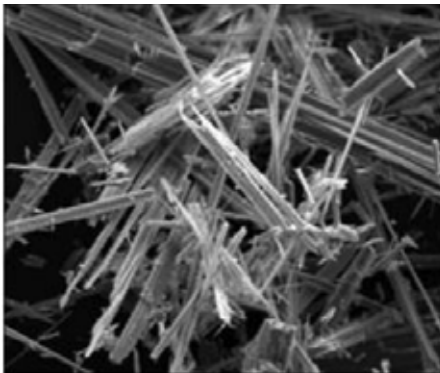
What is asbestos?

Asbestos is a mineral found underground and was obtained for use by open-cut mining. There are three mineralogical groups of asbestos:

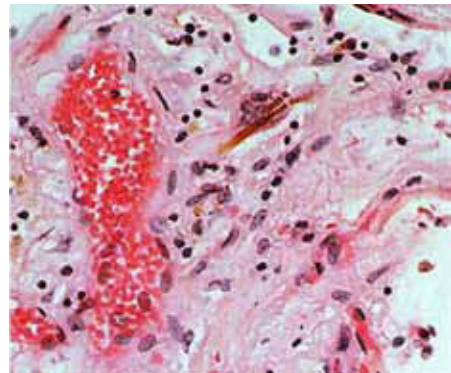
Chrysotile (white asbestos) - which has curved fibres

Amphibole made of Crocidolite (blue asbestos) - which also has curved fibres

Amosite (brown asbestos) - which has thin and straight fibres



Straight asbestos fibres



Asbestos fibres in the lung specimen

Asbestos fibres are 200 times thinner than human hair. Because they are durable, small and fine, they can easily be inhaled or swallowed. Once the fibres come into contact with mesothelial cells, the cells seem to be particularly susceptible to asbestos-mediated cellular injury, as compared to other cells, such as bronchial epithelium, in the lungs.

The mechanism by which asbestos fibres induce diseases is not yet completely understood.

Reference:

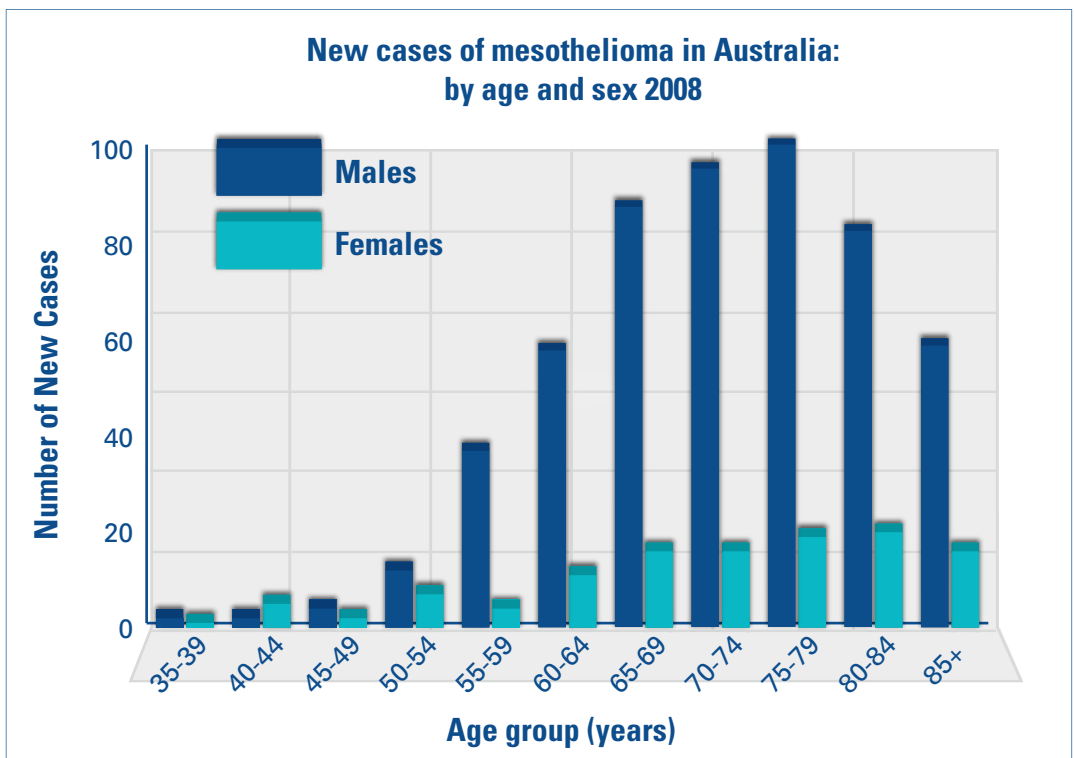
The Asbestos Diseases Research Institute (ADRI). Website: www.adri.org.au

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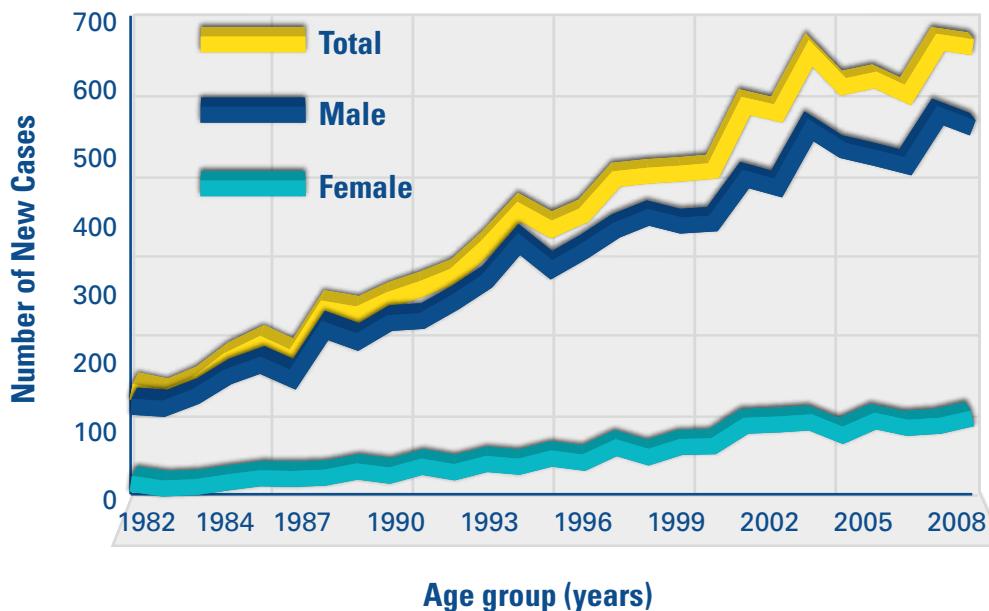
Incidence of Mesothelioma in Australia

Text books say that mesothelioma is a rare disease, but this is not the case in our practice in Australia. The following graphs demonstrate the incidence as reported by Safe Work Australia in its 2011 report: Mesothelioma In Australia Incidence 1982 to 2007, Deaths 1997 to 2008. www.safeworkaustralia.gov.au

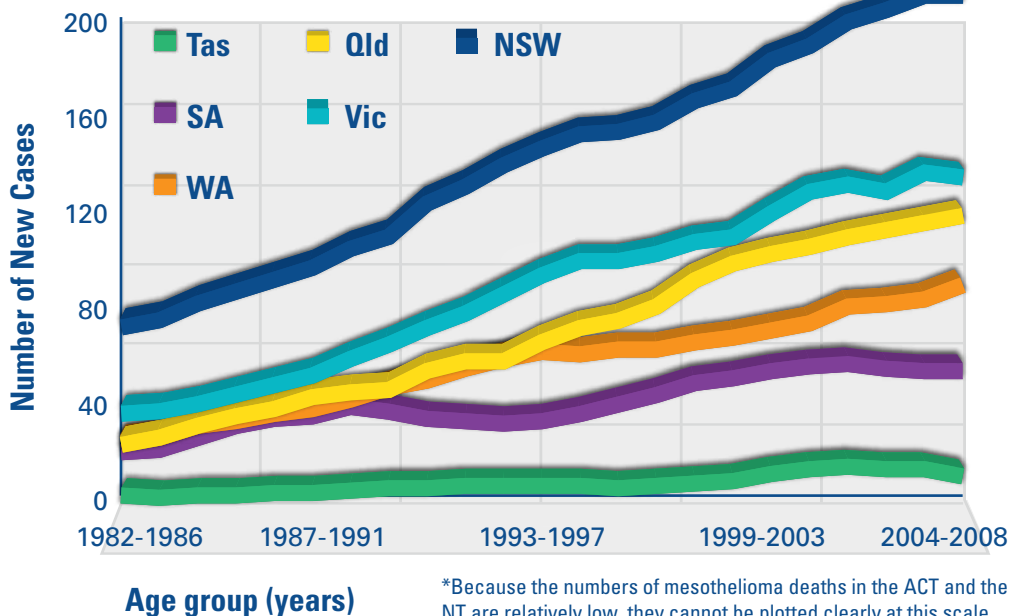
The incidence of mesothelioma continues to rise and officials are unsure when it will peak - some authorities suggest around 2030 – however, there is concern that there will be more than one peak, as people who renovated their homes (home renovators) during the 1980s and beyond may have been unknowingly exposed to fibres.



**New cases of mesothelioma in Australia:
year of diagnosis by sex, 1982 to 2008**



**New cases of mesothelioma in Australia:
five-year rolling average number of cases by State or Territory*
1982–1986 to 2003–2007**



*Because the numbers of mesothelioma deaths in the ACT and the NT are relatively low, they cannot be plotted clearly at this scale.

What is Mesothelioma?

Mesothelioma is a cancer that occurs in the lining of organs of the chest cavity, abdominal cavity and pericardium (lining of the heart). In the chest it occurs in the pleura - the thin membrane that forms over the lung and also lines the chest wall (ribs), the heart and the diaphragm. Mesothelioma in the chest is called malignant pleural mesothelioma or may be shortened to MPM.

Mesothelioma does not usually present as a single tumour but as multiple small tumours scattered throughout the pleura – herein lies one of the problems in treating this disease.

Malignant pleural mesothelioma is classified by cell type and there are three cell types – epithelioid, sarcomatoid and biphasic. Epithelioid mesothelioma is the most common type. Biphasic is a mixed tumour of both epithelioid and sarcomatoid type. Studies have shown that patients with epithelioid cell type might have a prospect of longer survival. Sarcomatoid mesothelioma is less common but more aggressive and survival rates are not as good.

The cell type is important when considering treatment options – in particular, suitability for trimodality treatment.



PART 2

The Patients Care and Treatment Journey

Introduction

The journey of each patient who receives care and treatment for mesothelioma is likely to be different, because the natural course of the disease can vary. Some people have slowly developing symptoms that suggest slow-growing disease activity, while others will have rapidly developing symptoms that reflect aggressive disease. Some people may have had unrecognised or undiagnosed symptoms for months or years before they lead to definitive action. How people respond to these situations varies greatly, just as the course of diagnosis and treatment offered may vary. A journey with mesothelioma is an individualised experience and therefore the suggested treatment will be tailored to each patient's clinical situation. Treatment is likely to involve a number of specialised health professionals at different times – a team that is called a multi-disciplinary team. Each member provides expert care related to their specialty.

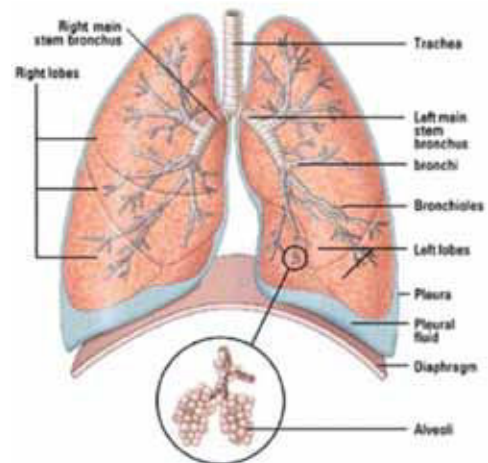
This document outlines the course of care and treatment that patients are likely to take when referred to the surgeons of the Sydney Cardiothoracic Surgeons Practice. It is a guide for patients and their family who present with symptoms that may result in a diagnosis of mesothelioma. Patients are referred to the surgeons because their symptoms are considered best managed by surgery. Surgery plays a key function in managing fluid that accumulates in the pleural space and may have already re-accumulated or is likely to re-accumulate, even after it has been drained. The cause or diagnosis is not usually known at this time, but there may be a history of exposure to asbestos (can be many years previously) and therefore the suspicion of an asbestos related problem is raised.

The surgeon is one member of a multi-disciplinary team of clinical experts providing advice, treatment and care. It is no coincidence that surgery is where most people begin their journey, but as mentioned above, patients move between clinicians and medical disciplines as situations change.

Medical help is sought because patients experience some or all of the following symptoms: breathlessness, chest pain, tiredness, weight-loss, and reduced appetite. While not all patients with these symptoms have mesothelioma, a suspicion is based on clinical symptoms and a history of asbestos exposure, proven by a tissue diagnosis.

Presentation

The most common symptom of MPM is breathlessness – a sensation that you cannot get your breath no matter what you do. It usually feels worse when lying down. Patients initially seek help, either directly from an emergency department, or via a general practitioner and a respiratory physician. The sensation of breathlessness is caused by fluid collecting in the pleural space, between the lung and the ribs, which is normally lubricated with a very small amount of fluid (about 20 mls). The fluid build-up is called a pleural effusion and it does several things. It makes the normal physiological act of breathing by moving the diaphragm up and down very difficult because the weight of water has to be raised and lowered with each breathe. Also, the air sacs, called alveolar, where respiratory gas exchange takes place are compressed by the sheer weight of the water. Water is heavier than air.



Pain related to involvement of the pleura, called pleuritic pain, can also be a presenting symptom though this is less common. The pain is related to tumour involving nerve endings.

The first medical action after a physical examination and listening to the chest is usually a chest x-ray to assess the presence or otherwise of fluid collecting in the pleural space, a so called pleural effusion. At this time some fluid can be drained off. This may be done in a skilled radiology department or by other arrangements. Patients generally notice immediate relief from the shortness of breath but invariably the fluid re-accumulates, necessitating further drainage.

Meeting the surgeon

As mentioned above, the most frequent symptom at presentation is shortness of breath caused by fluid in the pleural space. The surgeon will explain how surgery aims to relieve this distressing symptom by controlling the fluid by a talc pleurodesis, optimising lung re-expansion and taking tissue for analysis

and diagnosis. The type of operation offered depends on the overall health status of the patient and on the outcome the surgeon expects is achievable once considering a number of important questions. What is the chance of the lung re-expanding after the fluid is removed? How much disease is there? Can the patient withstand a general anaesthetic and surgery?

