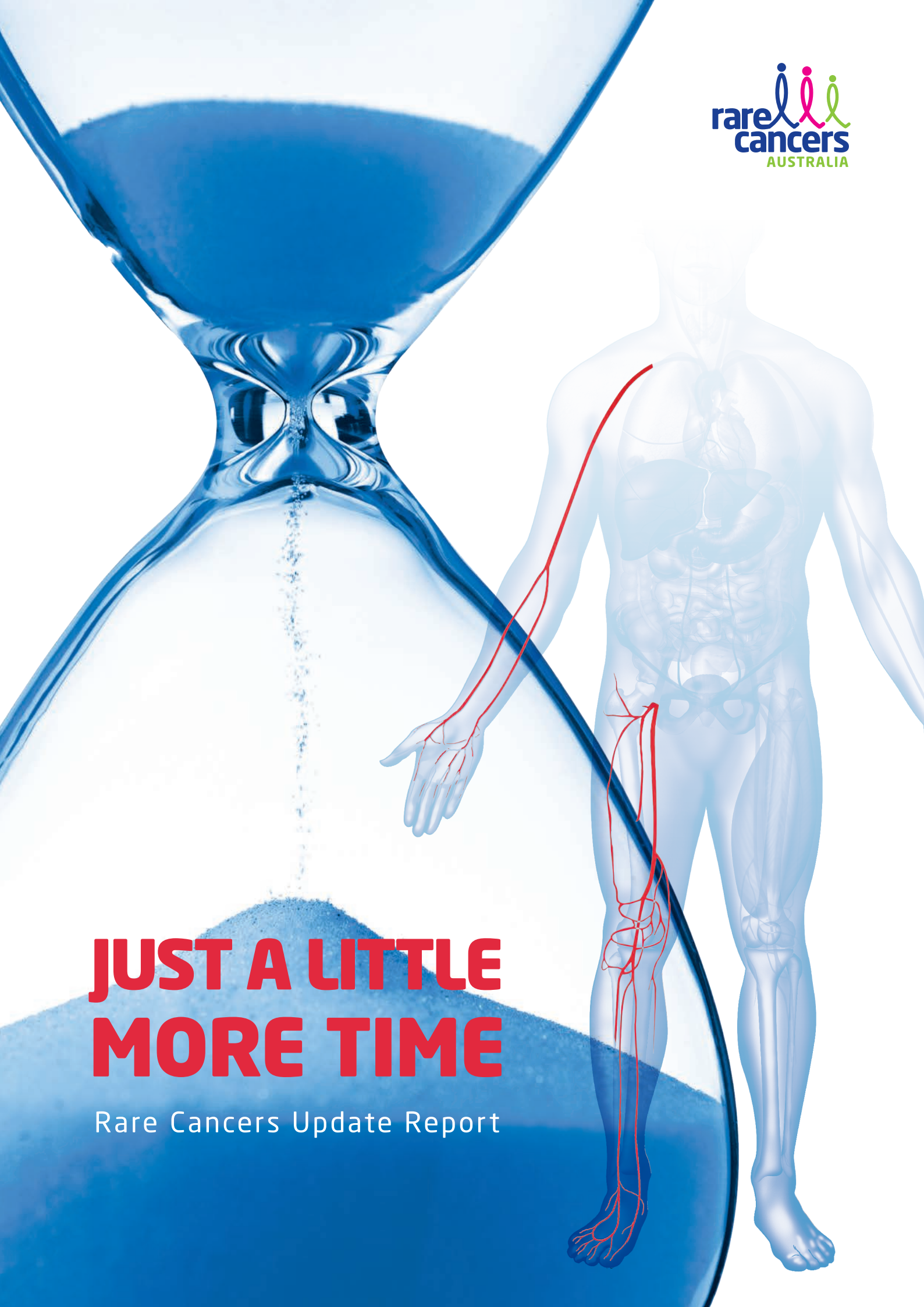
rare cancers AUSTRALIA



JUST A LITTLE MORE TIME

Rare Cancers Update Report

Acknowledgements

This report has been prepared by Rare Cancers Australia Ltd (RCA) to provide an update to the 2014 Just a Little More Time – Rare Cancers Baseline Report. RCA gratefully acknowledges Plum Stone's work in the preparation of the report and would also like to acknowledge the critical research contributions made by Nash Chance and Eliza Mitchell. This updated report provides an evidence-based understanding of the current state of research, diagnosis and treatment of super rare, rare and less common and common cancers in Australia.

The document draws on data publicly available from a number of organisations including the Australian Institute of Health and Welfare (AIHW), Cancer Australia and the Pharmaceutical Benefits Scheme.

All support is gratefully received and has been given without pre-condition or editorial input.

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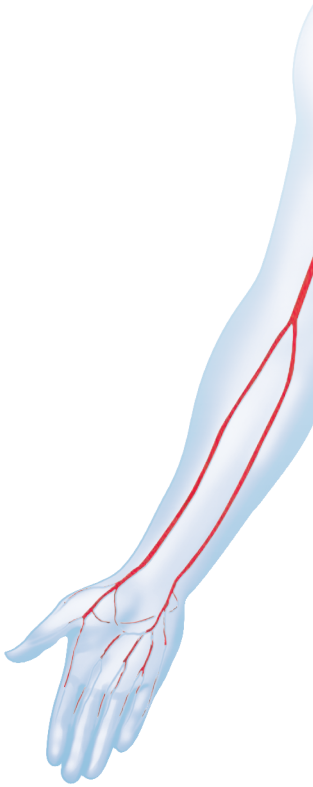
Our thanks also to Charlene Vien from RCA for her inspired and tireless work in designing and laying out this report. We are very grateful and appreciative.

Finally our thanks to Belynda Simpson from Bel-Art for her graphic design and report preparation. We hope this report, like its predecessor provides a reference point from which we, as a society, can clearly identify, understand and address the issues facing rare cancer patients in Australia today.

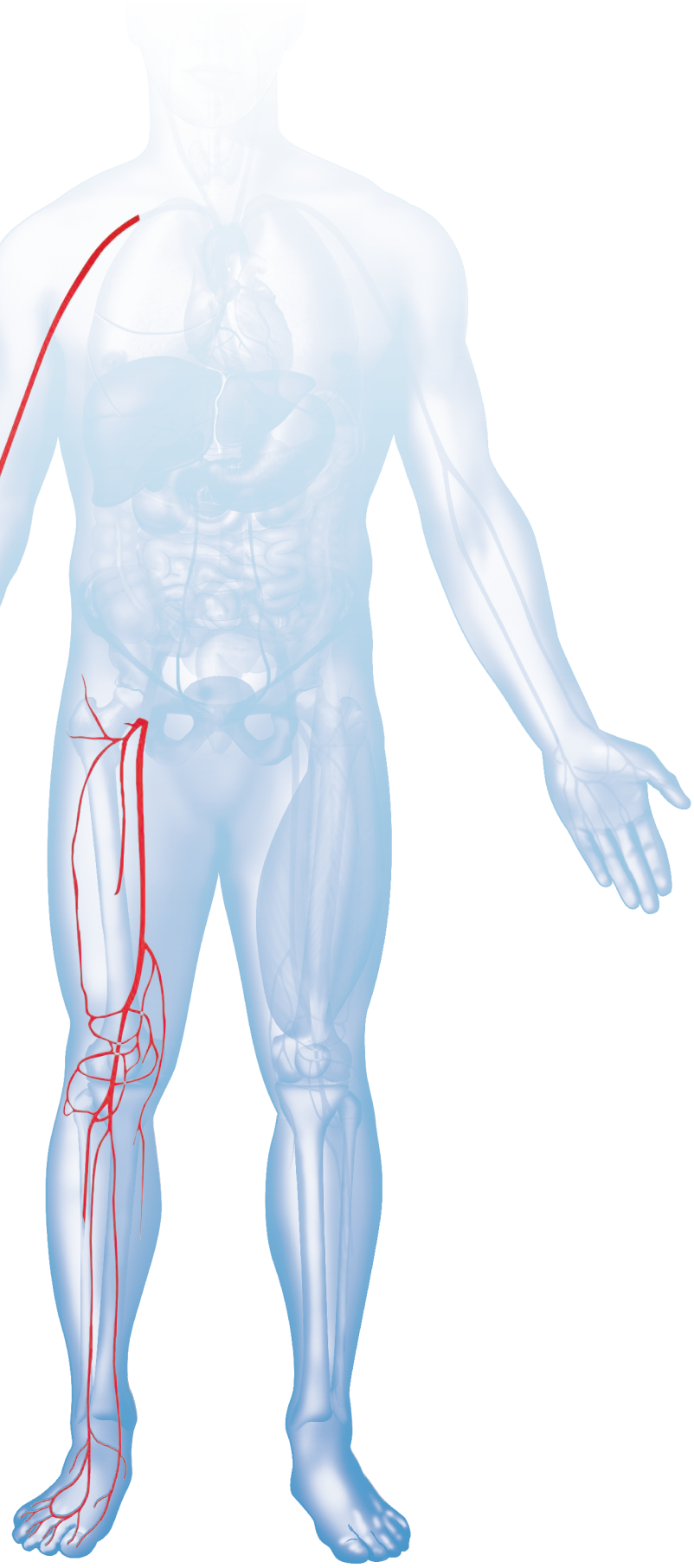


About Rare Cancers Australia

RCA is an Australian Charity established with the specific goal of **Disrupting Cancer.** Cancer has, since time immemorial, been a scourge of the human condition and none more so than those variations of cancer that are rare or less common. The patient experience for those diagnosed with one of these cancers can be an extremely distressing and isolating experience.

RCA is committed to:

- Building comprehensive support and information programs for patients;
- Providing financial support to patients to assist in dealing with the treatment and consequences of their disease;
- Ensuring our health system is reformed to provide equality of care and treatment to patients regardless of disease rarity;
- Working collaboratively with researchers to ensure rare cancer research is adequately prioritised and funded; and
- Using 21st Technology to improve the patient interface with the health system.



Patient Stories

Foreword

The title of this second Rare Cancers Australia's report, *Just a Little More Time,* signifies one of the most important themes for Australian cancer patients. Not only is it about the time that patients have, and how critical that time is to each and every patient; it is also about the pace of change in research and treatment, and the adaptation to that change of the health system as it tries to bring those timelines together in a way that meets patients' needs.

Recent advances in medical research have led to a shift in understanding the molecular biology of cancers and created new therapies to treat accordingly. As a result, for the first time we have a real opportunity to improve the outcomes for rare cancer patients across Australia.

On the one hand, molecular mapping of cancers has shown that even common cancers are actually composed of multiple subtypes, each of which would meet the definition of a rare and less common (RLC) cancer. On the other hand, many RLC cancers share the same molecular nature, which makes them more common. More importantly, many treatments developed in common cancers have been shown to work in RLC varieties.

So it is not as if the solutions don't exist—we just need to work out how to apply these solutions where they are most needed for RLC cancers; there are two key barriers here.

The first is how to increase the participation of patients with RLC cancers in medical research. There is excellent evidence to show that participation in clinical trials is associated with better outcomes. The future of health care depends on breakthrough discoveries and treatments that come from research. Moreover, governments use the information from trials to decide to fund new drugs. It is critical that patients with RLC cancers have access to clinical trials, and that government, academics, clinicians and the pharmaceutical industry work together to develop trials for RLC, as well as the more common cancers.

The second is how to ensure rare cancer patients have access to the best available medicines; those that offer the best quality of life and hope for more time. Too many patients, including many of my own sarcoma patients, are being left to fund expensive medical treatments for themselves simply because statistically they have been deemed insignificant.

As this report shows, our efforts to date have left behind the Australians with RLC cancers and we don't need to. Because of the way in which we can now genetically classify cancers using emerging technology, we no longer have to think about cancer treatment anatomically, and can ultimately now treat patients according to their genetics.

So the challenge now is not to science but to our health systems and our ability to translate our knowledge. With accelerating knowledge and exploration of all the potential opportunities, of genomics in particular, in what is essentially a genetic disease, we can expect over the next 20 to 30 years to see the same improvements in rare cancers as we've seen in common cancers, in the past 30 years.

While from a social equity point of view the challenge is to take rare cancers and provide patients with access to science and reasoned hope; through partnership, through collaboration, and through working together to arrest this problem. Our only limitations are our will and imagination.

Professor David Thomas
Director and Division Head, Genomic Cancer Medicine
Garvan Institute of Medical Research

Executive Summary

In 2014 Rare Cancers Australia (RCA) published the first of the *Just a Little More Time* series with a view to 'kick-starting research, policy discussion and action to improve the lives and outcomes of Australians living with rare and less common (RLC) cancers'.

Over the past two years we have seen an increasing awareness in government, the media and the general public of the issues facing Australian rare cancer patients and yet the day-to-day lives of these patients has not yet improved measurably.

Since the first report was launched two years ago, RCA has set up '**Sick or Treat**', a crowd-funding initiative, to support patients with rare cancers to pay for treatments, that are not listed on the PBS, for their cancer. That RCA needed to establish Sick or Treat is itself indicative of the current state of access to cancer medicines for Australian rare cancer patients. RCA hopes that one day, in the not so distant future, rare cancer patients in Australia will not have a need for initiatives such as Sick or Treat to help them fund the life saving treatments they urgently need.

As our research has shown this plague of cancer that we face will not restrict itself to the aged and infirm. In every age group (Baby Boomers, Gen X, Gen Y, and our children) rare and less common cancers are either our biggest killer, or close to it.

- RLC cancers are the major cause of death in children under 15 years of age in Australia, accounting for 79 per annum, or one child death every 4 days.
- RLC cancers kill 1 young 'Gen Y' Australian every day.
- RLC cancers are the most common cause of death in 40-59 year olds causing more than double the impact of coronary heart disease alone.
- RLC cancers are also the most common cause of death in 60-69 year olds in Australia, more than the impact of coronary heart disease and lung cancer combined.

In this age of medical research, the greater understanding of the molecular biology of cancers and the advent of immunotherapies for treating cancer, we have an enormous opportunity to improve the outcomes of rare cancer patients across all ages. We can, and we must, do better.

The simple cost of doing nothing to improve outcomes for RLC cancers is too high and the challenge therefore, is to find a mechanism whereby:

- Research funding is increased and specifically directed to encourage and drive research into rare cancers and rare molecular sub-types; and
- Rare cancer patients can receive equitable and fair access to medicines that have reasonable, proven safety and efficacy for those diseases.

Rare cancers represent a major diagnostic as well as therapeutic challenge and they represent a major source of discrimination among patients[1]. It is time we took the action necessary so that we can give these Australian patients the resources, support, treatment they need and most importantly provide them all with **"just a little more time"**.

