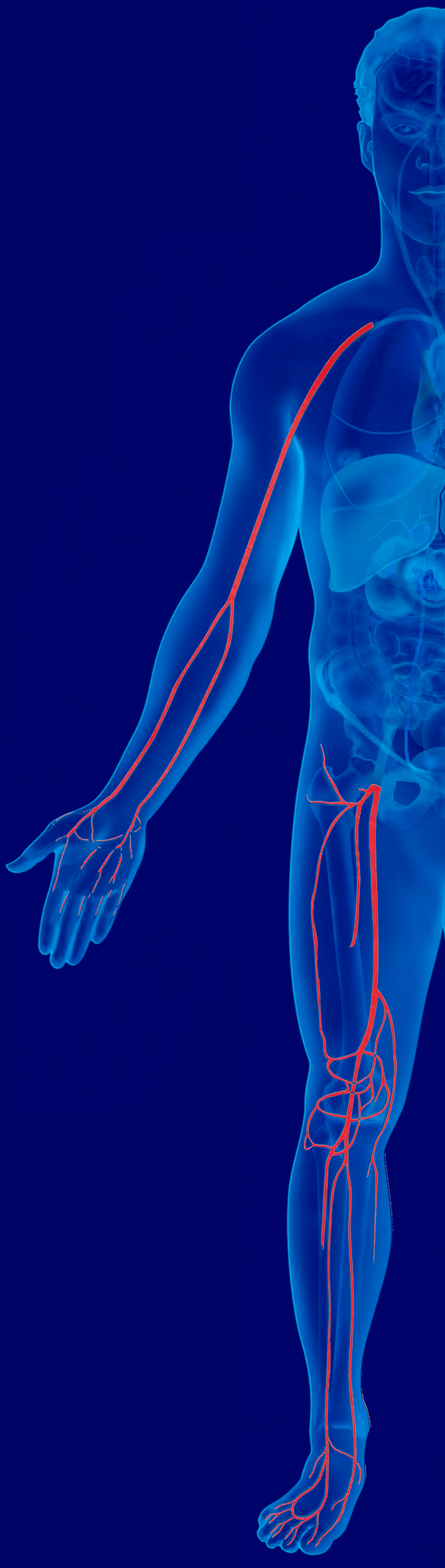
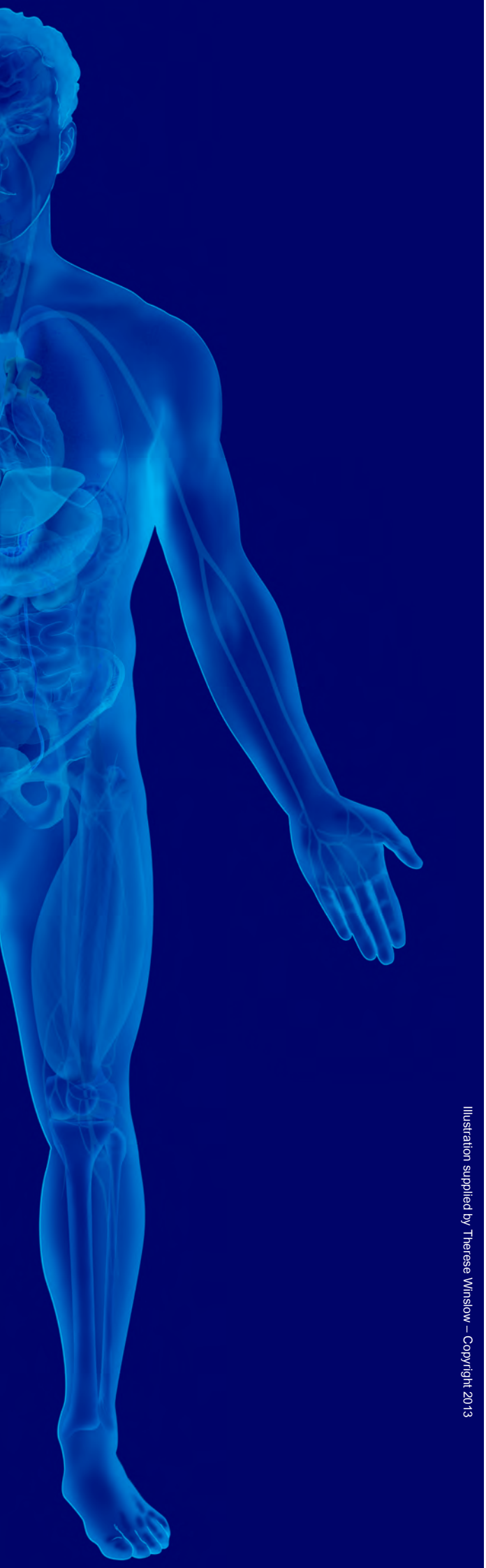
rare cancers AUSTRALIA

A FOCUS ON LESS COMMON CANCERS



JUST A LITTLE MORE TIME

rare cancers baseline report



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acknowledgements

This report has been prepared by Rare Cancers Australia Ltd (RCA) to provide an evidence based understanding of the current state of research, diagnosis and treatment into rare and less common cancers in Australia. The document draws on data supplied by a number of organisations including the Australian Institute of Health and Welfare (AIHW), Cancer Australia, The Pharmaceutical Benefits Scheme (PBS), Australian businesses and the pharmaceutical Industry.

We would particularly like to thank Mark Scott and his cancer team at the AIHW and the team at Cancer Australia. We would also like to acknowledge and thank those who provided financial and in kind support that assists RCA with our day to day operation and the creation of this report.

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Our thanks also to Charlene Vien and Belynda Simpson for their kind, generous and brilliant work in designing and laying out this report. We are very grateful and appreciative.

about rare cancers australia

Rare Cancers Australia (RCA) is a charity whose purpose is to improve awareness, support and treatment of Australians with rare and less common (RLC) cancers. RCA's goals can be summarised as follows:

- Patient Community - Develop an online community to ensure patients better understand their disease and communicate with others sharing their challenge;
- Fundraising - Provide an avenue for family and friends to raise money to assist with treatment costs;
- Treatment - Ensure the best treatments in the world are available and affordable to Australia cancer patients; and
- Early Diagnosis - Raise awareness with both the public and medical profession of rare and less common cancers.

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foreword

It is salutary to think that without the work of Rare Cancers Australia, spear-headed by Kate and Richard Vines, we would not have at our finger-tips, their graph showing the crux of the problem: that only 12.6% of research funding is spent on rare and less common cancers and yet, or perhaps because of that, these cancers account for 45% of cancer deaths. A story too often told, when faced with a rare cancer diagnosis, Kate and Richard not only had to face their mortality, but found there were few answers from the medical fraternity to guide their path through this devastating experience. Their response was to try to change this for others. Their determination to seek out answers, from the most appropriate national resources, revealed gaps relating to nearly all aspects of rare cancer information and management.

For me, as a clinician scientist, the frustrations are manifold:

i) By the time the diagnosis is made, often after months of distressing investigations and little information for the patient and their family, a rare cancer diagnosis usually means that there is little evidence to guide decision making;
ii) As we currently only have single "molecular tests" rather than multiple (or panel testing) which is required when the cause of the cancer is unknown, no funding for molecular testing is available to shed light on whether we can match this cancer to other cancers for which we have developed treatment paths; and
iii) If a reasonable hypothesis can be made as to a targeted treatment that may "match" the cancer, it likely will not come within the PBS-guidelines for use of that drug.

Frustrating for me, devastating for the Australian patient concerned and their family and carers. As clearly outlined in this brief report, our national effort in rare and less common cancers is limited and the current and predicted failure rates of our management belie the advances we hope we have made in treating cancer in the last forty years. The increasing incidence of these cancers, accompanied by a lack of improvement in survival, as shown in this report, indicate an increasing problem. We have the capacity to change this. As we have in other areas, such as childhood cancers and blood cancers, we need to streamline our approaches and utilise the current genomics explosion in technology to redress the balance of basic and clinical research in favour of the rare and less common cancers. Some rare cancers may be more challenging than childhood or blood cancers, less likely to have a strong "driver" or dependence that can easily be targeted. But not looking will guarantee failure. Providing support for those enduring such a diagnosis is one thing, changing the future of these Australians should be the next.

Associate Professor Clare Scott,
Principal Investigator, CART-WHEEL.org, BioGrid Australia
Cancer Council Victoria Sir Edward Dunlop Research Fellow,
Walter and Eliza Hall Institute of Medical Research

executive summary

Cancer is the leading cause of the total burden of disease and injury in Australia, accounting for approximately 19 per cent of the total disease burden in 2012.[1] Despite a decline in cancer mortality and an increase in survival over time, **1 in 2 Australians will be diagnosed with cancer and 1 in 5 will die from it before the age of 85**.[2]

'Rare cancers' are defined as those with an incidence of less than 6 per 100,000 Australians per annum and **'less common'** cancers as those with an incidence of between 6 and 12 (inclusive) per 100,000 Australians per annum.

Every year there are over 42,000 diagnoses of rare and less common (RLC) cancers and around 22,000 deaths here in Australia and there is very little available, from patient support to new treatment options, for RLC cancer patients. **Rare and less common cancers account for seven per cent of the total burden of disease in Australia.**

For those Australians with rare or less common cancers we have, for the most part, failed dismally. While it is true that we have made some excellent progress in common cancers over the last 20 years, survival rates in many rare cancers have only improved marginally if at all, our research funding into rare cancers remains disappointingly and disproportionately low, as does the money we spend on government funded treatments for these patients. **It is hard to believe that in the 20 years from 1990 to 2009 with all the advances in medical science and technology, that we have achieved so little for this group of patients and their families.**

During the last twenty years Australia has seen the introduction of screening programs for common cancers (breast, prostate and bowel), awareness programs (lung cancer and melanoma) and significant monies allocated to both research through National Health and Medical Research Council (NHMRC) and treatment through the Pharmaceutical Benefits Scheme (PBS). The impact of these programs and funding has been significant for common cancers, but in contrast RLC cancers have not been given attention and resources and there has been little or no improvement.

Unsurprisingly, the survival rates for many RLC cancers are very low when compared with rates for the more common cancers indicating at least in part, that we are much better at diagnosing and treating common cancers.

This report calls on the Australian Government to take action for improving outcomes for rare and less common cancer patients, to review existing mechanisms and improve research, diagnostics and access to medicines for rare and less common cancers.

Without targeted mechanisms specifically designed to address the prevention, diagnosis, and treatment of RLC cancers we cannot hope to have an impact on mortality or improve patient outcomes in the coming years.

1 Australian Institute of Health and Welfare & Australasian Association of Cancer Registries 2012. Cancer in Australia: an overview, 2012. Cancer series no. 74. Cat. no. CAN 70. Canberra: AIHW.
2 Australian Institute of Health and Welfare & Australasian Association of Cancer Registries 2012. Cancer in Australia: an overview, 2012. Cancer series no. 74. Cat. no. CAN 70. Canberra: AIHW.

introduction

“If only.... I had a little bit more time”

These are the words of Zach Sobiech, an American teenager who died recently of a very rare osteosarcoma. In the last few months of Zach’s life he penned a song called ‘Clouds’ that contained the above phrase. The phrase and the song perfectly capture the hope and acceptance of all cancer patients and emphasises the need for us to do better and give everyone at the very least... “a little more time”.

Zach, like so many of those with rare and less common cancers didn’t get the time he was hoping for, but his words should inspire us all to work harder and smarter to achieve Zach’s dream for others.

For those Australians with rare or less common cancers we have, for the most part, failed dismally. Over 20 years, survival rates in many rare cancers have only improved marginally if at all, 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). It is hard to believe that in the 20 years from 1990 to 2009 with all the advances in medical science and technology, that we have achieved so little for this group of patients and their families.