Where there is a diagnosis of mesothelioma, a known asbestos history, or a high suspicion of an asbestos-related diagnosis, the patient and family will be invited to consent to giving tissue for asbestos related research which is being conducted at the Asbestos Diseases Research Institute (ADRI) at Concord Hospital. The surgeon and the nurse case manager will assist with this process.

What surgery will be offered?

The surgeon uses all available imaging and a clinical assessment of the patient before discussing with the patient the most appropriate surgical procedure for their individual situation. The aims of surgery are to:

1. Control the recurring fluid, thereby relieving symptoms.
2. Re-expand the 'trapped' lung to enable better lung functioning.
3. Take tissue for a diagnosis (and research if permitted).
4. *Assess the cancer for suitability for trimodality treatment.*

There are three surgical options for controlling the fluid:

1. Keyhole surgery, consisting of thoracoscopy, pleural biopsy, decortication and talc pleurodesis, or if this fails or is not a suitable first option, consider
2. Open surgery consisting of thoracotomy, debulking pleurectomy and decortication, or
3. Pleural drainage alone, by inserting an indwelling drain.

Patient Journey

Thoracoscopy - control symptoms

- Control fluid – talc pleurodesis
- Optimise lung re-expansion
- Tissue for diagnosis
- Assess for treatment / suitable for trimodality therapy

OPERATION: Video-assisted thoracoscopy (VAT), with pleural biopsy, pulmonary decortication and talc pleurodesis.

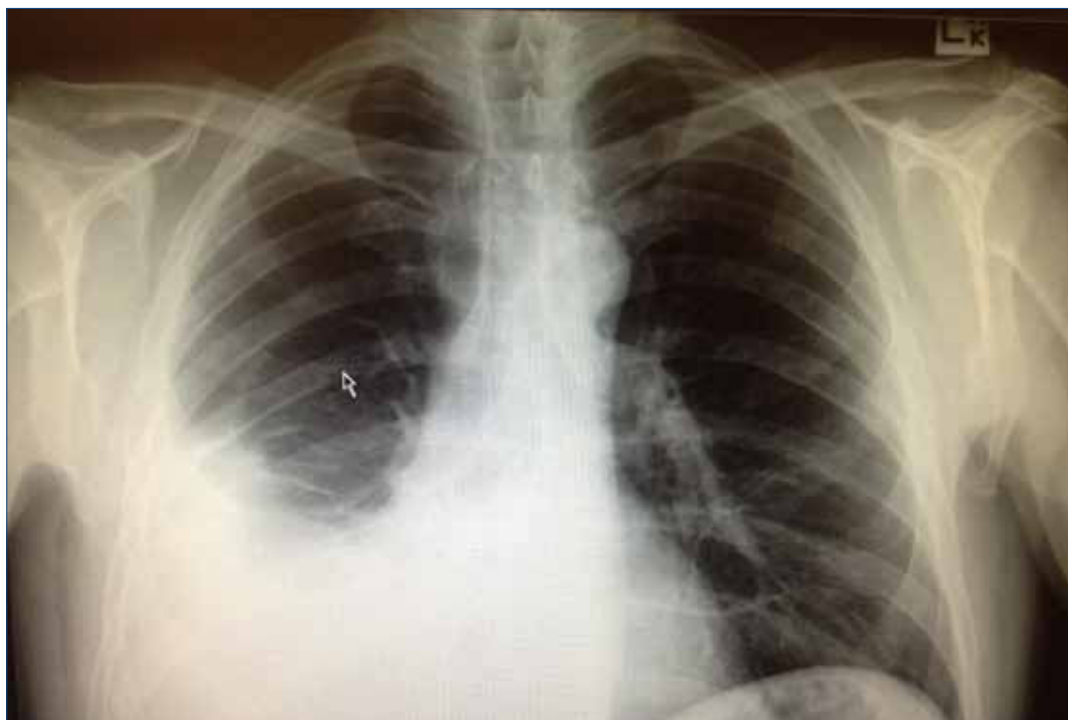
This procedure requires a general anaesthetic using a special breathing tube, and a camera to be inserted between the ribs into the pleural space, via portholes. The fluid contained between the two pleura is drained and the lung surface is “freed-up” (decortication) to allow the lung tissue to expand to its full potential. Samples of the pleura are taken (biopsy) and tissue is sent to the pathologist for analysis and diagnosis (and ADRI research if consent has been obtained). Sterile talc powder is instilled into the pleural space. It irritates and causes inflammation between the lung lining (visceral pleura) and chest wall lining (parietal pleura), so that they fuse together (a process called pleurodesis). This reduces fluid production by obliterating the pleural space.

During this operation, the surgeon inspects and assesses the pleura for volume of disease to help future treatment planning.

At the completion of the procedure, the anaesthetist re-inflates the operated lung to its maximum capacity. One chest drain is left in place, exiting one of the portholes. It is attached to a closed chest drain connected to a suction system for a minimum 48 hours. The suction makes the pressure between the pleura more negative and enhances the irritation process, to optimise adhesion between the lung and chest wall. The pleurodesis process can be quite painful, with a “burning” sensation during the first 24 hours. Pain is relieved using

a patient-controlled pain device (PCA) that is available for use immediately after the operation and remains until the drain is removed. Once the drain is removed, the intravenous pain relief is changed to tablet form and the patient is encouraged to mobilise independently and prepare for discharge from hospital. The procedure takes up to one hour and the hospital stay is between three and five days. Physical functioning should improve within two or three weeks of the surgery and the patient should return to a good level of functioning, if starting from a reasonable level of fitness.

It is therefore, normal to expect residual soreness around the front and lower part of the chest, to where nerve endings track because the nerves between the ribs are compressed during surgery. Once over the immediate post-operative recovery period and with symptoms that led to presentation and diagnosis under control, a period of increasing wellness (while being aware of the disease) is expected. What is unknown is how long this period might be, until the disease becomes active and symptoms intensify.



Chest X-ray demonstrating a right pleural effusion
- fluid collecting in the right pleural space

VAT Thoracoscopy (Keyhole) Surgery Is Best For:

- Obtaining tissue for diagnosis.
- Controlling pleural fluid - therefore eliminating repeated painful pleural taps.
- Assisting lung re-expansion, providing the lung is not encased or trapped and preventing re-expansion.
- Patients whose lung can be easily re-expanded.
- Patients who do not require an open operation.
- *Assessing the pleura for future treatment planning - consider potential need for open surgery or suitability for trimodality therapy.*

Why obtain a diagnosis and how is this done?

Taking tissue for a diagnosis at the time of surgery is useful, because diagnosis of mesothelioma can be difficult. It is common that fluid taken at the time of initial and subsequent drainage does not identify diagnostic cancer cells. Fluid in the pleural space will contain tumour cells, but they can be difficult to diagnose by cytology. A more reliable diagnosis is then obtained from a biopsy or sample of lung or pleural tissue. When there is uncertainty about a diagnosis, biopsies may be sent to another expert pathologist for assessment. This will delay the definitive diagnosis and add to the anxiety of the patient and family, but is an important step. On some occasions, despite clinical signs of disease, an accurate history of exposure to asbestos, and good amounts of tissue, a pathologist cannot make a diagnosis.

When and how will I be told the diagnosis?

There is no easy way to tell a patient about a diagnosis of mesothelioma and no one wants to hear the news, even when the malignancy has been suspected. Other issues that add to this dilemma are brief hospital admissions, as well as the distance patients travel for care. Patients are not usually kept in hospital until the diagnostic report is in print, however, the possibility of a diagnosis of mesothelioma is usually raised prior to surgery and there may have been ongoing conversations after the surgery and during recovery. The timing of being told the diagnosis depends on how quickly the results are returned.

Communication takes place between the surgeon, the patient and their relatives. The surgeon will discuss the diagnosis and what it means in terms of survival, recovery, treatment and compensation. This may be as a face-to-face conversation, or over the telephone. The nurse case manager follows-up the surgeon's communication with a visit or phone call to support the patient, provide clarity and understanding about the information, provide introductory compensation information, discuss preferences for ongoing treatment and offer ongoing support. Liaison is also established with a lung care coordinator in the patient's home area (by phone or email). Pathology results are forwarded to the General Practitioner (GP) and the referring specialist.

If the results are delayed or not available, or the patient lives outside Sydney, an appointment is made by the case manager for the patient to have an early

post-operative visit to their referring specialist, to discuss the diagnosis and co-ordinate ongoing management in the local area.

I have a diagnosis of mesothelioma. What now?

Professionals don't like giving bad news and patients don't like receiving it, but the journey continues. There will be physical and emotional issues to deal with, along with taking care of some practical administrative things. Having good support of a partner, family and friends will make this time more bearable.

Allow time to make a physical recovery from the surgery. This will take two to three weeks. Try to eat well, get quality rest and do regular exercise within your capacity. Over time the significance of a new diagnosis of an incurable disease will become clear. All sorts of emotions will be felt by all involved and many tears will be shed during this time – this is normal. Some common fears of patients relate to the cancer diagnosis itself, the fear of losing a life partner and family, feelings of anger about the diagnosis, as well as concerns and worry about work and financial support. Professional counselling support can be arranged as required – if this has not been done, please ask for it.

Many patients want to get on with a treatment. They want it started yesterday. Before exploring disease specific treatments, there needs to be an understanding of why a diagnosis is important and to address some of the practical things related to it.

Why is it important to diagnose mesothelioma?

A diagnosis is important, because it enables discussion about appropriate treatment options and the potential for compensation. Treatment options are discussed later in this document.

With respect to compensation in NSW, a diagnosis of mesothelioma, must, by law, be reported to the Dust Diseases Board (DDB). The DDB is administered as part of the NSW Workers Compensation Act 1942. Persons who can prove they were exposed to, and acquired, an asbestos-related disease in their workplace in NSW may be entitled to compensation under the DDB. Malignant pleural mesothelioma is nearly always a result of being exposed to, and therefore inhaling, asbestos fibres. Most often, this has occurred in a workplace, but

exposure can also be as a result of secondary handling of the dust fibres e.g., washing clothes worn by a person who worked with asbestos, or sweeping-up after another person has used asbestos. Exposure can also occur unknowingly and a common situation can be during home renovating. A number of patients cannot identify any place or time of exposure.

Patients may also consider taking civil action against a company that manufactured the product containing asbestos, to which they were exposed. They should explore their eligibility with a legal representative experienced in handling these matters.

While many patients balk at the thought of compensation or taking civil action, or both, successful claims may result in significant financial security. A vital factor affecting a claim is the pathological evidence of disease obtained more often from pleural tissue (biopsy) and less often from pleural fluid cytology analysis alone.

If exposure to asbestos occurred outside NSW, seek help by contacting that State's legislative body re compensation.

If exposure occurred outside Australia, seek advice from a legal representative experienced in handling asbestos claims – try using your local experts as a starting point.

If you are unsure of any of the above, contact a legal representative experienced in handling asbestos claims and ask them to guide you about your rights to any claim.

Summary: what do I do about compensation?

Phone the Dust Diseases Board (DDB) and explain that you have a new diagnosis of mesothelioma and would like to explore and understand what this means for you.

Phone a lawyer experienced in handling asbestos-related legal claims. Explain to them that you have a new diagnosis of mesothelioma and ask for their help to explore and understand what this means for you.

I have had keyhole surgery and have a diagnosis - what now?

Patients are reviewed by the surgeon or the referring specialist two to four weeks after initial surgery (with a new chest x-ray) to make sure the fluid has been controlled and the lung has remained re-expanded. If there are no symptoms or signs of residual fluid, this is the time to explore treatment options.

If, however, the fluid has re-accumulated and symptoms re-present, it would be considered that thoracoscopy and keyhole surgery has been ineffective. This may be because there is extensive tumour trapping or encasing the lung so that it is not able to re-expand. In this instance, a return consultation with the surgeon will be made, who will consider further surgery if appropriate.

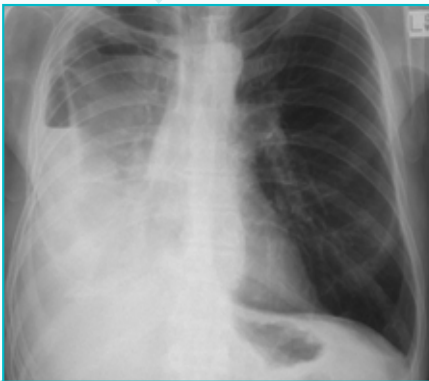
If fluid re-accumulates – what can be done?

Re-accumulation of fluid usually means the symptom of breathlessness returns and worsens. The fluid needs to be controlled, so ask to be returned to the surgeon for further assessment and consideration of other surgical options. The most common surgical procedure is open surgery via thoracotomy.

Patient Journey

Thoracoscopy - control symptoms

- Control fluid – talc pleurodesis
- Optimise lung re-expansion
- Tissue for diagnosis
- Assess for treatment / Suitable for Trimodality therapy



Fails
(fluid re-accumulates)



Pleurectomy / Decortication
via thoracotomy
or simple drainage

OPERATION: Open surgery - thoracotomy with parietal pleurectomy and pulmonary decortication.

Open surgery is offered when keyhole surgery fails to control fluid or at first presentation the surgeon believes keyhole surgery will not achieve a good result. It requires a general anaesthetic and placement of a double lumen breathing tube. The chest is opened via the back and between the ribs (posterior thoracotomy).

The pleural fluid is drained and a tissue is taken for the biopsy. All specimens are sent to the pathologist for analysis. The bulk of the tumour is reduced by a technique called pleurectomy. The pleura encased with tumour are peeled off the chest wall (parietal pleurectomy) and the lung (decortication). Not all of the pleura is removed – only that which has the worst bulky disease and is safe to remove.

The lung that had been trapped down by the fluid and tumour, is freed by a technique called decortication, whereby bands are peeled from the lung surface. The lung is re-inflated and re-expanded mechanically by the anaesthetist. The inflammatory action of the decortication process causes a pleurodesis.

The decortication exposes the patient to a greater risk of air leaking from the lung surface - called air-leak. Two chest drainage tubes are left in place at the end of the procedure for a minimum of 48 hours, as for thoracoscopy. The procedure takes about two hours.

Open surgery is more painful immediately post-operatively and there is more long-term pain and discomfort, but it may give better long-term symptom control and improved lung function, where thoracoscopy and pleurodesis failed to provide an optimal result.