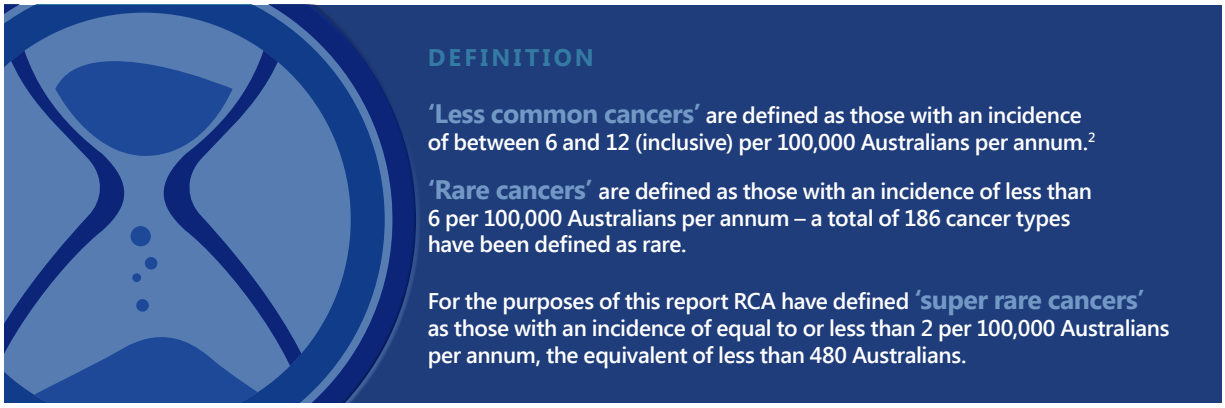
1 Dei Tos AP, Classifying rare cancers. What is a rare cancer? Asia-Pac J Clin Oncol 2015; 11 (Suppl. 4): 63-103

Introduction

In 2014 Rare Cancers Australia (RCA) published the first of the *Just a Little More Time* series with a view to 'kick-starting research, policy discussion and action to improve the lives and outcomes of Australians living with rare and less common (RLC) cancers'.

Over the past two years we have seen an increasing awareness in government, the media and the general public of the issues facing Australian rare cancer patients and yet the day-to-day lives of these patients has not yet improved measurably.

In this, the 2016, report we have again reviewed the available cancer data and investigated the disparities that exist for incidence, mortality and survival across the cancer spectrum. We have also looked at a new definition for super rare cancers, and the burden of disease that RLC cancers pose across all ages in Australia.



** A full list of those cancers that can be classified as super rare and rare or less common can be found in Appendix 1.*

The first report demonstrated that over the past 20 years, survival rates in many RLC cancers have only improved marginally, if at all; and our research funding into rare cancers remains disappointingly and disproportionately low, as does the money we spend on treatments for these patients through the Pharmaceutical Benefits Scheme (PBS).[3]

So what has changed in the past two years for these patients and how do patients with rare and super rare cancers fare comparatively to patients with more common cancers?

What's Changed?

The 2014-15 Senate Inquiry into the availability of new, innovative and specialist cancer drugs in Australia raised some of the critical issues for Australian cancer patients at a Federal Government level and made a number of recommendations for changes to the way that Australians access cancer medicines in Australia.

While the Senate Committee noted that investment in cancer detection and screening, and investments in medical research have led to dramatic advances in the way cancer is treated and will be treated in the future, the Committee also raised concerns that Australian patients continue to face significant delays and expenses in accessing new cancer drugs.[4]

2 Gatta et al., Rare Cancers are not so rare: The rare cancer burden in Europe. European Journal of Cancer 47, 2493-2511 (2011).
3 Rare Cancers Australia 2014, Just a Little More Time: Rare Cancers Baseline Report
4 Community Affairs References Committee, Availability of new, innovative and specialist cancer drugs in Australia, September 2015. Accessed 5th January 2016 http://www.aph.gov.au/Parliamentary_Business/Committees/Senate/Community_Affairs/Cancer_Drugs/Report

The Committee recommendations included asking the Government:

1. To initiate a comprehensive review of the system for the registration and subsidisation of medicines;
2. To commission a review of current data collection mechanisms for cancer medicines; and
3. To establish a Steering Committee to examine the feasibility of establishing a national register of cancer medicines.

Please note that at the time of this report going to print the Government's response to the Senate Inquiry has not been released, however we greatly look forward to its recommendations, which we hope will improve the lives of Australian cancer patients.

In the past two years, we have also seen Government initiated reviews into the Therapeutic Goods Administration, the Pharmaceutical Benefits Advisory Committee Guidelines, the Life Saving Drugs Program and others, and we also eagerly await the positive impacts those reviews will have on Australian patients.

At the same time we have also seen the emergence of both targeted therapies and immuno-oncology treatments being both registered and funded through the PBS for common cancers. Where targeted approaches aim to inhibit molecular pathways that are crucial for tumour growth and maintenance; immunotherapies endeavour to stimulate a host immune response that delivers long-lived tumour destruction.[5] These medicines, which work on differing cancer mechanisms, offer both opportunities for patients and challenges for our regulatory system.

These new medicines clearly have application across a range of cancers, many of which are sufficiently rare that there is neither the patient population nor the commercial opportunity for extensive traditional clinical trials to be conducted or funded. This has led to the emergence of 'basket trials' where a trial is focused on treating patients with a shared rare mutation with a drug that may respond to the mutated pathway, rather than focusing on a tumour's anatomical location.

This type of trial represents an important step forward in how we conduct trials for small patient populations and urgent work is required to ensure our regulatory mechanisms can cope and accept this new type of trial data.

Rare cancers represent a major diagnostic as well as therapeutic challenge and they represent a major source of discrimination among patients.[6]

5 Vannerman, M. and Dranoff, G. 2012. Combining immunotherapy and targeted therapies in cancer treatment. Nature Reviews Cancer 12, 237-251 (April 2012).

6 Dei Tos AP, Classifying rare cancers. What is a rare cancer? Asia-Pac J Clin Oncol 2015; 11 (Suppl. 4): 63-103

Sick or Treat

Although six medicines have been successfully listed on the PBS for rare cancers (not including rare blood cancers) and made available to Australian patients since 2010, the number of rare cancer patients having to self fund their own treatments, or go without, has continued to increase. In response to this need in October 2014 RCA launched '**Sick or Treat**', a crowd-funding initiative, to support patients with rare cancers to pay for treatments that are not listed on the PBS for their cancer.

Since October 2014 Sick or Treat has helped more than 30 patients; it's their stories you will read throughout this report. Some patients have been lucky and no longer require Sick or Treat to help with funding because they have had their treatments approved by the Pharmaceutical Benefits Advisory Committee (PBAC), and subsequently listed on the PBS. As a result, treatments for these patients are therefore now available for a maximum of $37.70 per month.

The majority of rare cancer patients, however, have not been so lucky. They continue to have to find many thousands of dollars per month to pay for treatments that their clinicians have prescribed, based on best available medical evidence, and which may be working but which will **never** be listed on the PBS for their rare or super rare cancer.

In the first three days of 2016 RCA received three patient requests to join Sick or Treat, a worrying way to start 2016, and still the numbers continue to rise. In total Sick or Treat has (at time of publishing) raised over $1.2 million to help 30 Australians suffering from a rare or less common cancer; Australians who may otherwise not have had *'just a little more time'*.

That RCA needed to establish Sick or Treat is itself indicative of the current state of access to cancer medicines for Australian rare cancer patients. RCA hopes that one day, in the not so distant future, rare cancer patients in Australia will not have a need for initiatives such as Sick or Treat to help them fund the life saving treatments they urgently need.

In order to achieve this goal RCA calls on all stakeholders to work together to provide an affirmative action plan for rare cancer patients in Australia. Without concerted action for 'rare' in research, diagnostics and treatment, Australia could be confronting over 30,000 deaths from RLC Cancers by 2020 and over 40,000 by 2030.

As our research has shown this plague of cancer that we face will not restrict itself to the aged and infirm. In every age group (Baby Boomers, Gen X, Gen Y, and our children) rare and less common cancers are either our biggest killer, or close to it. **We can, and we must, do better.**

12 year old

Angus

Relapsed Hodgkin's Lymphoma

A cancer diagnosis in your child is every parent's worst nightmare. It became our nightmare. In Autumn 2013, at 10 years of age, Angus pointed out a large lump in his neck. We were immediately concerned so saw our GP straight away, we were reassured that it was nothing. After 6 weeks, the lumps were still there so we returned to the GP. An appointment was then scheduled at the Children's Hospital for removal of a node. Our post-surgery follow up was a surreal experience. We arrived at the clinic with our beautiful, seemingly healthy son, blonde hair, tanned skin and a big smile. The surgeon smiled at us in the waiting room while she saw to other tasks before calling us in. We assumed that everything must be okay.

"A cancer diagnosis in your child is every parent's worst nightmare."

"We have the pathology from the lymph node we removed. Angus has Hodgkin's Lymphoma." Lymphoma. Is that cancer? I looked at my husband. He was visibly shaken. I looked at Angus. For the first time, Angus was completely unaware of what was unfolding around him. The surgeon then said, "I have arranged for the oncologist to join us."

"It's treatable", "It's curable", and it was to some degree; two months of awful, painful, sickening chemotherapy and Angus was given the all clear. By Christmas his hair was coming back and in January we took our regular camping holiday down the coast. We were lucky and extremely grateful. I had once said to our oncologist that this cancer was a good one to get, to which he replied, "There is no such thing as a good cancer".

We went back to our normal, crazy life, there is no other type with 4 active boys. Angus topped off 2014 by being elected school captain. Nothing could stop him from pursuing his goals.

However, things didn't go as we had hoped. More lumps appeared in Angus' neck in late January. I tried to reassure him and myself that it was just from the cold he recently had, but the panicked look on the faces of our oncology team told a different story. Pathology on a lymph node confirmed that Angus was one of the very few in whom Hodgkin's Lymphoma recurs.

2015 has been a tricky year. His assorted treatments have included high dose combination chemotherapy, radiotherapy and a stem cell transplant resulting in 30 days in hospital isolation, not easy for a social 11-year-old. He says he likes radiotherapy, heaps easier than chemo according to Angus.

Then the post treatment scan. No active cancer. We are so incredibly grateful, but, we are not done. Angus' cancer, after one relapse, has a recurrence rate in the order of 40-50%. So our journey and battle with cancer continues.

CANCER STATISTICS

The most common statistics for measuring cancer are incidence, mortality and survival, which may be defined as follows:

Incidence refers to the number of people who will get a particular type of cancer each year, and is often expressed as the number of cancer cases per 100,000 in a given population;

Mortality statistics refer to the number of people who have died from a particular cancer in a given year. On their own, mortality statistics provide little information with respect to the cancer and its progression, however, when presented as a ratio of incidence it provides a reliable proxy for survival rates when actual survival statistics are not available; and

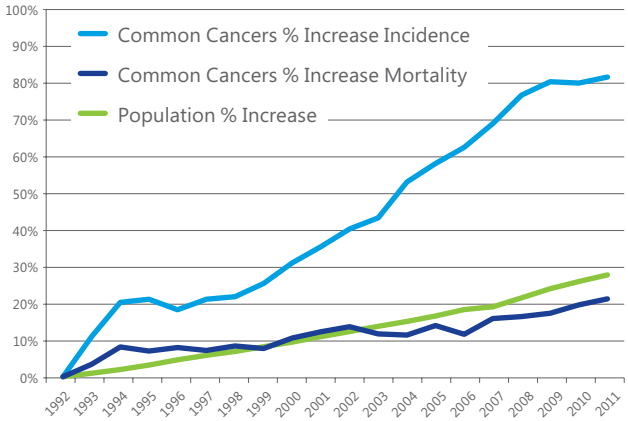
Relative survival is an average measure of the probability of being alive relative to the 'average' Australian of the same sex and age, at a specified interval after a diagnosis of cancer (usually five years).[7]

Cancer is one of the major causes of illness in Australia and has a significant social and economic impact on individuals, families and the community.[8]

In 2014 an Australian Institute of Health and Welfare (AIHW) report[9] estimated the following for **all cancers**:

- **130,470** people would be diagnosed with cancer in 2016;
- **47,380** would die from cancer in 2016;
- The age-standardised incidence rate was **467 per 100,000**; and
- The WHO estimated that cancer would contribute between **16-19 per cent** of the burden of disease in Australia.[10]

Common Cancers



Graph 1: *Incidence and mortality rates for common cancers compared to population change since 1992*

The successes we've seen over the past 20 years for common cancer patients are significant. While incidence rates have increased, as a result of increased surveillance and screening, mortality rates have decreased due to improvements in early diagnosis and treatment. As a result, patients today diagnosed with a common cancer have a much higher chance of survival than they did in the early 1990s.

7 Australian Institute of Health and Welfare. Interpreting Cancer Data, accessed on 12th January 2016 http://www.aihw.gov.au/cancer/data/interpreting
8 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.
9 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.
10 WHO 2014. Global health estimates for 2000-2012: disease burden. Geneva: WHO.

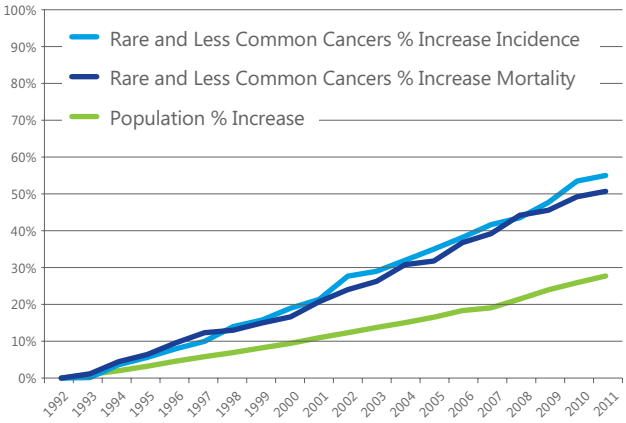
For example, improvements in the outcomes for bowel cancer patients, with survival rates increasing from 46.1% in 1982, to 66.9% in 2007-11, are in large part due to the improved screening that has led to a 300 per cent increase in diagnosis. Improvements in early diagnosis, coupled with the availability of innovative treatments have meant that for many common cancers we have had a significant impact on reducing mortality rates and increasing survival.

Despite the actual number of deaths for all cancers increasing, the mortality rate for **all cancers** fell by 20 per cent between 1982 and 2014.[11] Data in this report, however, demonstrates that while it is true that significant advances have been made for common cancers it is not the case for rare and less common cancers or super rare cancers.

