

Spring 2023

San Doctor

collaborating with our GPs
to provide coordinated
community care



Message from Brett Goods, Chief Executive Officer

We are excited to share the knowledge and expertise from a range of our specialists in this new edition of San Doctor with you.

We are also elated to share that in the 2022 Bureau of Health Information (BHI) Outpatient Cancer Clinics Survey the San leads NSW in providing an excellent cancer outpatient experience. This is the sixth year in a row we have achieved these outstanding results.

We are enormously proud of all the teams involved in these achievements and the fantastic support and care they consistently give our community.

Inside this issue you'll find details on Paediatric Pulmonary Hypertension, high blood pressure in the arteries of the lungs. You'll also read about our ongoing Inguinal Hernia Repair Project and Stereotactic Radiosurgery which uses radiation beams on tumours, veins that have developed differently or other abnormalities with extreme precision.

I hope this edition of San Doctor provides some insight into the amazing successes coming from the San.

Brett Goods, CEO
Chief Executive Officer
Adventist HealthCare Limited

AN ARTICLE
FEATURING

**Dr Cecelia
Gzell**

Stereotactic Radiosurgery

Stereotactic Radiosurgery (SRS) has emerged as an increasingly valued modality for Radiation Oncologists and is a treatment that is of particular interest to Dr Cecelia Gzell. She has specialist expertise in the use of stereotactic radiosurgery and stereotactic radiation therapy techniques. Her clinical experience covers a broad range of solid tumour malignancies with a special interest in brain cancer, primary and secondary central nervous system (CNS) neoplasms, and benign intracranial and base of skull lesions.

Stereotactic Radiosurgery (SRS)

Stereotactic Radiosurgery is a very precise form of therapeutic radiation that can be used to treat abnormalities in the brain, spine, neck, and other parts of the body.

Around 50 years ago, Stereotactic Radiosurgery was pioneered as a less invasive alternative to standard brain surgery. Despite what its name implies, it is a non-invasive procedure which does not require an incision or opening, rather it uses 3D imaging to target the affected area with high doses of radiation with minimal impact to the surrounding healthy tissue.

"It's a very precise, small field of radiation, often where we need to protect normal structures that are close by such as the brainstem or the optic apparatus. It's a very technical form of radiation, which we primarily use to treat brain tumours, both malignant and benign. But we can also use it to treat abnormalities like arteriovenous malformations (AVMs) or trigeminal neuralgia," said Dr Gzell.

Who is eligible for SRS?

Stereotactic radiosurgery has increasingly become a common approach for patients who have smaller brain tumours. Other candidates may also be those patients whose disease is not surgically accessible.

"Normally, the main sort of exclusion criteria would be the volume or size of tumour that needs to be treated, however if for instance a patient has a large tumour of more than three centimetres in diameter, it does not preclude them from undergoing SRS. Instead of between one and five treatments, that patient may undergo a longer course of treatment but done utilising the precise set-up of stereotactic treatment," Dr Gzell said.

In cases where adjacent structures need protection, a precision-based technique can still be used, however we might adjust the dose delivered per treatment. Stereotactic radiotherapy (SRT) may be used instead. "It means we are giving the more standard fractionation

which often is five or six weeks of treatment, but we are using the advantage of the precise technique with the extra mask, the extra set-up precision to make sure we can deliver the high dose of radiation and protect those normal structures."

"GPs can refer patients directly, otherwise patients may be referred via either a surgeon or a Medical Oncologist, or other specialist (such as Endocrinology, ENT surgery, Ophthalmology, Neurosurgery). If GPs are unsure, we can consult with them and provide advice on how best to proceed."

Typically, most patients with primary tumours and brain metastasis will be referred for the treatment through the Neuro-oncology multi-disciplinary team. Patients with non-cancerous tumours can be referred either directly via a neurosurgeon, GP or endocrinologist.

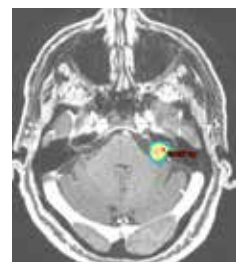
Benefits of SRS treatment

Typically, stereotactic radiosurgery is performed as an outpatient procedure, and usually only takes between 30 minutes to an hour.

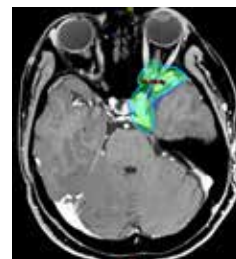
It can be given as either a single treatment or a short series of treatments. This results in greater patient comfort and limited side effects.

"The option of SRS means that patients now have the chance to not only get the treatment they need, but have a better quality of life," said Dr Gzell.

Additionally, if treatment is needed close to a critical structure such as the brainstem or the optic apparatus, the treatment can be delivered in a way that protects those structures. "The nature of the treatment we are delivering means the dose drops off very rapidly to ensure we protect any adjacent structures."



Target for SRS to Left vestibular schwannoma

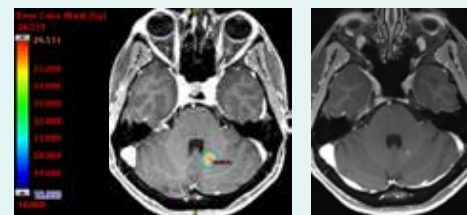


SRT target for a left optic nerve and sphenoid wing meningioma

Treating Tumours

1. Brain Metastases

Since its conception SRS has emerged as a leading method of treating brain metastases in a range of clinical scenarios including as definitive treatment alone for patients with a limited number of BMs and as an adjuvant treatment method in a pre and post operative setting.