The systems and processes for more common cancers and diseases have addressed many of the previously unmet patient needs; what is of serious concern is that support for RLC cancers is inadequate and this must be urgently addressed.

This report examines the incidence or occurrence of these cancers, mortality figures, survival trends and our funding of research and treatment. Interwoven with these facts and figures is the real story of Australians with rare and less common cancers.

It perhaps says a great deal about these “Forgotten Cancers” that much of this information was difficult to obtain and previously uncollated. Rare Cancers Australia has prepared this report with the goal of kick starting research, policy discussion and action to improve the lives and outcomes of Australians living with a rare or less common cancer.

In short, let’s give them all at least “just a little more time”

background

Cancer is the leading cause of the total burden of disease and injury in Australia, accounting for approximately 19 per cent of the total disease burden in 2012.[3] Despite a decline in cancer mortality and an increase in survival over time, **1 in 2 Australians will develop cancer and 1 in 5 will die from it before the age of 85**.[4] Given that most Australians have either direct or indirect experience of cancer the very utterance of the 'C' word brings our own mortality into immediate and sharp focus.

While cancer is often thought of as one disease it is not; cancer is hundreds of different diseases that manifests differently in individual patients and therefore represents a health challenge of great complexity.

According to the Australian Institute of Health and Welfare, in 2009 the five most commonly diagnosed cancers in Australia were prostate cancer (19,438 cases), bowel cancer (14,410 cases), breast cancer (13,778 cases), melanoma (11,545 cases) and lung cancer (10,193 cases).[5]

As distinct from these common cancers every year there are over 42,000 diagnoses of rare and less common (RLC) cancers and around 22,000 deaths. For these Australians with an RLC cancer there is very little available, from patient support through to new treatement options. **Rare and less common cancers account for seven per cent of the total burden of disease in Australia.**

definition

'Rare cancers' are defined as those with an incidence of less than 6 per 100,000 Australians per annum and **'less common'** cancers as those with an incidence of between 6 and 12 (inclusive) per 100,000 Australians per annum.

See Appendix 1 for details.

As the media and community leap on each and every story about new treatments, new drugs and implicitly new hope, it would be easy to think we are on the verge of 'curing cancer'. While it is true that we have made some excellent progress in common cancers it is essentially the case that for RLC cancers we are not significantly further advanced than we were 20 years ago.

This report specifically looks at RLC cancers and the current treatment environment for patients living with the diagnosis of an RLC cancer in Australia.

the future

Rare and less common cancers account for over 22,000 deaths in Australia each year, and the rate of increase is currently **twice that of the population growth**. So what can we expect in the near future?

- Over 75% of RLC deaths occur in Australians 50 years and over.
- Overall survival for RLC Cancers as a group is largely unchanged since 1990
- Australia is ageing rapidly and the number of Australians over 50 is expected to double by 2030

Without concerted action in research, diagnostics and treatment Australia could be confronting over 30,000 deaths from RLC Cancers by 2020 and over 40,000 by 2030. The health, economic and social cost of inaction in the fight against these cancers is simply breathtaking.

3 Australian Institute of Health and Welfare & Australasian Association of Cancer Registries 2012. Cancer in Australia: an overview, 2012. Cancer series no. 74. Cat. no. CAN 70. Canberra: AIHW.
4 Australian Institute of Health and Welfare & Australasian Association of Cancer Registries 2012. Cancer in Australia: an overview, 2012. Cancer series no. 74. Cat. no. CAN 70. Canberra: AIHW.
5 Australian Institute of Health and Welfare & Australasian Association of Cancer Registries 2012. Cancer in Australia: an overview, 2012. Cancer series no. 74. Cat. no. CAN 70. Canberra: AIHW.

aayaan

My wife Akshata and I migrated to Darwin Australia in 2009. Our son Aayaan was born in June 2011 in Mangalore, India. When Aayaan was born, I noticed a squint in his eye however a routine check up with a local GP in India, failed to notice any abnormality. The squint disappeared over time and our family resettled back in Darwin in February 2012.

Days passed and slowly a white pupil began to appear in his eyes. Early on it was not that visible but gradually the pupil became more and more distinct. Aayaan was examined by a number of doctors for routine check ups and vaccinations, none of whom raised any concerns about his eye.

Despite assurances from the doctors, we were still very concerned and decided to seek answers from the internet. Our research led us to believe our son had retinoblastoma, a rare cancer of the eye. This new found information played on our minds and we were confused and extremely worried.

It was not until March 2013, armed with a little more knowledge that we sort the advice of another doctor. After an eye check up, the abnormality in his eye was diagnosed as a beauty spot. I was somewhat reassured that all was fine with Aayaan, but my wife was not satisfied with the doctor's assessment.

We travelled back to India and were able to get a referral for Aayaan to see a specialist. We finally got the confirmation we had feared, a diagnosis of retinoblastoma was given. The tumor had spread across the optic nerve and surgery was scheduled immediately to reduce any further complications. Aayaan was 22 months old at the time.

The whole process was very frustrating and traumatic for my family. We have raised a complaint with AHPRA about the GP's lack of knowledge, evident in his diagnosis of the symptom as a beauty spot. It appears that doctors and professionals in the field are unaware of this rare cancer, forcing parents and those affected to make their own diagnosis. This is not the ideal situation. I have been inspired to help others learn from my experience in the hope of creating community awareness of this rare cancer.

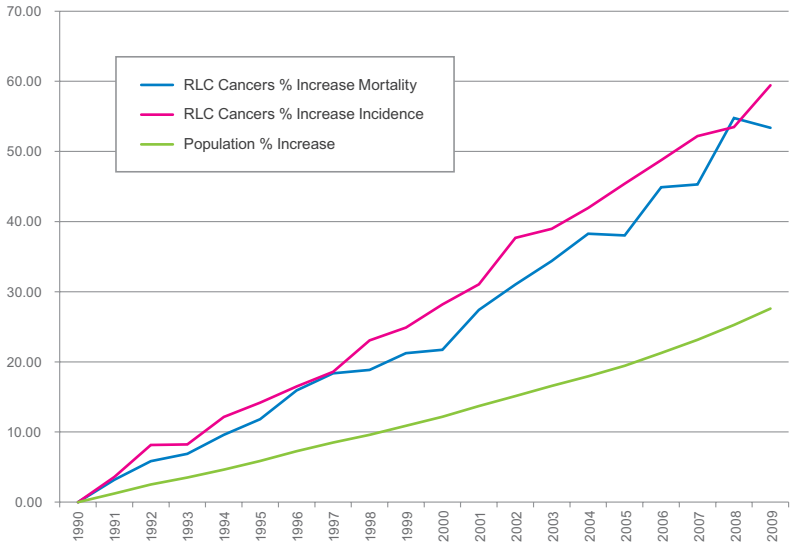
common and rare and less common cancer statistics

The most common statistics for measuring cancer outcomes are incidence, prevalence, survival (see next section) and mortality.

- **Incidence** refers to the number of people who will get a particular type of cancer each year, and as with the definition of RLC cancers on page 7, it is often expressed as the number of cancer cases per 100,000 in a given population;
- **Prevalence** refers to all the people still living who have ever had a cancer diagnosis at a given time. This is different to incidence (which counts new cases) as it includes both old and new diagnoses, as well as those patients who have been cured. Although a useful measure prevalence has not been used in this report as it can underestimate the impact of some cancers with very short survival (ref: European RareCare Project 2011); and
- **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 some information with respect to the cancer and its progression, however , when presented as a ratio to incidence it provides a reliable proxy for survival rates when actual survival statistics are not available.

To begin to understand the difference that exists in the diagnosis, treatment, support and outcomes for RLC patients versus patients suffering from a common cancer it was found to be instructive to consider these cancers collectively.

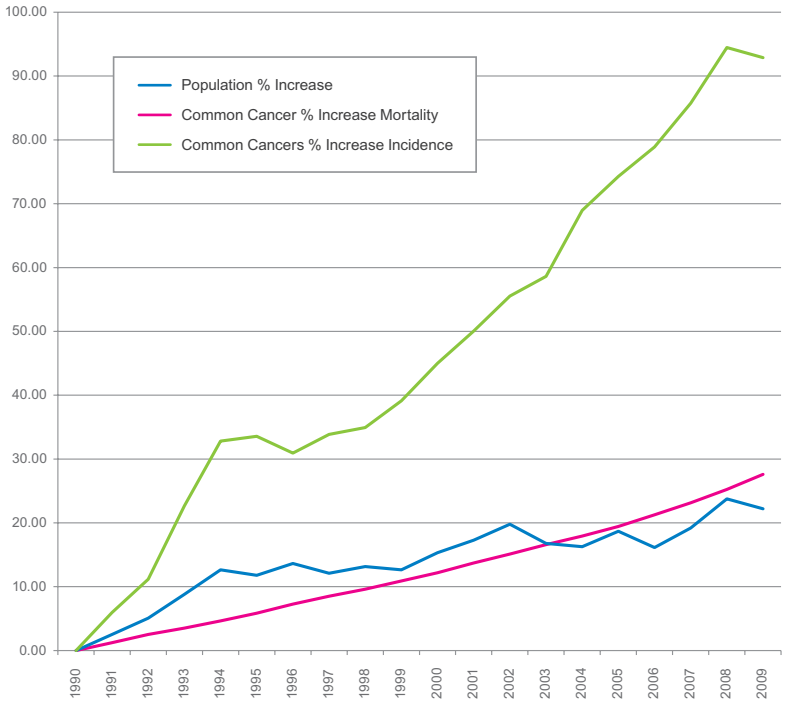
Graph 1 shows the increase in both the incidence and the mortality for RLC cancers in Australia during the period 1990 – 2009 and compares it to the rate of population increase during that time. The most striking aspect of this graph is that both the incidence and mortality rates of rare and less common cancers are increasing at over **twice** the rate of Australia's population growth.



Graph 1: Mortality and incidence of RLC cancers and population growth, as a percentage increase over the 1990 figures

common and rare and less common cancer statistics

By comparison, the graph below shows the same data, over the same time period, for common cancers when compared to population growth. The difference is readily observable; while the reported incidence of these cancers has increased at a rate of over three times the population growth (greater than that of RLC cancers) the mortality rates are significantly lower.

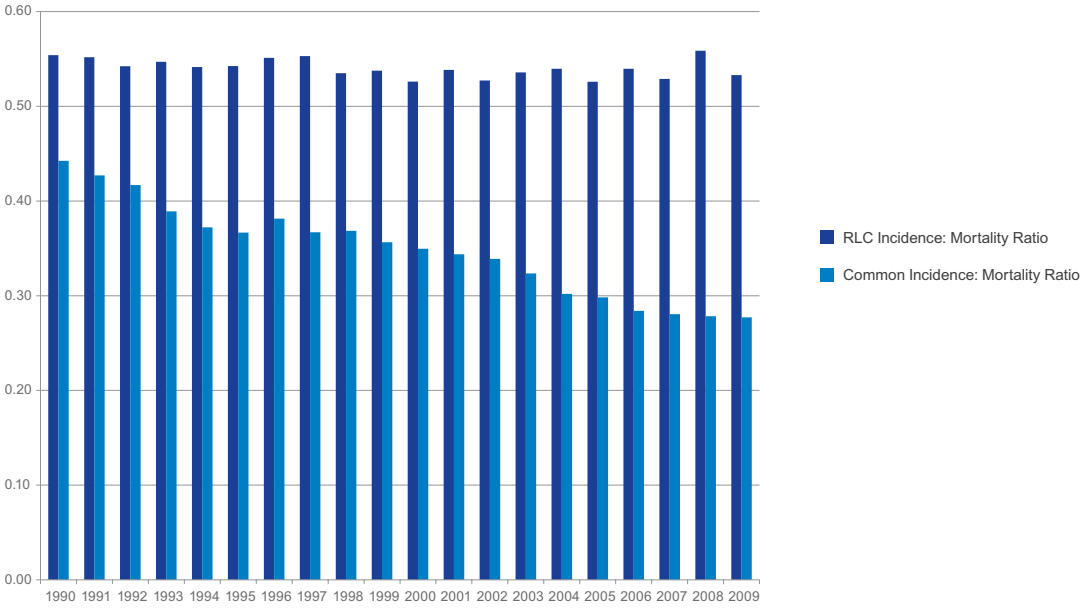


Graph 2: Mortality and incidence of common cancers expressed as a percentage increase over the 1990 figures

*It should be noted that the increase in incidence seen above for common cancers can in large part be attributed to the advent of improved screening for prostate cancer which has led to a 300% increase in diagnosis, and that the rate of increase in mortality from common cancers is also significantly inflated due to a 90% increase in female deaths from lung cancer.