Thoracotomy (open) surgery is best for:

- Obtaining tissue for diagnosis. If not previously done.
- Poor control of fluid and symptoms after thoracoscopy and pleurodesis.
- Debulking tumours and assisting lung re-expansion when the lung has been trapped or encased with tumour and unable to re-expand.

- *Trimodality therapy patients, who at operation for EPP, are found to have more disease than expected and therefore do not proceed to EPP.*

When thoracoscopy and thoracotomy is not an option then consider an indwelling pleural drain.

When surgery via thoracoscopy and thoracotomy fails, or is not safe because of unacceptable risks related to a general anaesthetic, or considered to be inappropriate for an individual patient, fluid may be controlled using an indwelling pleural drain.

An indwelling drain is a special tube inserted into the pleural space. It is intended to stay in place and be connected to a drainage system when symptoms of breathlessness become troublesome. The drainage system is designed for intermittent drainage whereby patients connect to a bottle and drain as needed. This may be once, or twice or three times a week, according to the amount of fluid and time frame that it re accumulates in. Over time, the pleural space may obliterate, may stop producing fluid and the body undergo self pleurodesis. If this takes place then, there will be no fluid to drain, symptoms might abate and the system will no long be required.

The drainage system can be managed by the patient, a supportive and confident carer, by a community nurse or a combination of all the above.

The chest drain is inserted under a local anaesthetic either by a cardiothoracic surgeon or an experienced interventional radiologist. There are two systems available (Pleurx and Rocket). This service uses the Pleurx drain manufactured by Care Fusion.



Pleurx drain and drainage bottle

Early referral to a cardiothoracic surgeon, when first presenting with a pleural effusion, may reduce the need for an indwelling pleural drain if a surgical pleurodesis can be achieved.

All patients are referred to palliative care and community nursing once the tube is inserted, to facilitate home management and avoid hospitalisation for painful drainages.

What are the treatment options for mesothelioma?

Currently, good news about a diagnosis and treatment for mesothelioma is sparse, but there are patients living quality time for longer periods on treatment (and some not on treatment) and there is always hope that new treatment options will become available in the future. Researchers are busy in Sydney at ADRI, with The Baird Institute, as well as nationally and internationally looking for breakthroughs in knowledge and understanding about mesothelioma cells and how they can be manipulated by treatments. See ADRI website for more information.

While there is no cure, there is a standard treatment available. This is chemotherapy provided by a medical oncology team.

A very small number of patients may be offered trimodality therapy with the hope of extending the time they live free of disease after treatment, but unfortunately most people have advanced disease at the time of diagnosis or are excluded from this option because of age, physical fitness and other illnesses such as heart disease.

Radiotherapy has a smaller treatment role.

Supportive and palliative care are key services in providing care to the patient with mesothelioma especially at a time where symptoms are difficult to control or when the disease process is becoming uncontrollable and not responding to standard treatment.

Chemotherapy - medical oncology

Chemotherapy is a systemic treatment. Cytotoxic drugs are administered to circulate through all the body's cells via the blood system and the lymphatic system. The drugs are prescribed by a medical oncologist and administered by specialist nurses, who are pivotal to a medical oncology team. The patient is referred to a medical oncologist in a hospital convenient to the patient, for an in-depth discussion about this treatment before commencing.

There is scientific and anecdotal evidence that some patients with mesothelioma respond well to chemotherapy drugs. The treatment contains two drugs - the chemotherapy agent Alimta (pemetrexed disodium), combined with a platinum agent such as cisplatin or carboplatin.

All patients diagnosed with mesothelioma are encouraged, and have a right, to discuss the role of chemotherapy with a medical oncologist. We offer all patients a consultation with a medical oncologist, to discuss the pros and cons of treatment, drugs used, their effects and side-effects, along with the practical issues and possible timing of a course of chemotherapy.

The best time to start chemotherapy is not clear, but most clinicians believe that if a patient presents with symptoms and there is evidence of active disease on a chest X-ray or CT scan, it is worth commencing treatment early. Some patients prefer to delay the treatment while they feel well without any, or with limited symptoms, always with the option of having treatment later if/when needed. Some patients prefer not to have any chemotherapy.

Chemotherapy is a specialised, but well developed treatment, given in an outpatient setting in a chemotherapy suite. The treatment is administered in "cycles". The specific facts of this treatment are presented at a first consultation, where a comprehensive discussion is had about what drugs are offered, why they are used, when and how they are given, the expected effect, as well as the potential side-effects. If a decision is reached to proceed with treatment, the patient will be well supported by a multi-disciplinary oncology team including specialist nurses, social workers, counsellors, a dietitian and an occupational therapist. The patient's response to treatment will be monitored very closely. Treatment duration depends on how the disease responds to the drugs and how the patient feels during and after the treatment.

What is the role of radiotherapy?

Radiotherapy is a treatment provided by another medical specialty, called radiation oncology. Radiation beams are directed to a specific site where cancer cells are, or have been, present. The role of radiotherapy in treating mesothelioma has been limited. There are a number of reasons for this. Mesothelioma cells have not been sensitive to radiotherapy, resulting in poor responses to treatment. One of the challenges is that it is difficult to deliver the required optimal (big) doses of radiation to a large area (the lung and chest wall), without administering toxic doses that will damage surrounding tissue and organs.

Some centers give radiotherapy in small doses in an attempt to prevent the spread of mesothelioma along a needle or drainage tube track. There is no proven benefit from this. Radiotherapy can be beneficial in providing symptomatic relief from the discomfort when tracking of the disease to external tissues does occur, e.g. along needle biopsy ports, chest tube sites. Pain from diffuse pleural disease may also be relieved by radiotherapy.

A new method of delivery is being trialed in Victoria, using a technique called IMRT/PET. This delivers radiation to targeted areas, thus allowing doses to fluctuate within the chest anatomy. This treatment can be given without removing the lung.

Prior to any radiation treatment beginning, patients and relatives meet with a radiation oncologist to discuss why the treatment is offered, how it will be administered, the timing and duration and, of course, potential side-effects. Radiation therapy is prescribed by a radiation oncologist, delivered by a radiation technologist and patients are supported by specialist radiation oncology nurses and members of a multi-disciplinary team that includes a dietitian, social worker and counsellor – psychologist.

I have had chemotherapy. Radiotherapy may not be useful. What next?

There is always hope that tumours will respond to chemotherapy treatment and allow patients to have a period of quality living. Unfortunately, some patients with aggressive disease may not achieve this.

Where chemotherapy has initially been useful in controlling symptoms and the patient has time off the treatment but the symptoms recur, there may be an option to have further chemotherapy. This decision is made between the oncologist and the patient. If retreatment fails, there may be an option to receive other chemotherapy called 2nd line chemotherapy

If the tumours have not responded to chemotherapy and/or the side-effects are intolerable, the treatment is likely to be stopped. Decisions about treatment will always be based on discussion between the oncologist and the patient.

What we do know, is that all mesothelioma patients need and want to feel supported by their medical and multi-disciplinary teams. An important member of the team is the palliative care service who provides ongoing symptom management through what is also called best supportive care.

Best supportive care and palliative care

It is well known that mesothelioma cannot be cured, but it is our aim for all patients to receive the best care and support available as they absorb and make sense of information about the diagnosis, have symptoms relieved and get on with treatment. At some point after diagnosis, treatment and a period of living well, with minimal or no symptoms, the disease will become active, recur and symptoms will again become a problem. What is not known is how long the window of “wellness” will last after diagnosis and the fluid is controlled. We do know, however, that all patients benefit from the support and expert knowledge of a community-based palliative care team.

Palliative care is not just something that happens when someone is dying. Palliative care is about looking after the patient and family, with the goal of relieving symptoms and improving patient comfort. Of course, the team becomes more involved when the disease progresses and death becomes

more imminent. A vital function of palliative care is to provide sensitive support to patients and families on all matters related to living with a terminal cancer and facing dying. The team consists of specially trained medical, nursing, allied health staff, community nurses and the GP, experienced in recognising medical and non-medical issues and finding ways to resolve them.

Palliative care is also valuable to patients who might be living quite well with their disease, but who have symptoms that are difficult to manage or relieve. Early referral to a service enables links and relationships to be established in a controlled situation, rather than at a time of crisis. It also ensures that patients have access to timely expert help to manage medical and practical problems of daily life. Each major hospital has a palliative care service.

The World Health Organization defines palliative care as “an approach that improves the quality of life of patients and their families facing the problem associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual”.

(Palliative Care, NSW)

How and who will monitor my disease and progress?

The state of disease needs to be monitored once a diagnosis is made, compensation issues have been sorted, and a decision is made about treatment. A key player in performing that function is the local General Practitioner (GP) so establishing a good rapport with the GP, who can liaise with other specialists is valuable. In most instances, chemotherapy will be the main treatment, so the medical oncologist will coordinate ongoing appointments and appropriate imaging and communicate the results to the GP and other specialists.

It is important to be aware of new developing symptoms, and to seek medical assessment of these as early as possible. The most common method of assessing how the body is reacting to therapy is by CT scanning. The need for,

and timing of tests depends on the reason for them, and possible changes to treatment that may arise from the test results. More testing does not necessarily mean better care.

Early referral to palliative care is advised to ensure support is provided when symptoms become less manageable.

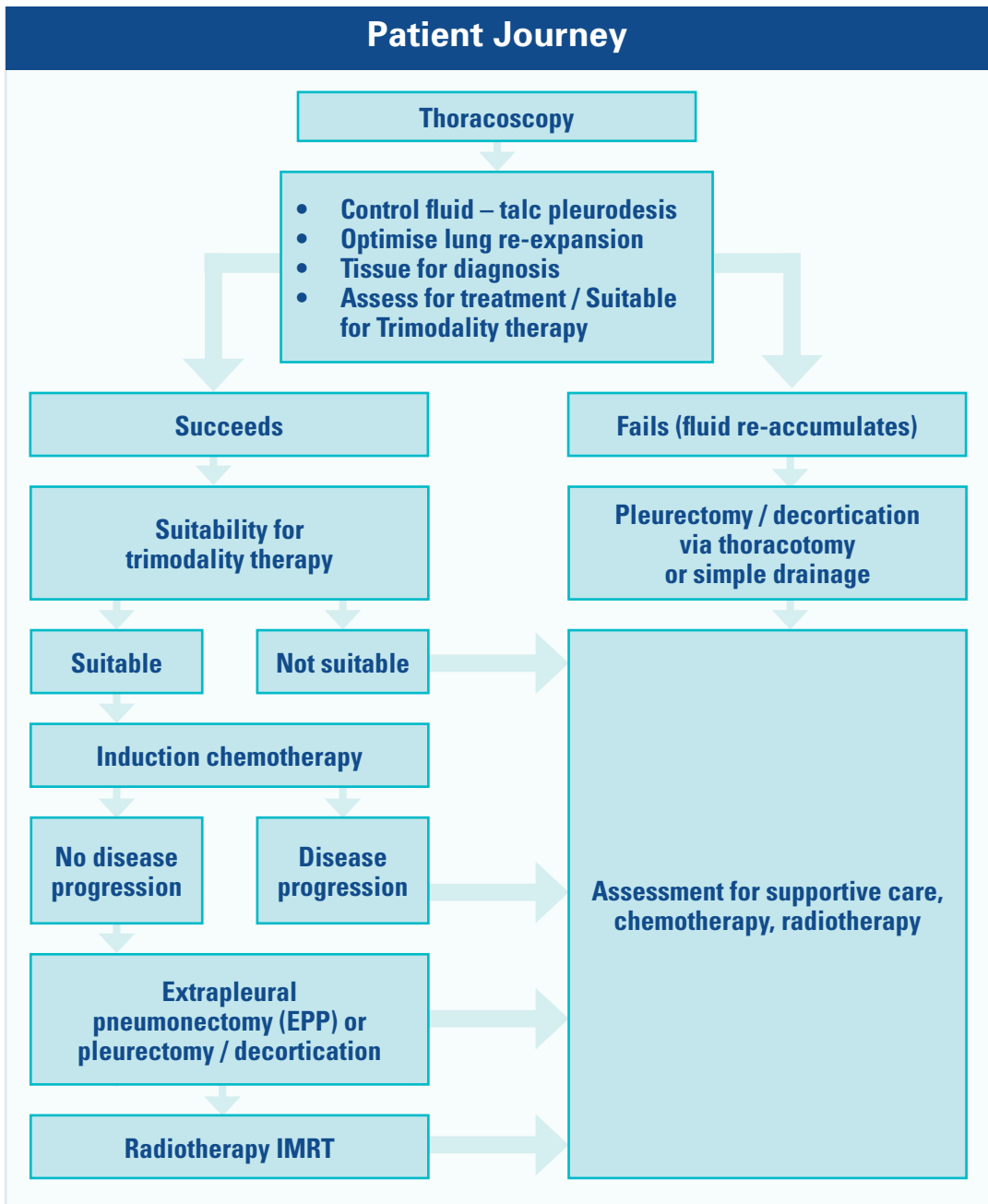
I have been told I might be suitable for trimodality therapy. What is this?

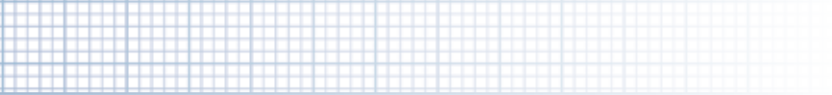
A very small number of patients diagnosed with mesothelioma may be considered suitable for trimodality therapy. Trimodality therapy consists of a sequence of three treatments. It begins with induction chemotherapy lasting three months, followed by radical surgery and recovery that spans six to eight weeks, before starting radical radiotherapy, consisting of six weeks of treatment.

A decision about suitability for this regime can only be made by the consultants providing the treatment and, in particular, the surgeon. Every bit of clinical information is used to ensure the best course of treatment is offered for each individual patient, but despite all the excellent knowledge and skills, sometimes what is best for the patient is to not proceed with the trimodality therapy.

- Not every patient will be suitable for radical surgery after completing chemotherapy. This is a decision made by the surgeon based on the progress of the mesothelioma.
- Not every patient will automatically have radical surgery at the time of the planned operation. The surgeon will make this decision at the time based on the operative findings.
- Not every patient will have radical radiotherapy after surgery if he or she is not physically fit to proceed, or it is not recommended based on the pathology results. This decision is made by the surgeon and the radiation oncologist.

Patient Journey





PART 3

Trimodality Therapy

Trimodality therapy comprises of three sequential treatments beginning with induction chemotherapy that spans 3 months, radical surgery (where the complete lung is removed) and recovery that spans 6-8 weeks, prior to 6 weeks of radical radiotherapy.