Rare and Less Common Cancers

As a comparative example, it is estimated that in 2014:

- **42,000 people** were diagnosed with an RLC cancer;
- **24,000 patients died** from an RLC cancer; and
- RLC cancers contributed to seven per cent of the total burden of disease in Australia.[12]

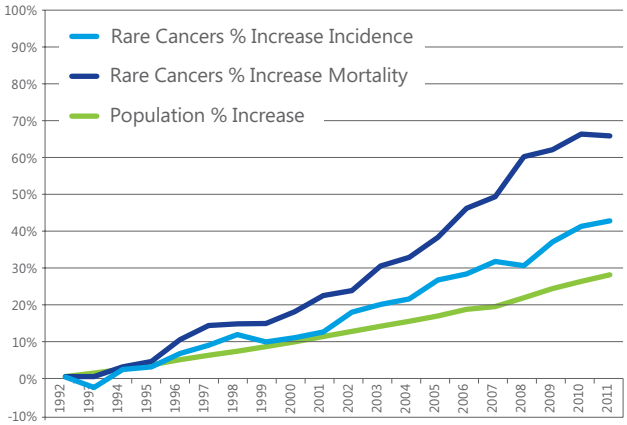


Graph 2: *Incidence and mortality rates for rare and less common cancers compared to population change since 1992*

As distinct from common cancers the percentage increase in incidence and mortality for RLC cancers occur at roughly the same rate, i.e. twice the rate of population increase. While we have seen increases in incidence for common cancers, we have also seen dramatic reductions in mortality due to early diagnosis and improved treatments, but this has not been the case for RLC cancers where diagnosis remains slow, and treatment availability limited.

The same effect is even more devastating in rare cancer diagnoses.

Rare Cancers



Graph 3: *Incidence and mortality rates for rare and less common cancers compared to population change since 1992*

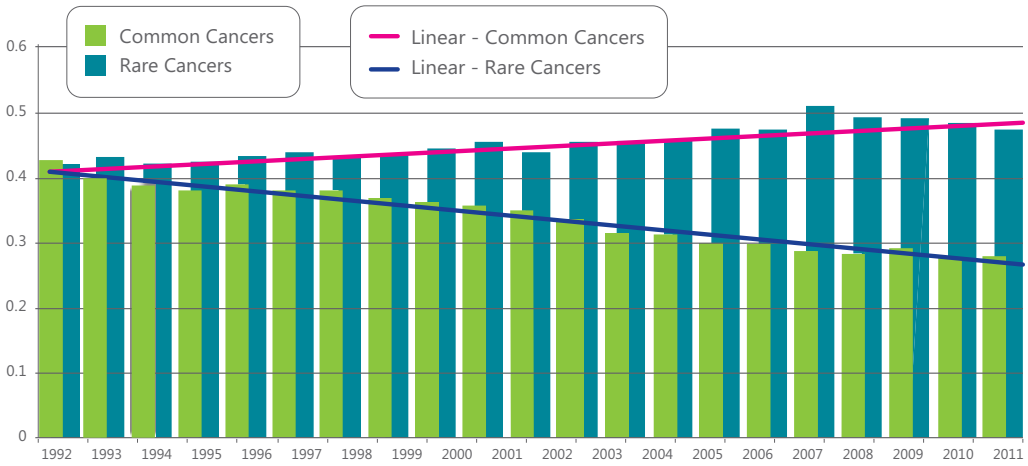
Australian patients diagnosed with a rare cancer face the greatest challenge of all. In the past 20 years we have made significant advances in reducing the mortality rates for common cancers compared to incidence; and we have even seen incidence and mortality rates increase at roughly the same rate for RLC cancers. **But for rare cancer patients the increase in mortality rates far outstrip the rising incidence rates.**

We need to recognise that a rare cancer diagnosis is often accompanied by a very poor prognosis and as our population ages we will continue to see the worsening impact of these very neglected and under treated cancers.

11 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.
12 Rare Cancers Australia, Just a Little More Time: Rare Cancers Baseline Report 2013

Incidence to Mortality Ratio Comparison

Indeed, looking at the incidence to mortality ratio of common cancers compared to rare cancers, the first has decreased since 1992, i.e. there are fewer deaths per common cancer diagnosis, while it has increased for rare patients, i.e. there are more deaths per rare cancer diagnosis.



Graph 4: *Incidence to mortality ratios for common cancers versus those of rare cancers.*

Unlike common cancers, we have not improved screening and other diagnostics for rare cancers in the past 25 years and hence our clinical ability to recognise new patients when they present with these cancers remains limited.

As a result rare cancer patients are commonly misdiagnosed, and patients may face long delays before the correct diagnosis is achieved and the appropriate treatment prescribed; anecdotally we are aware of a number of patients who have been misdiagnosed for more than a year, as they have contacted RCA directly for help. As long as patients with rare cancer are misdiagnosed, and treatment is delayed, the incidence to mortality ratios for these patients will continue to increase markedly.

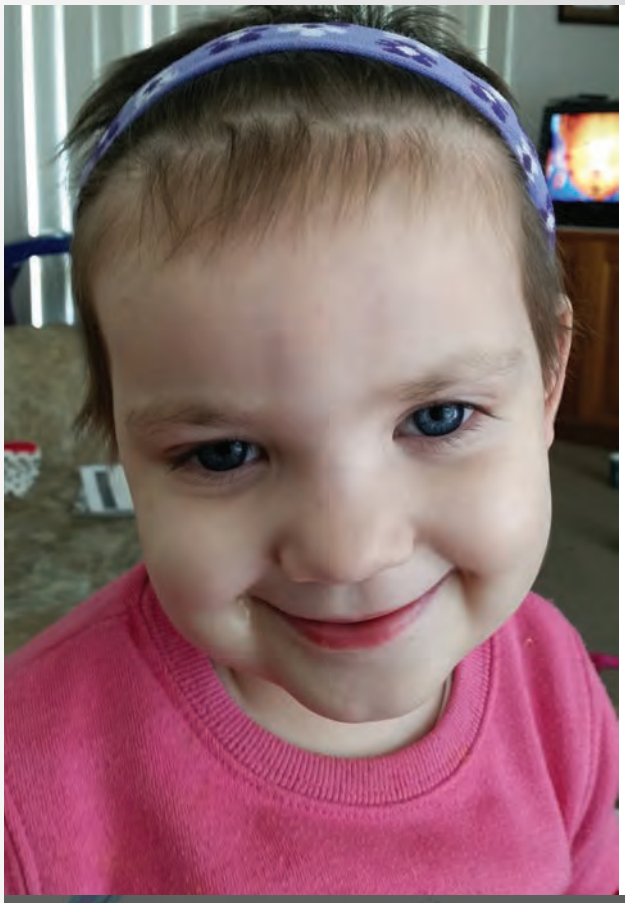
The difference between rare cancers and rare genetic subtypes

Advances in medical research have improved our understanding of the individuality of cancer. Where once a cancer was defined by its anatomical location, or cellular behaviour, cancers may now be categorised according to their molecular pathology.

This means that while there remain a number of 'discreet' rare cancers such as cancer of the mouth, oesophagus, larynx, and mesothelioma, we are now increasingly able to define more common cancers by their genetic abnormalities through molecular diagnosis, and discover rare subtypes such as HER2-positive stomach cancer, ALK+ non-Hodgkins lymphoma and ROS1 non-small cell lung cancer.

The molecular-level understanding of these cancers now enables researchers, and pharmaceutical companies, to target specific medicines to these genetic abnormalities and thereby increase the benefit of treatment to individual patients.

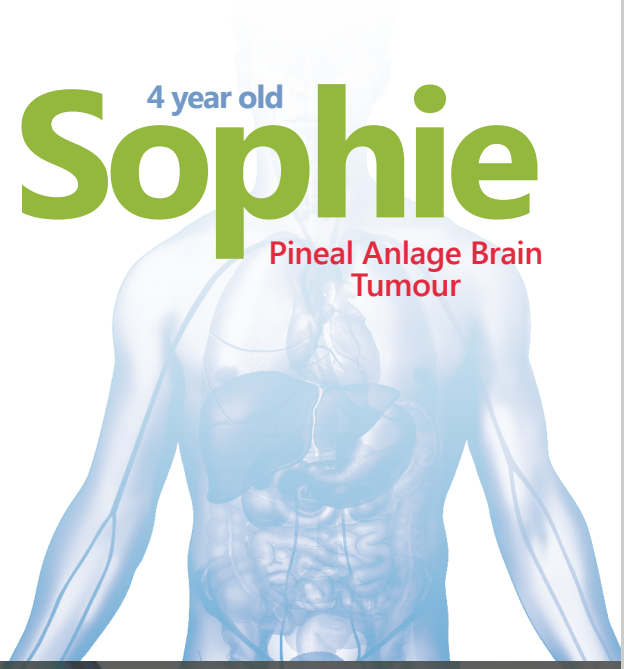
With every advance in our understanding of the individuality of cancer we see a growing number of cancers that we know to be rare or have rare definable subgroups.



4 year old

Sophie

Pineal Anlage Brain Tumour



Sophie was two and half years old when we took her to various doctors and specialists as she had been unwell on and off for weeks. We knew something wasn't right but they just kept saying that it was a virus. One morning, whilst eating breakfast, Sophie suffered a massive fit. That's when our world come tumbling down.

"They didn't know what they were dealing with as it was very rare."

She was taken to John Hunter Hospital for emergency surgery to release the pressure in her brain. Doctors there told us she had something in her brain that looked like a tumour. They tried to get it out but failed. She was transferred to Westmead Hospital and her tumour was successfully removed.

We were initially told that it was non-cancerous, only to find out four days later that it was and that it was bad. We were told that our daughter may not survive. Our Sophie went through ten months of intense chemo. She was stronger than the adults. They didn't know what they were dealing with, as it was very rare. She got really sick with the chemo and we nearly lost her. She was on nasal feeds and a few months after, lost all her strength and had to learn to sit, stand walk, play and talk again. This was extremely hard for us and for her.

She is still starting over with speech and is very weak in her muscles but manages to walk a little on her own. Sophie will need a lot of special treatment with her feet, she will never be able to look up anymore and she can no longer focus properly. In the last few months she has also had two emergency surgeries as her shunt has been blocked. Our children are deeply affected by this. We keep battling on. It's tough on my husband as he has health issues including chronic fatigue, chronic depression, bi-polar and back problems. He gets very upset that he can't provide for us but I tell him that we're just happy to have him. We always say there are others worse off and that helps to get us through the days.

Sophie is our miracle child.

Burden of Disease

Cancer has a major impact on the Australian community and is the leading contributor to the burden of disease. It is estimated that in 2012, cancer represented 19% of the burden of all diseases in Australia. By comparison, cardiovascular disease contributed to 16% of the burden of disease, whilst nervous system and sense organ disorders accounted for 14% of the burden of disease and mental disorders accounted for 13% of the burden of disease. However, in terms of health care expenditure, in 2008–09, cancer and other neoplasms accounted for $5 billion or 7% of total recurrent health spending.[13]

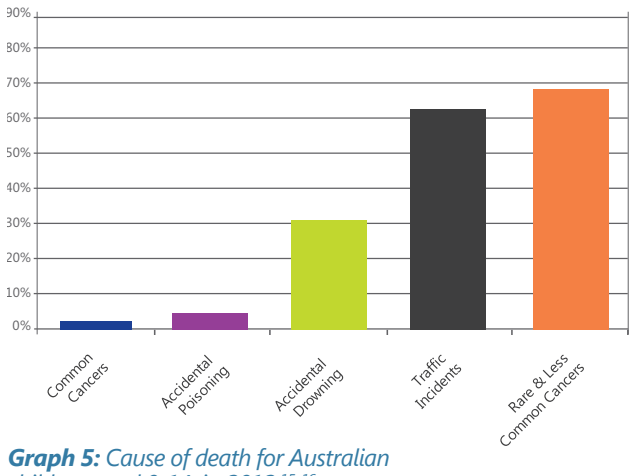
Age Specifics of Rare and Less Common Cancers

Cancer is usually considered a disease of older people, with mortality rates increasing with age for most cancers. Indeed, the AIHW estimates that in 2015 the risk of being diagnosed with cancer by the age of 85 was one in two for men, and one in three for women.[14]

One of the key characteristics of RLC cancers is that they do not tend to follow this pattern; while RLC cancers also affect older patients they disproportionately place a burden on our children and young families.

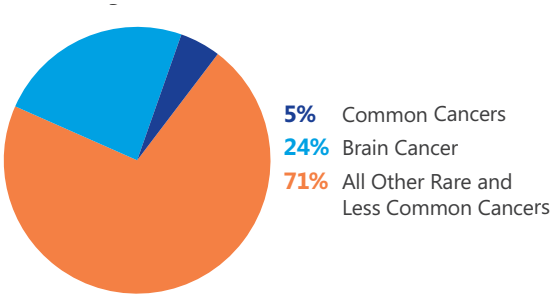
**Please note that in the interest of remaining true to the available data sets we have used the terms 'Gen Y', 'Gen X' and 'Baby Boomer' in this chapter to represent the corresponding age categories – this has provided an approximation of four-five years in most age groups.*

Childhood Cancers



***Graph 5:** Cause of death for Australian children aged 0-14, in 2012* [15, 16]

RLC cancers are the major cause of death in children under 15 years of age in Australia, accounting for 79 per annum, or one child death every 4 days. (**Please note that road death data includes children aged 1-16*).



***Graph 6:** Total cancer deaths of Australian children aged 0-14, in 2012*

95% of cancer deaths reported in children under 15 are caused by RLC cancers. **However, it is important to note that all cancers in this age group are considered rare, with less than 1% of all cancers in Australia occurring in this age group.**

While leukaemia are most commonly diagnosed cancer in children (see Blood and Lymphatic Cancer section), it is brain cancer that accounts for the highest mortality – causing 1 in 4 cancer deaths in this age group.

One of the major issues faced for childhood cancer patients is the lack of availability of approved treatments. As with rare cancers generally the barriers that arise due to the need for large randomised clinical trials and the very small and quite varied population groups present significant challenges. It is not however, acceptable to put the lives of these vulnerable children in the "too hard" basket.

13 Cancer Australia, 2015. Cancer Research in Australia: an overview of funding initiatives to support cancer research capacity in Australia 2006 to 2011.
14 Australian Institute of Health and Welfare. All cancers in Australia. http://www.aihw.gov.au/cancers/all-cancers/ Accessed January, 2016.
15 Department of Infrastructure and Transport, 2013. Road Deaths Australia Statistical Summary
16 Mindframe, Facts and Stats about Suicide in Australia, http://www.mindframe-media.info/for-media/reporting-suicide/facts-and-stats, accessed January, 2016

22 year old

Laurence

Osteosarcoma

At 19 years of age, my life drastically changed. I received the shocking news that the bad pain and swelling in my knee wasn't a ligament injury, but an extremely rare and highly aggressive form of bone cancer called Osteosarcoma. The cancer was not confined to my knee but had metastasized and spread to my lungs by the time of diagnosis. I was studying a physics degree at the time and working and living a healthy, active life which included sports and martial arts. The chance of developing my type of cancer is 3 in 1,000,000.