Finally, looking at the difference in the mortality to incidence ratios for each group of cancers it is clear that the patient outcomes and survival are greatly improved for those diagnosed with a common cancer than those patients with a RLC cancer. For example, mortality has increased at the same rate as incidence for RLC cancers, but has steadily decreased for common cancers, therefore the chances of survival are improved for patients with a common cancer.

These overall figures highlight a crisis in RLC cancers compared to common cancers. Although there are similarities between both groups, with the incidence rates continuing to increase at more than twice the rate of population growth, there is a significant difference in the incidence to mortality ratio for common cancers compared to that of RLC cancers. In other words, over the past 20 years we have made advances in treating common cancers but have not noticeably improved outcomes for Australians with RLC cancers.



Graph 3: Mortality to incidence ratio for both common and RLC cancers

what is the difference?

During the twenty years under examination Australia has seen the introduction of screening programs for common cancers (breast, prostate and bowel), awareness programs (lung cancer and melanoma) and significant monies allocated to both research through the NHMRC and treatment through the PBS. The impact of these programs and funding has been significant for common cancers, but in contrast RLC cancers have not been given attention and resources and there has been little or no improvement.

Without similar mechanisms specifically designed to address the prevention, diagnosis, and treatment of RLC cancers we cannot hope to have an impact on mortality or on improving patient outcomes in the future.

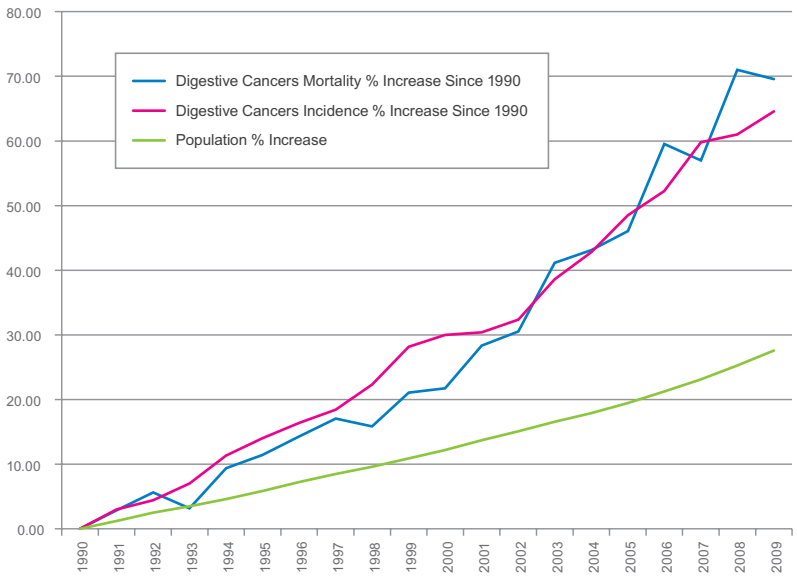
common and rare and less common cancer statistics

cancer specific examples

• *digestive cancers*

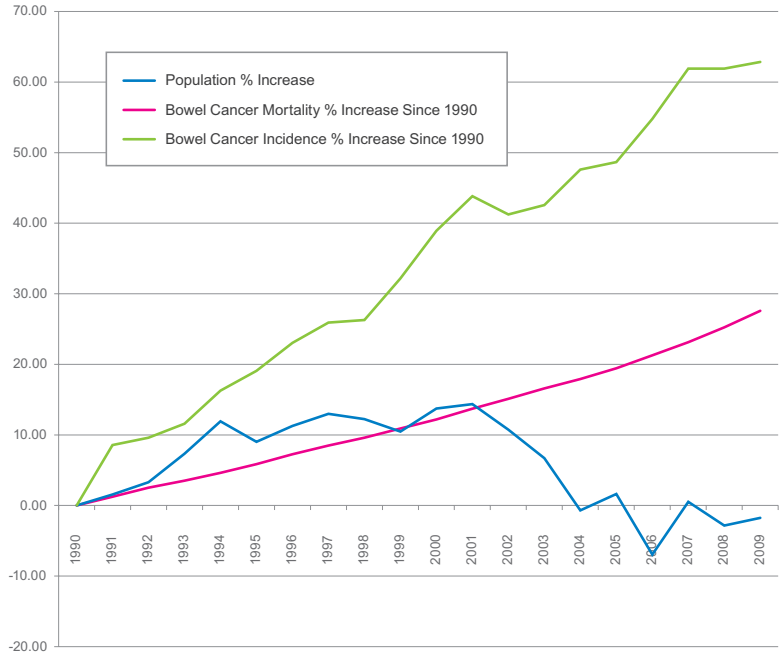
As an example of the impact of targeted programs on cancer we can compare the digestive cancers as a group, to bowel cancer, a common digestive cancer that has received significant support and funding from government. For the purposes of this report the group 'digestive cancers' includes cancers of the oesophagus, stomach, small intestine, anus and anal canal, liver, gallbladder, biliary tract, pancreas and other digestive organs (excluding bowel cancers).

Since 1990 incidence and mortality of digestive cancers other than bowel cancer, have increased by 65% and 70% respectively compared to a population increase over the same period of just 27.4%.



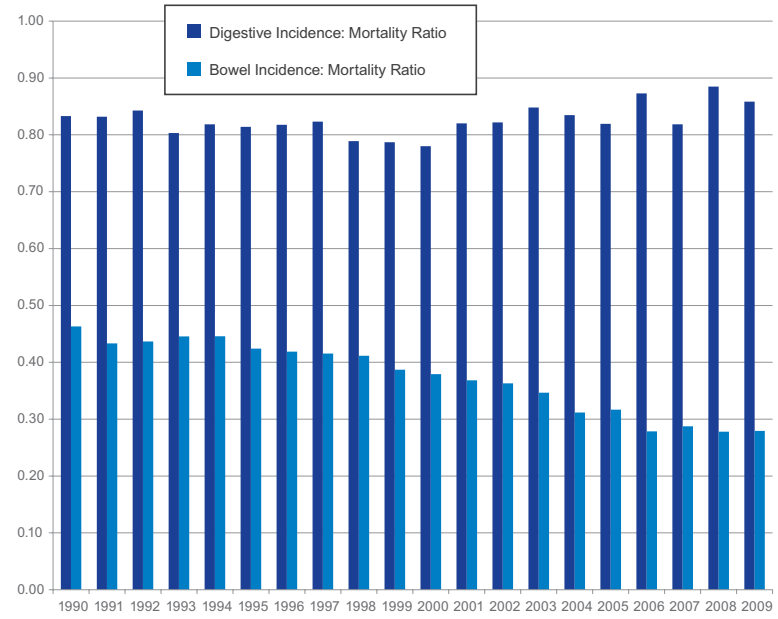
Graph 4: Mortality and incidence of digestive cancers and population growth, as a percentage increase over the 1990 figure

By contrast over the past 20 years bowel cancer (Graph 5), showed an increase in incidence of over 60% but **an absolute reduction in mortality of 2%.** Screening is critical for early diagnosis and improvements in awareness and the uptake of screening in Australia means that we are now diagnosing more patients with bowel cancer, earlier. Bowel cancer is one of the most curable types of cancer if detected early, therefore screening programs identifying new cancer patients earlier, allows treatment to commence, in turn reducing mortality and greatly improves patient outcomes.



Graph 5: Mortality and incidence of bowel cancers expressed as a percentage increase over the 1990 figures

To complete the picture, a similar pattern is also seen for the comparison of incidence to mortality ratios, confirming that the improvements in screening and treatment are having an impact on patient outcomes.



Graph 6: Mortality to incidence ratios for both digestive and bowel cancers

This is a classic example of what can be achieved with screening programs, research and effective treatment programs. The 5 year survival rate of bowel cancer has improved from 48.2% to 66.0% over the past 20 years, a gain of 18% whereas stomach cancer, improved less than 10% (the highest of the other digestive cancers) and now has 5 year survival of 26.7%.

ivana

On the 17 March, 2011, I was flat out at work preparing to go to New York when the hospital called and told me they needed to see me immediately to discuss my biopsy results. I really did not think anything of it. I advised them I would be in after work for the results, however the doctor was insistent and asked that I go immediately. My doctor saw me and I noticed it was odd that he could not look me in the eye. He muttered something that I thought sounded like "You have got cancer." My reaction was one of shock and disbelief, my head started to spin. The official diagnosis was Mucosal Melanoma, a very rare form of cancer. This is the day that cancer came to live with me.

I had to go home to tell my family and cancel my trip. I was advised that I needed urgent treatment. This type of cancer can be fatal and needed to be removed as soon as possible. Telling my husband and children was heart breaking. The looks on their faces was of utter disbelief and shock. How could their mum and wife be sick? I have always been the strong one, their rock. No one slept that night and the next week went by in a blur, doctor's appointments, scans, MRI's, Pet scans and blood tests.

On Tuesday March 30, I was in hospital having a craniofacial and tracheotomy operation. Six weeks later I commenced radiation five days a week for next two months. The operation was a success and they removed all the cancer. Radiation was to ensure the destruction of anything suspect that was not visible to the eye. I was so proud of myself that I went back to work full time within four months of being diagnosed.

In early 2013 I started to get my symptoms back, a PET scan was ordered. My specialist had a devastated look on his face when I went to see him for the results. Mucosal melanoma. I was back on the merry go round, scans, appointments and then surgery again. They were unable to remove all the cancer in the operation and were also unsure how they could treat it. Radiation was a no go, maybe Yervoy, but that was expensive as it wasn't on the PBS. The Oncologists were going to have a meeting to discuss the limited options I had. Without treatment I would have about 6-12 months to live, so this time it was really serious.

It was decided that my only option was to go on a trial drug called PD1. My melanoma was growing at an alarming rate 6 cms in 1 week. My tumour shrunk 52% in the first scan and is currently stable. I will be having treatment for a long time. My life expectancy is uncertain. Having cancer is emotionally and physically draining. There is so much uncertainly. Not knowing if I will see my children marry or have grandchildren, can I plan a holiday? I have already cancelled three. Emotionally, physically and financially having cancer especially a rare one, leaves you with a lot of uncertainty that is very taxing.

common and rare and less common cancer statistics

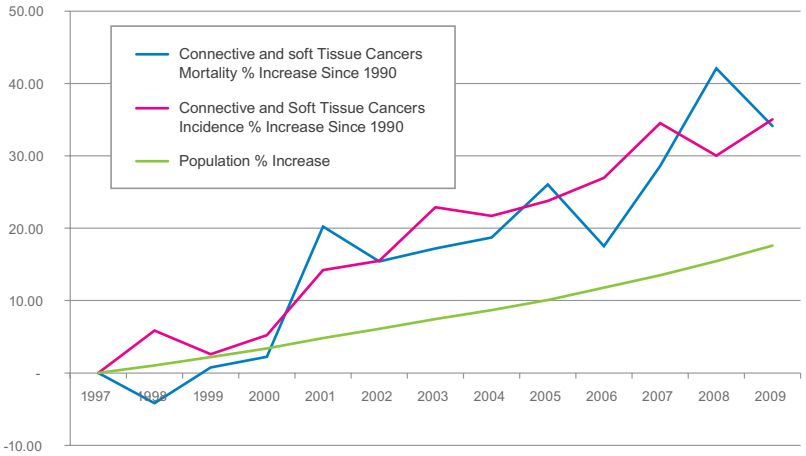
• *eye, brain and central nervous system*

Similar patterns are seen in this group of cancers, which includes cancers of the eye (including retinoblastoma), meninges, brain and spinal cord; while mortality has increased at a similar rate to population growth there has been very little improvement in either survival or the incidence to mortality ratio.