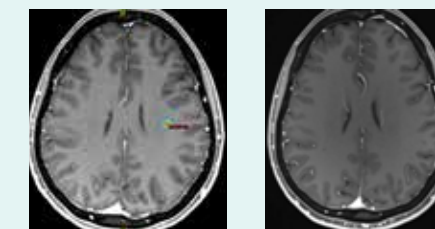
Target in Left cerebellum (single treatment SRS)

5 months later lesion Left cerebellum reduced by >50%

"The technology has really evolved, particularly in the last five years, and we know a lot more about giving SRS to patients who have BM's. Whereas we used to decide based on a numerical criterion for instance that a patient would have less than four to five lesions, we have moved away from that, and it is more about if the tumours are well spread apart and small in size, we can treat up to 10 fairly comfortably," said Dr Gzell.

2. Benign Tumours

In terms of benign tumours, we can treat meningiomas, vestibular schwannomas, glomus tumours (which do actually go below the brain and into the neck in a lot of cases), as well as pituitary adenomas with SRS. "We treat a lot of those, but some other non-tumour related indications are arteriovenous malformations, trigeminal neuralgia and then functional neurological disorders, like patients with essential tremor."



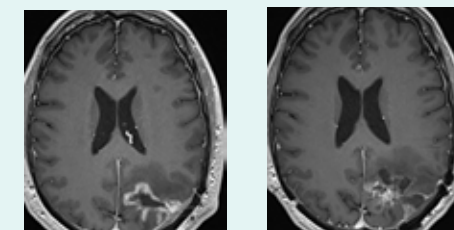
Second target in same patient in Lt frontal lobe

5 months later lesion in Left frontal lobe resolved

3. Radiation Necrosis

Radiation necrosis is an inflammatory, treatment-related reaction that typically occurs greater than six months after delivery of radiation treatment. It is usually self-limiting. Routine surveillance imaging using MRI means in most cases the inflammatory change is detected early. An increased incidence of radiation necrosis is seen in patients having concomitant immunotherapy or targeted therapy, but the number of patients that have symptomatic radiation necrosis has not increased. Typically, symptomatic cases represent less than 10% of all treated patients. Symptoms are often mild and respond to a short course of steroid medication.

"In the past, when we started using this type of treatment, we would ask Medical Oncologists to stop systemic therapies for at least a couple of weeks. This is a practice we have moved away from, particularly when patients have had immunotherapy or targeted therapies," said Dr Gzell. "In fact, there is emerging evidence that these newer therapies may be synergistic with radiation and the optimum period may be starting these therapies within a week of SRS."

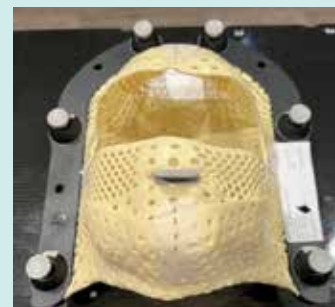


Asymptomatic radiation necrosis after SRS one year earlier (image from 2019)

Evolution of radiation necrosis over time (image from 2023), patient remains asymptomatic

Stereotactic Radiosurgery at ICON at the San

"At ICON cancer centre at the San we are working with cutting edge radiation technology, purpose built for the facility," said Dr Gzell. "in the last year we have introduced a new quality assurance system of patient position verification using surface guidance. We are one of the only centres in the world, and certainly in the southern hemisphere that has implemented this practice for cranial stereotactic radiosurgery patients."



Surface-guidance SRS mask

Stereotactic Radiosurgery requires submillimetre spatial accuracy. The accuracy depends on the alignment of the beams as well as on the positioning and immobilisation of the patient. Recently Dr Gzell has implemented the use of open face masks which are more comfortable for the patients than the closed mask set-up. "Previously, patients' eyes and nose would be covered by the masks, whereas now we use a mask with an opening across the eyes. It's more comfortable for the patient. We then use a blue light to monitor every pixel in that area to see if there is any shift. We constantly monitor this throughout the treatment and will stop if there is any movement. It's a very safe and very effective way of monitoring a patients position throughout the treatment," said Dr Gzell.



Dr Cecelia Gzell

BMedSc(Syd), BMed(Newcastle), FRANZCR, PhD

Dr Cecelia Gzell is an experienced radiation oncologist who consults and treats patients at Icon Cancer Centre, Wahroonga and Concord. She obtained her Bachelor of Medical Science (BMedSc) at the University of Sydney and Bachelor of Medicine (BMed) at the University of Newcastle. Dr Gzell subsequently undertook specialist radiation oncology training at the Royal Prince Alfred Hospital prior to becoming a fellow of the Royal Australian and New Zealand College of Radiologists (RANZCR) in 2011. She later completed a post-graduate neuro-oncology fellowship at Royal North Shore Hospital with research and publications in the field of high-grade gliomas. This research provided the foundations for her PhD focusing on the use of radiation therapy in optimising outcomes of patients with glioblastoma.

She has specialist expertise in the use of stereotactic radiosurgery and stereotactic radiation therapy techniques. Her clinical experience covers a broad range of solid tumour malignancies with a special interest in brain cancer, primary and secondary central nervous system (CNS) neoplasms and benign intracranial and base of skull lesions.

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AN ARTICLE
BY**A/Prof Stephen
Pillinger**

Impressive early results – project evaluates robotic hernia repairs

The first complete review of data from a project analysing robotic inguinal hernia repair shows this approach has a number of significant benefits for patients and, with an experienced team, it is an efficient use of robotic surgery resources.

“The outcomes with robotic hernia repair are well documented: patients can generally expect less pain, fewer complications, a shorter stay in hospital, and a quicker return to work and normal activities,” said A/Prof Stephen Pillinger, Head of Robotic Colorectal Surgery at the San, and Principle Investigator for this project. “Despite these benefits, the established dogma is that the robotic approach for inguinal hernia repair is cost prohibitive. However there is a lack of really good evidence to support that view. The purpose of this project is to prove we can run a high-volume robotic inguinal hernia service that is beneficial for our patients as well as being efficient and economical. This is the first time a project of this nature has been done in Australia.”