"The chance of developing my type of cancer is 3 in 1,000,000."

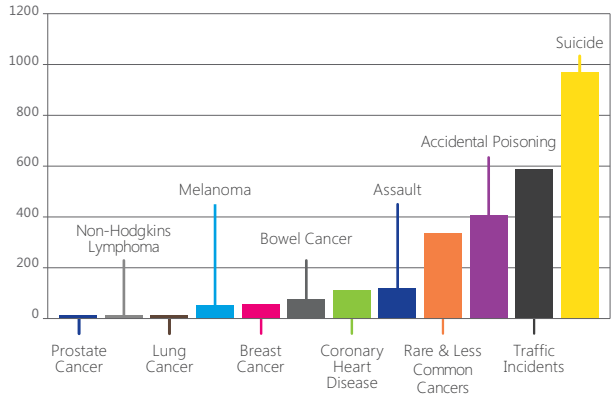
It has been just over three long years since my diagnosis and I have undergone 23 chemotherapy cycles with highly toxic drugs, 7 months of trial drugs, 4 major surgical procedures including removal of my entire cancerous knee and replacement with a metal mega-prosthesis. I have had surgery to remove metastases from my lungs, at least 12 chest drains for collapsed lungs and 18 radiotherapy treatments. I endure unbearable pain from a tumour that is now encroaching into my spinal cord and could potentially cause me to become paraplegic in the near future if something further is not done to stop its growth.

I have recently undergone further chemotherapy to try to shrink the tumours but this hasn't been as successful as hoped.

The next option for me is immunotherapy that would hopefully decrease the debilitating pain and breathlessness. As my cancer is so rare and under-researched, these medicines are not government funded by the PBS (Pharmaceutical Benefits Scheme). It is a considerable amount of money that is extremely difficult for me to pay as I am unable to work because of my condition. I wish that money did not have to be part of this.

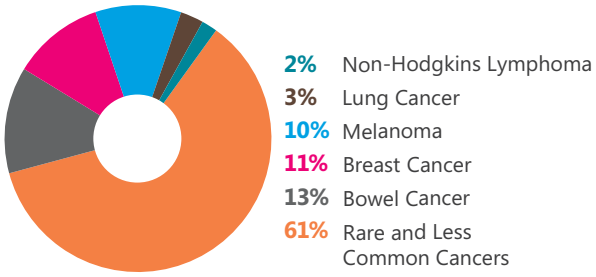
Laurence sadly lost his final battle on 27 February 2016.

Gen Y Cancers



***Graph 7:** Cause of death for Australians aged 20-39, in 2012* [17,18]

The most common causes of death in this young age group are suicide, traffic and poisoning accidents. RLC cancers are the fourth most common cause of death in 20-39 year olds in Australia, and the most common cause of disease related death.



***Graph 8:** Total cancer deaths of Australians aged 20-39, in 2012*

Among all cancer deaths in this young age group of 20-39 year olds, RLC cancers account for 61% of all cancer deaths.

Indeed RLC cancers kill 1 young Australian every day.

Matthew

27 year old

Osteosarcoma

"Tumours kept appearing in places that defied all scientific knowledge and record."

In 2014, Matt had just been promoted to an overseas position. After being there six months he experienced increasing back pain and was diagnosed with a spinal tumour. He was immediately flown back to Sydney and underwent exhaustive tests before having surgery (June 2014). He was diagnosed with Angio Matous Fibrous Histiocytoma Sarcoma. Less than 500 reported cases world wide. He was informed that surgery was all that was needed as there was a less than 1% chance of metastasizing.

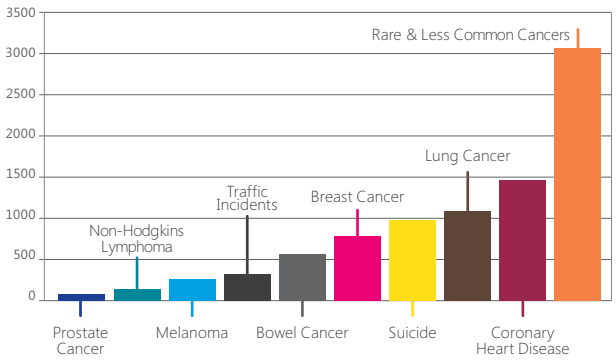
Unfortunately for Matt he was one of the unlucky ones. Two more surgeries and radiation followed, however, another tumour appeared. The next surgery saw him losing one major organ and 40% of another, rendering him extremely weak. He lost an enormous amount of weight. Matt had an extremely aggressive cancer. Chemotherapy failed and tumours kept appearing in places that defied all scientific knowledge and record. Tumour samples were sent to Boston, America for advice only to find out that they had never seen this before and could not offer any assistance.

Further invasive surgeries followed and the following scans revealed even more new sites of disease. In total, Matt endured 35 rounds of radiation, 9 surgeries and 13 rounds of chemotherapy. Frustrated by the system, his Mother took him to Germany in September for Biological targeted therapy and testing. Matt has had some encouraging results. This is the first time Matt has received positive news throughout his cancer journey.

As we now know, he is possibly one of only 2-3 people worldwide to be inflicted with his type of cancer, thus there is no set treatment protocol here in Australia. He is a generous and kind young man, a highly motivated individual whose life has been unbelievably altered in his prime, in a very short period of time.

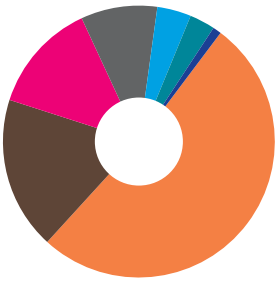
17 Department of Infrastructure and Transport, 2013. Road Deaths Australia Statistical Summary
18 Mindframe, Facts and Stats about Suicide in Australia, http://www.mindframe-media.info/for-media/reporting-suicide/facts-and-stats, accessed January, 2016

Gen X Cancers



***Graph 9:** Cause of death for Australians aged 40-59, in 2012* [19, 20]

RLC cancers are the most common cause of death in 40-59 year olds in Australia, with more than 3,000 deaths per annum; more than double the impact of coronary heart disease alone.



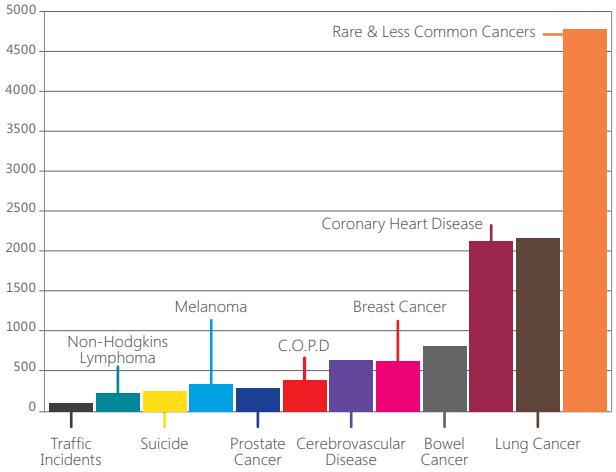
1% Prostate Cancer
3% Non-Hodgkins Lymphoma
4% Melanoma
9% Bowel Cancer
13% Breast Cancer
18% Lung Cancer
52% Rare and Less Common Cancers

***Graph 10:** Total cancer deaths of Australians aged 40-59, in 2012*

RLC cancers are once again the most common cause of cancer death in the 40-59 year old age group, accounting for 52% of all cancer deaths.

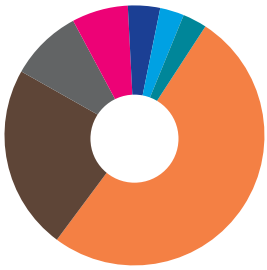
Other age-related cancers have increasing impact in this age group; with prostate cancer causing 1% of deaths and deaths by other cancers such as lung and melanoma both on the rise.

Baby Boomers



***Graph 11:** Cause of death for Australians aged 60-69, in 2012* [21, 22]

Finally, RLC cancers are also the most common cause of death in 60-69 year olds in Australia, with more than 4,600 deaths per annum; more than the impact of coronary heart disease and lung cancer combined.



3% Non-Hodgkins Lymphoma
3% Melanoma
4% Prostate Cancer
7% Breast Cancer
9% Bowel Cancer
23% Lung Cancers
51% Rare and Less Common Cancers

***Graph 12:** Total cancer deaths of Australians aged 60-69 in 2012*

RLC cancers remain the most common cause of cancer death in the 60-69 year old age group, accounting for 51% of deaths.

As the Australian population continues to age, the actual numbers of RLC cancer patients in this age group is set to continue to increase.

While common cancer patients may be offered some hope by our improving ability to diagnose and treat their cancers, RLC cancer patients remain destined for the same outcomes as the early 1990s. Examples of our impact on specific cancer examples are discussed in the following chapters.

19 Department of Infrastructure and Transport, 2013. Road Deaths Australia Statistical Summary
20 Mindframe, Facts and Stats about Suicide in Australia, http://www.mindframe-media.info/for-media/reporting-suicide/facts-and-stats, accessed January, 2016

21 Department of Infrastructure and Transport, 2013. Road Deaths Australia Statistical Summary
22 Mindframe, Facts and Stats about Suicide in Australia, http://www.mindframe-media.info/for-media/reporting-suicide/facts-and-stats, accessed January, 2016

66 year old

Gerry

Adenocarcinoma of the stomach

I have been married to Georgette for 43 years, we have 3 adult children and 8 beautiful grandkids. We migrated to Australia in 1978. I undertook many roles including Parish Ministry, chaplaincies in aged care, the S.A Police Department and an extensive chaplaincy at the Royal Adelaide Hospital. My final placement, which I hoped would lead to retirement in my late 60s was in Parish Ministry once again.

Probably the most challenging role was hospital chaplaincy, and it is with some irony looking back that I spent many hours with cancer and palliative care patients. Both of these I found an absolute privilege and so often felt that others gave me more than I was able to give them. Georgette was also an oncology nurse for some 10 years.

On 17 February 2015, I was diagnosed with an adeno-carcinoma of the stomach, which had encroached into the lower oesophagus and lymph nodes close to the stomach. The initial hope was that I could have 9 weeks of chemotherapy, followed by a complete gastrectomy, then further chemo. Unfortunately, the cancer was too aggressive and had invaded my peritoneum. Since then, I have undergone a gruelling 25 rounds of chemo. But the cancer continues to march on.

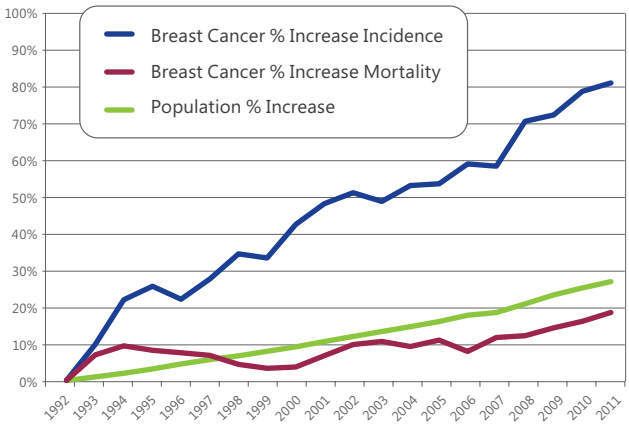
"...while our past experience may have educated us about some aspects of cancer, it does not equip us for the reality of the experience."

The disease has forced me into early retirement and Georgette has taken leave without pay to give me maximum care and support and to also spend time together. Like many others I guess, this whole event has caused us to focus on what is really important to us and we value more and more the love, care and compassion of our family and friends.

As committed Christians our faith continues to sustain us and we have learned that while our past experience may have educated us about some aspects of cancer, it does not equip us for the reality of the experience. Like many others, we seek to manage day to day and it is certainly not easy. One thing that has helped us enormously is to have a grateful heart and a thankful spirit for all that we have. The deep desire of our hearts is to be healthy and to be together for as long as possible.

Cancer Specific Examples

Breast, Cervical and Ovarian Cancers



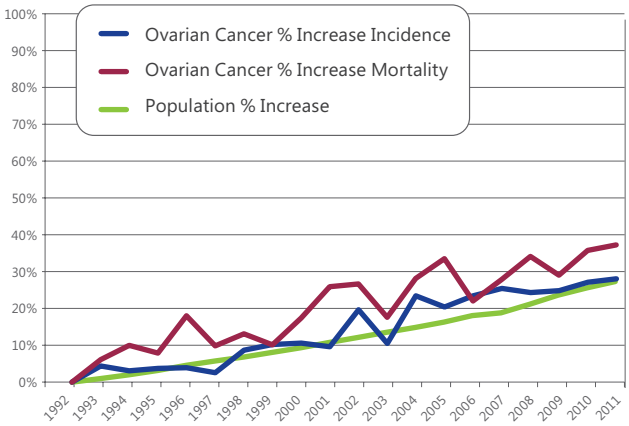
Graph 13: *Incidence and mortality rates for breast cancer compared to population change since 1992*

> *The AIHW estimates that in 2016 there will be 16,080 patients diagnosed with breast cancer; five-year relative survival at diagnosis for breast cancer patients is 89.6%. The age-standardised incidence rate for breast cancer is 116 per 100,000.*[23]

Breast cancer is the most common cancer in Australian women. Between 1992 and 1994, the incidence of breast cancer increased sharply from 98 new cases of breast cancer per 100,000 females to 114 per 100,000. This observed increase corresponded with the introduction of the national breast cancer screening program, known today as BreastScreen Australia, in 1991.[24]

Due to significant investments in research, diagnostics, and treatments we have been able to significantly reduce breast cancer mortality over the past 20 years. There are however a small subset of breast cancers that are rare subtypes, where natural history is less well defined and for which there is a much smaller evidence base.[25]

For example, the incidence figure given above includes 150 male patients who will be diagnosed this year, with a 2015 age-standardised incidence of 1.1 per 100,000.[26] Until recently male breast cancer patients were not able to access the same medication as female breast cancer patients via the PBS. But in 2015, an application by the Medical Oncology Group of Australia, enabled the PBS listings of five breast cancer drugs to be amended to ensure that men were also granted subsidised access.