Specifically over the 20 year period the 5 year survival rate in brain cancer only increased by 1.8% and remains at an alarmingly low 21.9%. In other words, only 1 in 5 Australians diagnosed with brain cancer are alive after 5 years and we are as yet unable to improve these outcomes.

• *connective and soft tissue cancers*

A significant proportion of soft tissue sarcomas occur in young Australians and these patients are often confronted with the prospect of radical and debilitating surgery as their best option. These cancer types include Mesothelioma and various soft tissue sarcomas.



**The data here is presented from 1997 rather than 1990 as it is only from that year that the group was established to include Mesothelioma. Prior to 1997 Mesothelioma was included elsewhere in overall statistics.*

As with many other rare cancers this cancer group is increasing at **twice** the rate of population growth and the mortality to incidence ratio is stable indicating that there is no change in overall survival prospects for Australians with these cancers.

Graph 7: Mortality and Incidence of connective and soft tissue cancers and population growth, as a percentage increase over the 1997 figure.

• *female reproductive cancers*

As with digestive and bowel cancers this group of cancers allows us the opportunity to examine the impact of active community engagement combined with government prioritisation and increased spending in research and treatment.

The results are very encouraging. Despite a significant increase in breast cancer incidence (83%) the mortality growth rate of 15% has been much less than population growth. The net result is that breast cancer survival has increased by 17.5% over the period and the incidence to mortality ratio has also improved.

If we compare these figures to other female reproductive cancers, also the subject of substantial community focus, we see that while the mortality rate has also been contained survival rates, in most cases, have increased by less than half that of breast cancer. The significant difference between breast and other female reproductive cancers can also be seen in the areas of research and PBS funding. Breast cancer receives a very high proportion of research funding in Australia and over 10 times the treatment funding through the PBS when compared with all other female reproductive cancers combined.

common and rare and less common cancer statistics

survival rates

Survival from cancer is a key indicator of cancer prognosis, control and treatment.[6] Survival rates for different cancer types are expressed as the probability of a cancer patient surviving for a specific period when compared with the rest of the population. For example, someone with a 90% relative survival over 5 years has a 90% chance of being alive after 5 years when compared to the population as a whole. Population-based survival statistics are essential for monitoring progress in cancer control and highlighting areas of improvement and need.[7]

Survival rates differ significantly between cancers and also between age groups. The survival rates expressed in this report are calculated using the date of diagnosis and provide insight into how effective we are at treating various cancers once they are identified.

Unsurprisingly, the survival rates for many RLC cancers are very low when compared with rates for the more common cancers indicating that we are much better at diagnosing and treating common cancers.

A glance at the improvements made in survival rates across a selection of cancers over the past 20 years can be seen on the opposite page.

As can be readily seen the rare and less common cancers have shown only small improvements over this period, with the exception of blood cancers. Advocacy for increased research and treatment for blood cancers has been highly successful resulting in these cancers being well funded and delivering significant increases in survival.

The common cancers have also had dramatic increases in survival, particularly breast, prostate and bowel cancers. Lung cancer has had neither significant research nor treatment funding during this period and perhaps not surprisingly there is little increase in survival. There have however been significant investments in education around smoking and preventative initiatives that have had a positive impact on male mortality however this has not impacted female mortality, which has significantly increased during this period.

6 Australian Institute of Health and Welfare 2012. Cancer survival and prevalence in Australia: period estimates from 1982 to 2010. Cancer Series no. 69. Cat. no. CAN 65. Canberra: AIHW.
7 Australian Institute of Health and Welfare 2012. Cancer survival and prevalence in Australia: period estimates from 1982 to 2010. Cancer Series no. 69. Cat. no. CAN 65. Canberra: AIHW.

Cancer Type	5 Year Survival Rate % for period 1982 - 1987	5 Year Survival Rate % for period 2006 - 2010	Change in % Survival Rate between 1987 - 2010
Bladder	67.9	57.5	-10.4
Lip	93.4	92.5	-0.9
Mesothelioma	5.5	6.2	0.7
Larynx	63.2	64	0.8
Brain	20.1	21.9	1.8
Pancreas	3	5.2	2.2
Chronic Lymphotic Leukemia	70.3	72.6	2.3
Cervix	68	72.1	4.1
Melanoma	85.8	90.7	4.9
Lung	8.7	14.1	5.4
Oesophagus	9.8	16	6.2
Testis	90.7	97.6	6.9
Uterus	74.7	82	7.3
Liver	7	15.5	8.5
Stomach	17.2	26.7	9.5
Gallbladder	10.1	19.9	9.8
Unknown Primary site	5.6	15.7	10.1
Ovary	32.4	43.3	10.9
Thyroid	83.5	95.8	12.3
Tongue	48.2	61.5	13.3
Acute Myeloid Leukemia	9.9	23.6	13.7
Hodgkin Lymphoma	71.5	87.2	15.7
Myeloma	26.3	43.4	17.1
Breast (Females)	71.9	89.4	17.5
Bowel	48	66.2	18.2
Non Hodgkins Lymphoma	46.6	70.6	24
Kidney	47.4	71.9	24.5
Prostate	58.2	92	33.8

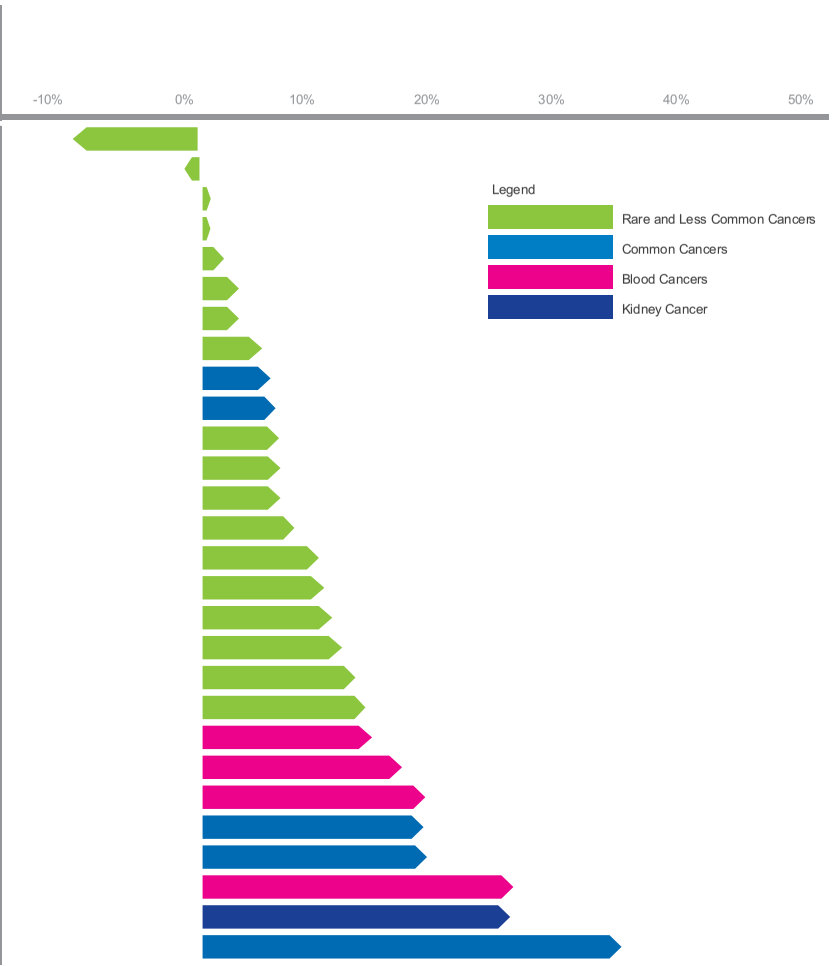


IMAGE 1: Survival trends at a glance. Adapted from AIHW Survival Trends at a glance, Australia, 1982–1987 to 2006–20108

**A note on interpreting IMAGE 1: Each cancer is represented as an arrow where the base of the arrow aligns with survival percentage as it was in the period 1982 – 1986. The tip of the arrow aligns each cancer's survival percentage as measured during the period 2006 – 2010. Consequently, we can see that the horizontal positioning of the arrow correlates to each cancers survival (lower survival are to the left) while the length of the arrow indicates the improvement in survival from 1986 to 2010.*

***Only bladder cancer showed a significant decrease in survival, which may be partly related to changes in coding and age at diagnosis.Kidney Cancer is highlighted in purple- as it is a less common cancer that has benefitted from the introduction of new funded treatments in recent years. It is also possible that changes to bladder cancer coding may have also positively impacted Kidney Cancer survival statistics.*

8 Australian Institute of Health and Welfare 2012. Cancer survival and prevalence in Australia: period estimates from 1982 to 2010. Cancer Series no. 69. Cat. no. CAN 65. Canberra: AIHW.

renee

My name is Renee. I was diagnosed with brain cancer – Anaplastic Astrocytoma (grade 3 Astrocytoma) in 2010, 12 weeks after my husband left me. I was 36 and my children were aged 3 and 4.

I encountered a lot of unexpected expenses before I was finally diagnosed. I had five MRI scans and several specialist appointments, including interstate travel as I was unable to get into a neurosurgeon locally (due to long waiting lists). By the time I went to surgery I was already more than $2000 out of pocket after Medicare and private health insurance rebates. This was a huge strain as I was no longer working and my ex-husband was not contributing to the mortgage.

My craniotomy to remove the tumour went well and the pathology returned as Grade 3 Astrocytoma. My neurosurgeon advised that the standard post-surgery treatment for my type of cancer was 6 weeks of daily radiation, followed by 6 months of chemotherapy (Temozolomide).

I came through the radiation relatively well, though I then had to face the chemotherapy stage of my treatment. My medical oncologist advised that Temozolomide was not available to me under the Pharmaceutical Benefits Scheme (PBS). While Temozolomide was approved for grade 4 astrocytomas and recurrent grade 3 astrocytomas, it had not yet completed phase 3 trials for grade 3 astrocytomas, which are relatively less frequently diagnosed in Australia each year. My medical oncologist advised that my treatment would cost around $3000 per month and I would require 6-12 months of treatment, costing just under $40,000.