Early findings

The early results from the first three months of the project have been very positive. Importantly, costs are the same for robotic hernia repair as for any other method of hernia repair. “There is no added cost,” noted A/Prof Pillinger. “Historically people were concerned robotic surgery was very expensive for patients. The San was the first hospital in Australia to implement a no-extra-cost model for robotic surgery for patients. We have continued this policy for the lifetime of the robotic hernia repair project, so robotic inguinal hernia repair costs exactly the same as patients would be charged for any other modality of hernia repair.”

“The project also clearly shows that robotic hernia repair is now available as day-case surgery: 97% of patients are going home the same day. And even in this early phase of the project, efficiencies in the operating theatres are also being undertaken, which is one of the key objectives of the study,” he added.

Patient experience

Rob* suffered for a number of years with a troublesome right inguinal hernia. “I had a lump a few centimetres in size sticking out of my groin and it was very painful. Sometimes I had to walk along with my hand in my pocket to press on the hernia to hold it in, and it was a bit embarrassing.”

“I’d been to the GP a couple of times over the years and he had referred me to have the hernia operation. But I’m self-employed, and taking time off work at the bakery to get this done was very difficult. It was another 12-18 months down the track when I went back to the GP. By then it was super painful and I knew I had to get something done as soon as possible.”

The GP referred Rob to a Sydney Adventist Hospital specialist, who talked through all the options of the various approaches to hernia repair surgery. “I heard about this project. I was keen to get the hernia fixed, and happy to take part in the robotics project,” Rob said. As it turned out, scans revealed that Rob not only had the large hernia in the right groin, he also had a hernia in the left groin, and both needed repair.

“I went into hospital at 6.30am to have the surgery, and was home by 6pm that night. I was up walking around before leaving hospital. The next morning I had a bit of pain when I rolled over, but was otherwise fine. By day five I felt well enough to go into the bakery for a short time – not to work, just to check on things. Honestly, if I’d known a few years ago that this type of surgery was around, I’d have had it done then. I had surgery in August and six weeks later I was back to playing golf.”

Selection criteria

There are three groups of patients that may be suitable for robotic hernia repair, with the first two groups being assessed for inclusion in the study:

- Patients who have had no prior lower abdominal surgery, previous repair, recurrence or pelvic radiotherapy
- Patients who have had previous inguinal hernia repair with recurrence
- Patients who have had previous abdominal surgery and have complex or large incisional hernias that require component separation/abdominal wall reconstruction.

While there is significant interest around robotic hernia repair, not every patient is suitable for robotic surgery. Each patient will be carefully assessed, and their hernia surgery will be done using the most appropriate approach for their individual needs. The project will analyse the outcomes of up to 200 patients over 12 months, with results expected to be published mid-2024.

Six surgeons are participating in the robotic hernia repair study. “Robotics is a very accepted and sought after technology now,” said A/Prof Pillinger. “It is the next evolution in surgical care and, with it, minimally-invasive surgery keeps getting better and better. This project will continue into 2024, however I don’t think robotic hernia repair will have a finite period – I think it’s likely this approach to hernia repair will be a service that will be ongoing.”

**Patient’s full name withheld for privacy.*



A/Prof Stephen Pillinger

MBChB, FRACS

Dr Pillinger is a Consultant Colon & Rectal Surgeon, having commenced Consultant practice in 2005. The major focus of his practice is benign and malignant colorectal pathology and has particular expertise in minimally invasive Laparoscopic and Robotic Surgery, Transanal Endoscopic Microsurgery, diagnostic and interventional colonoscopy. He has trained in Robotic surgery in the US and South Korea and is one of the busiest Robotic Colorectal Surgeons in NSW.

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AN ARTICLE
FEATURINGDr Chris
Rogan

A new minimally invasive option for knee arthritis – genicular artery embolisation

Knee osteoarthritis:

Osteoarthritis of the knee is an extremely common condition which affects millions of Australians and can negatively impact quality of life and the ability to stay active. Patients may obtain relief through lifestyle measures such as weight management, exercise and adjunctive therapies including thermal therapy, massage and mobilisation, physiotherapy as well as use of nonsteroidal anti-inflammatory drugs and joint injections.

Despite the above, there is a large population of patients who still suffer persistent pain daily but have few options available other than to continue daily analgesia use +/- occasional intra-articular corticosteroid injections for flares and possibly other intra-articular injections such as hyaluronic acid and PRP.

Patients may have to endure pain for many years before the knee has significant enough degenerative changes to warrant a knee replacement.

Orthopaedic surgeons generally like to delay knee replacement until there is advanced degenerative changes on the imaging studies and the patient is requesting surgery.

Until recently, there has been a dearth of options for the many patients in this scenario which may persist for decades, however there have been recent advances and publications in embolisation therapy which offer a

minimally invasive option which may be available for these patients.

Osteoarthritis pain as an inflammatory problem:

Traditionally osteoarthritis has been thought of as a wear and tear physical degeneration condition however inflammatory mediators produced by thickening of the synovium (synovial hypertrophy) are a major pain generator in the osteoarthritic knee. Suppression of this inflammatory component may currently be achieved by NSAIDs and cortisone injections but the effect is temporary.

Embolisation is a minimally invasive technique designed to reduce blood flow to thickened synovium and in so doing induce reduction in synovial hypertrophy which reduces the local inflammatory environment in the knee.

Embolisation in the knee is known to be safe and effective at reducing synovial hypertrophy in patients with troubling haemarthrosis which occasionally occurs after knee replacement surgery and in haemophilia. The same technique (with modification) is now being applied to reduce synovial hypertrophy in the osteoarthritic knee, decreasing local inflammation and pain.