Graph 14: *Incidence and mortality rates for ovarian cancer compared to population change since 1992*

> *The AIHW estimates that in 2016 there will be 1,480 patients diagnosed with ovarian cancer; five-year relative survival at diagnosis for ovarian cancer patients is 43%. The age-standardised incidence rate for ovarian cancer is 5.4 per 100,000.*[27]

By comparison, despite being the sixth most common cause of cancer-related death in women in Australia, no screening programs are available for ovarian cancer and incidence continues to increase at the same rate as population increase. The AIHW estimates that in 2016 there will be 1,480 patients diagnosed with ovarian cancer; it is also estimated that there will be 1,040 ovarian cancer deaths in the same year.[28]

When ovarian cancer is detected at an early 'localised' stage, when the cancer is confined to the ovary, up to 93% of women are likely to survive more than five years. However, only about 15% of all cases are diagnosed at this stage,[29] and as a result the average five-year survival remains low at 43%.[30]

A further example of the impact of screening, and the introduction of preventative measures such as vaccination, is seen when comparing the outcomes incidence and mortality of cervical and ovarian cancers.

23 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.
24 Australian Institute of Health and Welfare. Interpreting Cancer Data, accessed on 12th January 2016 http://www.aihw.gov.au/cancer/data/interpreting/
25 Mann B, Rare Subtypes of Breast Cancer. Asia-Pac J Clin Oncol 2015; 11 (Suppl. 4): 63-103
26 Cancer Australia. Breast Cancer in Australia, accessed on 22nd January 2016 http://canceraustralia.gov.au/affected-cancer/cancer-types/breast-cancer/breast-cancer-statistics
27 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.
28 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.
29 World Ovarian CancerDay. 5 Facts Everyone Should Know about Ovarian Cancer, accessed on 12th January 2016 http://ovariancancerday.org/about-ovarian/5-facts-everyone-should-know-about-ovarian-cancer/
30 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.

37 year old

Louisa

Osteosarcoma

On 23 June 2014, just two days before my son's 13th birthday, I was given the devastating news that the extreme pain I was feeling in my shoulder was not caused by something muscular but by Osteosarcoma, a rare form of cancer.

"It's so heartbreaking to be given the hope of a treatment that could change your life expectancy but then wonder how on earth you will afford it."

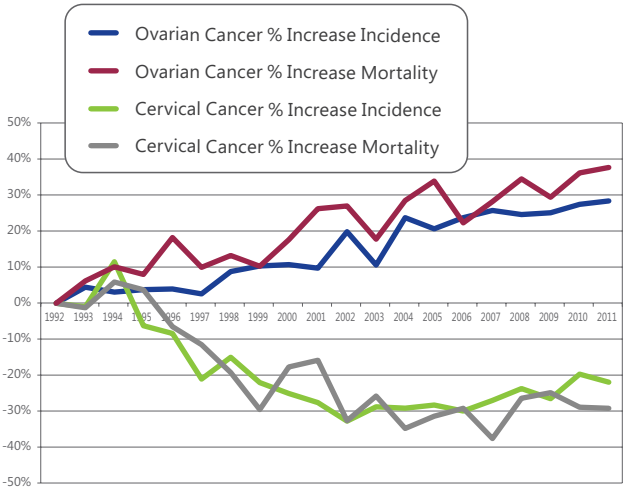
Not long after this devastating diagnosis, I commenced ten months of chemotherapy treatment. So far I have been given 5 different kinds of chemo, some have helped to slow the progress of my cancer, but have unfortunately done a lot of damage to the rest of my body and were therefore not sustainable. Others did nothing to stop my cancer.

Recently I have been given the chance to try a new treatment. There have been some real successes with this treatment for other patients with advance melanoma, my oncologist believes it might work for sarcomas too. This is a drug trial of one and hopefully if I get some success with it, others with sarcoma may get the chance to trial it too. Unfortunately, as it is not on the PBS, it is prohibitively expensive.

It's so heartbreaking to be given the hope of a treatment that could change your life expectancy but then wonder how on earth you will afford it.

I'm really hoping that this new treatment will extend my life. My greatest wish is for more time to watch my precious children grow up and more time to grow old with my husband, the way I had anticipated on our wedding day. I have so much to live for and I so desperately want to live.

Louisa sadly lost her final battle on 11 November 2015



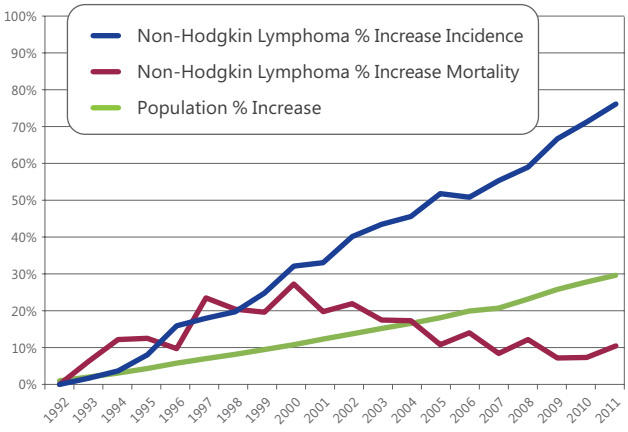
Graph 15: *The difference between screening and prevention has on cancer incidence and mortality for cervical and ovarian cancers*

The AIHW estimates that in 2016 there will be 905 patients diagnosed with cervical cancer; five year relative survival at diagnosis is 71.9%. The age-standardised incidence rate for cervical cancer is 3.5 per 100,000.[31]

Unlike ovarian cancer, there have been major advances in cervical cancer in the past 20 years. The current National Cervical Screening Program was introduced in 1991, and in 2007 the Government introduced the free National Human Papillomavirus Virus (HPV) Vaccination Program, using Gardasil, for school girls (boys were included in 2013). As of 1 May 2017 the National Cervical Screening Program (Pap test) will be replaced by an improved primary HPV test.

Both breast cancer and cervical cancer offer great hope for what is achievable in cancer prevention and treatment and, in different ways, can be seen as the 'gold standard' in terms of what is possible for improving outcomes for Australian cancer patients. The demonstrable effect of preventative interventions, early diagnostic tests and improved access to treatment on the incidence and mortality of breast and cervical cancers is unfortunately not yet replicable across all cancers.

Blood and Lymphatic Cancers



Graph 16: *Incidence and mortality rates for Non-Hodgkin Lymphoma compared to population change since 1992*

The AIHW estimates that there will be 5,200 patients diagnosed with Non-Hodgkin lymphoma in 2016, an estimated 1,445 deaths and a five-year relative survival rate of 72.1%. The age-standardised incidence for Non-Hodgkin lymphoma is 19.1 per 100,000.[32]

Non-Hodgkins Lymphomas (NHL) are cancers of the lymphatic system and are the most common form of blood cancer diagnosed annually. In the late 1990s a new biological therapy was introduced, called rituximab, which when coupled with chemotherapy is credited with greatly improving the outcomes for NHL patients; hence the significant reduction in mortality rates since 2000.

There are 43 types of NHL, and as such although classified as a common cancer, many patients have very rare subtypes. As with most rare cancers, patients diagnosed with any of these rare subtypes can face the tragic situation where there are treatments available that will save or extend their life but access is denied. Australians urgently need a more flexible and responsive way of ensuring all patients receive optimum care not just those with common conditions.

31 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.
32 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.

28 year old

Jess

Anaplastic Large Cell Lymphoma ALK+

Just over a year ago I was a normal, happy 25-year-old. I was a legal secretary at a top law firm in Sydney and was planning the trip of a lifetime to Europe with my husband. I was focused on health, fitness and nutrition. On the weekends, I'd hang out with friends, have date nights with my husband and cuddle up with my baby; a cavalier named Bailey.

"I have so much still left to accomplish, so much of the world I still want to see."

I started noticing a pain in my back which got increasingly worse. Eventually I saw a specialist who explained it could be a strain or an infection. They ordered an MRI just to be sure. The results from this scan, changed my life forever. They found tumours near my spine. At first it didn't hit me, I just felt numb. It didn't become real until I had to call my husband and tell him. I could tell from his shaky voice and how he stumbled through his sentences that he was completely gutted, as were my family and friends.

A biopsy was ordered and after an agonising one and a half weeks, I got my diagnosis of Non-Hodgkin's Anaplastic Large Cell Lymphoma (ALK positive). I was told that my cancer was rare and very aggressive.

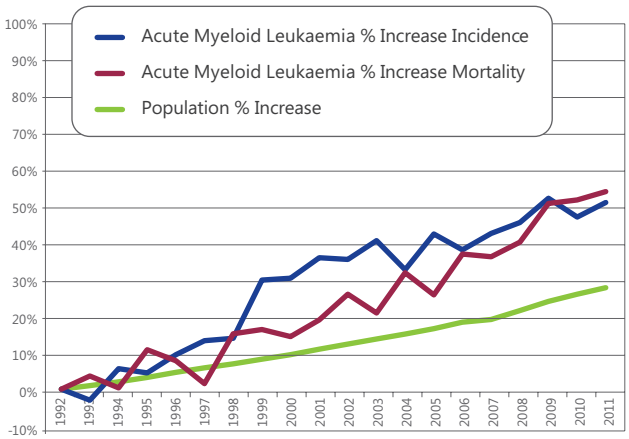
After a few rounds of chemo, the cancer began to subside. Then one morning, I was running late and in my haste, I hurt my hip, the pain gradually got worse. By the time I got in to see the doctor, my leg had swollen to the size of a tree trunk. I was told that there was a patch of cancer in my left hip, which was not only immune to the chemo, but had actually grown.

I was then placed on radiotherapy. In the following weeks, I noticed lumps in my neck, chest and abdomen. Then in my legs and arms. The doctor ordered a PET scan and the news was not good; the cancer had spread throughout my body for a second time. I was gutted.

Once again I was in excruciating pain and spent more time in hospital. I tried a new treatment which initially seemed to work, but after six weeks I started getting headaches. At first, I thought they were possible side effects to the medication, however the pain got steadily worse. I was unable to keep food down and so my husband took me to the hospital. As I was sitting in emergency, I had a psychotic episode where I exploded in anger and didn't recognise any of my family members. The lymphoma had caused severe swelling in my brain and I had narrowly escaped death. My husband calls it the worst and most traumatic night of his life. After that, I required lumbar punctures every ten days, where doctors inject chemo into the spinal fluid.

Later on I was told that I was in remission for the first time in my journey. My stem cell transplant had been arranged after an overseas donor was found and I was to go through with the procedure in a month's time. That was until the cancer came back in my left hip. It had mutated so the drug became ineffective; it's very conniving! My stem cell transplant has been put on hold until I'm back in remission.

I just want to be a normal young woman again. I want to grow old with my husband and spend time with Bailey. I also want to help others who are going through this same horrible and terrifying journey. I have so much still left to accomplish, so much of the world I still want to see.



Graph 17: *Incidence and mortality rates for Acute Myeloid Leukaemia compared to population change since 1992*

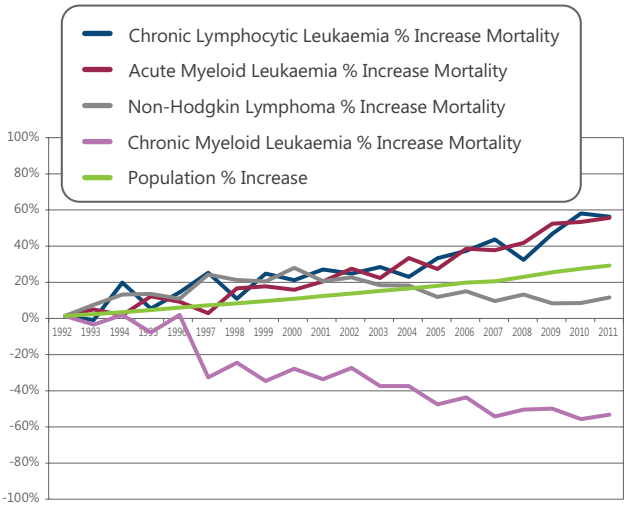
The AIHW estimates that there will be 1,070 patients diagnosed with acute myeloid leukaemia (AML) in 2016, an estimated 980 AML deaths and a five-year relative survival rate of 24.5%. The age-standardised incidence for AML is 3.8 per 100,000.[33]

Adult acute myeloid leukaemia (AML) is a rare cancer of the blood and bone marrow and is the most common type of acute leukaemia in adults. Since 1992 both the incidence and mortality for AML have increased at twice the rate of population growth.

There are 8 subtypes of AML, and like NHL, a patient's prognosis depends on the genetic make up of their cancer. Unlike NHL there have been no significant breakthrough treatments identified to treat AML and as a result, prognosis remains poor.

Prognosis does however vary with age. AML treatment success is largely age specific with many children responding well to standard care, which includes in most cases a stem cell transplant. Older AML patients, however face much lower survival rates.

In recent years targeted therapies are becoming available and these bring new hope for the treatment of AML. Ensuring their availability to AML patients in Australia remains a major challenge.



Graph 18: *Mortality rates for haematological cancers including Non-Hodgkin Lymphoma compared to population change since 1992*

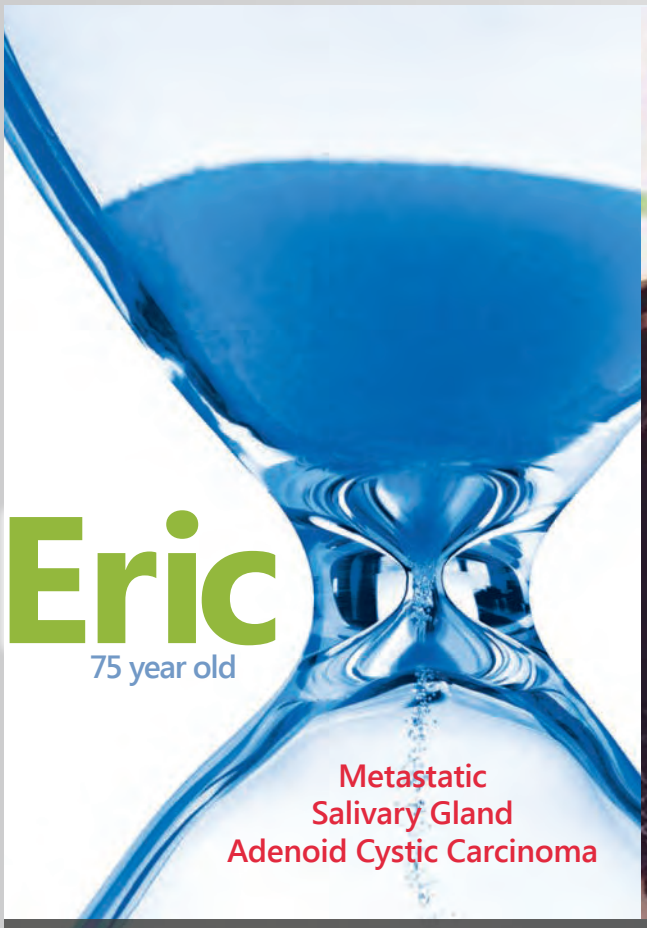
The AIHW estimates that in 2016 there will be 1,360 patients diagnosed with chronic lymphocytic leukaemia (CLL); five-year relative survival at diagnosis for ovarian cancer patients is 76.7%. The age-standardised incidence for CLL is 4.7 per 100,000.[34]

Without significant breakthrough therapies for blood and lymphatic cancers it appears that little progress is being made to reduce mortality rates. The very nature of these cancers, and their many rare subtypes makes them especially difficult to treat with one breakthrough, but very susceptible to highly targeted medicines.