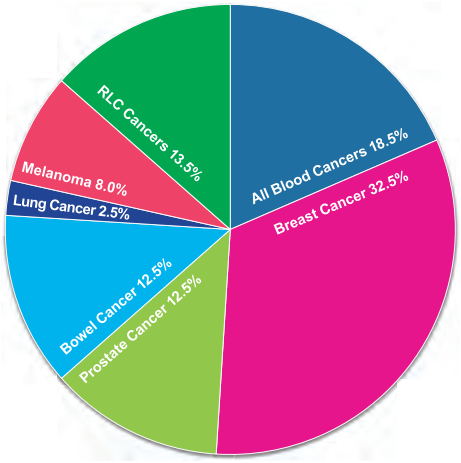
At that point I had no method of raising that money. My situation with my ex-husband did not allow me to borrow against any equity in our home, nor could I sell my home. I contacted the drug company to apply for compassionate funding, which did not happen. I cannot overestimate the trauma my whole extended family felt through this period. Aunts, uncles and grandparents were asked to dip into retirement savings. At the twelfth hour, my treatment was funded by the Canberra Hospital Foundation Fund, so I only needed to pay around $200 each month. The whole process of trying to raise money from extended family and friends was extremely stressful, humiliating and ultimately unfair.

In 2011 I spoke on behalf of the Brain Tumour Alliance Australia, at a Senate Committee into PBS funding. I strongly believe that being diagnosed with a rare cancer does not mean I should be denied best standard treatment, seemingly just because of some arbitrary red tape.

funding cancer research, treatment and care

research

Australia is a leader in cancer research and invests over A$1bn each year through public and private sources. An audit conducted by Cancer Australia in 2005[9] provides some indication of the focus and direction of tumour specific research in Australia at that time; Cancer Australia are expected to release a study of 2012 funding in the near future.



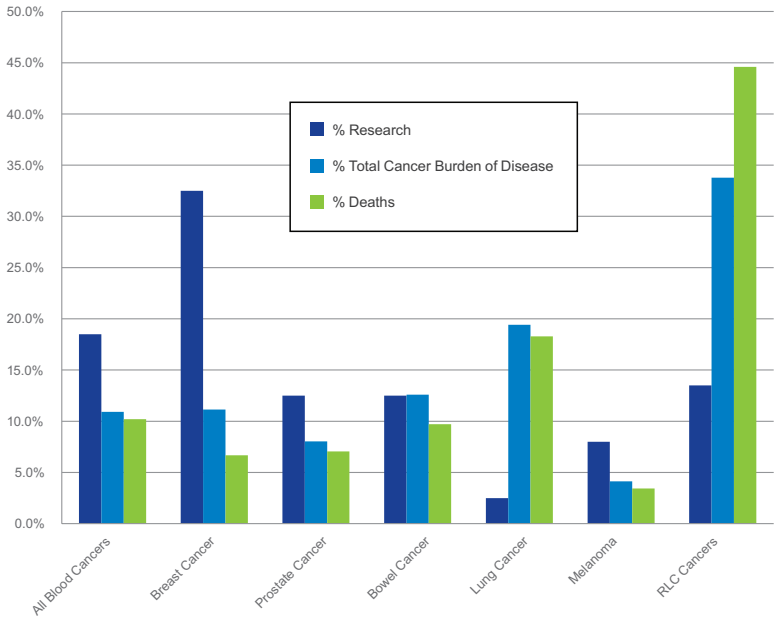
Graph 8: Pie chart representing the breakup of research funding by cancer type (2005 National Audit).

Anecdotal information from researchers suggest that while there may have been some changes in these ratios with more funding going to less common cancers such as pancreatic and cancer of unknown primary, there is still a dearth of funding provided to the vast majority of RLC cancers.

the impact of research funding

When we look across the spectrum of cancers it is clear that a correlation exists between research spend, burden of disease and mortality. While Australian research is only a small part of all global cancer research, with the possible exception of melanoma, there is no reason to believe our overall focus on common cancers would not be replicated throughout the global research community.

Lack of research into RLC cancers has two direct impacts; the first and most obvious is that without research, there is no likelihood of improved treatments and potentially cures, **the second and perhaps less obvious, is that without research we will not develop the knowledge to design screening tests or early diagnosis mechanisms.** Early diagnosis is highly significant in improving patient survival and our experience, as seen in many of the patient stories, is that many Australians with RLC cancers had their outcomes compromised by late diagnosis.



Given the neglect of rare and less common cancer research when compared to burden of disease and mortality we must take action to encourage the research community to increase activity related to these cancers.

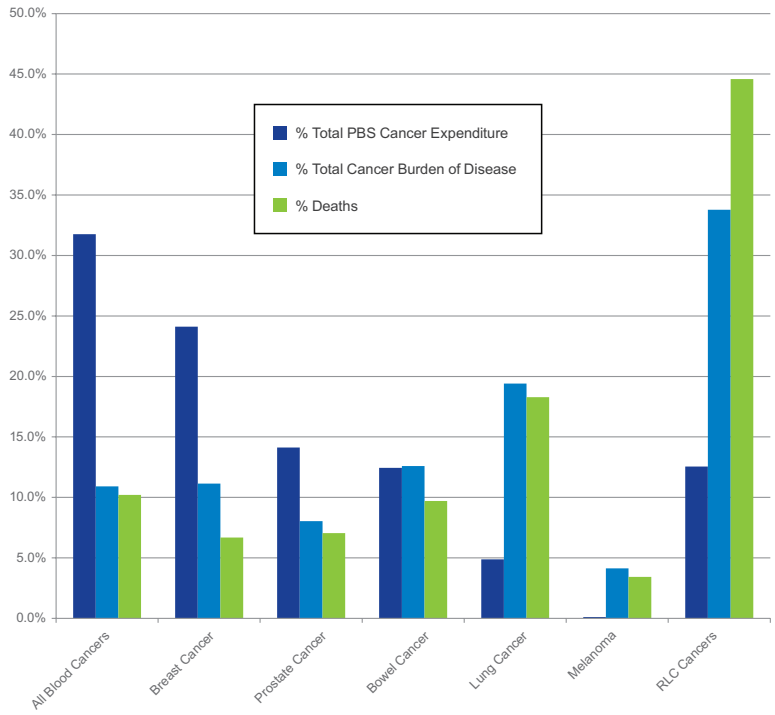
Graph 9: Percentage cancer research expenditure (per annum) vs the percentage burden of disease and deaths

9 Cancer Australia 2005. Cancer Research in Australia: An overview of cancer research projects and research programs in Australia 2003 to 2005.

funding cancer research, treatment and care

treatment funding for rare and less common cancers

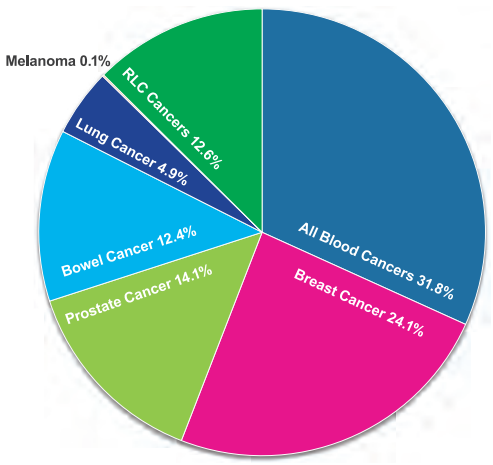
Rare and less common cancers account for almost 50% of all cancer deaths and yet funding for these cancers for both research and treatment does not reflect their true burden. Examining the funding provided to each of the main cancer groups through the PBS shows that the proportion of funds allocated to RLC cancers is significantly lower than that allocated to common cancers. This is most readily seen when comparing the percentage of funds allocated to each cancer type to their burden of disease and mortality. The support for all cancers is important and of significant value to patients; we must now urgently address the growing need and priority for rare cancers.



Graph 10: Percentage PBS cancer expenditure (per annum) vs the percentage burden of disease and deaths associated with each cancer.

Despite RLC cancers causing 33.8% of the burden of disease and 45% of all cancer deaths, they only receive 12.6% of PBS cancer funding. As an example, cancers of the eye, brain and central nervous system cause over 1,200 deaths in Australia each year but there is a very small allocation of PBS spending to approved treatment (approx. $20 million per annum).

As we have seen (graph 10) the cancers that have previously had a low survival but have improved significantly over time are the blood cancers, particularly the myelomas and lymphomas. It is these cancers, together with the common cancers that have the most significant spend through the PBS.



Graph 11: Breakdown of total PBS cancer expenditure (in YEAR 2012 Est. $1 billion) by cancer type.

For a number of reasons, including lack of patient advocacy, media awareness and political pressure, the research, drugs and treatments available to Australians with a RLC cancer are funded at a much lower level than the common cancers and in too many cases, no treatments are available at all. As a result of this funding discrepancy Australians with these cancers are forced to rely on the older, more damaging cytotoxic chemotherapies often without high level evidence of benefit rather than having access to new targeted, more specific therapies which would dramatically improve patient outcomes if currently matched to the cancer in question.

bruce

My name is Suzie Ward and in September 2012, my husband Bruce, died of several cancers. One of which the doctors had never seen before. Married for 38 years, wonderful father to David and Andrew, father in law to Camilla and Sally (who lost their fathers when they were too young) and grandfather to Olivia, Lucas, Daisy and Kingsley, who was born a few weeks prior to Bruce's death (a little joy in our lives during a horrendous few months).

In May 2012, his health and wellbeing began to deteriorate very rapidly. Our nightmare had begun. After a blood test, scan and colonoscopy we realised that our world was about to be tipped upside down.

Our surgeon stated that he could not operate on the rectal cancer until it had been shrunk by radiation. A visit to the radiation oncologist told us that Bruce should start treatment immediately. The cancer was at stage 4. In hindsight, we never stood a chance but we were not going down that track, we were going to find a way forward, researching all alternatives to fight and rid Bruce of the cancer that had unfairly invaded his body.

At the same time as the radiation, Bruce was on oral chemo daily and once a week, was given chemo intravenously. There were several trips to emergency sparked by frightfully high temperatures and heart rates. It was during this horrific time that they realised, the spot on his liver (a secondary cancer) had grown massively. An operation was scheduled immediately to remove 80% of his liver.

Post operation they realised that the mixture of chemo given was antagonistic to another secondary cancer they had discovered in his anus. This was a rare cancer, one they had never seen before or had any experience with.

During these months (less than 4 months), Bruce experienced horrific pain. One cannot imagine what he went through despite the efforts to get the pain under control. It was just not happening. He was never in the running to get on top of either cancer. These cancers defied all knowledge for all the excellent care that he was given. Bruce believed very strongly in the importance of getting to the root cause of a problem. This was ironic as during this period we were constantly fighting symptoms and never got to the bottom of why he had these two hideous cancers.

This was a fight that we as a family were never going to win. It beat us at every turn emotionally and physically. As a family we went through every agonising day with him. Financially it drained us.

In his own words, Bruce wrote the following note to a friend:

> *"…this cancer has decided to be difficult. The two specialists/surgeons sat on my bed at 6:00 this evening and with tears in their eyes told me they have never seen a cancer behave like this one. The bottom line is that in all probability it will take me out by this weekend".*

We have to do better. There is a huge amount of funding going into fighting cancers of all types and they are making great inroads with the funding that is being distributed, but we believe very strongly that there needs to be more dollars directed towards finding out the root cause of these rare cancers and if doing autopsies will enable researchers to be better informed then this must be the way forward. Please do not let other families endure what we have had to.

ian

Hope, love and horizons and a thank you to all the oncology,endocrine staff at RPA and my family and friends. Hope and good is easier to believe in when you have people around believing in it too. The story of Ian,Vicky (50) , Ella, (20) Phoebe (16) and Hetty (13).

If we could take one common thread in our journey so far, it would be hope. At almost every stage, we have had hope. The hopes of course have been ever changing with our altering horizons but we have never given up hope, we always see it in some form or another.

14th May 2012 was a life changing day. A day when life lost its security and its certainty. We were without a map and the compass no longer worked. Ian came home and said they were almost sure that he had thyroid cancer. Not so bad, thyroid cancer is very curable and we should be hopeful as we knew. We waited for the biopsy results. The result came in. Ana-plastic Thyroid cancer, the doctor was realistic and gave us her opinion on his prognosis. 3-5 years, we thought he could beat this, they are wrong we told ourselves we would show them.