Genicular artery embolisation

A few years back, at Royal Prince Alfred Hospital, we published the outcomes of embolisation for knee

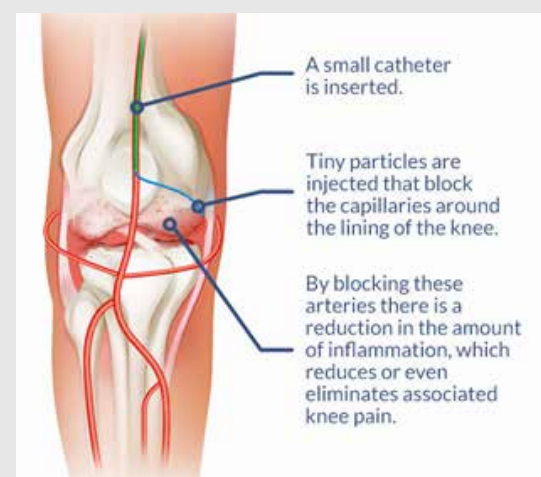
haemarthrosis in the Australian context and this is well established therapy for this uncommon condition after knee replacement and in patients with haemophilia.

Over the last 10 years, especially in Japan and the USA, genicular artery embolisation has been offered for knee osteoarthritis and the studies to date have shown a low risk profile for this day procedure and a substantial reduction in knee pain out to at least 24 months. I have personally spoken with key opinion leaders from around the world in interventional radiology and there is significant optimism about the future scope of this therapy.

The procedure:

GAE is performed as a day procedure under local anaesthetic +/- light conscious sedation. After the access site is numbed, a thin flexible catheter is introduced into the lower limb artery and the blood supply to the synovium is mapped with attention to identifying the abnormal increased vasculature seen in the symptomatic osteoarthritic knee. Generally this **may** be predominated in either the medial or lateral compartment and often the treatment is focused to the more symptomatic compartment.

A very thin (less than 1 mm) flexible microcatheter is steered into the genicular artery which supplies the abnormal synovium and microscopic particles designed to reduce blood flow are trickled into the vessels to reduce blood flow



(embolism) back down towards physiologic levels. Following introduction of the embolic agent, blood vessels are again mapped on completion and the catheter is removed. The patient can be discharged home that afternoon.

There is requirement for a couple of days off strenuous exercise and heavy lifting to minimise bruising risk and there may be a temporary flare of knee arthritis pain for a few days. Potential complications described in literature include bruising in the groin as well as a temporary skin rash but generally the risk profile is very low. There is a requirement for a couple of days off strenuous exercise and heavy lifting to minimise bruising risk.

A new field of minimally invasive MSK treatment?

Momentum is building internationally for a new field of what is called MSK embolotherapy because the same technique described above in the knee has been shown to also have some benefit in other chronic inflammatory pathologies affecting tendons and joints.

Conditions where this has shown to be potentially beneficial include:

- Frozen shoulder
- Achilles tendinopathy,
- Golfer's and tennis elbow.

However data is still in the early days for applications outside of the knee joint. As president of the Interventional Radiology Society of Australasia I am in early discussions to establish a national registry to better capture and track outcomes and data in this emerging area.

Patient evaluation:

Patients are generally screened for suitability for the procedure based on their level of knee pain, and a questionnaire regarding the function pre and post procedure is normally administered.

All patients will need an x-ray to exclude advanced degenerative changes which are much more appropriately treated with surgery and generally an MRI is helpful to exclude any acute knee injuries and assess the state of the degenerative changes in more detail.

I like to work collaboratively with orthopaedic surgeons where appropriate and will normally liaise or refer to discuss in cases where patients may have other pathology causing pain or more advanced changes that are more suitable for consideration for surgery.

There are also other contexts in which genicular artery embolisation may prove useful but are not yet well established. Areas of research include obvious potential in monoarticular inflammatory conditions. There is also a role in the 10-20% of patients who have persistent knee pain after total knee replacement.



Dr Chris Rogan

MBBS (Hons1), BSc (Med), FRANZCR, EBIR

Chris Rogan is a specialist interventional radiologist using imaging guidance and cutting edge minimally invasive techniques to perform pinhole procedures. He is current president of the Interventional Radiology Society of Australasia and a visiting medical officer at Sydney Adventist Hospital and Chris O'Brien Lifehouse with special interest in embolisation procedures.

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Patients you may want to consider referring to discuss an embolisation procedure:

- Troubled daily by knee osteoarthritis pain.
- X-ray knee shows relatively intact knee joint with early degenerative changes.
- X-ray does not show severe advanced changes "bone on bone" which may be best treated surgically.
- The patient may have already seen a surgeon and not currently be a candidate for surgery.

AN ARTICLE
FEATURING

A/Prof Ross Grant

The San is one of two sites where a new study – the Brain Enhancement Study – will investigate whether lifestyle factors can prevent or slow cognitive decline.

Participants wanted for Brain Enhancement Study

This research will be of great interest to those impacted by cognitive impairment, dementia, or Alzheimer's disease. The study will be conducted by the Australasian Research Institute (ARI) which is affiliated with the Sydney Adventist Hospital, Sanitarium Health Food Company and Avondale University.