The Sydney Cancer Centre is the only center in Australia that has an established treatment programme and has published results.

The best way to treat mesothelioma is still a research mystery and what guides practice in medicine and surgery are studies called randomised controlled trials (RCTs). There are, however, no RCTs to guide best treatment for mesothelioma. Trimodality therapy is one treatment option offered to a small number of patients, because only a few present with early disease and are considered potential candidates for this difficult three-stage treatment regime.

Surgery is just one part of the treatment. There is general consensus amongst practitioners that if radical surgery is to be considered, it should be considered as part of trimodality therapy – not as a stand-alone treatment.

Induction chemotherapy

Trimodality therapy starts with induction chemotherapy. Patients receive the same chemotherapy as other mesothelioma patients would be offered, but in this setting they are given a minimum of three cycles. At completion of the 3 cycles, the disease is assessed using PET/CT imaging. If the response satisfies the surgeons requirements, a rest period of four to six weeks is taken before proceeding to surgery. If there has not been a suitable response to chemotherapy, surgery would not be recommended. The patient would return to medical oncology care, to consider ongoing chemotherapy and further assessments.

This treatment period offers time for several important milestones. It provides living evidence of the natural history of the disease. For example, if the disease progresses in this treatment time, this is evidence that surgery is not the appropriate option. It provides the patient and family with time to get a good understanding of the nature and prognosis of the disease, while having a standard treatment. It provides time for medico-legal compensation issues to be initiated.

Most patients tolerate chemotherapy reasonably well. They remain active and most do not lose a lot of weight. A strong message from past patients is that the possibility of radical surgery is an incentive to endure chemotherapy and its side effects. Before seeing the surgeon, a PET scan is completed to assess the response to treatment. It is also used to exclude the presence of other sites of cancer, as this would preclude proceeding to radical surgery.

Radical Surgery

Once chemotherapy is completed arrangements are made for a return visit to the surgeon (with a new PET scan) about 3-4 weeks after the final chemotherapy cycle. Providing the PET scan confirms no new disease or better still a reduction in disease bulk from the chemotherapy, the surgeon is likely to offer to proceed with the surgery. The visit to the surgeon will involve a frank and open discussion about why and how the surgery is done, the risks involved with the procedure, and issues relating to removing a complete lung and recovery. Patients also spend time talking with the thoracic case manager about the expected immediate and long-term post-operative recovery. Radical surgery is a very challenging procedure and the recovery process is enhanced when there is good support from a partner/carer. Patients and carers are encouraged to speak with another patient and carer to hear about their experience. They are also encouraged to attend a support group meeting (where practicable) before making their final decision to proceed. Several conversations with the surgeon and case manager may be required, before reaching a decision to either proceed with or decline surgery. If there is any doubt, surgery will not proceed. It is vital that all persons are feeling positive about the surgery.

The Operation

The operation is described as: Radical right or left pleuropneumonectomy with en bloc resection of hemidiaphragm and pericardium, formal mediastinal lymph node dissection, then reconstruction of diaphragm and pericardium with Gortex mesh.

The chest is opened via an extended posterior lateral thoracotomy (a longer than usual opening on the back). If a previous keyhole or open operation has been performed, the ribs, skin and tissues surrounding the old drain sites are

excised. The lung, chest wall (parietal pleura), hemi-diaphragm (that half on the operation side) and pericardium (lining over the heart) is removed in one piece (en bloc). The lymph nodes that drain the lung area are removed. All tissue is sent for pathological analysis. Any defects over the hemi-diaphragm and pericardium (heart) are closed with a layer of gortex mesh. The wounds are closed and one chest drain is left in place. The patient is meticulously managed in a highly specialised intensive care unit and must participate in a rigorous post-operative physiotherapy programme. Recovery from this surgery is difficult and there is a potential for many things to go wrong. Early detection by staff, who painstakingly monitor all aspects of hospital care, means that critical events can be minimised. Following discharge, recovery at home is slow, with a focus on rehabilitation in readiness for the next round of treatment – radiotherapy- a six-week course of treatment starting six to eight weeks after surgery.

The usual hospital stay is 10-14 days. Some patients are discharged directly to home, while others benefit from a week or so in a concentrated rehabilitation centre. The hospital stay varies according to the type and duration of complications.

Radical surgery is only for:

- Patients known to have early (limited) disease. Most would have had keyhole surgery for this to be known as well as CT and PET imaging.
- Patients who have had a good clinical response to chemotherapy and have no signs of disease progression.
- Patients with no evidence of lymph node involvement, or other cancer disease on PET scan.
- Patients deemed able to live independently with one lung.
- Patients deemed medically and physically fit to withstand the challenges of this surgery.
- Patients with carers who accept the physical and mental challenges associated with recovery.
- Patients who wish to proceed after they have had frank and open discussions with the surgeon and thoracic case manager.

Radical Radiotherapy – IMRT

Radical radiotherapy is the third arm of trimodality therapy. It is called IMRT that stands for intensity modulated radiotherapy. The aim is to treat any residual tumour cells present after chemotherapy and surgery and proven in the histopathology report. One may ask why use it, if in the palliative setting it has not been beneficial? Radiotherapy is most effective when the tumour cell burden is lowest which is immediately after surgery. It is adding another layer of protection. There is clinical evidence of improved survival when all three modes of therapy are used, as compared to single treatments.

The process begins with the radiation oncologist explaining the aims, methods, timing, side effects and expected outcomes of the treatment. A second visit involves planning the treatment. Here special imaging is taken and used to develop an individualised prescription of therapy. A couple of weeks later the treatment will start.

Radiotherapy begins six to eight weeks after radical pleuropneumectomy (if the patient is fit enough) and is designed to inhibit local recurrence of disease. This is an aggressive treatment, in which for six weeks, Monday to Friday, the patient comes to the centre, changes into a gown, has the treatment, changes back into street clothing, is reviewed by the radiotherapy staff for any developing side-effects and returns home to be ready for the next day's treatment.

It is an accumulative treatment in which side effects develop as treatment progresses. Not until two to three weeks after completion of the treatment, do patients begin to feel recovery taking place. Some issues that have challenged patients and carers have been difficulty with eating good quantities of food, nausea and reflux, external skin irritation from the ray beams, lethargy, tiredness and exhaustion. Anti-nausea medications are usually prescribed from day one to lessen the impact of nausea.

The radiation oncologist reviews each patient each week. Blood tests and treatment side effects are checked regularly by the team of doctors, registrars, technicians and nurses. RPAH has a dedicated radiation specialist who has treated all the EPP patients and so has wide experience in delivering this therapy and dealing with side effects from this treatment.

A downside of this experience is that the treatment can only be given at RPAH, Sydney. Before treatment, it is imperative that the patient and carer plan how to get to and from the hospital. If the patient is Sydney-based, a roster of drivers is useful. Country and outer Sydney-based patients will need to relocate to Sydney for the treatment.

These issues will be discussed with the case manager before the first appointment and with the radiation oncologist at the initial consultation. The team is used to dealing with the needs of country patients. If the patient is covered by the Dust Diseases Board, it is advisable to speak with them about assistance with accommodation during this treatment.

If I am considered for radical surgery, what can I expect in recovery?

Deciding to have radical surgery can be a very difficult decision and it is important that patients and carers be aware of the duration and nature of recovery. Recovery is both physically and emotionally challenging. For some patients it can be extremely slow, following a hospital stay of 10 -14 days. Recovery impacts on many aspects of family, social and work dynamics.

Patients are not expected to return to their "normal" level of physical activity. In time, however, between 6 -12 months after surgery, it is expected they will begin to appreciate a satisfying quality of life. No two patient recoveries have been the same, as each experience is individual. One essential requirement of recovery is a good support team.

There have been a small number of patients who unfortunately did not return home following radical surgery. Some have complications that require delayed hospitalisation, some require further surgery, but most have periods of "good" quality living before the disease relapses. Survival can be a short time to a long time but the goal of trimodality surgery is to gain as long as possible with good quality. Currently, there are a number of long survivors. One male is alive and well nine years after surgery and other survivors range from 8 years down to current surgery. Current survivors have achieved many activities related to travel and just enjoying life. These include golf, outback touring, local and international travel, visiting Antarctica, as well as enjoying family weddings and births of new family members.

In Hospital Experience of Surgery and Recovery.

All EPP patients are transferred to the intensive care unit (ICU) from the operating theatre where intense monitoring continues through the first and second 24-hour periods. The ICU stay is between two and four days.

A number of significant clinical issues need to be considered. These are outlined below under separate headings.

Operation

The lung, rib lining (pleura), lining around the heart (pericardium) and lining of the diaphragm are removed. Gortex is used to reconstruct the pericardium and diaphragm.



Wound

Access to the chest is gained through a cut on the back (see picture). This wound is closed with a dissolvable suture under the skin. There is one suture to remove from the drainage tube site.

Left extended thoracotomy and pleuropneumonectomy wound

Body's response to removing one lung

The body's blood is circulated through both lungs for oxygenation and nourishment. When one lung is removed, all blood is pumped by the heart through the remaining lung. This changed pumping arrangement places extra work on the remaining lung and the heart. ICU staff monitor very closely the body's response to these changes. Any change in heart and lung function have the potential to impact on kidney function so all of these organs are monitored. Monitoring includes heart rate and function, respiratory rate and function by recording oxygen saturations and by daily chest x-rays. Renal function is monitored by blood testing and measuring hourly the amount of urine excreted.

A urinary catheter is inserted while the patient is in the theatre and under anaesthetic for this purpose. The careful monitoring aims to prevent the heart and lung from being overworked and over-hydrated with fluid. Initially this is done by limiting the amount of fluid given by intravenous infusion and then once oral intake is permitted, limiting the fluids taken by mouth. An accurate method of assessing how much fluid is circulating within the body is via a CVP line. This is an intravenous line inserted while under anaesthetic, in the front of the chest near the neck. It tells how much fluid is circulating through the body. It normally remains in place two to three days.

Patients who have the right lung removed will wake with a naso gastric (NG) tube. The tube exits the nose but is placed into the stomach to drain gastric fluid and air. This system prevents gas bubbles from collecting in the stomach and pressing on the raised left diaphragm, thus reducing the respiratory capacity of the left lung. It is usually removed the morning after surgery, before oral intake is allowed.

Changes to the chest with the lung removed

The space that contained the lung is called the pneumonectomy space. Immediately after surgery, one chest drain will be placed into this space and will remain for 12 – 36 hours. The space slowly fills with old blood and “normal” body fluid. A daily chest x-ray monitors the rate of filling. The fluid will stay in the space. Over about three months, the fluid becomes thickened, gelatinous and then solid, but stays in its sealed sterile environment.

The centre alignment of the chest will move towards the side where the lung was removed, as there is no lung providing a counterbalance. This is normal and will stay this way even after discharge. This will be evident on x-rays in the short and long term.

The space between the ribs will shrink, as there is no lung making the spaces open and close (the breathing effect). This is normal. Regular checks and adjustments to posture will prevent extensive shoulder drooping.

Shortness of breath/breathlessness

Removing a lung reduces one’s lung capacity by roughly half, thereby reducing the oxygen reserve, and a consequence is that supply might not keep up with

demand. Any physical activity after EPP requires a higher use of oxygen – activities such as showering, washing hair, talking on the telephone, climbing stairs and just moving, all use increased oxygen. The sensation of shortness of breath that feels like you are “running out of air” is experienced frequently and can be quite frightening initially. In time, the feeling is less frightening as patients learn to be comfortable with their reduced capacity and make adjustments to cope. In time, one learns to trust that the changed respiratory system will not fail – they will not run out of air. Patients learn to SLOW DOWN and adopt manageable activities. Enrolling in pulmonary rehabilitation is highly recommended - see physiotherapy section.

Fluid retention

Retaining body fluid after any major surgery is normal, and this is the case after radical lung surgery. Initially, the hands and fingers and face will be noticeably puffy. As this fluid absorbs, fluid is more noticeable around the chest area, abdomen, scrotum as well as thighs, legs and ankles. Fluid retention early in recovery is related to the physiological pumping changes and its impact on the cardiac system. Several things can be done to monitor and control fluid retention and prevent fluid overload. First, there is a need to exclude that it is related to the pumping of the heart - hence the detailed cardiac assessment prior to surgery. Secondly, fluids are restricted, so that no more than 1200-1500 mls per 24 hour period is taken. The amount depends on body mass. A daily weigh provides a measure for fluid retention. An assessment of trunk, legs and ankles for swelling as well as monitoring respiratory rate and level of breathlessness provide important information about the body's state of fluid balance or imbalance. A diuretic will be prescribed, so that you urinate (wee) the fluid off if excessive fluid is retained. Frequent walking will assist fluid distribution.

Physiotherapy

After surgery, the physiotherapy team are key players in patient recovery. Medical staff will monitor the physiological changes that take place when one lung is removed. The physiotherapy staff will guide and supervise the work needed to keep the remaining lung healthy. This will begin at the pre-operative

meeting, then immediately on return from the operating theatre and thereafter until discharge from hospital.

Patients are expected to get out of bed the day following surgery. The physiotherapy staff will guide this activity according to medical progress. Patients are asked to work hard and will sometimes want to be left alone. Physiotherapists, however, have valuable experience in getting EPP patients back on their feet and home.

Enrolment in a pulmonary rehabilitation programme after discharge, to optimise physical and respiratory recovery is strongly encouraged. Recovery will be slow, but over time patients become confident of their new respiratory capacity and adjust to new limitations related to living with one lung. (See booklet – Living with one Lung, resource information). Some patients may benefit from a period of time in an intensive rehabilitation programme in a nearby private rehabilitation facility before returning home. This provides time to consolidate their exercise regime and build confidence with their changed respiratory and circulatory systems.

Pain relief

Adequate relief from pain is vital for a good recovery. Immediately after surgery, relief from pain is provided by the theatre and recovery room staff but once awake patients are connected to a PCA machine to self administer pain medication. The machine is usually loaded with a narcotic drug called Fentanyl. Once stable, oral pain relief replaces the PCA. Staff monitor pain levels closely and frequently to ensure appropriate tablet medication is given, so that physiotherapy exercising and movement is ongoing with comfort. It is vitally important for patients to be established on a “good” routine of tablet pain relief before discharge.

Please refer to the pain management section within the “Preparing for Lung Surgery” handbook.

Position in bed

Positioning in bed is important to optimise expansion of the remaining lung, and to help remove secretions that have accumulated during anaesthesia. Therefore, it is critical **to not lie on the good lung after pneumonectomy.**

Patients are best positioned on their back with their head elevated so that the air entry is maximised and the remaining lung is not compressed by the weight of the body and made to work harder. Some patients find sleeping in a bed difficult and are more comfortable in a chair. Later, before discharge, and with good pain management it may be possible for patients to lie on the side of the operation.