We were hopeful that they had made the wrong diagnosis. We told the children, we encouraged hope. We told the truth. This was going to be a battle and we all had to fight. It was like we all got on a boat together and left the safety of shore. We were no longer like other families, we were out in an unfamiliar sea.

They removed the tumour, they took the lymph nodes, they did a PET scan there was no more cancer in his body. We were hopeful for a cure, thinking they had got it early.

Chemotherapy, powerful radiotherapy and one determined patient, the odds were surely in our favour. He could overcome this. It would be a terrible year, but we would look back and it would something that had strengthened our family and brought us closer together. We were hopeful. Ian had a life to live beyond this. We had a life to live with him.

Chemo and radio came and went. Family life strained, tempers were short, but hope was always high. The cough arrived, nothing much that worried any of us. He always got a cough at this time of the year. The cough stayed. The X-ray showed pneumonia. A few days in hospital, some antibiotics and he would be home again. Though in actual fact, it was a few weeks in hospital, a lot of pain and a new diagnosis. It had metastasised to his lung. We were all very frightened, but we were hopeful that this expensive new drug would give him 18 months or more. We hoped for time and our horizons altered.

He came home and we just coped with his friend and his enemy morphine, with the one hand it took away the pain and with the other it took away Ian, it took away Dad. The sense of humour disappeared and sadness replaced it. We were hopeful, he would respond to the drug and the cancer would shrink and the pain would subside. The scans in December showed a 50 percent reduction in the cancer. We were right to hope! We could have a good Christmas. Christmas crept closer, so did the pain in his back. We thought he just had a bad back. All the tests came back with nothing. We could be hopeful for good Christmas. We tried to have the best Christmas we could, this wasn't his last, we hoped.

The CT scans showed the cancer had been having a good Christmas too. It had wrapped his lung and not in paper. They kept him in, I tried to be hopeful and then I realised I had to change my horizons and be hopeful for only the four of us. The horizon for him was a different kind. I hoped now for him that I would be with him when he died and that he knew we loved him forever. I hoped my children would survive this. I hoped that I was up to this parenting task.

He died on the 24 January 2013, in my arms and with his twin and cousins beside him. I hope with all my heart that he knew he was loved forever. My children are brave, strong and supportive. I now hope that from all of this sadness, loss and love, something will blossom. Yes I am still hopeful; there will be a cure, some time in the near future.

conclusion

It is 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. We must now make changes to the way we approach cancer prevention, diagnosis, treatment if we are to impact the outcomes of these rarer cancers, which includes over 150 different cancers.

Research has shown that the allocation of resources to research and funded treatments through the PBS positively impacts survival of cancer patients. The lack of focus on RLC cancers manifests in poor survival and consequently much higher mortality.

Addressing the discrepancies for RLC cancers compared with common cancers needs to occur at the highest level, and we need the Australian Government to take action. We need to improve outcomes for rare and less common cancer patients and to do so we must review existing mechanisms and improve research, diagnostics and access to medicines for rare and less common cancers.

Without similar mechanisms created for tackling common cancers, i.e. those specifically designed to address the prevention, diagnosis, and treatment, we cannot hope to have an impact on mortality or on improving patient outcomes for RLC patients in the future.

It's time to act and we urgently need:

- Improved research to identify new approaches for screening and treatment;
- Improved screening for early and correct diagnosis;
- Increased availability of PBS funded treatments; and
- Equal access to care and treatment regardless of cancer type.

Rare and less common cancers present great challenges to our health system. Each disease is complex, unique and potentially lethal. Together they represent one of Australia's largest and growing health problems. Our experience with common cancers gives us hope and confidence that with government and community focus we can have a major impact. These cancers manifest themselves in aggressive and painful disease and we owe it to our fellow Australians to do everything we can to give them dignity and support and most importantly at the very least "just a little more time".

appendix 1

rare and less common cancer codes

Rare and less common cancer group	ICD – 10 Codes
Malignant: lip, oral cavity and pharynx	C00–C14
Malignant: digestive organs excluding colon and rectum	C15–C17, C21–C26
Malignant: respiratory and intrathoracic organs excluding lung and trachea (1)	C30–C32, C37–C39
Malignant: bone and articular cartilage	C40–C41
Malignant: skin excluding melanoma	C44
Malignant: mesothelial, connective and other soft tissue (1)	C45–C49
Malignant: female genital organs	C51–C58
Malignant: male genital organs excluding prostate	C60, C62–C63
Malignant: urinary tract	C64–C68
Malignant: eye, brain and other parts of central nervous system	C69–C72
Malignant: thyroid and other endocrine glands	C73–C75
Malignant: ill-defined, secondary and unspecified sites	C76–C80
Malignant: haematological excluding non-Hodgkin lymphoma	C81, C88–C96, D45–D46, D47.1, D47.3
Malignant: independent (primary) multiple sites	C97

Cancers considered as common and therefore not included in this report other than for comparative purposes are

ICD 10 Code	Cancer Description
C18 – C20	Bowel Cancer
C33 – C34	Lung Cancer
C43	Melanoma
C50	Breast Cancer
C61	Prostate Cancer
C82 – C85	Non-Hodgkins Lymphoma

appendix 2

anna

My name is Kate and my mother's name is Anna.

In February 2008, my mum complained of severe pain in her abdomen. After rushing her to the emergency department we were told she had a blocked bile duct and gall stones. Her gall bladder was removed, a stent inserted, and we took home a nice container of pebbles.

A couple of months later, mum got sick again. She turned yellow with jaundice. By the end of that month she could not keep food or liquids down and her stomach became very distended. She was told she had a blocked duodenum. This was operated on but did not fix the problem. She was soon returned back to hospital and was then diagnosed with pancreatic cancer. She immediately underwent a Whipple's procedure. Mum started the usual chemo regimen of Gemcitabine and 5-FU.

During a break from chemo in January 2009, our family went on an amazing overseas trip to the UK and Europe. We visited family in England and then travelled to France, Switzerland and Italy. As wonderful as it was, there was always the underlying knowledge and sadness that this was the last time we would be doing these things with mum.

Life continued on upon our return. Mum sought alternative medicine to complement the traditional methods. She tried massage, Reiki, and meditation. Mum and dad also attended a week long cancer retreat. She told us about the "Rainbow Ritual" that she had learned. Every time I see a rainbow now, I know mum is there.

Tumours were later found in her liver. She decided not to continue with any more chemo. I sent her scans to top medical centres in France, The Netherlands, the UK and the US. We were offered no hope. In June she underwent surgery in Sydney to have radioactive beads inserted into the tumours in her liver. A new television show on radical medical procedures had gone into production and a television crew started following our story. It was great distraction for mum. It never went to air. I have a copy of what was filmed, but cannot bring myself to watch it.

The beads made her very tired and mum continued to decline. Our visits from Brisbane to Newcastle became more regular. On each visit we could see an obvious decline. Having two friends who also lost a parent each to pancreatic cancer, I was well versed in the signs to watch. Her liver became enlarged and she became very jaundiced. Her feet became swollen and started to turn purple. In mid September we visited again.

The decline in mum since the two weeks prior was startling. Both my brothers and their partners came to stay that weekend too. I think mum had been waiting for all of us to be together. That weekend I told her that everything good in me came from her. I'm not sure if she understood or if she was just being cheeky but her response was "I know".

Mum passed away in her beautiful home, which was filled with family and love, in the early hours of September 21, 2009. She was 62 years old. The period of time from when she first fell ill to when she passed away was 19 months.

The years since have been a struggle for me. My partner and I got married in 2011. We have since been trying, without success, to have a baby. These are times when a girl needs her Mum. There is nothing I wouldn't give to have her by my side. Losing her has been the most amazing lesson in life. My one goal now is to make her proud.

avner

My husband Avner started to feel unwell from the day we moved into our new home. Over the next couple of months he experienced pain, back ache, and was diagnosed with pneumonia. His GP initially sent him to a chiropractor, suspecting he had hurt himself during the move. When it became clear that this was not the cause, he sent him for a series of tests. It was a frustrating time as there was definitely something wrong, but the tests were not showing it. One last test was ordered by a specialist.

The test came back and gave us our first indication of what was to come. Avner was sent for a biopsy on his liver, the first one was unsuccessful so a second one was required. On the 28 September, Avner got a call and found out he had incurable/inoperable pancreatic cancer. We were in shock as we tried to come to terms with this diagnosis. We hardly knew what the pancreas did, let alone that the pancreas is the organ of the most lethal cancer (relative to how many people are affected by it). How could it be? We believed that Avner led a healthy life – was active, and had a balanced diet with lots of fruit and vegetables. He drank, but never to excess and did not smoke.

It took a couple of weeks to see an oncologist, but the care shown and the competence the oncologist had, made the wait worthwhile. Avner began chemotherapy (gemcitabine) and almost instantly this helped and the pain eased. We took back control of our lives and enjoyed time with friends and family. Avner even got to pursue his passion for photography. It was in a way, the most magical time – getting to spend so much time together, enjoying the simple pleasures of life and really getting to see the incredible kindness and goodness in so many people.

Sadly the chemo became resistant to the cancer and so the treatment was changed to a second type of chemo. This was not as easy to tolerate and was more of an intrusion on Avner – being hooked up to a pump for several days at a time. It did however, continue to help with the pain.

After a short few months, Avner again began to experience pain and had severe problems with constipation. This second round had also become resistant to the cancer. He had to go into hospital where they did everything possible to try and control the intense pain and get the constipation fixed. Nothing worked. Avner's oncologist suggested that a third round of chemo might help. He explained that most oncologists only gave one round of chemo for pancreatic cancer, few gave two rounds and it was a very rare occurrence for a third round to be given. Chemo had helped Avner's pain when nothing else seemed to, so we agreed to give it a go. It worked, the pain eased and the constipation was fixed. Just like the other two types of chemo, this one also became resistant. There were no other options, but to try and manage the pain as best as possible.

In spite of his weakened state, Avner was keen to do something to help right the wrong that had been the history of pancreatic cancer over the preceding decades - with barely any improvement in survival rates over the last 50 years. With friends and many great supporters, we set up the Avner Nahmani Pancreatic Cancer Research Fund (which became the Avner Nahmani Pancreatic Cancer Foundation in July 2010). Avner died on 30 October 2008, he didn't get to see how this has grown, but would have been amazed and humbled by it all. I miss Avner so much, he was the love of my life, but I am so happy to be doing something to try and make a difference.

jane

I went to a yoga session in October 2011 (when I was 31) and then in the following days I felt pain in my right side. I thought perhaps I had pulled a muscle but after a few days, the pain got sharper so I went to a GP. A mass in my pelvic area was identified measuring 18cm x 16cm x 10cm. The GP said that it was most likely a cyst and an ultrasound was ordered. The results of the ultrasound suggested a fibroid, a mass that is often benign. At this point I called my mum, not having any family in Melbourne, she immediately booked a flight over from Adelaide and we started the process of arranging surgery with a gynaecological surgeon.

I got an MRI done the following day and surgery was scheduled. However a couple of days before the scheduled operation, the surgeon called and told me that the surgery was cancelled. The MRI scan revealed that an oncologist was required as it may be cancerous.

Surgery was lined up for a date that was about two weeks away (about three and a half weeks since the initial GP appointment). This was a tough wait. I wanted the melon sized and possibly cancerous tumour inside me removed quickly. Further, the pain was increasing and escalated to the point where I went into emergency two days before the scheduled operation and was operated on a day before the scheduled date.