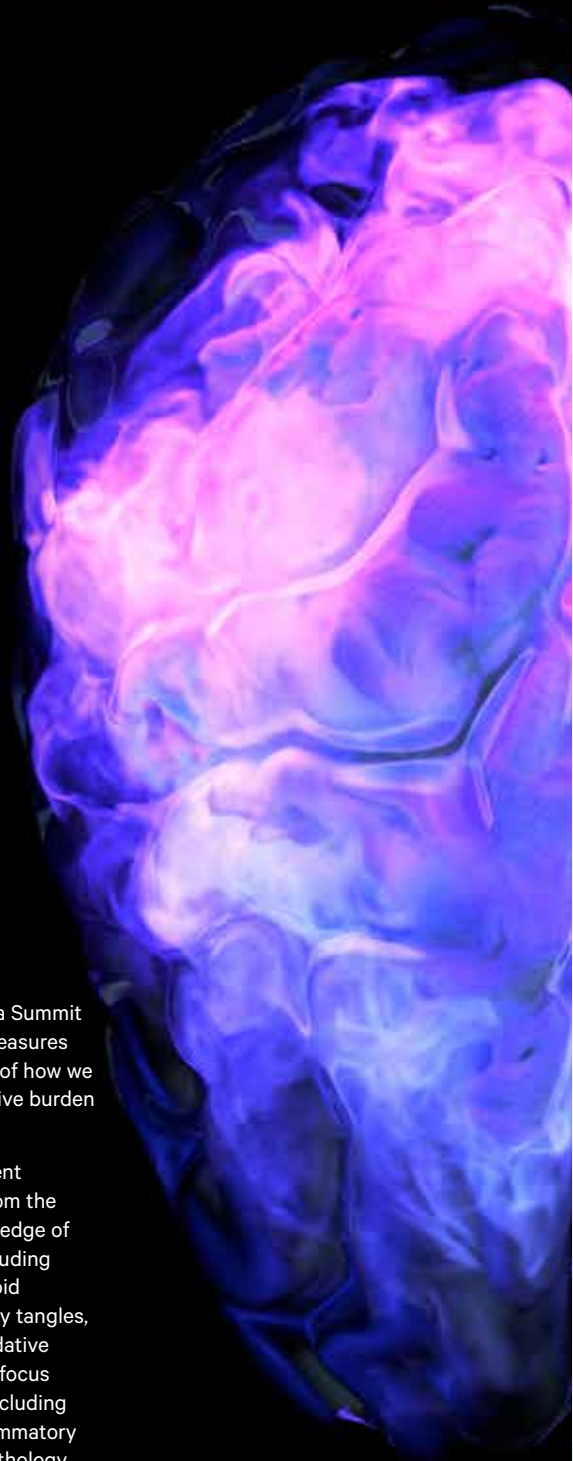
"We are inviting people between 65-85 years of age who have been diagnosed with mild cognitive impairment to participate in a six-month study," said A/Prof Ross Grant, CEO of ARI and chief investigator of this study. "We are recruiting people who have mild cognitive impairment (MCI) before they track to frank dementia. Not everybody with MCI ends up with dementia, but a significant proportion do. So that's the group we want to study and, ideally, move them off that track: at the very least slow it, ideally stop it, and if there's any potential to reverse it even a little bit, that would be fantastic."

Current treatment approaches for Alzheimer's disease can be categorised into two basic groups – symptomatic and disease modifying. "Disease-modifying drugs aim to prevent the progress of cognitive decline by targeting the causal elements or mechanisms of degeneration," said A/Prof Grant. "Billions of dollars are spent on developing potential drug therapies for diseases such as Alzheimer's, and most have failed. While some such drugs may modestly slow the initial rate of cognitive decline, none halt disease progression or restore neural networks – and thus do not restore any functional loss. The side-effect profile of these drugs often results in poor compliance. While no drug can effectively prevent disease progression, emerging evidence indicates that lifestyle-based interventions may at least stabilise the cognition. The World Health Organisation recognised back in 2013 that we need to do something more effective than just chase after drug therapies. At the same

time, the G8 Dementia Summit advocated lifestyle measures as being a major part of how we help reduce the massive burden of dementia."

The Brain Enhancement Study (BES) draws from the latest scientific knowledge of cognitive decline, including the role of beta-amyloid plaques, neurofibrillary tangles, inflammation and oxidative stress. The study will focus on multiple factors, including the participant's inflammatory markers and other pathology, exposure to toxins, diet/nutrition, sleep, stress, genetics, exercise/activity, brain training, and social connection.

The Brain Enhancement Study is a multi-modal randomised-controlled study. "Our research is focused on the pathology – what's actually going on in the brain – driven by our lifestyles. For a number of years, we've known the association between certain lifestyle factors and the impact these have on our brains and body and, ultimately, the risk of disease," said A/Prof Grant.



"Both clinical and scientific evidence suggests that a lifestyle-based, multimodal intervention that stimulates numerous biochemical pathways known to reduce inflammatory or oxidative processes and promote repair, neurogenesis and general brain health, may effectively prevent mild cognitive impairment progressing to frank dementia or Alzheimer's disease," said A/Prof Grant. "We will also be investigating the potential for lifestyle measures to reverse, to some degree, cognitive decline in at least early disease."

The Brain Enhancement Study builds on a successful pilot study ARI conducted in 2019 which showed that lifestyle interventions improved cognition, reduced inflammation, lowered blood pressure, and resulted in improvements – both clinically (for e.g., pathology results) and anecdotally – in many areas of participant's health and wellbeing.

Outline of the study

GPs or specialists refer patients with diagnosed MCI to take part in the study. If participants have diabetes, it needs to be well controlled, and any sleep issues or depression needs to be investigated by the GP prior to joining the study.

Participants will do the study with a 'study-buddy' – a partner or carer. The study starts with a one-week intensive, which includes a comprehensive assessment of lifestyle and nutrition and pathology tests. Over six months, participants will receive regular one-on-one coaching and support from study facilitators to guide them through the program. Progress reports will be sent to the participant's doctor, and at the end of six months, those who have been in the non-treatment group will have the opportunity to cross-over (i.e., enrol) in the treatment group.

The Brain Enhancement Study is inviting expressions of interest by potential participants to become part of the study. The study will commence in early 2024 and will involve a six-month commitment from participants. The study will take place at two sites: at the San (Wahroonga) and at Avondale University (Lake Macquarie). "The ARI Lifestyle Medicine and Research centre on the San campus has been conducting

research for 20 years," said A/Prof Grant. "We've always had an interest in the biochemical basis of lifestyle medicine – what is it about lifestyle that drives us towards health or disease. Our expertise is particularly within the medical sciences, clinical biochemistry and neurochemistry."

"We have three goals in Brain Enhancement Study: one, to remove or eliminate anything that damages the brain; two, to supply the brain with everything it needs to function and repair; and three, to stimulate the brain's activity," added A/Prof Grant. "Seven distinct strategies – based on lifestyle factors – will be used during this study to address those three goals. The aim is to investigate whether a personalised, multimodal intervention will – in comparison to standard care (control group) – cause significant differences in cognition, everyday function, mood, and biochemical markers of brain health."