Nutrition

No food or fluid is taken after surgery until the surgeon gives approval so oral intake usually begins day 1 or 2 post operation. As mentioned above, fluids will be restricted. This is really important. There is no restriction on food, but choose that which is easily digested. Appetite issues or lack of appetite are common, but a dietician will visit and arrange supplements. Symptoms such as reflux, burping and nausea are common and will be managed on an individual basis.

Day-by-day progress

Each day will be a challenge, so focus on getting through one hour and one day at a time. Remember, good relief from pain will allow effective physiotherapy. Patients feel exhausted, but will be expected to exercise (within the bounds of safety), so be guided by the experienced physiotherapy and nursing staff. Activities like personal hygiene can be paced through the day e.g., shave - rest, shower – rest, teeth and hair - rest.

Aim to take small, but frequent steps and rest well between hard work.

Look for improvements week-by-week rather than day-by-day.

By discharge, patients will be moving independently, managing personal hygiene, either alone or with minimal assistance, and be able to walk slowly up a flight of stairs.

Once home and settled, set goals and timeframes that are realistic and achievable.

Further information follows in Part 5 in the form of patient personal experiences reported during questionnaires and interviews conducted during 2010 and 2011.

Long-term Recovery

There are four stages of recovery:

1. Getting over the chemotherapy.

Patients (and their family) will endure almost anything so they can move forward and have radical surgery. After chemotherapy, most patients look well, not malnourished, mostly with their hair intact, tired and may be struggling with appetite. They are encouraged to take added supplements like Sustagen and maintain a good level of exercise, in readiness for surgery planned four to six weeks after the chemotherapy. The importance of maintaining physical exercise during this time cannot be more highly stressed.

2. Getting over surgery.

It takes time, a long time, it is frustrating. Patients struggle to maintain weight and take adequate amounts of food. The appetite is poor and there may be left-over effects of nausea from the chemotherapy. New issues related to a distorted gastrointestinal tract and reflux may be problematic. Pain may be an issue, as well as the overwhelming effect of breathlessness and fatigue – patients want to lie down, but need to exercise. A disciplined approach to exercise and progressing positively is useful. Carers are tired. They struggle to provide food that the patient likes, even though they requested it two hours earlier. Patients ask “Will I ever feel better”? There is in their mind also the need to start radiotherapy. Not every patient has a challenging recovery, but every patient is different and most do come across a few obstacles.

3. Getting over radiotherapy.

After six weeks of Monday to Friday treatment in RPAH Radiation Oncology Department and seeing the specialist every Friday, patients feel exhausted. They have organised and used a transport roster or they and a carer have returned home after staying in Sydney for the duration – perhaps getting home on the weekends. There may be feelings like: “My chest is sore. I can’t eat and I am losing weight. Why did I do this?” But then things start to get better.

4. The next six months.

Treatment behind and a holiday planned, it all seems to have been the right thing to do. Slowly, slowly, you begin to gain energy, gain weight, lose some tiredness, become more active and start to enjoy living again. The timescale varies for each patient but patients do learn to live with the limitations of having one lung. There is nothing you cannot try to do.

Follow up

An early appointment with the GP is required within the first week of discharge from hospital. This visit will acquaint the GP with the current state of recovery. They will continue medical treatment commenced in hospital, check wounds, renew prescriptions, and deal with day to day symptoms that might present.

If a Respiratory Consultant was involved with care prior to commencing trimodality therapy, it is ideal to have a return visit to them so that they can monitor on an ongoing basis the function of your remaining lung. The timing of this will fluctuate but 1 – 3 months after surgery is ideal depending on the timing of other appointments.

An initial consultation with the Radiation Oncologist will be arranged prior to discharge. This is ideally around 4 weeks after surgery but the timing is dependant on recovery and place of residence: Sydney city, country or even interstate. The timing of appointments and treatment will be tailored to individual needs.

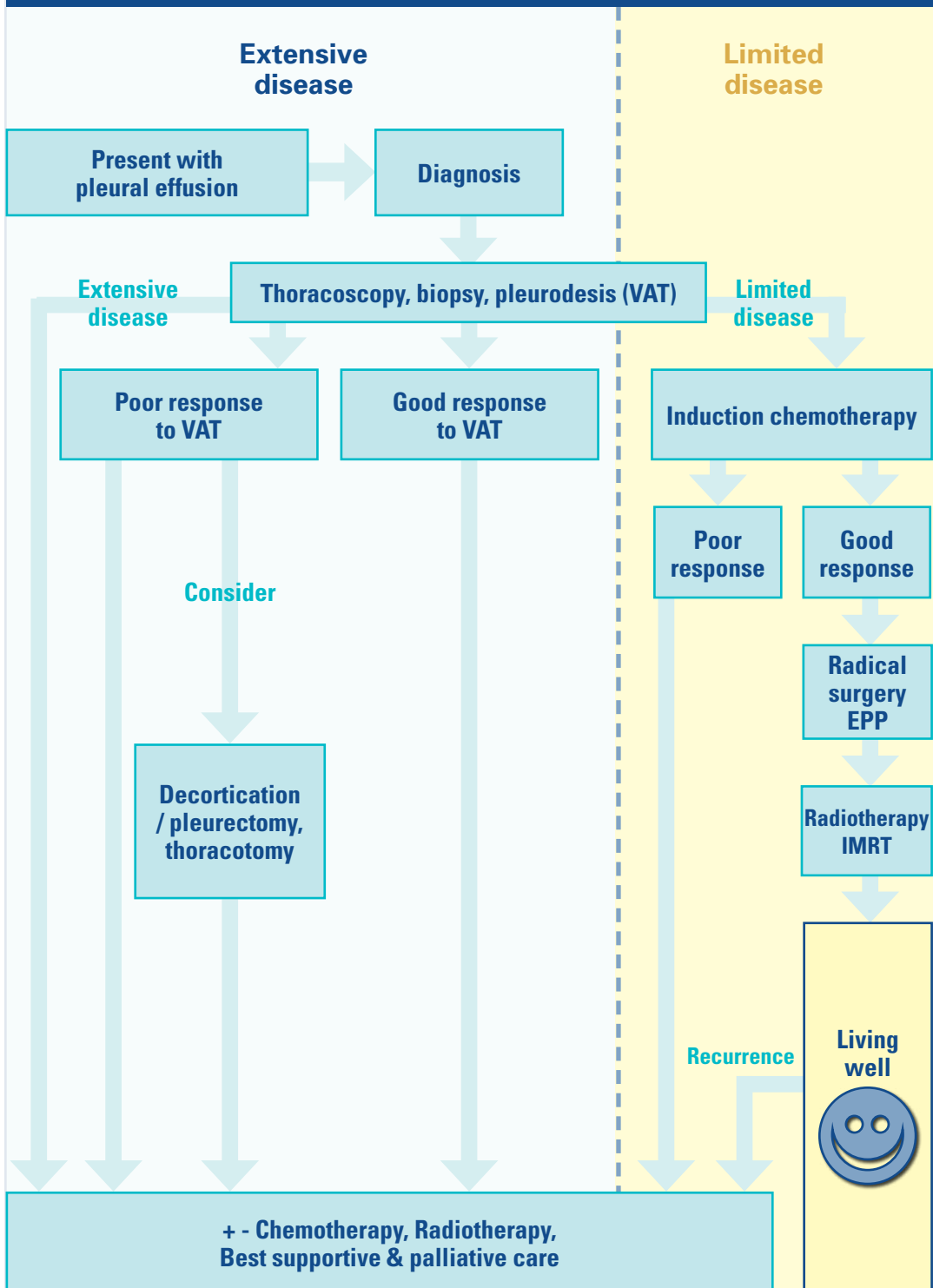
A visit to the surgeon is arranged around 6 weeks post surgery and before radiotherapy commences. If problems arise in between this time, the surgeon can be seen earlier.

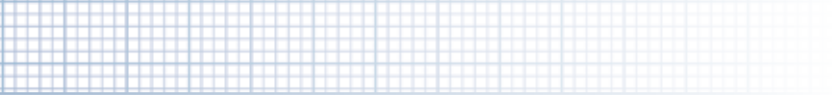
Once trimodality treatment is completed, a baseline CT scan will be completed 3 months after the radiotherapy, and then at 6 monthly intervals, and as required depending on clinical need.

Follow-up visits can alternate between radiation oncology and the surgeon to limit the number of medical appointments however any EPP patient can be reviewed at any time if the need arises.

Surveillance PET scanning is not required, however, a PET scan will be ordered where there is a specific clinical need.

Algorithm for Mesothelioma Management





PART 4

Review of our Radical Treatment Experience – Publications and Research

EPP has been offered to patients since the early 1990s, but became a more accepted option of treatment in the mid 2000s. Radical surgery became part of trimodality therapy from 2008, but prior to that, patients went straight to surgery from diagnosis. A review of the outcomes of treatment and prognostic factors of the first 70 patients who had EPP performed by Prof McCaughan between 1994 and 2008 was completed by Dr Tristan Yan and published in 2009. The abstract, introduction and conclusion of this publication are included here.

Title

Extrapleural pneumonectomy for malignant pleural mesothelioma: Outcomes of treatment and prognostic factors.

Authors

Tristan D. Yan, BSc(Med), MBBS, PhD, Michael Boyer, MBBS, PhD, FRACP, Mo Mo Tin, MBBS, Daniel Wong, MBBS, Catherine Kennedy, RM, RA, Jocelyn McLean, RN, MN, Paul G. Bannon, MBBS, PhD, FRACS and Brian C. McCaughan, MBBS, FRACS

J Thoracic Cardiovascular Surgery 2009;138:619-624

Abstract

Objective: This study aimed to evaluate the peri-operative and long-term outcomes associated with extrapleural pneumonectomy for patients with malignant pleural mesothelioma.

Methods: From October 1994 to April 2008, 70 patients were selected for extrapleural pneumonectomy. Univariate analysis was performed using the Kaplan–Meier method and compared using the log-rank test. Multivariate analysis with entering and removing limits of P less than .10 and P greater than .05, respectively, was used. The prognostic factors included age, gender, side of disease, asbestos exposure, histology, positron emission tomography, date of surgery, neoadjuvant chemotherapy, completeness of cytoreduction, lymph node involvement, peri-operative morbidity, adjuvant radiotherapy, and pemetrexed-based chemotherapy.

Results: The mean age of patients was 55 years (standard deviation $\frac{1}{4}$ 10). Fifty-eight patients had epithelial tumors. Six patients received neoadjuvant chemotherapy, 28 patients received adjuvant radiotherapy, and 16 patients received post-operative pemetrexed-based chemotherapy. Forty-four patients had no lymph node involvement. The peri-operative morbidity and mortality were 37 per cent and 5.7 per cent, respectively. Complications included hemothorax (n $\frac{1}{4}$ 7), atrial fibrillation (n $\frac{1}{4}$ 6), empyema (n $\frac{1}{4}$ 4), bronchopulmonary fistula (n $\frac{1}{4}$ 3), right-sided heart failure (n $\frac{1}{4}$ 2), pneumonia (n $\frac{1}{4}$ 1), constrictive pericarditis (n $\frac{1}{4}$ 1), acute pulmonary oedema (n $\frac{1}{4}$ 1), small bowel herniation (n $\frac{1}{4}$ 1), and disseminated intravascular coagulopathy (n $\frac{1}{4}$ 1). The median survival was 20 months, with a three-year survival of 30 per cent. Asbestos exposure, negative lymph node involvement and receipt of adjuvant radiation or post-operative pemetrexed-based chemotherapy were associated with improved survival on both univariate and multivariate analyses.

Conclusion: The present study supports the use of extrapleural pneumonectomy-based multimodal therapy in carefully selected patients with malignant pleural mesothelioma.



PART 5

Quality of Life Studies & Support Group

The study by Yan et al, led to a need to learn more about the recovery of patients after EPP, in terms of the quality of life of survivors. The main reason for this was so that new patients considering this option of treatment could be better informed about their likely journey. The ultimate aim of any ongoing study was to produce an information book for new patients based on fact, clinical evidence, and experiential information. The results of this quality of life work were presented in poster format to the 14th World Lung Cancer Conference in Amsterdam, July 2011. The study was approved by the RPAH Human Ethics Committee. A review of the study follows.

Title

Extrapleural pneumonectomy (EPP) for malignant pleural mesothelioma: what is the quality of life of survivors?

Authors

McLean, J, McCaughan, B.C

Astract

The study reported the quality of life of survivors from the Yan study, as well as those who had had EPP after 2009, so the population consisted of patients who were living without disease, as well as those living with recurrent disease, up until the end of March 2011.

Methodology:

There were two components to the study - one quantitative, one qualitative.

The quantitative component measured the quality of life of survivors who had had EPP, using two recognised cancer questionnaires – the European Organization of Research and Treatment of Cancer (EORTC) QLQ-C30 and LC-13 and the McGill Quality of Life Questionnaire (MQOL). The QLQ-C30 provided a core measurement of Global Health, Functional Scales and Symptom Scales. The QLQ-LC13 provided specific lung-related symptom scales, some of which were related to chemotherapy. The MQOL questionnaire was chosen to measure the subjective wellbeing of patients, i.e., what they believed their quality of life was.

The qualitative component of open interviews was conducted over several months. Patients and a carer were invited to talk about their experience of radical surgery and recovery in the setting of a diagnosis of malignant pleural mesothelioma. The interviews were transcribed into text and then, using a phenomenological methodology, the text was scrutinised looking for recurring themes that would interpret patients' lived experience of recovery following this surgery.

Study Population

The study populations were survivors who had EPP between January 2004 and December 2010. One surgeon performed all of the operations. One case manager was involved with the care of all of the patients.

The first mailing of questionnaires measured the QOL of 13 patients during July 2010.

The second mailing of questionnaires measured most of that same group of patients, as well as those who had surgery in 2010, with the total being 17 patients at the end of March 2011.

The characteristics of the total study population are listed in **Table 1**.

Table 1

Characteristics of study population in March 2011		
Sex	Male	15
	Female	2
Age at time of surgery	Median	62
	Range	40-72 years
Side of surgery	Left	5
	Right	12
Induction Chemotherapy++	Yes	12
	No	5
Radiotherapy completed**	Yes	13
	No	4
Patients with Recurrent Disease (Months from Sx to recurrence)	Total	5
	Range	17-40 months
Months from Surgery to March 2011	Mean	33 months
	Range	6 - 87 months
Deaths during study (all from progressive disease)	Male	2
	Female	1

** 2 patients did not have radiotherapy, as there was no microscopic disease after chemotherapy

2 patients were not well enough to have radiotherapy

1 patient had treatment in NZ

Radical radiotherapy was commenced in 2004. One specialist has treated all of the patients, except the patient from NZ.