Going into surgery it was discussed that depending on what they find I may have to lose my fertility which was daunting as I had not had any children yet. The tumour was encapsulated and the entire tumour was removed in one piece. It was confirmed then and there that it was cancerous. Along with my left ovary and fallopian tube, they also removed my appendix and some lymph nodes to be safe.

In mid January of 2012, after all the different tests and scans to search for a primary source, we got a final diagnosis (3 months after the initial GP appointment) of a rare Mucinous Ovarian Cancer. As it was encapsulated and removed intact, there was no spread and likely to be a surgical cure, for which lucky is an understatement! Chemotherapy was not required.

The oncologists had advised I should think about family planning sooner rather than later and referred me to the fertility clinic. Unfortunately I have a low ovarian reserve whether this is due to the cancer, surgery or just unrelated is uncertain. My husband and I are embarking on IVF. The oncologist also advised that later on down the track I should consider an early hysterectomy to be on the safe side.

I have since been asked to be part of Cart-wheel, a rare cancer study. I was very happy to be involved and help contribute to better outcomes for people diagnosed with this rare cancer.

I had a lot of support from family and friends. Cancer has such a profound effect on family and together we have shared many highs and lows throughout this process. I live with worry, with uncertainty but certainly feel blessed and lucky that I remain cancer free for two years.

jenna

After finishing high school in 2006, I started feeling vaguely unwell, this continued for 3 months, worsening over time. In March 2007, I was diagnosed with a rare liver cancer called Fibrolamellar Hepatocellular Carcinoma. I had physically deteriorated to a mere shadow of my former self. At the time of diagnosis, I had been at uni for a number of weeks and decided to defer my studies until I had recovered. I was told this cancer was advanced, that it usually strikes young adults and that it had been growing undetected in my liver for many years. The 15cm tumour in the left lobe of my liver had squashed my stomach and I required urgent surgery to remove it. Once diagnosed, things moved very quickly which thankfully did not allow me to ponder all the possibilities.

On the 3 of April 2007, the day before my 18th birthday, I underwent major abdominal surgery to remove the tumour and was told afterwards that lymph nodes were also removed and they had tested positive for cancer. I was surprised at how long and painful the recovery was. My liver specialist referred me to an oncologist who advised that chemo was not an option. Liver cancer was a surgical disease and usually does not respond to treatment.

In September that year, a tumour was found on the right side of my liver, two spots on my right lung and several affected lymph nodes in my abdomen were discovered. This was devastating. I underwent abdominal surgery for a second time in November 2007. I was then referred to an oncologist and was prescribed an oral chemo drug. As it turns out, I was severely allergic to this drug and had to stop taking it. In August 2008, I started another oral chemo which kept the disease stable. In early 2009, I had two surgeries on my lungs to remove metastases but my liver has remained clear since my second liver surgery.

In 2010 I resumed uni part-time, I am now back studying business full-time and am enjoying life. Thankfully in March of 2013 my oncologist took me off treatment as the disease had remained stable for so long. Since then my energy has returned and I am feeling as healthy as ever. Even though I continue to have tests and scans on a regular basis, I really appreciate life and feel that I have overcome something that a lot of people in the medical profession doubted I would. I am forever grateful to the doctors and nursing staff who have taken care of me over the past seven years and all those who continue to. What I have gone through has really opened my eyes as to what people with rare cancers have to deal with in relation to limited options for treatment and the lack of research for these cancers.

john

My journey with cancer started with a trip to an emergency centre in Adelaide for a totally different illness. After feeling very ill back in 2007, I decided to go to the emergency centre at Flinders Medical Centre to seek medical assistance. During this visit, a number of tests were conducted over a 3-4 day period. An ultra sound over my abdomen revealed a large mass on my right adrenal gland.

I was informed that there was only a 1 in a million chance that the mass was malignant. After a number of further tests, it was found to be more than likely pheochromocytomas, a common benign tumour found on the adrenal gland. I was told not to worry and that a CT scan would be arranged to assess the growth of the tumour. At this stage no surgery was required.

While I was told not to worry, I did. I had a growth in my body. What was this all about? How did this occur? Was it a cancer? The CT scan was conducted some six months later. It revealed that the tumour had grown considerably and required surgery.

I was sent to a surgeon that specialised in treating patients with breast cancer. So I sat in a room with a number of distressed ladies with their partners, husbands and family members. At this stage I had no inkling of what the future would hold. I was going in for surgery to remove a benign tumour. In hind sight, I should have been very worried about the future. The majority of the women in the waiting room, if not all, would have a full recovery and their cancer would not recur.

After the operation and a test of the mass, it was revealed that I had won a lottery that I did not want to win. I was one of those unlucky few diagnosed with a rare cancer. At the time I was thinking that the cancer would be cured, that life would go on as normal. After all with modern science, the majority of cancers are curable these days. I was sent to an oncologist who informed me that there was no cure for the cancer I had, that there was a 20% chance of living for another 5 years. I remember going into a cold sweat. I think panic set in, there was no compassion just a statement, mate your days are numbered.

The future was uncertain, there was going to be a lot of emotional strain and financial consequences. I have not worked for the last 2 years and with the current diagnosis will most probably not go back to work. At the time I did not appreciate the gravity of the situation. It was all very surreal. I did not understand the difference in research funding for rare cancers compared with the more common cancers and the impact this was to have on our future.

As I write this I feel well but I know that currently there is no cure for this cancer and that I possibly do not have much longer to live, unless through participating in a trial, I am lucky enough to be cured through some ground breaking research. Research, such as that of Professor Stan Sidhu of Sydney North Shore Hospital. Unfortunately though, this research is at a standstill due to ethical questions. This has been the case for several months.

In May, 2010, I had a relapse. Cancers were removed by surgery, only to reoccur again in October 2011. Following this diagnosis I participated in a trial in Melbourne at the Austin Hospital. I was on the trial for 12 months, the treatment slowed the growth of the cancer for several months but unfortunately the cancers grew substantially over the last few months of the trial and I was subsequently taken off.

In summary, my journey with this rare cancer, along with my family has been one of frustration and a feeling that the experts are hindered by the lack of research that has been done on this cancer. In saying this, there are a number of people doing ground breaking research in this area, but are hindered by the funding that is available and the bureaucracy that exists. I live in hope that a cure can be found in the near future.

margaret

My name is Margaret Emery and I am from Highbury, a suburb of Adelaide, in South Australia. This is my story.

In 1989 I was asked by the Oral Surgeon treating my son, then 14, for cyst and teeth problems, if I would take him to the Women's and Children's Hospital in Adelaide to assist with research. I can vividly remember walking into the Professor's room and him telling me "You have Gorlin Syndrome". Hey, wait a minute, I am here for my son, not for me. My large head and facial features were clear evidence for him to tell me the news. After extensive tests and x-rays, it was confirmed both of us had the condition. What a wonderful 40th birthday present!

At the time I was diagnosed, I had just started seeing a dermatologist about a rash that had developed on my legs. Yes, they were skin cancers. Appointments with my dermatologist, spanning 23 years, has seen me have every kind of treatment possible and the number of skin cancers removed would be in the thousands. I also go to an eye specialist where I have had numerous cysts removed from my eyelids, and he keeps a check on my eyes. I have regular pap smears and internal ultrasounds, but thankfully to date, have not had any problems.

About three years ago I had a large cyst on my lower jaw. My dentist referred me to a specialist, who believed that the growth had probably been lying dormant for about five years and had gone undetected. I have had the cyst removed and regular check ups have revealed the healing process was successful and there have been no further jaw cysts.

As there is supposedly no history of skin cancers or jaw cysts in my family history, I have been classed as the carrier, passing it on to my son. I am very fortunate to have a fantastic team of medical people assisting me on my journey with Gorlin Syndrome. I also cherish my Adelaide circle of Gorlin friends and value their encouragement, inspiration, humour, courage, support and friendship.

mark

I have a very large tumour located mostly on the left side of my brain which affects my speech and movement on the right half of my body as well as my balance. I was diagnosed with a grade II oligoastrocytoma in March 2007 when my wife Susan was 7 months pregnant with our first child. We had one week to come to terms with the diagnosis, choose a neurosurgeon, book in for surgery, phone friends and relatives and get all of our affairs in order.

I woke up from surgery with the entire right side of my body paralysed. I could not move my right leg or arm, had difficulty talking and even swallowing. I was in hospital for nearly two months for rehabilitation but I managed to leave just before Susan went into hospital to give birth to our baby girl. I was still having frequent seizures and suffering severe fatigue so it was a difficult time for all of us but having a child shifted some of the focus.

I started chemotherapy in August 2007. Temozolomide (Temodol) was not available on PBS for Grade II tumours and it was 'expected' that I would have PCV (Procarbazine, CCNU, and Vincristine) but I also had an issue with weak veins and I was worried that the injections would be a disaster. So we paid for the Temodol ourselves. It cost around $120,000 for this cycle which took a huge toll on our finances. A cycle is 28 days, 5 days on and 23 days off. That equates to nearly $4,000 per tablet.

I was also concerned about the drug Dilantin which I had been given to control my seizures. It is a very old drug and requires regular blood tests for the INR levels and I needed constant changes to the dose. I managed to transition to Keppra with the help of my neurologist but there was more cost involved.

I suffered a progression of my tumour towards the end of 2012 and lost the ability to speak. I also had trouble understanding anyone if there was more than one person talking at once like social gatherings or busy shopping centres. I went straight back on to Temodal but this time it was covered by the PBS because my tumour had become an anaplastic Grade III. I also had nearly 8 weeks of radio-therapy, 5 days per week which I chose to have in Sydney due to the more advanced equipment. This meant that I had to be away from my family during the week but I managed to go home to Canberra on weekends.

I have been very fortunate. While becoming disabled was a disappointment, I vowed to continue to carry on with my normal life as much as possible. Remaining positive is essential.

Becoming a parent after leaving hospital has taught me a lot about dealing with negative emotions. The self management of emotions is vital as there have been many highs and lows since my initial treatment.

There were many stressful events during 2013 that were triggered by the progression of the tumour but we managed to get through it. I no longer work but assume an active role in raising my daughter and I also engage in voluntary work to keep engaged and active.

noelle

My name is Patrick Michaelson. My wife Noelle was given her diagnosis of multiple myeloma on 14 February 2007. We were already aware that there was a problem when Noelle was ill on a flight home from an Alaskin Cruise in August 2006. By the time we received the diagnosis she was already stage 3. The haematologist told us that he knew earlier but did not want to spoil our Christmas. Shortly afterwards we were consulting a new oncologist/haematologist who was responsible in conjunction with Noelle's fighting spirit, for her survival, often against seemingly impossible odds, over the next six years.

At the time of diagnosis I was in full-time employment at sea and Noelle, a retired school teacher, ran a flourishing literacy consultancy. Her business stopped at once. I took all of my outstanding leave, followed by indefinite leave of absence. It soon became clear that I would need to be a full-time carer so I retired. The most immediate non-medical effect was the loss of two incomes. I had a pension but you do not lose over 60% of your income overnight without noticing it.

Over succeeding years various regimens were applied and with several hospitalisations and a stem cell transplant in March 2010, the myeloma was kept more or less at bay. These regimens included thalidomide, revlimid and velcade, often in conjunction with other drugs.

The diagnosing haematologist had stated that Noelle was too old to be considered for a stem cell transplant, she was 65 at the time. A course of treatment was started, which while appropriate to those circumstances, I have often heard at various seminars to be toxic to stem cells. When our new haematologist said that she was not too old there was some difficulty in harvesting sufficient cells for just one attempt at a transplant. Noelle had a long stay in hospital for the transplant as a result of an infection acquired there.