"We invite GPs, geriatricians, neurologists and other specialists to refer patients with mild cognitive impairment to take part in this study which, we believe, has the potential to change the course of how we treat and prevent cognitive impairment in the future."

For more information and to refer your patients to participate in this trial, please contact:

Project Coordinator
☎ +61 2 9480 9601

Participation Criteria:

- Between age 65-85.
- Have trouble with their memory and/or known mild cognitive impairment.
- Are willing to change their lifestyle for six months.
- Can complete the study with a loved one or carer.



A/Prof Ross Grant

BEd(Sc), MAppSc(clin chem),
PhD(neuropharm)

A/Prof Ross Grant is a clinical biochemist/biochemical pharmacologist and CEO of the Australasian Research Institute at Sydney Adventist Hospital. A/Prof Grant has contributed to over 100 academic publications in his research area which focusses on understanding how a person's range of Lifestyle factors changes their body's biochemistry and immune activity driving it toward either health or disease. In particular the influence of the constellation of lifestyle factors on redox balance (i.e. oxidative stress) and NAD+ metabolism and how these influence cellular degeneration, particularly in the brain and central nervous system.

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AN ARTICLE
BY

Dr David Youssef

Paediatric Pulmonary Hypertension: A Comprehensive Guide for General Practitioners



Paediatric Pulmonary Hypertension (PH) is a rare but serious condition that affects the lungs and heart in children and can lead to significant morbidity and mortality if not diagnosed and managed promptly. Current estimates suggest a PH prevalence of ~1% of the global population. General practitioners play a crucial role in identifying early symptoms, making timely referrals, and supporting affected families. In this article, we will provide a comprehensive overview of paediatric PH, including its causes, symptoms, diagnosis, and management strategies.

Understanding Paediatric Pulmonary Hypertension

Paediatric PH involves increased blood pressure in the pulmonary arteries, the vessels responsible for carrying blood from the heart to the lungs. This elevated pressure can strain the heart and lead to various complications, including heart failure. Pulmonary hypertension is defined as a mean pulmonary artery pressure (mPAP) > 20mmHg at rest (reference ESC guidelines). This figure is based on studies assessing the upper limit of normal pulmonary artery pressures in healthy individuals.

Classifications

Pulmonary Hypertension is classified into one of 5 categories based on the underlying aetiology. These have been summarised below and are based on the WHO Group classifications.

Group 1 Pulmonary arterial hypertension – rare
<ul style="list-style-type: none"> Idiopathic pulmonary hypertension Associated conditions including porto-pulmonary hypertension, congenital heart disease and connective tissue disease.
Group 2 Pulmonary hypertension associated with left heart disease – very common
<ul style="list-style-type: none"> Includes mitral valve disease and aortic stenosis
Group 3 Pulmonary hypertension related to lung disease and hypoxemia – common
<ul style="list-style-type: none"> COPD
Group 4 Pulmonary hypertension associated with chronic pulmonary artery obstructions – rare
Group 5 Pulmonary hypertension with unclear and/or multifactorial mechanisms – rare

Groups 1, 3 and 5 are the most common seen in infants and children whilst Groups 2 and 4 are less commonly seen.

Risk factors for pulmonary hypertension include prematurity, family history of PH, and certain medical conditions like connective tissue diseases.

Recognizing the Symptoms

Early recognition of symptoms is crucial for timely intervention. Common signs of paediatric PH include:

- Shortness of breath, especially during physical activities
- Fatigue, even after mild exertion
- Palpitations
- Haemoptysis
- Weight gain due to fluid retention
- Fainting spells (syncope)
- Cyanosis
- Difficulty feeding in infants
- Hoarseness (dysphonia) due to compression of the left laryngeal nerve

Diagnosis and Evaluation

General practitioners should have a low threshold for suspecting PH in children presenting with these symptoms. Diagnosis typically involves:

- Physical Examination: Listening for abnormal heart sounds or murmurs.
- Imaging Tests: Echocardiography to assess the heart's structure and function.
- Electrocardiogram (ECG): To detect abnormal heart rhythms.
- Blood Tests: Including NT-proBNP levels, which are often elevated in PH.
- Referral to Paediatric Heart Centre for Right HeartCatheterization: A definitive test to measure pulmonary artery pressures - this is the gold standard.



Dr David Youssef

B.Pharm, M.B.B.S, D.Child Health (USyd), FRACP

Dr David Youssef is an experience and well-respected paediatric cardiologist and pulmonary hypertension specialist at The Sans, specialising in Paediatric Echocardiography, 3-Dimensional Imaging and Paediatric Pulmonary Hypertension.

Dr Youssef has consistently demonstrated exceptional dedication to patient care, embodying qualities such as altruism, compassion, integrity and leadership especially when supporting those in greater need.

Management and Treatment

Once diagnosed, a multidisciplinary approach involving paediatric cardiologists, respiratory physicians, and PH specialists is essential. Treatment strategies may include:

Medications:

- Pulmonary vasodilators such as prostacyclin, endothelin receptor antagonists, and phosphodiesterase-5 inhibitors can help relax pulmonary arteries, reducing pressure.
- Oxygen Therapy: Supplemental oxygen can improve oxygen levels in the blood, easing symptoms.
- Surgery: In some cases, surgical intervention to correct underlying heart defects might be necessary.

Lifestyle Modifications:

- Encouraging a healthy lifestyle, including regular exercise (as tolerated) and a balanced diet, can support overall well-being.

The Role of General Practitioners

General practitioners serve as the first point of contact for many patients. By maintaining a high index of suspicion for paediatric PH, promptly referring suspected cases to specialists, and educating families about the condition, GPs significantly contribute to improved outcomes and quality of life for affected children.

Conclusion

Paediatric Pulmonary Hypertension is a complex condition that requires early detection and specialised care. As frontline healthcare providers, General Practitioners play a pivotal role in this process. By remaining vigilant, staying informed about the latest diagnostic and treatment modalities, and collaborating closely with specialists, GPs can make a significant difference in the lives of children affected by this challenging condition. Timely intervention and ongoing support can greatly enhance the prognosis and overall well-being of young patients with PH, offering them a chance at a healthier, more vibrant future.