Chronic Myeloid Leukaemia (CML) was the first cancer to be identified by specific genetic change and this allowed researchers to develop the ability to map targets. CML has since become the model for the development of treatments for all cancers.

In the past six years there have been a number of new drugs come through successful TGA and PBAC approvals for these RLC blood cancers but, because the majority of blood cancers are so heterogeneous and therefore rare, we have a long way to go to deliver treatments for all patients.

33 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.
34 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.



Eric

75 year old

Metastatic
Salivary Gland
Adenoid Cystic Carcinoma

Six months ago the world changed as we knew it. Our Dad had cancer and what was even worse was that it was a rare one, Adenoid Cystic Carcinoma (ACC).

It was easy to ignore the back pain, mysterious lumps appearing all over his skin and a persistent cough that never went away. However, it all came to a head a couple of months ago. He'd gone from someone with mild back pain a week prior, to someone pale as a ghost who couldn't even walk. I took him to emergency and he had back surgery the very next day. There was a large tumour that had wrapped itself around his spine. While they successfully removed most of the tumour, they then found that the cancer he had 23 years ago, had been growing inside of him since 1993.

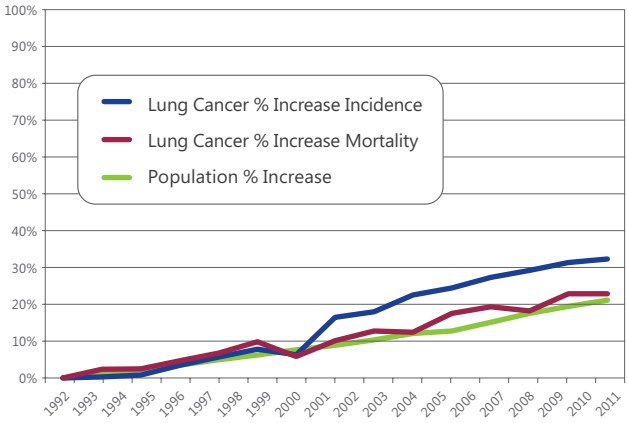
Dad's prognosis isn't great. He has advanced Stage IV terminal cancer that presents all over his skin, in his lungs and other major organs. Additionally, he has another tumour which has presented behind his leg.

"It's important not to let these things define your life, or what's left of it, but it's hard to let go of someone when there could be something that can save them."

As days go by, his tumour is becoming more aggressive. I am so hopeful treatment is going to work so that my dad can be there in some of the most crucial events of our lives; birthdays, holidays, Christmas, family gatherings and most importantly, being there at each of our weddings. It's important not to let these things define your life, or what's left of it, but it's hard to let go of someone when there could be something that can save them. We have so much to be thankful for, wonderful friends and family, memories that will last a life time, but most of all that we've had such a wonderful dad for such a long time.

My father has helped so many people over the years and has asked for nothing in return, and all I can do in return is help to save his life, or prolong it. I hope to help my father receive treatment in a bid to save his life, before it's too late.

Respiratory Cancers



Graph 19: *Incidence and mortality rates for lung cancer compared to population change since 1992*

The AIHW estimates that there will be 12,200 patients diagnosed with lung cancer in 2016, an estimated 8,960 deaths and a five-year relative survival rate of 14.3%. The age-standardised incidence for lung cancer is 42.5 per 100,000.[35]

Lung cancers present a different side to the improvements in common cancers story. While lung cancer treatments have come a long way in the past 20 years, i.e. there was almost nothing previously available for these patients, these improvements pale in comparison to other common cancers. However, where previously treatment benefits were judged by improved survival measured in weeks and months, it is now possible to measure survival in years.

For people with lung cancer, the diagnosis of lung cancer may be compounded by feelings of guilt, shame, distress and isolation.[36] A recent Cancer Australia audit of cancer research investment showed only five per cent of tumour-specific research funding was spent on investigating lung cancer, despite it being Australia's number one cancer killer[37] (see Funding Cancer Research for more information). Lung cancer can occur in both smokers and non-smokers, none of whom should suffer the added burden of stigma due to their disease, let alone face delayed improvements in outcomes because of below average research funding.

Today, with the introduction of preventative health programmes, genetic testing and identification biomarkers we may finally be seeing an impact on mortality rates. Targeted treatments for rare lung cancers (discussed in more details later in this report, see p29) are emerging and the first of these was made available, via the PBS, to Australian patients last year; the first immunotherapy for lung cancer is also due to be considered at the March 2016 PBAC meeting.

35 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.

36 Cancer Australia. Stigma and nihilism and the lung cancer diagnosis. Accessed February 2016: https://canceraustralia.gov.au/affected-cancer/cancer-types/lung-cancer/stigma-and-nihilism-and-lung-cancer-diagnosis.

37 Cancer Research in Australia: an overview of funding to cancer research projects and research programs in Australia 2006 to 2011

"With two young kids and a beautiful wife, time means everything to me."

Five years ago I met my wife Kellie. We made a wonderful home together and welcomed two beautiful children, our 3 year old daughter and 11 month old son.

Shortly after I turned 36, I developed a cough which was diagnosed as pneumonia. I had several courses of antibiotics with no results. The next tests moved very quickly and within days I was sitting across from a doctor, with Kellie by my side and our 7 month old son on her lap, being told I had Stage 4 lung cancer and given 6 months to live. I was in shock. I had never smoked, rarely drank and had a healthy diet.

Unfortunately, my first gene test results came back showing my cancer's gene mutation was not the common mutation. However, some targeted therapies have been developed only recently and have proved effective at stopping or slowing the progression of the cancer even in these cases, not a cure but time. With two young kids and a beautiful wife, time means everything to me.

So I began my chemotherapy treatment and just prior to Christmas I got my first test results which were better than expected. A reduction in my primary tumour, lymph nodes and bone lesions, the chemotherapy was working.

Early January 2015, I commenced my second course of treatment. I had responded well to chemotherapy, with few side effects, but in mid January I started to feel nauseous, bloated and short of breath. The change was quite sudden, tests were ordered and the results were devastating. My original tumours had not grown significantly but new cancer had developed. Fluid had spread to my ankles, stomach, heart and lungs. The doctor said he had not seen such a rapid reversal in test results in many years. I spent 5 days in intensive care.

The journey has been hard. I desperately want to fight this, I have so much to live for, so many milestones to celebrate. My family want me to fight this, they need me to fight this.

42 year old

Tanya

Mesothelioma

"I spent the next 10 days in hospital while tests continued to be done. I was then sent home with a pneumonia diagnosis."

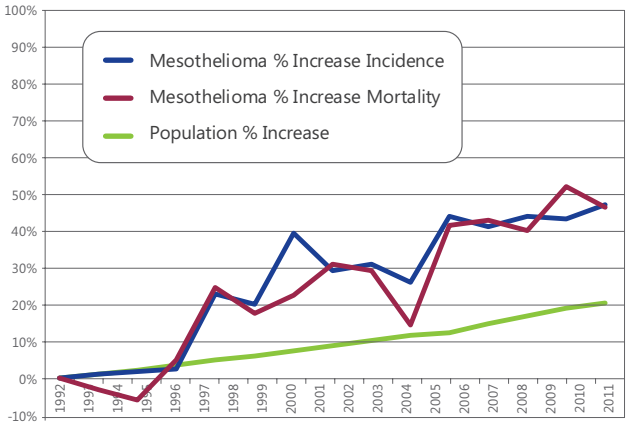
I have a husband and three children. I worked as a school teacher and prior to that as a fitness instructor/personal trainer. I lived a very healthy lifestyle. My life was busy, but nothing could have prepared me for what was to come.

18 months prior to my diagnosis, I began having sharp pains in the left side of my chest. The pains increased in frequency and duration and breathing became very challenging. I was turned away from the hospital on a number of occasions, until finally in June 2012, a doctor listened. A scan showed a large amount of fluid pressing on my lung. Two litres of blood stained fluid was drained. I spent the next 10 days in hospital and was then sent home with a pneumonia diagnosis.

As the months passed, I didn't get any better so I saw a respiratory specialist. I demanded a biopsy as I knew something was definitely wrong. The results indicated advanced mesothelioma with about nine months to live. So my treatment began, it consisted of harsh chemotherapy, radical surgery and radiation therapy. I went on to have scans every six months, for the next 18 months, which were all clear of any mesothelioma.

Until earlier this year I was okay, but then the symptoms came back. Unfortunately I was diagnosed again with pleural mesothelioma, this time it was my right side. I have undergone chemotherapy, however, it has failed to shrink the tumours.

Tanya lost her battle with cancer on 23 September 2015.



***Graph 20:** Incidence and mortality rates for mesothelioma compared to population change since 1992*

The AIHW estimates that there will be 830 patients diagnosed with mesothelioma in 2016, an estimated 755 deaths and a five-year relative survival rate of 5.8%. The age-standardised incidence for mesothelioma is 2.8 per 100,000.[38]

Mesothelioma is a rare type of cancer in which malignant cells are found in the lining of the chest or abdomen. Exposure to airborne asbestos particles increases one's risk of developing malignant mesothelioma.

In 2000 incidence rates are shown to increase as a result of the number of patients with asbestos related mesothelioma developing symptoms and improvements in our ability to diagnose those patients. The simultaneous increase in mortality also occurs at this time, as there was no effective treatment available. Mesothelioma really is a neglected disease, even now there is only one treatment available and outcomes remain poor with a five-year relative survival of just 5.8%.

In fact, mesothelioma remains universally fatal, with a median survival of approximately 9–12 months.[39] Unfortunately most patients present with advanced disease, and palliative cytotoxic chemotherapy has been the mainstay of treatment for the past 15 years, prolonging survival modestly in selected patients.[40] More focus on mesothelioma has occurred in the past 10 years, and there are now a small number of companies, which have treatments going through trials. Immunotherapies currently provide the most promise for new treatment advances and these currently provide the best hope for patients.

38 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.

39 Musk AW, Olsen N, Alfonso H, et al. Predicting survival in malignant mesothelioma. Eur Respir J 2011; 38: 1420-1424.

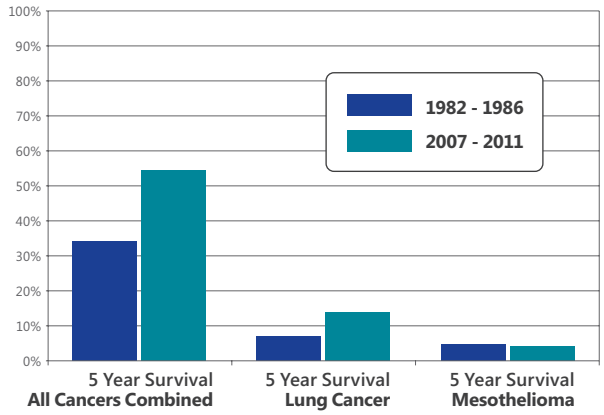
40 Vogelzang NJ, Rusthoven JJ, Symanowski J, et al. Phase III study of pemetrexed in combination with cisplatin versus cisplatin alone in patients with malignant pleural mesothelioma. J Clin Oncol 2003: 21; 2636-2644.

Survival Rates

Improvements in survival rates over time are key to demonstrating the success interventions such as introducing screening, improving diagnosis, access to treatment and other preventative activities have for cancer patients.

The following graph shows the significant improvements in survival that have been achieved for all cancers between 1982-86 and 2007-2011. These advances are largely due to the successes in early diagnosis and treatment for cancers such as breast cancer (72.2% to 89.6%) and prostate cancer (56.6% to 93.2%).

While still low lung cancer survival has almost doubled (8.4% to 14.3%) while mesothelioma patients have seen a marginal decrease survival over the same period (5.9% to 5.8%).



Graph 21: *Five-year survival for all cancers compared to lung cancer and mesothelioma*

Improvements in overall survival are shown to be directly related to research funding. For example, in 2006 – 2011 funding for breast cancer research was $201m, bowel cancer research was $195m and prostate cancer research funding was $108m, while funding for lung cancer research was significantly lower at $44m.[41]

It is not surprising that the survival rates of rare and super rare cancers are significantly lower than those of common cancers, but the fact that we have made so little improvement for these patients remains disappointing. If we are to have any chance of increasing survival rates for all cancers, it is critical that we address the failings of current diagnosis and treatment for rare and super rare cancer patients.

41 Cancer Australia, 2015. Cancer Research in Australia: an overview of funding initiatives to support cancer research capacity in Australia 2006 to 2011.

NSCLC ROS1+

"The prognosis for this cancer is generally dismal, measured in months for advanced stage cancer."

My world shattered just days after my 34th birthday when I was diagnosed with advanced lung cancer; but I'm not about to let this diagnosis define my whole life. I aim to live with cancer with love and thankfulness in my heart.

You see, I have so much to be thankful for. God has given me a loving husband who is my rock and best friend, an amazing 3 year-old daughter who is our everything, a wonderful extended family that I could not have picked better, beautiful friends and colleagues. I also have a dream job as a social justice lawyer that I am deeply passionate about.

It was easy to miss the signs, I had a cough for two months prior to my diagnosis, but had dismissed it as a post-viral cough. As the days went by, further testing showed I had developed a rare form of lung cancer caused by the mutation of the ROS1 gene, a mutation that only 1-2% of non-small cell lung cancer patients have and one often found in young never-smokers like myself.

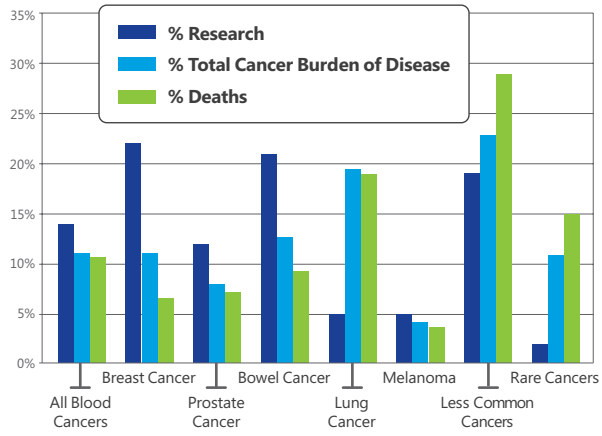
The prognosis for this cancer is generally dismal, measured in months for advanced stage cancer.