When velcade was prescribed, PBS would finance it for a period of eleven months. Four months into the programme a positive report from the prescribing doctor was required if it was to continue that support for the remaining seven months. Only one period of support was permitted. Encouraging results were obtained. The following year, velcade was prescribed again. This time, PBS would not provide assistance. Velcade therefore cost us $7,000 per month, which involved dipping into retirement funds, at least we had that. After three months and $21,000 later, the manufacturer agreed to fund ongoing provision of velcade. In July 2012 we were finally granted a carer's payment of $113 per fortnight.

Noelle became very ill in late January 2013 and passed away on February 11.

simone

For years I had constant aches and pains through my whole leg, I went and saw doctors, had X-rays and even had an ultrasound. Results came back without anything unusual. I went on with this pain and in 2011 I started to see an osteopath. It helped for a couple of hours but then the pain would come back.

On the 5 of December 2011, the night of my brother's birthday, I found a lump in my groin. It was sore to touch, I had my mum have a look at it and she was quite anxious so we went straight into emergency. The doctor checked it out and said if it has not gone away in 3 days, to get an ultrasound. I asked my osteo for his opinion and he said to me because I had lost a bit of weight it might just be swollen glands, so I ignored going to get an ultrasound.

A couple weeks go by and it started to get a bit bigger, at this point I thought I should probably get an ultrasound, how bad could it be? After the testing I went back to the doctors to get my results and I was told that they saw a mass and it could be lymphoma. I was referred to a lymphoma specialist and they had me take all different biopsies and tests, but nothing showed. They then referred me to Peter Mac and I was seen by another specialist who ordered a PET scan and yet another biopsy. A week later the PET scan came back and showed hot spots which usually means malignancy, but my biopsy came back normal. The specialist said he thinks its just inflamed bursitis and I probably have nothing to worry about but wanted to consult with others.

At this stage we are hearing different stories from everyone we had seen. The specialists then had a meeting and contacted me again and referred me to my now oncologist, who specialises in soft tissue. He had me take a few more tests and it came back showing a schwanomma. I had another test to check the status of the tumour, which came back as benign. I had the surgery in July 2012 to remove the benign tumours, which I thought was the end of it. A couple of weeks later at my follow up appointment, the most of my worries was seeing my scar, I had never been through anything big, never had an operation, I was healthy, so I thought. My oncologist begins to talk and I heard a word that I knew of, but it was like I was truly hearing it for the first time, cancer. I had three tumours in my groin/pelvis area but there was a fourth one hiding, it turns out I had a soft tissue sarcoma (Malignant peripheral nerve sheath tumour).

I completed 6 weeks of radiotherapy which then went onto a 12 hour wide margin operation to remove the iliosis muscle and the peripheral nerve in my left leg with a flap reconstruction and a nerve graph. I am now recovering, cancer free because of my amazing oncologist, his team, my plastic surgeon and his team. For the next 8 years I will continue to have regular MRI's and ultrasounds to make sure it stays this way.

sue

Hi my name is Suzanne Stewart; I am 48 years old, the mother of two children and live in Darwin. I have Metastatic Neuroendocrine Pancreatic Tumours (PNETs). My cancer has spread to my lymph nodes and my liver, I have been told that I am terminal and that I will die from this disease. This is a slow growing cancer but some of my tumours have become aggressive. I have been told to get my affairs in order. I have retired. I now cherish the time that I have to spend with my children, family and friends.

I was first diagnosed with NETs in October 2010 after the discovery of a mass in my pancreas. I underwent surgery and was told that 70% of these types of masses were cysts, I was confident that I would not have cancer. After being in hospital for 7 days, I got a call from my surgeon. He paused and said he was sorry to tell me that I had a very rare type of cancer, it was called a Neuroendocrine Tumour, the mass they took was positive as was the 18 lymph nodes.

I saw an oncologist and he told me that it was very rare, slow growing cancer and that they would need to do a special x-ray called a PET scan in Adelaide. After I had my PET scan, I was told that there was no sign of NETs in my body. I would need blood and urine tests every 3 months to monitor the situation. I was sceptical and requested a second opinion from Dr Michael at Peter Mac. After another PET scan, he also agreed that there were no NETs present.

I was very frustrated, scared and annoyed. I wanted to fight this disease; I knew it would come back. I had a mass with 18 lymph nodes that was all positive to cancer. I wanted chemotherapy and did not care how sick it would make me.

It was at my next review that the urine and blood tests aroused some suspicion. I was told that there was the possibility that the PNETs had returned. I was devastated. My son was in his first year of university, my daughter had just turned 16 and their father had died when they were young. It was then that I questioned why I had to wait for it to return. I feared about how far it had spread, if they could stop it and if my children would lose their mother as well.

After some consultations I was prescribed Octreatide injections. The day had finally arrived I was starting treatment; I was fighting this disease even if it was just the symptoms. I was now a patient. Oh I loved that injection I felt so much better. My symptoms settled and I was able to function better than I had in months.

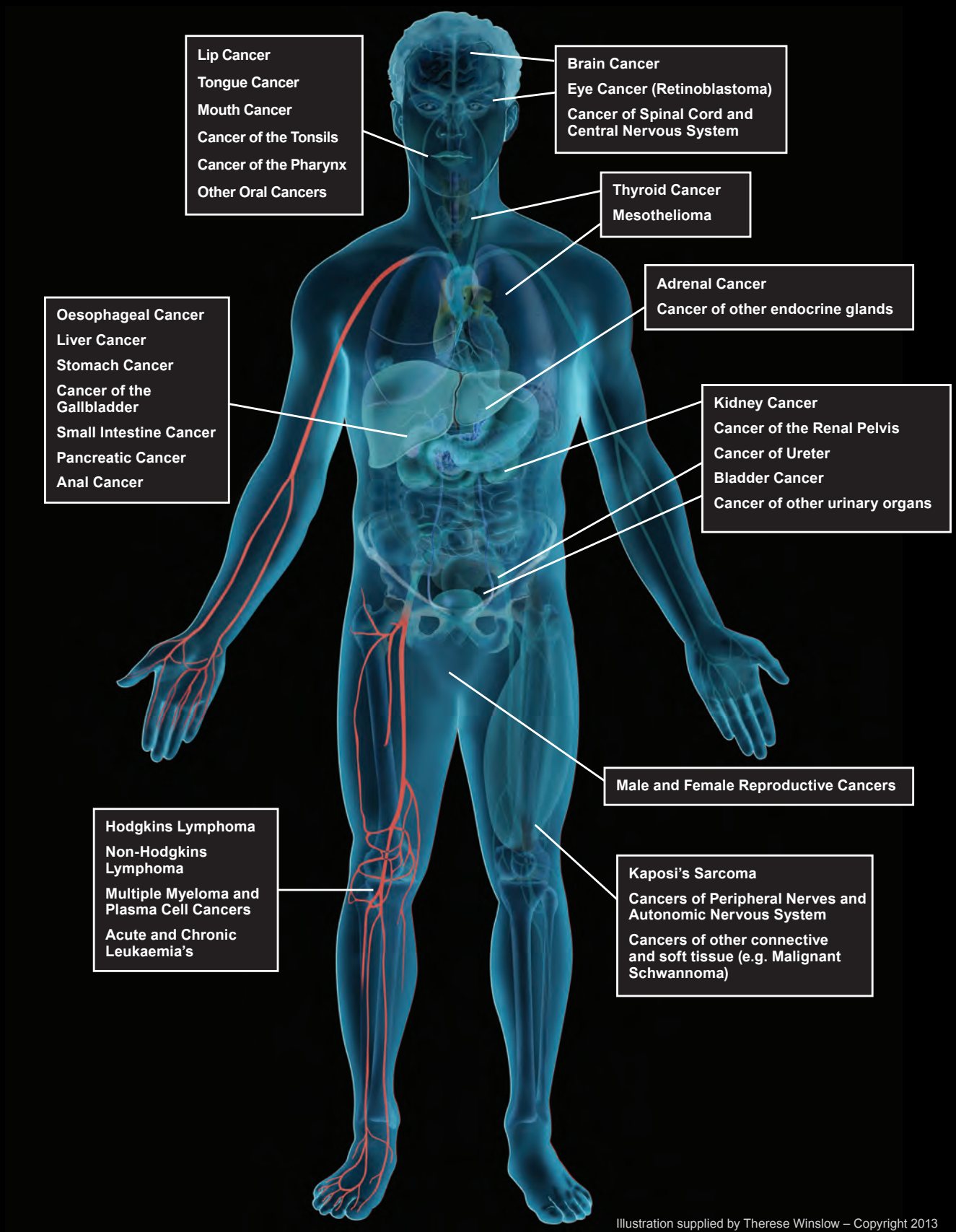
After a few months on the injections, a PET scan was ordered for me. The scan revealed that I had a tumour on my aortic artery, my left renal artery and there were three maybe four tumours in my liver. I was told to put my affairs in order and that maybe the team at Peter Mac could assess me for a trial drug called Everolimus.

I started the trial and at my last review, I was stable. I must acknowledge the Staff within the NT Department of Health especially the Oncologist Dr Naryan and CNM of the Allan Walker Cancer centre in Darwin. All of whom after sharing my journey with rare cancer, are now working to improve patient outcomes. This includes mental health, identifying support options, integrating rare cancer patients with more common diseases through activities and initiating cancer care plans.

About six months after I was diagnosed with PNETs, I was introduced to the Unicorn Foundation. I was extremely grateful to finally find an avenue to be able to hear other people's stories, treatment, issues and very importantly how to cope with living with the disease. Without the Unicorn foundation, I am not sure how I would have coped over the last few years.

appendix 3

body map of rare cancers



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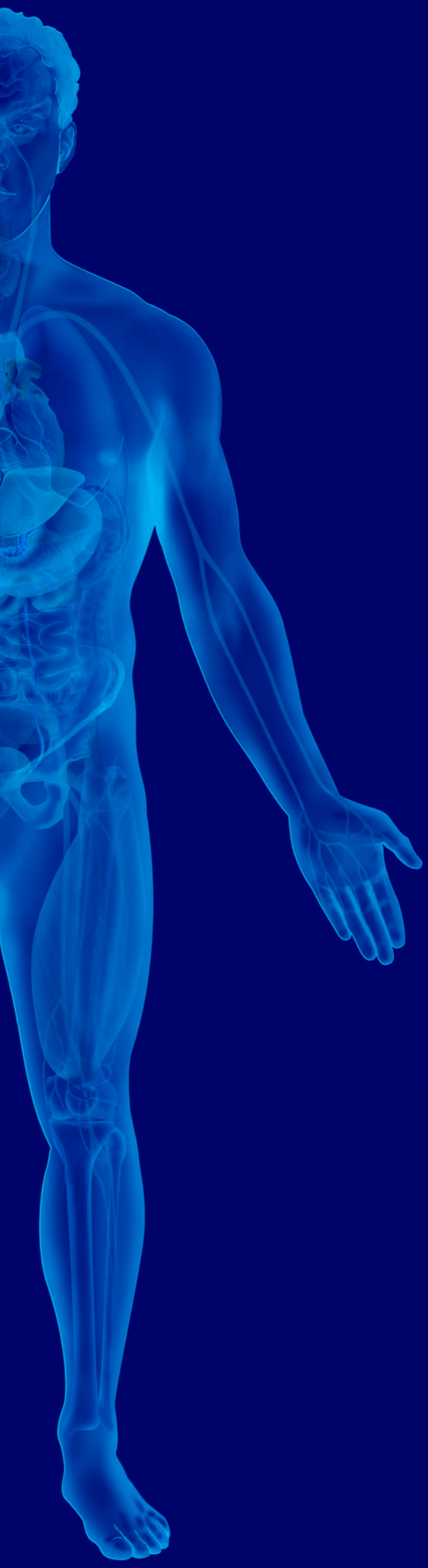

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