Reference: <https://www.escardio.org/Guidelines/Clinical-Practice-Guidelines/Pulmonary-Hypertension-Guidelines-on-Diagnosis-and-Treatment-of>

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ELIA Lifestyle Article



The undeniable significance of lifestyle factors in disease prevention is well-known to medical practitioners. As part of the learned mantra, ‘lifestyle measures’ is commonly written in quality clinical notes. Brief advice is provided, referrals are made to allied health specialists, compliance is dreamed of, and clinicians hope for a coordinated approach.

Evidence-based guidelines and consensus statements from the vast majority of organisations dealing with chronic disease acknowledge the prominent role that nutrition, physical activity and smoking cessation play in the prevention and treatment of chronic disease. There remains, however, a disconnect between our knowledge base and clinical outcomes, with global statistics in 2023 showing that we are far from achieving our goals with the ever-increasing burden of chronic disease.

What exactly is Lifestyle Medicine?

Lifestyle Medicine dates back to Hippocrates as a strong advocate for a healthy diet and physical exercise to prevent disease. Yet it has only been officially recognised in the medical field recently, with the American College of Lifestyle Medicine founded in 2004, and the Australasian Society of Lifestyle Medicine opening in 2015. To ensure global standardisation of the field, the International Board Certification of Lifestyle Medicine was established in 2016. Lifestyle Medicine models of care are now widely used in the UK and the USA, including the Mayo Clinic, Cleveland Clinic and Loma Linda University Hospital, and are beginning to gain traction in Australia, especially through primary care.

Lifestyle Medicine is a medical specialty that uses evidence-based therapeutic interventions as the primary modality to prevent and treat chronic diseases such as cardiovascular diseases, type 2 diabetes, and obesity. It is delivered by a team of clinicians trained and certified in this specialty, and when used intensively, can lead to disease prevention, treatment and remission. Treatment focuses on six well-studied pillars: whole food plant-based eating, regular physical activity, restorative sleep, stress management, avoidance of risky substances and positive social connections.

Benefits of each pillar:



1. Healthful Eating

Nutrition plays a significant part in lifestyle habits and practices that effect nearly every chronic disease. There is undeniable evidence and consensus for the role nutrition plays in cardiovascular disease, obesity, diabetes, and cancer, among many other conditions. In general, a whole food plant-predominant diet made up of whole grains, legumes, fruits, vegetables, nuts and seeds, one that minimises or eliminates processed foods, is optimal for individual and planetary health. According to the EAT-Lancet Commission on Food, Planet, Health, prescribing a sustainable, healthy whole food plant-based diet would reduce the burden of chronic disease globally and prevent close to 11 million deaths per year, while also protecting planetary health.



2. Physical Activity

Exercise is undeniably an effective medicine, but unfortunately, it has been estimated that less than 40% of physicians regularly counsel their patients on the importance of increasing physical activity. Regular physical activity has been specifically demonstrated to reduce risk of CVD, type 2 diabetes, metabolic syndrome, obesity, and even certain types of cancer. Increasing physical activity can decrease the risk of depression and anxiety and has been shown to be equally as effective as medication in treating major depression. It also plays a vital role in managing insulin resistance and diabetes, increasing bone density and reducing the risk of cognitive decline.



3. Sleep

Poor sleep quality is linked to chronic diseases including cardiometabolic disorders, obesity, and inflammation, as well as mental health conditions. Studies show that even relatively small sleep disturbances from neighborhood noise can impose psychological distress, mental fatigue, depression and anxiety, all of which can amplify other health-related problems and impact physical health. By prescribing 7–9 hours of restorative sleep, health professionals acknowledge the importance of sleep to prevent chronic disease, build resilience, and help individuals establish a more robust immune system through which to combat illness.



4. Avoid Risky Substances

Use of tobacco, alcohol, and non-prescription medications are inextricably associated with an increased risk of chronic diseases and death. The WHO particularly cautions against the use of tobacco as the single greatest avoidable risk factor for cancer mortality which kills approximately 6 million people each year. Alcohol use is a known risk factor for many types of cancer including cancer of the oral cavity, pharynx, larynx, esophagus, liver, colorectum and breast, with the risk increasing in relation to the amount of alcohol consumed.



5. Manage Stress

Over 75% of primary care patient visits are now attributed to stress-related complaints. Chronic stress and the associated inflammation are linked to the development and progression of cardiovascular dysfunctions, diabetes, cancer, autoimmune syndromes and mental illnesses such as depression and anxiety disorders. Counselling patients in stress management through therapeutic interventions such as cognitive-based therapy, mindfulness, physical activity, medication, green spaces, nature and park prescriptions, may prevent or reduce the onset, severity, duration, and/or overall burden of stress-related illnesses.



6. Relationships

Social connections and relationships have an indisputable impact on our physical, mental and emotional health. Research shows that the single most important predictor of human happiness and long life is having strong social connections. Health-related measures like blood pressure and heart rate have demonstrated improvement even with short positive social interactions.

Lifestyle Medicine practitioners are not ignorant to the difficulties of behavioural change but acknowledge their impact on short and long-term health and quality of life. Educating patients about the influence and power of diet and lifestyle changes to transform their current and long-term health does not guarantee translation into behaviour change. For this reason, Lifestyle Medicine clinical skills have a greater emphasis on motivational interviewing and professional skills surrounding behaviour change. Health coaching is a powerful tool which facilitates a shift in motivation from extrinsic to intrinsic during lifestyle behaviour change.

Conclusion

The exciting promises of Lifestyle Medicine in preventing and reversing disease and maximising health and lifespan can only be fulfilled if clinicians and systems are proficient in assisting patients to set goals, have clear action plans, and assist them in navigating obstacles along their health journey with accountability and support.