However, I hope to be there to send my daughter off on her first day of school with tears in my eyes, I hope to celebrate many more birthdays with the people I love. Most of all, I hope to share many more years of tears and laughter with my husband, the love of my life, as we navigate this journey together.

FUNDING CANCER RESEARCH

The 2015 Audit of Cancer Research in Australia reported that between 2006 and 2011, the Australian Government (including the NHMRC) provided $1.03bn (or 58% of $1.77bn total funding) for cancer research. Of the $350m spent annually on cancer research only a negligible 2% of that going to solid rare tumours.[42]

The Audit report noted that while breast cancer, colorectal cancer, haematological cancers and genitourinary cancers received the highest levels of funding in Australia the proportional funding to research in many cancers was low compared to incidence, mortality and burden of disease on the Australian population. Those cancers included lung, lymphoma, pancreas, oesophagus, kidney, stomach, bladder, myeloma and cancer of unknown primary.[43]



Graph 22: *Percentage cancer research expenditure (per annum) versus percentage burden of disease and deaths*

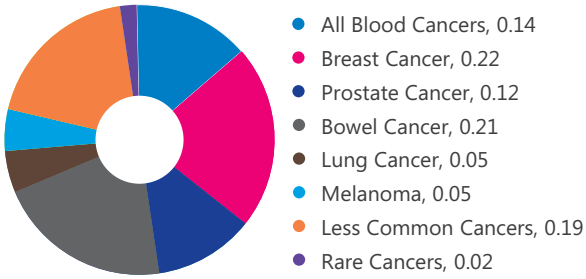
While the Audit report showed a slight increase in research funding for less common cancers, it demonstrated a continuing strong focus on common cancers within the Australian Research Community. Indeed it even recommended that '*Research funding investment in Australia could be prioritised for cancers which have a high impact (incidence and mortality) and burden of disease – disability-adjusted life years (DALYs)*'.[44]

The impact of this neglect of rare cancer research is significant in a number of ways. The first and most obvious being that if "you don't look you don't find" meaning that without focussed research we are unlikely to find and evaluate worthwhile treatments.

Equally important however, is that without research we do not build up centres of knowledge and clinical excellence that are critical to providing the best possible standard of care for patients with specific rare cancers. **The establishment of properly funded centres for rare cancer research is now an urgent priority.**

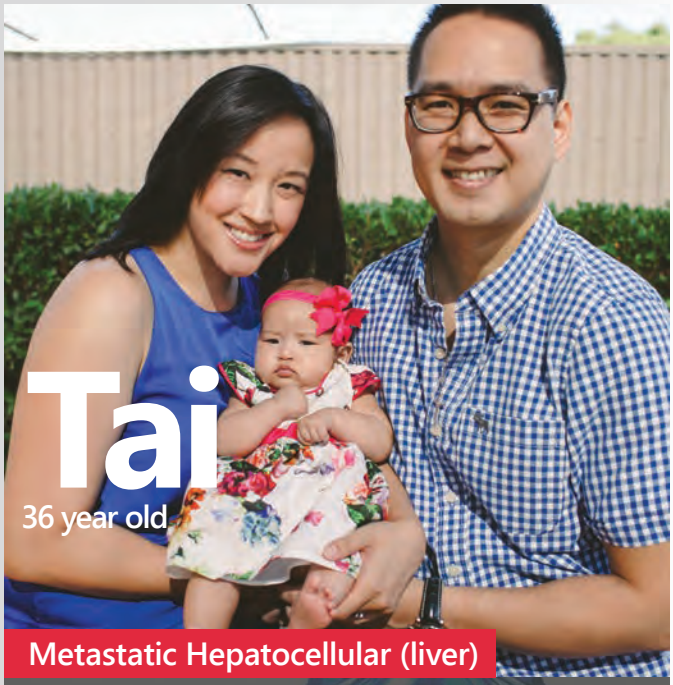
In the 2014 Budget the Government announced that it would create a Medical Research Future Fund (MRFF), to deliver additional Commonwealth funding for medical research and innovation into the future. In August 2015 the Bill to pass the MRFF into law was passed.

This new source of research funding presents an ideal opportunity for Government to take affirmative action and specifically target areas of neglect such as rare cancer research, which remains woefully low compared to combined incidence and mortality.



Graph 23: *Pie chart representing the division of research funding by cancer type*

42 Cancer Australia, 2015. Cancer Research in Australia: an overview of funding initiatives to support cancer research capacity in Australia 2006 to 2011.
43 Cancer Australia, 2015. Cancer Research in Australia: an overview of funding initiatives to support cancer research capacity in Australia 2006 to 2011.
44 Cancer Australia, 2015. Cancer Research in Australia: an overview of funding initiatives to support cancer research capacity in Australia 2006 to 2011.



Metastatic Hepatocellular (liver)

"It felt like it was one thing after another and we were just trying to buy him time to make it to the birth."

I met my husband Tai 6 years ago. We were married on December 6, 2013 and one week before our first anniversary my husband was diagnosed with a rare liver cancer. I was also 31 weeks pregnant.

The night our future became unclear, I was out with a friend. My husband called me to come home as he said he couldn't breathe. I hurried home to find him keeled over. The pain continued the next day. We went to emergency where a diagnosis would shatter our lives. A 13 centimetre mass in the liver. The pain was caused by the tumour bleeding. I was terrified. I thought the next time I'd be in hospital, we'd be welcoming a beautiful child, not potentially saying goodbye to my soul mate.

The doctors believed that his age and health would give him a high chance of shrinking the tumour. However, the CT scan showed that it had actually grown to 15 centimetres and had progressed into his lungs. It felt like it was one thing after another and we were just trying to buy him time to make it to the birth.

Six months ago we welcomed our darling daughter Kesiah. My husband was besotted. I was determined to help him achieve his one wish; for Kesiah to have one living memory of him before he dies. Our daughter is young, we know she won't remember. She will be told stories of how much her Daddy loved her. How much he wanted to see her grow and how much he wanted to stay. We know the odds are against us. We know it's going to take a one in a million chance. But we're not giving up.

Sadly Tai passed away on August 2, 2015 after battling his cancer for over 8 months.

Funding Cancer Treatment and Care

Patients with rare cancers are disadvantaged compared to other cancer patients because they have limited access to new, effective treatments.

Few new innovative treatments are reimbursed for rare cancers in Australia, mainly because of a difficulty reaching the clinical evidence threshold necessary to prove cost-effectiveness; controlled clinical trials are difficult to conduct due to the small patient base so the available clinical trials evidence is generally of low quality; and there are generally fewer drugs developed for these rarer diseases.

As previously discussed, even amongst the commonest cancers there are very small numbers of patients with rare genetic abnormalities. As oncology is being increasingly understood at the molecular level, the focus is shifting away from non-targeted cytotoxic agents towards newer targeted therapies which act on specific cancer cell growth control and survival mechanisms.

Targeted medicines to match the molecular diagnosis have brought more treatment options, improved patient quality of life and survival but have brought challenges to the regulatory and reimbursement processes but the benefits are not yet flowing to Australian cancer patients.

Imagine the following scenario

Two Australians, both aged 55, both employed in the same role for 40 years paying the same amount of tax and both diagnosed with cancer on the same day – one cancer is common and the other is rare.

Both patients consult the same oncologist who determines after consideration that they should both be treated with the same medicine. In the case of the patient with the common cancer his treatment is fully funded through the PBS.

The rare cancer patient, in contrast, must find $7,000 per month, sometimes more, to pay for his treatment simply because his disease is rare.

Examples of treatments for rare cancer patients in Australia

Identification of molecular drivers in individual tumours and treatment with effective targeted therapy is essential for personalised medicine in lung cancer (12,200 total cases per annum in Australia[45]). Rearrangements in the Anaplastic Lymphoma Kinase (ALK) gene are present in 3-5% of patients with non-small cell lung cancer (NSCLC), often in younger, never-smokers (see Corey's story on P24). These patients show high response rates and improved survival after treatment with ALK inhibitors, such as crizotinib (Xalkori) which was listed on the PBS for these patients in 2015.[46]

Other molecular alterations in NSCLC such as ROS1 gene rearrangements are present in 1-2% of lung cancer patients[47] (see Lillian's story on P26). Despite trials demonstrating impressive efficacy of the same inhibitors in ROS1 NSCLC, it is unlikely that it will ever be possible to conduct a phase III study in this very rare, molecularly defined population leading to challenges for the registration and regulatory approval of drugs for these populations (see Discussion).[48]

Another example that illustrates the issues with subgroups of patients is stomach cancer (a less common cancer with 2,240 total cases per annum in Australia). One in 10 patients (i.e. approximately 200 new patients/year) have increased levels of a growth factor receptor on the surface of cells known as HER2. This same growth factor receptor is present in a small percentage of breast cancers and when present, patients derive substantial benefit from the drug trastuzumab (Herceptin). While Herceptin has been available to Australian HER2-positive breast cancer patients since 2006, it was only recommended for HER2-positive stomach cancer in 2015; a tragic, and yet avoidable delay for many patients.

For many of RCA's Sick or Treat patients the scenario opposite is all too real. Most must face the difficult decision as to whether they can afford the treatment that their oncologist recommends as offering the best chance of survival, or try their chances with something older, that may not work as well, but which won't bankrupt them and their family.

Since 2010, the PBAC has made a total of 85 positive recommendations, 56 of these were for 'common cancers' or 'unspecified cancers'. Of the remaining 29 positive recommendations only six (7%) were for rare solid tumour cancers. In fact, two of those were made to expand the listing of 6 breast cancer drugs, which have all been available for around a decade, to other rare indications (notably male breast cancer and HER2-positive stomach cancer, see box).

Why the Challenges for Rare Cancers?

In most cases, a major barrier to the public reimbursement of therapies for the treatment of rare diseases is the available clinical evidence fails to reach the threshold necessary to prove cost-effectiveness. The supporting clinical evidence is often limited to data from clinical trials involving small numbers of patients. This is because the generation of meaningful comparative data requires the participation of many sites across multiple geographies. Such an approach is largely impracticable and unrealistic for rare diseases, as recruitment is often difficult and the costs associated with running such trials are disproportionate to the potential revenue that may flow to the manufacturer.

While the PBAC acknowledges the need for concessions to be made when considering the clinical evidence supporting reimbursement applications for rare diseases, the reasons cited for rejection of these drugs frequently include uncertainty regarding clinical effectiveness and therefore uncertain cost effectiveness.

An outright rejection means either the manufacturer gives up completely its attempts for reimbursement, or has to go through a lengthy and costly (both to the sponsor and the PBAC) resubmission process with no guarantees of success. **Either way the patients are the losers, as they may never gain access to needed treatment.**

45 Australian Institute of Health and Welfare 2014. Cancer in Australia: an overview 2014. Cancer series 90. Cat. No. CAN 88. Canberra: AIHW.
46 Solomon B (2015). ALK and ROS Positive Lung Cancers. Asia-Pac J Clin Oncol 2015; 11 (Suppl. 4): 63-103
47 Solomon B (2015). ALK and ROS Positive Lung Cancers. Asia-Pac J Clin Oncol 2015; 11 (Suppl. 4): 63-103
48 Solomon B (2015). ALK and ROS Positive Lung Cancers. Asia-Pac J Clin Oncol 2015; 11 (Suppl. 4): 63-103

Discussion

Since RCA wrote the first *Just a Little More Time* report in March 2014, there have been a number of opportunities, both in Australia and globally, to improve the lives of people living with rare cancers. Yet it is clear from the available evidence that what we have achieved in the past two years has had very little impact on the outcomes for Australian rare cancer patients and their families.

Improving research funding

Despite the Cancer Australia Audit of research funding demonstrating that total funding for rare and less common cancers has increased in recent years, the total funding required to close the gap between funding and the burden of disease and mortality caused by RLC cancers compared to common cancers remains significant.

Clinical trials into effectiveness of novel, targeted therapies, in small patient populations, require collaborative trial development and research which crosses traditional boundaries of trials currently being undertaken in Australia,[49] and the evidentiary requirements for regulators must also be made to be more flexible for rare and super rare cancers.

The Australian Government, through the NHMRC, and other Departments is the largest funder of cancer research in this country, and this funding is set to increase through the MRFF. Given the significant role that the Government has to play in funding this important work, RCA calls on the Government to take the lead through affirmative action to direct funding to specifically target areas of neglect such as rare cancer research.

Only by improving our investments in rare cancer research will we ever be able to deliver improvements to patients and reduce mortality rates for these otherwise neglected patients.

Improving healthcare networks

Although RCA has not been able to include data on healthcare networks in Australia in this report, we are acutely aware of the problem that rare cancer patients face in terms of receiving a prompt and accurate diagnosis. As we have seen through all the cancer specific examples, prompt and accurate diagnosis is critical to a patients overall health and prognosis, so it is essential that we take steps to address this challenge.

While RCA is working to create a Patient Support Programme that aims to address these challenges by identifying the right specialists and multi-disciplinary teams, all those involved in patient care must take a more proactive approach, to provide rare cancer patients across Australia with the best available care.

Improving treatment availability

Despite existing sympathies within the PBS towards 'orphan medicines', the majority of cancer drugs that may be effective for rare cancers are not orphan, for example the PD-1 inhibitors, because they are used and listed for more common cancers.

RCA envisages a framework might be developed for future PBAC submissions for common cancers to include the reimbursement of targeted oncology drugs for rare cancers. The framework would not be too far removed from the current framework for reimbursement submissions. However, it would allow consideration of the wider use of the targeted treatment beyond the common cancer indication.

We recommend that we now take action to ensure that medicines, that are not technically orphan, because of their use in common cancers, are treated as orphans when under consideration for much rarer indications (i.e. super rare cancers with an incidence of less than 2 per 100,000, equivalent to less than 500 patients, per annum).

Therefore when new, innovative treatments are considered by the PBAC for 'common' cancers, the PBAC may also include in its assessment, the super rare indications that may be identified as likely targets of the treatment, and the same flexibility for evidence is applied as though the treatment were orphan.

For example, when a treatment going through PBAC assessment for a more common indication, associated super rare indications are also included in the decision making process. For treatments for common cancers, such as melanoma (with an incidence of 12,640 in 2014), including less than 4-500 additional patients with super rare indications may be possible without creating a significant additional burden on the Budget.

The patients with super rare cancers will never have access to these newer highly targeted medicines, if we do not take action on their behalf.