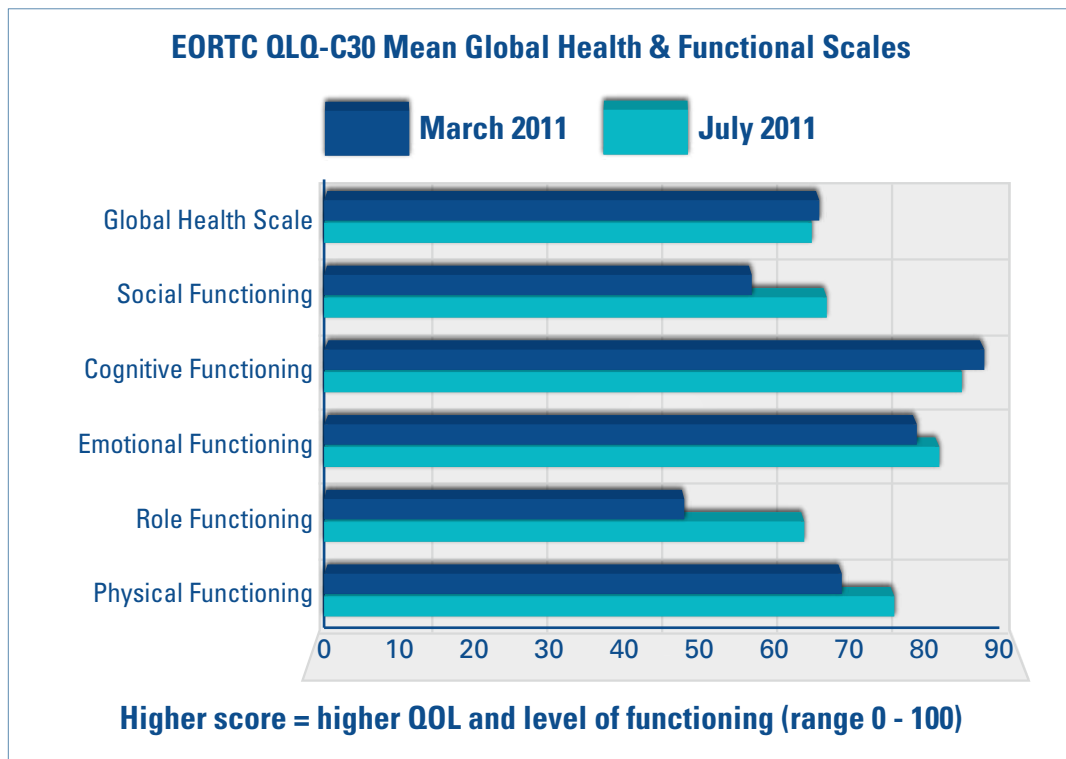
++ Induction chemotherapy became standard management in 2008, therefore patients prior to 2008 may not have induction chemotherapy.

Two quality of life questionnaires were chosen to measure quality of life domains that included global health, functional and symptom scales, lung cancer specific symptoms, as well as physical, psychological and social wellbeing.

EORTC QLQ-C30 (Cancer General)		EORTC QLQ-LC13 (Lung Cancer specific)	
Global Health		Dyspnoea	(3q)
Global health status	(2q)	Coughing	
		Haemoptysis	
Functional Scales		Sore mouth	
Physical functioning	(5q)	Dysphasia	
Role functioning	(2q)	Peripheral neuropathy	
Emotional functioning	(4q)	Alopecia	
Cognitive functioning	(2q)	Pain in chest	
Social functioning	(2q)	Pain in arm or shoulder	
		Pain in other parts	
Symptom scales		McGill (MQOL)	
Fatigue	(3q)	Physical symptoms	(3q)
Nausea and vomiting	(2q)	Physical wellbeing	(1q)
Pain	(2q)	Psychological wellbeing	(4q)
Dyspnoea	(1q)	Existential wellbeing	(6q)
Insomnia	(1q)	Support	(2q)
Appetite loss	(1q)		
Constipation	(1q)		
Diarrhoea	(1q)		
Financial difficulties	(1q)		

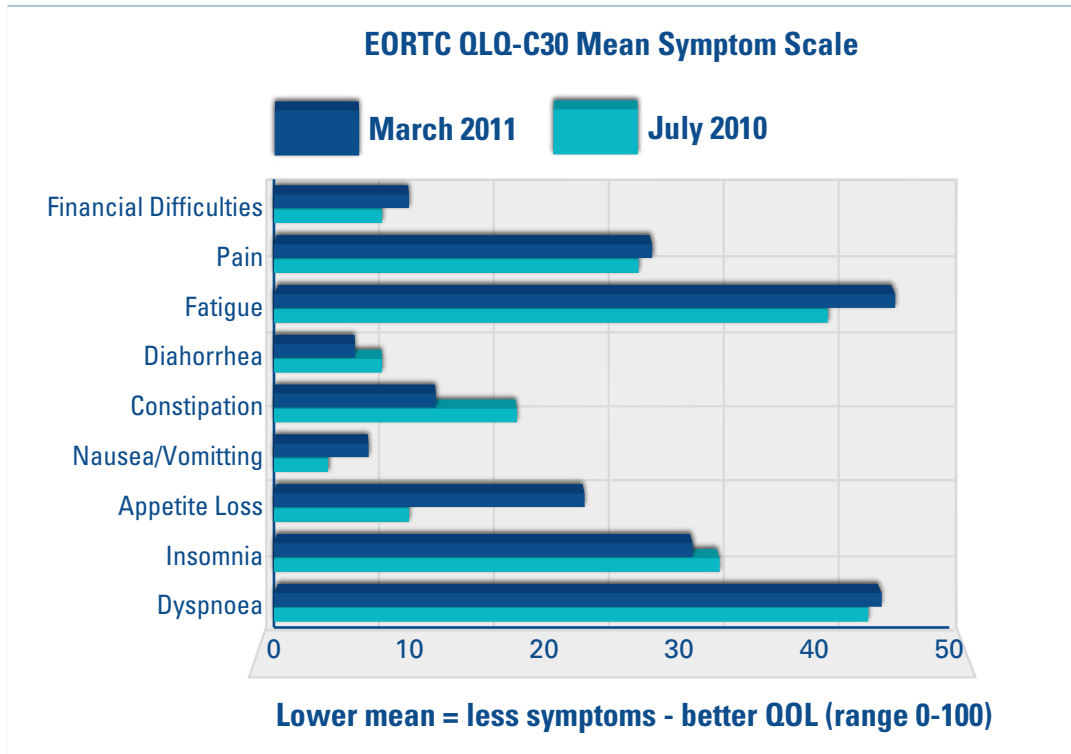
Results: EORTC QLQ-C30 Global Health & Functional Scale

There was no major difference between the two groups – July 2010 and March 2011 except for Role and Social Functioning. This difference may be related to three patients living with recurrent disease and dying in the period March -May 2011.



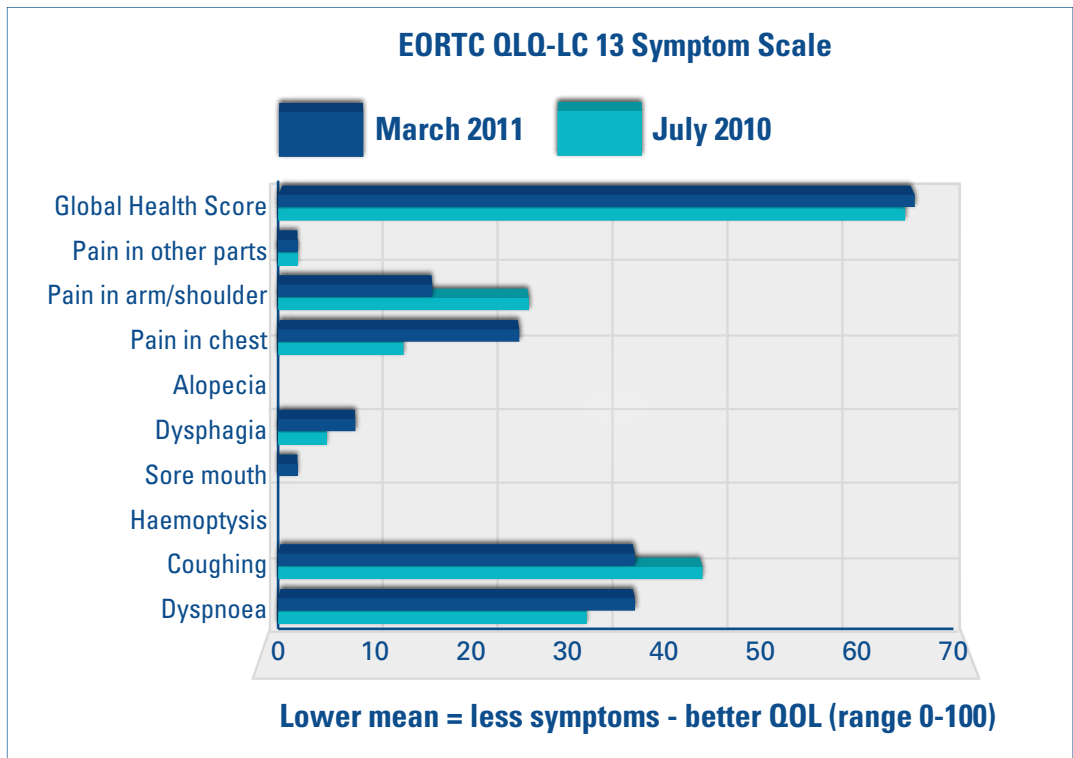
Results: EORTC QLQ-C30 Symptom Scale

There were similar rankings in both groups, July 2010 and March 2011. Fatigue and dyspnoea ranked highest and were therefore the most troublesome symptoms. Insomnia and pain were a problem and were similarly ranked.



Results: EORTC QLQ-L13 Symptom Scale (Lung Cancer Specific)

There were similar ratings between both groups, July 2010 and March 2011. Like the QLQ-C30 findings - dyspnoea and cough were the most troublesome symptoms. Pain was also an issue. Despite symptoms, the global health score ranked positively.



What Do These Results Tell Us?

In the context that there is a view held by health professionals that radical surgery is associated with increased morbidity and poor QOL, these results were compared with patients enrolled in three different studies for the purpose of seeing if the QOL of EPP patient is truly worse than other MPM patients. The comparative studies were as follows:

- Study 1 Patients with mesothelioma having chemotherapy. Nowak et al 2004 (patients with pleural mesothelioma undergoing combination chemotherapy).
- Study 2 Patients with mesothelioma unsuitable for surgery and before any medical treatment. Bottomley et al 2007 (histological proven unresectable MPM, not pre-treated with chemotherapy).
- Study 3 Patients with proven lung cancer (NSCLC) prior to potentially curative surgery. Kenny et al 2008 (Baseline QOL assessment of patients with clinical stage I or II NSCLC).

Results: EORTC QLQ-C30 Global Health Status and Functional Scales

The Global Health Status was similar across all groups.

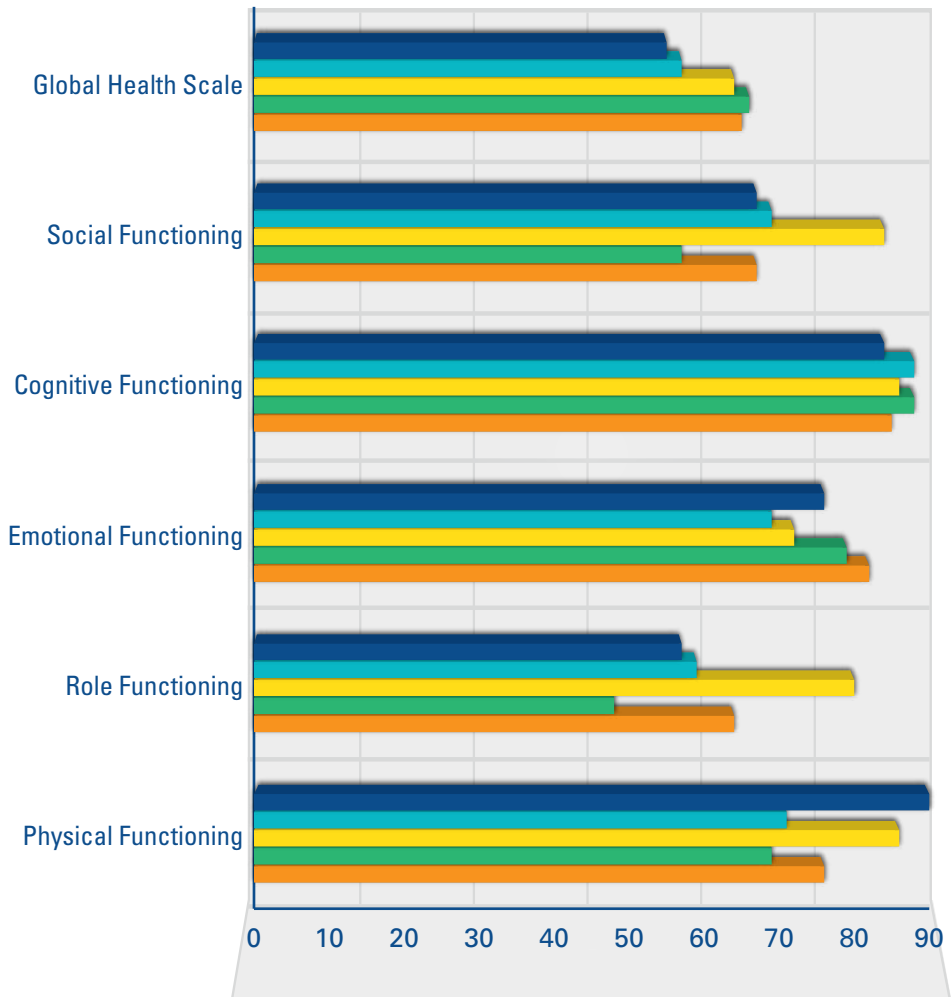
Cognitive functioning and emotional functioning was similar in all groups.

Social functioning and role functioning was better in group 3 (prior to surgery) - this is not an unexpected finding, given it was before potentially curative surgery.

Physical functioning was positive in all groups, but better in group 1 (during chemotherapy) where good responses to treatment should be associated with better functioning, and in group 3 (prior to surgery) for NSCLC.

EORTC QLQ-C30 Global Health Status and Functional Scales Comparison with other studies

- 1. MM During Chemo
- 2. MM Prior to Chemov
- 3. NSCLC prior to Sx
- EPP March 2011
- EPP July 2010



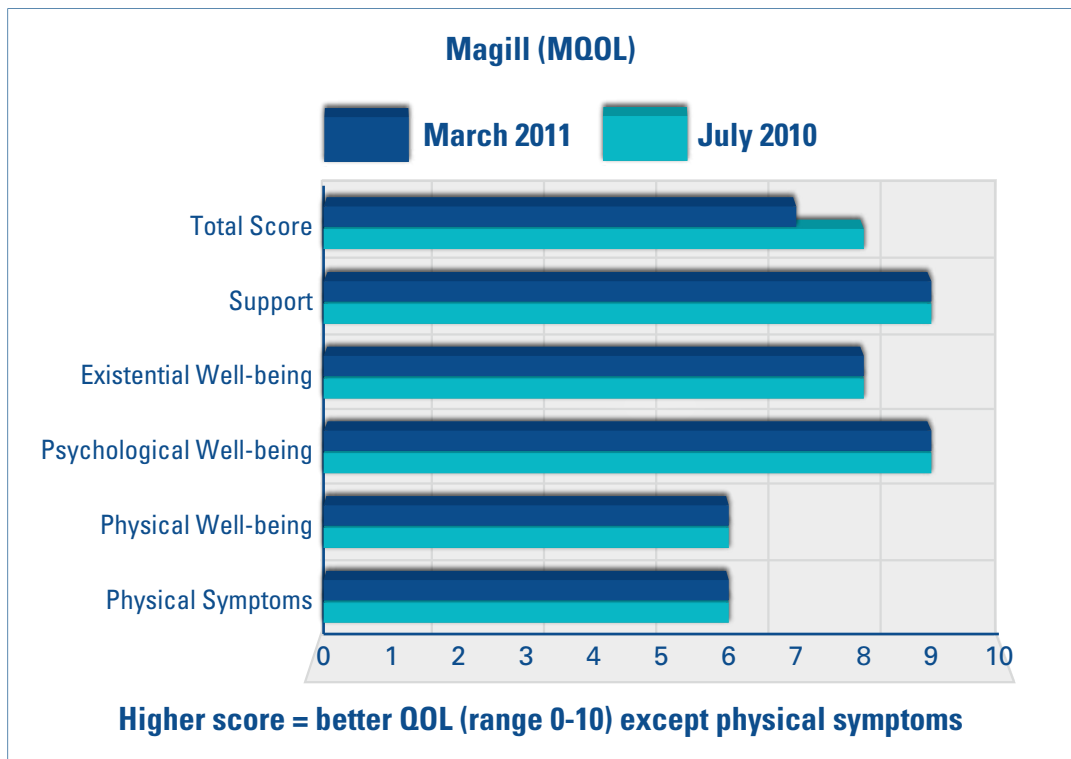
Results: MAGILL- MQOL

This is an instrument used more often in the palliative care setting, but was chosen to get a focus of experiential issues of living with a fatal disease.

Both groups recorded similar rankings, July 2010 and March 2011.

All patients reported a negative impact of physical symptoms.

Despite the physical symptoms, all other dimensions were ranked very positively.



Qualitative Component

The second part of the study was the qualitative component, where eight patients (each with a carer present) were interviewed between November 2010 and May 2011. There were seven males and one female. Three patients gave interviews but later died of recurrent disease during this period.

The interviews were transcribed into text. Each text was then read and re-read, looking for common themes. Statements were coded on the text and then categorised according to the message theme. The message from the surviving EPP patients is expressed in the following thematic statements.

1. I want to live longer; I will explore all options.
2. I made my decision and I have no regrets.
3. There were three treatments: chemotherapy, surgery and radiation – radiation was the most difficult.
4. Someone talk to me. Give me information. Give me hope.
5. Living with the limitations of one lung is manageable and acceptable: living with recurrent disease is intolerable.
6. Relationships: professional and personal. Some are developed, some are challenged.
7. Recurrence: Is this the end of the line?

For the purpose of understanding the real impact of the interviewee experiences, each statement will be further explained the interviewee dialogue will appear in *blue* print in the text.

1. I want to live longer; I will explore all options.

There was an overwhelming feeling from patients that they would do all in their power to live longer and, of course, their carers to have their partner for longer. This is a natural human desire, but when facing certain death, intelligent and motivated people, who were newly diagnosed with mesothelioma, searched for treatment options. Discovering radical surgery came from a variety of sources: the surgeon at the time of diagnosis, searching the internet, other patients,

family contacts, other physicians and surgeons, legal attorney, and TV shows. Prior to any surgery, all the patients were made aware of the pros, cons and risks of radical surgery, including the risk that they may not leave hospital alive, however, their desire to live longer outweighed their risk of the surgery.

“There was not any doubt. We had some discussions with the surgeon about other things and he said he had other people who had made a decision not to have the surgery, but I knew I was not just going to go through chemotherapy and waste what time I had left, so for us it was full steam ahead.”

The comment *“I could not sit back and be idle”* was a common sentiment.

Two patients considered travelling overseas for surgery, but on discovering that it could be performed in Australia, they were prepared to have it in Sydney. A number of the interviewed patients and carers asked why the option of radical surgery was not freely discussed with mesothelioma patients. The group in general was not aware of how few patients actually have this surgery in Australia.

2. I made my decision and I have no regrets.

Each patient had a unique experience of surgery and recovery. Some are currently living well without disease. Some have died of recurrent disease since their interview. No patient expressed any feeling of regret for having the surgery, as the following dialogue suggests.

“Yes, I would do it again. I don’t think I would risk not having it done. Maybe I might have waited a bit longer, but I wasn’t really painted that picture..... because [the surgeon] said if we can’t get them reduced, then I just can’t do it. When they were reduced, that was like the green flag I just got that green flag that I could have it done and went to for it.”

“It wasn’t a hard decision to have my lung out, and wasn’t something I took lightly, but I didn’t see that I had an option.”

“There is no parallel life to compare with so (I chose to) make a decision and then look forward, not back.”

“I have no regrets; surgery is a life line not a sentence.”

3. There were three treatments: chemotherapy, surgery and radiotherapy – radiotherapy was the most difficult.

Chemotherapy

Not all patients had chemotherapy. Those that did talked about it without much fuss. They didn't like it, but put up with the related ill-feeling and discomfort of treatment, knowing that they needed a good response from the drugs to be able to proceed to surgery.

"...the surgeon said that if the chemo works we will then talk about surgery and if we have surgery I have got to tell you that the mortality rate..."

Surgery

Surgery provided a wide range of experiences. Overall, patients had about a 2 week stay in hospital. Two interview patients had complications: chyle leak and pneumonectomy space infection, with a consequential prolonged recovery. The overall attitude, however, was that they would endure anything to have the tumors removed and a hopeful chance of prolonged living.

"I also had the attitude that if you have cancer you have it cut out, you get rid of it."

The hospital experience of surgery and recovery was for most not the most negative aspect of their recovery. They recalled pain and discomfort, but most were willing to endure discomfort for potential gain.

Patient: *"It was sore, there was no doubt about that, but it was well and truly medicated and it was discomfort rather than soreness. I cannot so clearly remember it now, but I do know that everyone was there (supported by staff), I had all the support I needed and the physio came in and got me moving, and I just did everything they wanted me to do. I opened my eyes in recovery and you (wife) were standing there."*

Carer: *"I had been bawling my eyes out."*

Follow-up care never stops.

Wife: *“Well, it ALL burst out, well here is something, we didn’t know that we could go back to hospital, ...his wound was leaking profusely, and um, they wouldn’t touch it, no one would touch it till Monday because it wasn’t their baby. We didn’t know, we had no idea we could go straight back to hospital. ... we were too naive to know, we just went to the local medical centre.*”

Patient: *“We didn’t know what it was initially, we did a bit of work. I was sitting on a chair out on the balcony of our old homestead and I could feel a lump developing there and I said, I am weeping. Local doctors in (suburb) run a seven-day day practice, but no, she (the doctor on duty) wouldn’t touch it. By Monday we got back to the hospital and were told by the young Irish sister then ...to come back anytime.*”

Radiotherapy

Patients were very expressive about their experience of radiation treatment. It was not an easy treatment.

“Radiation was worse. Radiation is hell on earth; I think it is barbaric.”

“Radiation seems to be the hardest thing to get over. Even the chemo you had two good weeks, you had one rotten week but one good one before you had the next one [treatment].”

Radiation-induced neuropathic pain had a catastrophic impact on the early recovery and quality of life of some patients.

“It was the radiation that really knocked me around. And in the last three weeks the pain in my left breast has just been excruciating. I feel like my QOL has been taken away ...I can’t do things that I used to be able to do, even just recently at home we have had work done which I just had to go and lay down and walk away from, because I couldn’t get around,.....it just felt like a huge part of my life had been taken away. I was looking forward to getting back, probably not to 100 per cent of where I was, but when the tablets [medication for neuropathic pain] take over and I start to feel pretty good then this is where I am heading for.” This patient has just returned from travel though Asia (May 2011).

Patients who did not live within the Sydney Metropolitan were required to relocate to Sydney for the six weeks of daily treatment. This resulted in a separation from support networks, as well as the comforts of home. Sydney

based patients had to cope with the daily grind of driving to and from treatment in city traffic up to 4 hours of driving a day.

“As far as having people around, like you and [Dr XRT] and [the surgeon] and the rest, even the nurses here, we could not have wanted any better than what we have got. Yes, I think we feel well supported down here (Sydney). Not so well at home (country) because I guess it is not the sort of illness that they have very much [experience with] at home.”

“He coped really well, wasn’t sick and I am a very lucky lady, we had a wonderful system with our friends, we had a roster system...we have a group called the retired teens. They just wrote a roster. I had support from both family and friends.”

The body became physically tired and maintaining a good dietary intake was an issue. Patients and carers became emotionally drained, but after treatment they focused on rehabilitation.

Pulmonary rehabilitation was helpful, in that it enabled patients to manage some of the physical limitations of having one lung. It provided a disciplined exercise regime and helped patients take the focus off themselves and onto other “sicker” people in the exercise room.

“They would get you doing a series of exercises and want you to get to a point of breathlessness, so you can then recover from that and they would keep upping the ante all the time and force you to do more and more, but they were good. They taught me how to breathe and how to recover.”

Good dietary advice was also imperative to recovery.

“But you know, I was strong enough to make myself eat and I did that all the time. I went to the chemist, I got big drums of protein foods, and, as revolting as it was, I had these drinks every day. I never went without them and I never let my weight go below 75kgs.”

4. Someone talk to me. Give me information. Give me hope.

The issue of information, or lack of information, about the disease and treatment options at the time of diagnosis was a common finding. In some cases, patients received no useful information from health professionals and went searching on their own. All they knew was that they had an incurable disease and a very limited lifespan.

“The whole family was in shock. I mean me particularly. There was nothing for me. I was going to lose my husband and I didn’t know how long I was going to have him and that went through my mind planning, “what am I going to do without him?”. Then we saw [the surgeon]. He gave us the cold hard facts. He doesn’t lie, but at least we knew that there was hope, that there was always hope, but prior to that, it was a hard couple of weeks.”

The Internet provided clinical information which was essential, but patients were very cautious about it. The Internet was bad news, at a time when they were most vulnerable (newly diagnosed).

“We had no information at all, and to get through that, that was horrific. There wasn’t a piece of paper and so we got on the Net and when we got on the Net it was all bad. There was no good news on the Net.”

The need for an EPP-specific support group became clear as the interviews progressed.

Partner: *“I think people need to have the opportunity to just chat to people.... most people won’t go to them [support groups], but if it was for a specific group of people..... I think that would be beneficial.”*

Patients were unaware that they are part of a *“small exclusive club”*.

Meeting another patient helped them to see they were like others and this made them feel OK about the physical limitations they were living with. Seeing other patients living well was inspirational and motivating and made them hopeful of being a long survivor.

“Hope is real when you are with a survivor.”

5. Living with the limitations of one lung is manageable and acceptable: living with recurrent disease is intolerable.

Living well and with quality was a priority and so patients stopped work. This enabled them to focus on their prolonged treatment course and rehabilitation, as well as the fact that they were not able to do the heavy physical work of their usual occupation. They insisted that living with one lung was not a major issue. They accepted there were physical limitations, but it was better than being dead.

"I have seen the arrival of two grandchildren and a daughter married in the last four years, when I was given 6-12 months to live and no operation."

A supporting carer was essential for patients to get through their treatments. Early in recovery some patients felt frustrated and demoralised, as they watched their partner struggle with home chores that they usually did.

"I feel for [my wife] and it is hard for me to be sitting in the house and see her out doing what I used to do around the property... and then she comes in absolutely exhausted and that is a concern."

Despite all of this, life took on something that resembled "normal". Patients travelled internationally, became active in family life, got back to "social" sport, as opposed to competitive sport. Golf was a popular leisure activity. Patients were keen to share their experience with prospective patients.

"For that 12 months after the treatment he was playing golf three times a week, 18 holes and walked the whole lot and tennis... and Thursday night he played with the boys."

"You do learn to regulate YOURSELF. When you first jump into something you think "I can do it", then you realize that you can't and get to know your limitations, like playing golf. I could and I will just walk at this pace. I would be alright, but if I tried to go at faster pace... so I would make sure I would regulate myself by having little breaks half-way along."

Limitations vary between patients.

Interviewer: *"Was the surgery what you anticipated?"*

Patient: *"Nothing like it. I thought I would come out and in a couple of months I would be back to normal. And then I realised I only have one lung AND it is*

the smaller one. They took the big one...I used to use the weights as well, and then.....that is the worst thing for me, it is the lack of breath."

Disciplined exercise paid off!

I found I couldn't just walk on the street, we lived on the bottom of a gully. I would get too exhausted, I had to sit on the neighbour's fence till I got my breathing back, so thought I would measure up and down the concrete slab around the house. So I measured how many laps I had to do to do a kilometre. I got the chalk and blackboard and I would mark off every lap. I had a chair and I would have a coffee and then go again. I would try to do 1 to 1½ kms at a time.

6. Relationships: professional and personal. Some are developed, some are challenged.

It is well known that an illness experience challenges existing personal relationships and sees the development of new professional ones. The researcher learned how vital rock-solid personal relationships were to EPP patient recovery.

"Nobody would have put up with me. There is no doubt about that."

Wife: *"He just complained about everything. No matter what I cooked him!"*

"Maybe I could have survived on my own, but it is much easier with a strong person."