49 Jackson P, et al. The genomic cancer clinical trial initiative – collaboration toward achieving common goals. Asia-Pac J Clin Oncol 2015; 11 (Suppl. 4): 63-103

CONCLUSION

In this age of medical research, the greater understanding of the molecular biology of cancers and the advent of immunotherapies for treating cancer, we have an enormous opportunity to improve the outcomes of rare cancer patients.

The simple cost of doing nothing to improve outcomes for RLC cancers is too high. This report clearly demonstrates that the burden of disease from RLC cancers affects everyone, from our children, our young families, all our working-age professionals, our parents and grandparents.

The challenge therefore, is to find a mechanism whereby:

- Research funding is increased and specifically directed to encourage and drive research into rare cancers and rare molecular sub-types;
- Health professionals are supported to be able to deliver improved diagnosis and care, either through multi-disciplinary teams or networks; and
- Rare cancers patients can receive equitable and fair access to medicines that have reasonable, proven safety and efficacy for those diseases.

We must ensure that rare cancer patients have access to the best available care, as directed by their treating clinician. This includes the PBAC considering whether the 'alternative' treatments available to these patients are suitable in today's treatment environment, and assessing whether it is ethical to provide rare cancer patients with 30 year-old, cytotoxic chemotherapy treatments, when newer, more effective, treatments are available.

It remains extremely disappointing that in the last 20 years when we have seen many advances in common cancers and other diseases, that we have seen very little, if any, progress in the treatment of rare and less common cancers.

Addressing the discrepancies for RLC cancers compared with common cancers needs to occur at the highest level, and we need the Australian Federal Government to take action.

Rare cancers represent a major diagnostic as well as therapeutic challenge and they represent a major source of discrimination among patients.[50] It is time we took the action necessary so that we can to give these Australian patients the resources, support and treatment they need and most importantly, provide them all with "just a little more time".

50 Dei Tos AP, Classifying rare cancers. What is a rare cancer? Asia-Pac J Clin Oncol 2015; 11 (Suppl. 4): 63-103

Appendix 1

ICD- 10 Cancer Codes

ICD Code	Incidence
C39 Other and ill-defined sites in the respiratory system and intrathoracic organs	0
C58 Placenta	6
C33 Trachea	11
C75 Other endocrine glands and related structures	24
C63 Other and unspecified male genital organs	28
C96 Other and unspecified cancers of lymphoid, haematopoietic and related tissue	29
C94 Other leukaemias of specified cell type	30
C76 Other and ill-defined sites	35
C47 Peripheral nerves and autonomic nervous system	38
C68 Other and unspecified urinary organs	41
C70 Meninges	41
C08 Other and unspecified major salivary glands	42
C72 Spinal cord, cranial nerves and other parts of central nervous system	49
C38 Heart, mediastinum and pleura	49
C37 Thymus	52
C13 Hypopharynx	56
C31 Accessory sinuses	68
C46 Kaposi sarcoma	69
C74 Adrenal gland	75
C93 Monocytic leukaemias	75
C95 Leukaemias of unspecified cell type	77
C52 Vagina	77
C14 Other and ill-defined sites in the lip, oral cavity and pharynx	83
C88 Immunoproliferative cancers	89
C12 Pyriform sinus	91
C10 Oropharynx	95
C55 Uterus, part unspecified	97
C60 Penis	108
C30 Nasal cavity and middle ear	109
C03 Gum	109
C40 Bone and articular cartilage of limbs	114
C41 Bone and articular cartilage of other and unspecified sites	115
C11 Nasopharynx	120
C05 Palate	130
C66 Ureter	142
C57 Other and unspecified female genital organs	149
C06 Other and unspecified parts of mouth	151
C04 Floor of mouth	181
C26 Other and ill-defined digestive organs	185
D45 Polycythaemia vera [WARNING: Incomplete time series]	196
C48 Retroperitoneum and peritoneum	203
Total for Super Rare (2.89% of all cancer diagnoses in Australia in 2011)	**3,439**

C07 Parotid gland	245
C01 Base of tongue	259
C69 Eye and adnexa	266
C65 Renal pelvis	273
C84 Peripheral and cutaneous T-cell lymphomas	275
C51 Vulva	318
C23 Gallbladder	339
C09 Tonsil	361
C21 Anus and anal canal	369
C02 Other and unspecified parts of tongue	430
C24 Other and unspecified parts of biliary tract	433
D47 Other cancers of lymphoid, haematopoietic and related tissue	440
C17 Small intestine	442
C32 Larynx	590
C49 Other connective and soft tissue	604
C81 Hodgkin lymphomas	606
C45 Mesothelioma	690
C62 Testis	732
C44 Skin - Non-melanoma, excluding BCC and SCC	769
C53 Cervix	801
C00 Lip	912
Total For Rare (8.55% of all cancer diagnoses in Australia in 2011)	**10,154**
Total for Rare and Super Rare (11.45% of all cancer diagnoses in Australia in 2011)	**13,593**
D46 Myelodysplastic syndromes [WARNING: Incomplete time series]	1,309
C56 Ovary	1,330
C15 Oesophagus	1,395
C22 Liver and intrahepatic bile ducts	1,446
C92 Myeloid leukaemias	1,457
C90 Multiple myeloma and other plasma cell cancers	1,533
C91 Lymphoid leukaemias	1,654
C71 Brain	1,724
C16 Stomach	2,093
C73 Thyroid gland	2,098
C54 Body of uterus	2,140
C67 Bladder	2,404
C25 Pancreas	2,748
C80 Unknown primary site	2,802
C64 Kidney, except renal pelvis	2,847
Total For Less Common (24.34% of all cancer diagnoses in Australia in 2011)	**28,981**
Total for RLC (35.86% of all cancer diagnoses in Australia n 2011)	**42,574**
C82, C83, C85 Non-Hodgkin Lymphomas	4,357
C34 Bronchus and lung	10,500
C43 Skin - Melanoma	11,570
C50 Breast	14,568
C18-20 Bowel	15,151
C61 Prostate	19,993
Total for Common (64.14% of all cancer diagnoses in Australia in 2011)	**76,138**
Total Diagnoses in 2011	**118,711**

47 year old

Vincent

Squamous Cell Carcinoma

I am the father of two amazing children. I was lucky enough as an 18 year old to receive a heart transplant, after contracting a virus that destroyed my heart tissue. It was a success and I was able to continue with my life and pursue further education and it was during my studies that I met my wonderful wife Sheryl. We live in a small town in NSW, where I spend my life looking after my kids and helping my local Lions Club make the town a better place.

In 2008, we discovered that skin cancer was a side effect of the drugs I was taking for my transplant. In 2011, I found a rapidly developing Squamous Cell Carcinoma on my hand which was removed. A year later, I discovered a lump underneath my arm which required a round of radiation therapy. To my delight, I was given the all clear from this and was cancer free.

"Without treatment, I was given at best, 6 months."

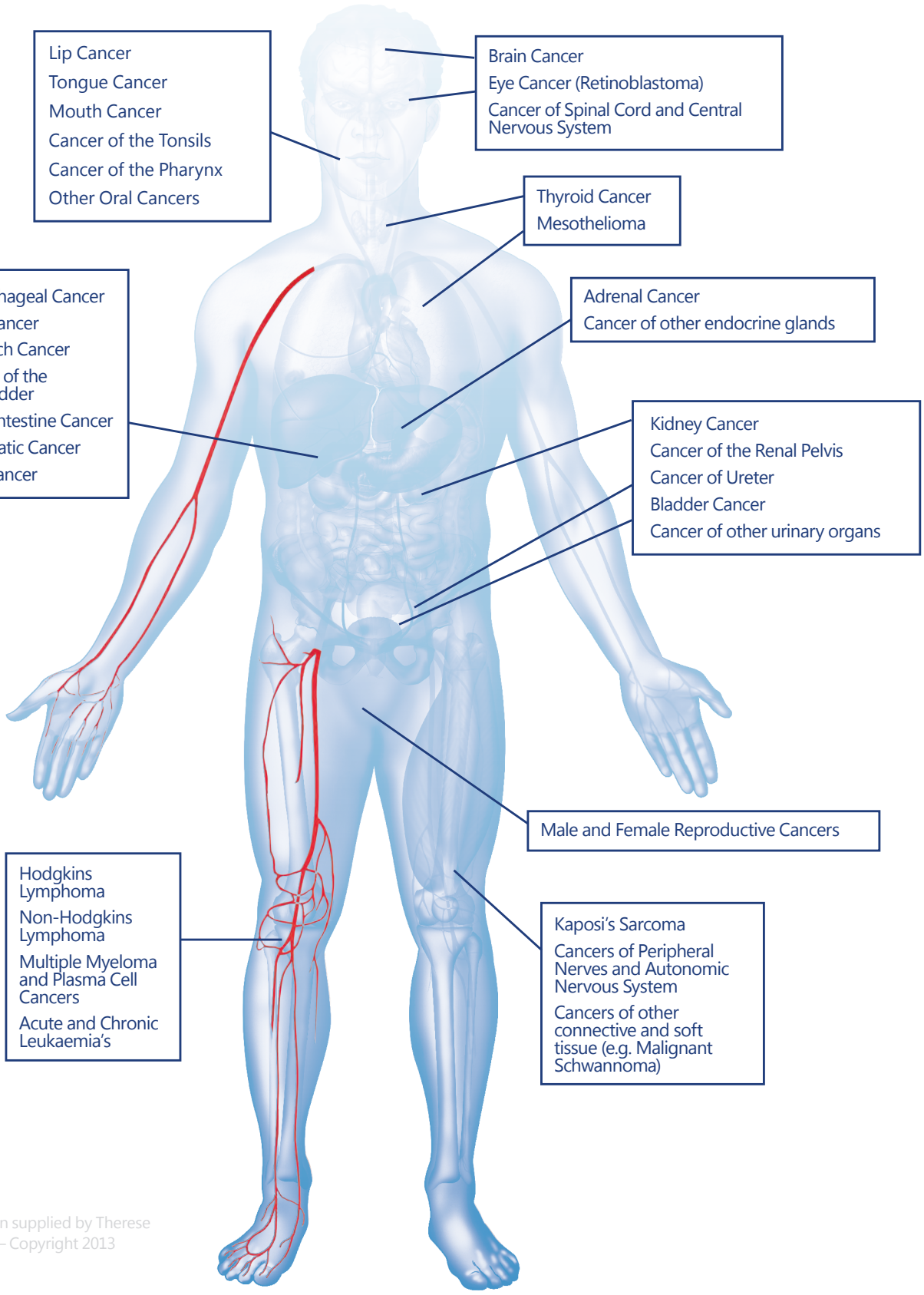
However, in July 2015, I found a lump in my leg. Tests revealed that the cancer had spread to my muscles, lungs and other places. Without treatment, I was given at best, 6 months.

I didn't want to accept this prognosis; I needed to fight this for my family.

Vincent sadly lost his battle with cancer on 1 February 2016.

Appendix 2

Body Map of Rare Cancers



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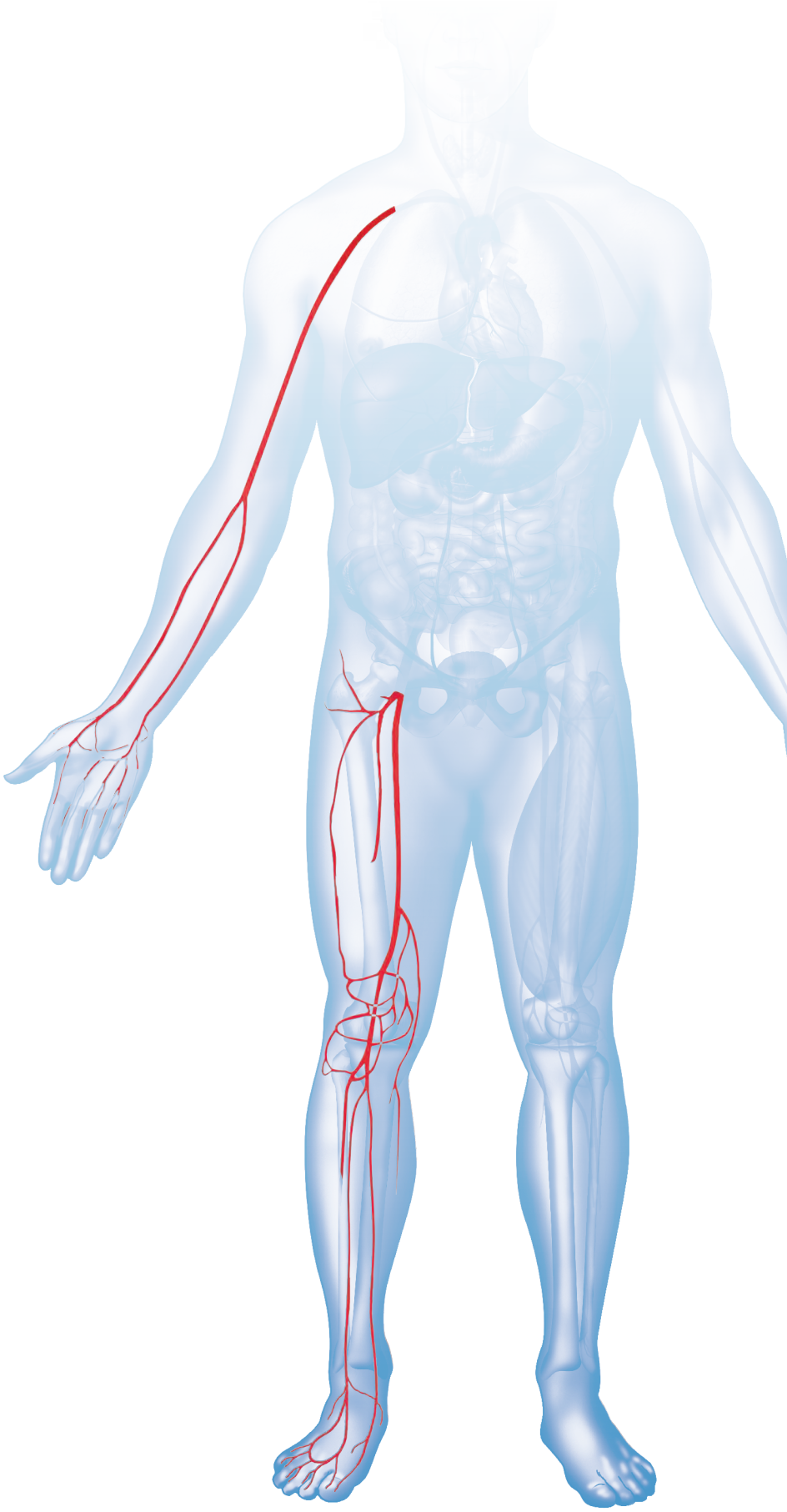

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