Patient/carer relationships are tested.

Patient: *"One of the things I have done which is not very good is, if the tablets are wearing down, I get very agitated at things."*

Carer: *"He gets short-tempered."*

Patient: *"I could not do it the other way around and be the carer. I would be hopeless."*

Carer: *"Sometimes I wish it was me, I can't bear to see him going through this and he doesn't deserve it. He is such a good person."*

Patient: *"The carer's job must be really hard though. I fluctuate up and down."*

Carer: *"I am just trying to make life easier for them, constantly working. I wanted to be a nurse, but my father wouldn't let me."*

There was always the lighter side of relationship issues.

Carer: *"So he didn't like my cooking and he couldn't get a good night's sleep ... he bought it up one time when we were in your office. He said, would I marry him. I said, "hey we have other things to worry about...(they are now married!)"*

Professional relationships were crucial and lasting.

Patient: *"... I knew that I had looked after myself ALL THE TIME and then I had this miserable disease and I was really pissed-off with it. Bum, and then I thought.*

Carer: *"... You're arriving in Sydney under the flight path and you go to the top surgeon in Sydney and to another specialist, who is one of the top five in the world and then you go to a top hospital.*

Patient: *"...I think that being in their hands was a very reassuring situation, especially when I met the team...I never had any qualms about the operation. I asked (surgeon) if he could give me a couple of years and he said ...so we have got our fingers crossed.*

The longest-survivor commented *"The professionals are moulded like Leggo"*.

"I feel that you need to have complete confidence in your case worker, your doctor, your oncologists and the whole group that are looking after you. But you were all part of a circle and not one had a different attitude. You were all consistent, it was all the same and that really helped us, before you have this operation you have to feel secure."

7. Recurrence: is this the end of the line?

All three patients who died after the interviews were completed, died from malignant ascites. What was most challenging for them was not having a plan about this stage of living with the disease. They accepted that they were dying and were grateful they had done everything possible to arrest the disease. They had had some element of control of their living while well, but then the

development of new symptoms and increased weakness took that control from them. Here is one conversation:

Interviewer: *“What was the first sign that things were going wrong?”*

Patient: *“Oh, last year. Last year I was on a golfing trip down atI wasn't quite sure what it was, I thought it might be the heart tablets, but it wasn't. Then I went up and got the fluid drained and [the surgeon] arranged the biopsy and we realised then that it had recurred.”*

Interviewer: *“What did you say to [your wife] then. What were your thoughts?”*

Patient: *“Oh, my thoughts were f... it has come back. (giggle). I just said to her, it's come back. It's been a bit longer [living since surgery] than I thought. That was all.”*

Interviewer: *“Do you have any regrets?”*

Patient: *“No regrets, no regrets. I have lived my life the way I want it, everything is set up, I am pretty right. Yep, they just won't give me Nembutal, that's all. But that is society, it's not the doctors.”*

Another patient commented:

“I had an early scan because I was complaining so much, but he just said it [chemotherapy] was not working [to control peritoneal fluid], so it is palliative care. That wasn't easy to take, but I was lucky. There were a series of articles that came out in the paper about palliative care that made me realise that it was not the end of the line. If I hadn't had that article I think I would have thought, Oh I am in Greenwich now to die.

No-one ever told me that something could be done [about the fluid], so I went back to the Internet and found something about a peritoneal drain or something. I had been to the local doctor and whined to him, I had been to the physio and had no luck, then I rang you.”

A brave lady said six weeks before she died:

“From the word go, when I found there was an option to delay the disease, I never thought I could be cured. I never believed I could be cured, that this would cure me, so all I wanted to do was delay the onset of what will happen

next, retard it, slow it. To me, it has done what I wanted it to do. Someone was diagnosed at the same time and she never had her lung out and she probably is in a better health situation than I am, but who is to say that? The disease is so fickle. Who is to say that I could be pushing up daisies and she could go on for another five years or vice versa, but no-one knows. You just have to have a little faith."

There was a resounding agreement amongst interviewees, that while the treatments were difficult and challenging and in some cases unbearable, they would not have changed their decision to have surgery. This study is a true account of how this patient group (and their carers) felt about their experience of surgery and recovery.

At the conclusion of the study a thank you morning tea was hosted by The Baird Institute, for those who had participated in the study as well as all other known survivors and their carers. Twelve patients attended, as well as ten partners. The meeting proved invaluable, as patients were encouraged by seeing and exchanging stories with fellow survivors. They were provided with tips on living with one lung, managing fatigue and coping with breathlessness, by a pulmonary rehabilitation physiotherapist. The group meet at Doltone House, Sylvania Waters – the premises of the late Biaggio Signorelli, who died of malignant pleural mesothelioma. It was agreed by all that these meetings needed to be ongoing.



PART 6

Support Group

Following the publication of our surgical results and the reporting of the quality of life of survivors, a request from survivors to meet other survivors and to share experiences led to a fruitful first meeting in March 2011. Following from this, a one year grant was obtained from the Comcare Asbestos Innovation Research Fund to develop a “well living programme”.

This programme ran through 2012 and was integrated into the support group. The programme was titled: Meeting the needs of patients following extrapleural pneumonectomy (EPP).

The support group meetings were very well attended and provided opportunities for survivors and carers to develop new and trusting friendships. Time spent sharing stories and experiences has been a powerful healing and motivational tool.

The Programme in brief.

The aim of the well living programme was to optimize the physical, emotional, and social functioning of patients and to assist carers to perform and maintain their continuous and vital role. The programme ran as 4 one day sessions (April, June, August, and November). A key requirement of the programme was to allocate time for support networking amongst participants. The group decided at their first meeting that patients would be called survivors, and carers called carers.

Goals

The goals set at the first meeting were as follows:

- Establish a meaningful support group for survivors and carers that had a structure that would ensure relevant information was provided.
- Assist survivors and carers to focus on living well after treatment.
- Address the negative aspects of the EPP quality of life study in particular issues related to role and social functioning and relief from symptoms of fatigue, pain, shortness of breath and insomnia.
- Enable networking amongst survivors and carers by way of open conversation and sharing of stories.

- Provide written information for new patients and carers considering this treatment option.
- Realise the potential of this group in relation to advocacy.
- Gain public recognition for what this group means and how it can impact on the wider community.
- Set a group goal – consider a significant activity involving walking – e.g. walk across the Sydney Harbour Bridge to the ADFA mesothelioma commemorative ceremony – November 2012

Well Living Programme Topics

The well living programme consisted of 4 single days that ran from 10 am until 3 pm. The subjects covered were carefully chosen and programmed to meet the needs of participants and included the following:

- Optimising living with one lung: addressing symptoms of breathlessness and fatigue.
- Physical and respiratory assessments: spirometry testing and 6 minute walk test.
- Mindfulness healing: an introduction to the role of mindfulness healing and meditation, gain an understanding about how disease impacts on the daily lives of survivors and their carers.
- Resilience: sharing and documenting skills and knowledge – with survivors.
- Resilience: sharing and documenting skills and knowledge – with carers.
- Resilience sessions feedback: the living document. See The Baird Institute website for the living document.
- Writing: fun with words – survivors.
- Music: healing and relaxation for carers. Reinforcing the notion of self care.
- Creating opportunities for self-attention and nurturing for carers.
- Pain management related to the why, where, when of pain after radical chest surgery.

- Eating for Wellbeing: A self management plan. Meeting nutritional needs is something most patients struggle with during treatment and recovery. Carers have the challenging task of providing the food.
- Relating science to the living experience: the role of Biobank and ADRI research.
- Walking is the best medicine: The meeting began with a 1.5 Kilometre walk around the harbour foreshore.
- What has everyone been up too. Participants have been active in advocacy work and shared their experiences with others.
- Programme in review. Goal for 2013 – walk the Sydney City to Surf 14km in August 2013.

The Future

Each meeting was evaluated. Results suggested the programme provided relevant information to those who attended. Many have responded and are optimizing living well. The support group continues.

The Comcare grant was extended so the programme could continue into 2013. The programme focused on physical exercise and was titled: “Walking is the Best Medicine”. The aim of this programme was to increase the overall level of fitness of survivors and carers and prepare them to participate, if they chose, in the 7 kilometre “Round the Bay Walk” on 4th August. This walk was chosen instead of the city to surf as the route would be more suitable to more survivors and their carers and families.

The Round the Bay walk was extremely successful. Six survivors and 58 other walkers comprising of carers, family, friends, research staff, and pets completed the walk on a beautiful spring like Sunday morning and concluded with hospitality provided by The Baird Institute.



Abbreviations

ADFA	Asbestos Diseases Foundation of Australia
ADRI	Asbestos Diseases Research Institute
CT scan	Computed Tomography
CVP	Central Venous Pressure
DDB	Dust Diseases Board
EORTC	European Organization of Research and Treatment of Cancer
EPP	Extrapleural Pneumonectomy
GP	General Practitioner
ICU	Intensive Care Unit
IMRT	Intensity Modulated Radiotherapy
MPM	Malignant Pleural Mesothelioma
MQOL	Magill Quality of Life
NSCLC	Non-small Cell Lung Cancer
PCA	Patient Controlled Analgesia
PET scan	Positron Emission Tomography
QOL	Quality of Life
RCT	Randomized Controlled Trial
VAT	Video Assisted Thoracoscopy

Glossary

Chemotherapy

Chemotherapy is a systemic cancer treatment using one or more cytotoxic antineoplastic drugs.

Decortication

Surgical removal of the surface layer of the lung. Also called visceral pleural decortication.

En-Bloc

Removing tissues in one piece – they remain connected to each other – as one mass.

Extrapleural Pneumonectomy

Removal of the lung, chest wall (parietal pleura), hemi-diaphragm and pericardium (lining over the heart) in one piece (en-bloc). Defects over the hemi-diaphragm and pericardium (heart) are closed with a layer of mesh.

Malignant Pleural Mesothelioma

Malignant Pleural Mesothelioma (MPM) is a cancer that occurs in the lining of organs of the chest cavity, abdominal cavity and pericardium (lining of the heart).

Palliative Care

Palliative care relieves symptoms and improves patient comfort when disease progression negates curative therapy and death becomes more imminent. Palliative care provides sensitive support to patients and families on all matters related to living with a terminal cancer.

Pleurectomy

Peeling pleura from the lining of the chest / rib cavity - called parietal pleurectomy.

Pleurodesis

Sterile talc powder is instilled into the pleural space. It irritates and causes inflammation between the lung lining (visceral pleura) and chest wall lining (parietal pleura), so that they fuse together.

Radiotherapy

Radiation beams are directed to a specific site where cancer cells are, or have been, present. IMRT is the process or technology used to deliver the beams to targeted areas within the body.

Thoracoscopy

Surgery is performed using instruments that are inserted into the pleural space via rib spaces under video assistance.

Thoracotomy

The chest cavity is entered via an incision on the back, and ribs are stretched open to enable manual access to the chest organs.

Trimodality Therapy

Trimodality therapy comprises three forms of treatment. It begins with chemotherapy, followed by radical surgery, and then radical radiotherapy.

Useful Websites & Resource Material

Sydney Cardiothoracic Surgeons

www.scts.com.au
Telephone (02) 9550 1933

Patient information booklets:

- Preparing for Lung Surgery.
- Living Life after Pneumonectomy.
- Pneumonectomy: What is it like to live with one lung?

The Baird Institute (TBI)

www.bairdinstitute.org.au
Telephone (02) 9550 2350

The Baird Institute is the research arm of the Sydney Cardiothoracic Surgeons Service and financially sponsors the support group for EPP patients. Any donations go directly to the work of TBI.

Asbestos Diseases Research Institute

www.adri.org.au
Phone (02) 9767 9800 or
email info@adri.org.au

ADRI is the world's first stand alone research facility dedicated to asbestos related diseases. It is located in the Bernie Banton Centre at Concord Hospital. A biobank or tissue bank is pivotal to the research being done at ADRI – you may be invited to give some tissue at the time of surgery. Coordinated the

Guidelines as follows:

Organising Committee. Guidelines for the Diagnosis and Treatment of Malignant Pleural Mesothelioma. Asbestos Diseases Research Institute: 2013.

Dust Diseases Board of NSW

www.ddb.nsw.gov.au
Phone 1800 550 02

The DDB statutory function is to administer the Workers Compensation (Dust Diseases) Act 1942-67. The liaison staff at the Board will answer questions and provide direction about compensation issues that arise from work place exposure to compensation. Do call them and ask questions.

Australian Mesothelioma Registry

www.mesothelioma-australia.com
Phone 1800 378 861

Check out the website for regular newsletters.

Sydney Cancer Centre / Support services www.sydneycancer.com.au

An established Psycho-oncology service provides support and counseling for patients, carers, and families coping with social, emotional and physical consequences of having a cancer diagnosis. You may contact the case manager who can make a referral to the service for you or a self referral will be accepted on (02) 9515 6677.

Asbestos Diseases Foundation of Australia (adfa) www.adfa.org.au

Telephone (02) 9637 8759

Free call 1800 006 196

This organisation provides helpful information about safe work practice of handling asbestos, and advice on who to contact should a person of the public suspect the presence of an asbestos product.

Asbestos Awareness www.asbestosawareness.com.au

If you have friends or colleagues or know of anybody thinking about renovating- please ask them to visit this website before starting.

Legal representatives experienced in asbestos related claims

Turner Freeman www.turnerfreeman.com.au Telephone (02) 8833 2500

Slater & Gordon www.slatergordon.com.au Telephone (02) 8267 0607

Maurcie Blackburn Cashman www.mbc.aus.net Telephone (02) 9261 1488

Reading

Organising Committee. Guidelines for the Diagnosis and Treatment of Malignant Pleural Mesothelioma. Asbestos Diseases Research Institute:2013

Pass H, Roy L and Vento S. (2005) 100 Questions and answers about Mesothelioma. Jones and Bartlett Publishers: Sudbury Massachusetts.

CD

Mesothelioma: Understanding, Managing, Living

Lung Foundation Australia www.lungfoundation.com.au

Telephone 1800 654 301

Cancer Council Victoria www.cancer.org.au

Telephone 131 120

SPONSORS

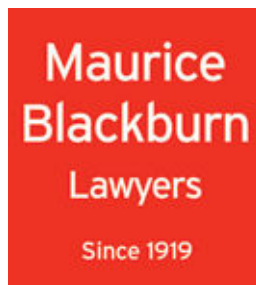
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**Sydney Cardiothoracic Surgeons
Royal Prince Alfred Hospital
Sydney Cancer Centre
Strathfield Private Hospital
The Baird Institute**