For this reason, Adventist Healthcare has established the new and innovative ELIA Lifestyle Medicine Centre at the Sydney Adventist Hospital, which has long maintained a wellbeing and health promotion focus for patients since it opened in 1903.

The ELIA Lifestyle Medicine Centre offers individual Lifestyle Medicine consultations, as well as tailored 12-week clinical programs. Our interdisciplinary team comprises of Lifestyle Medicine Physicians, a Dietitian, Exercise Physiologist, Registered Nurses, Health and Wellness Coaches, and psychological care.

Find out more about Lifestyle Medicine and how the ELIA Lifestyle Medicine Centre can assist your patients by calling (02) 9480 6140 or visit www.elialmc.com.

AN ARTICLE
BY **Dr Sam Steele**

Case Study: Cancer rehabilitation – reconditioning support for people with cancer

After experiencing niggling pain that didn't resolve with time, Richard Fleming* thought he was going to be booking in for a hip replacement. However when he met with his doctor for the scan results, it was a complete shock to find he not only had cancer but that it had already spread to his bones.

"I'm a broadcast engineer, and at that time I was travelling throughout Australia, New Zealand, Papua New Guinea and Fiji designing and building radio stations," said Richard. "With the pain I was experiencing in my hip, I thought I'd just damaged something along the way. It turned out I had prostate cancer which had spread to other areas including my spine."

That was six years ago, and Richard and his family have been through many ups and downs along the cancer journey. "I started cancer treatment which included chemotherapy and a new radiation treatment (Lu-PSMA theranostics) which targets a protein that only lives in prostate cancer cells. The treatments slowed the cancer for awhile, but then the tumours started to compress my spinal nerve column, causing extreme pain and difficulty walking."

Richard's oncologist, Professor Gavin Marx, referred him to Dr Sam Steele, a rehabilitation physician and palliative physician at the San. At particular points during cancer treatment, patients and their families may need more comprehensive support, and cancer rehabilitation can be part of that.

Richard was the first cancer patient to go through the San's inpatient cancer rehabilitation program, focused on reconditioning for people living with cancer.

Dr Steele said, "The transition from cancer being a short, life-limiting disease to being more of a chronic disease in many cases, has led us to consider the needs of cancer patients within a rehab environment. Cancer treatments are becoming more effective, which means people are now living longer with cancer. Cancer complications and the side-effects of chemotherapy, radiotherapy, or surgery can leave some patients deconditioned. They may benefit from some extra help and support through rehabilitation, to regain their function."

What does cancer rehab involve?

While cancer rehab works on similar principles to general rehabilitation, there are additional complexities that need to be taken into account for people who have an underlying cancer. "The goal of cancer rehabilitation is to improve function, improve symptoms associated with cancer treatment, improve quality of life and reduce morbidity," said Dr Steele.

Things that are most commonly addressed in cancer rehab:

- Pain management
- Strength and range of motion
- Overall conditioning – improving exercise tolerance
- Managing other symptoms associated with cancer, such as fatigue and lethargy
- Nutrition advice
- Problem solving, such as equipment and services that patients may need at home to assist with their day-to-day function, to make things easier for their them
- Psychological support – having the social worker and clinical psychologists involved for both the patient and their families/partners/carers
- General functional gains through mobility/gait aids
- The ability to attend daily chemotherapy or radiation treatments while at the same time having rehab.

For Richard, a very important element of his recovery was good pain control. "My pain fluctuated, depending on what stage of the treatment cycle I was in. At first the pain medications were just tablets taken orally. With time, I began to be in pain whether I was sitting, lying down or walking, and the dosage of pain meds was increased. It got to the point where the dose of pain tablets that were needed to control the pain were making my brain fuzzy. I kept falling asleep, and I was in a mental fog all the time," recalled Richard.

The insertion of an implantable pain pump – or intrathecal pump – was offered to Richard as an option to consider for pain control. "The intrathecal pump is inserted under the patient's skin and delivers pain relief directly into the fluid surrounding the spinal cord," said Dr Steele. "When you bathe the spinal cord in pain relief medications, you only need tiny doses. It doesn't have the same systemic side-effects of taking tablets orally, because it goes directly to the spinal cord."

Richard started to notice a difference as the pain pump was adjusted to meet his needs. "In my case, the amount of morphine required to keep me comfortable was around 1/100th of what I used to take orally. My brain fog lessened; my thinking became clearer."

After a number of admissions to hospital with various cancer-related infections and complications, Richard was referred to cancer rehab for a period of reconditioning. "Over time, my condition had become quite degraded. The purpose of spending time in rehabilitation was so that I could get up and walking again. My four-wheeled walking frame became my best friend."

"The rehab team adapted their program to me, and I'd get into the rehab gym first thing in the morning and work really hard. And then I'd work in my own room as well – trying to impress the physios with my progress so they'd let me go home."

"It has taken my wife and I quite a long time to come to grips with what the future is, coping with the eventuality that I may pass away a lot younger than I would have otherwise been expected to live. At least I got to three score years and ten, and I still have my sense of humour!" And indeed, he does. When talking about one of his treatments, Richard quipped about the cost: "We felt the cost was worthwhile, because it was a targeted treatment (targeted specifically at prostate cancer cells). Plus, I didn't really want a new BMW to replace my Nissan Xtrail anyway!" he said with a laugh.

"Keeping a sense of humour is very important to me," added Richard. "I know the cancer journey is very hard on the whole family. My wife and I do sometimes have a cry together, but I usually try to laugh at anything I can."

Richard also brings joy to others by designing children's toys on his computer and giving the designs to 'The Shed' in his retirement village to be made into toys. "At the moment I sit out on the balcony, sandpapering and painting 1930s-style police cars and pull-along crickets – all made out of beautiful natural Australian timbers. I feel I can still take part, and if I can still pass on a smile – to my family and to others – that's really important to me."



*Richard Fleming
Mr Fleming has since passed away but this article has been published with the permission of his family.



Dr Sam Steele

MBBS, B AppSc (Physiotherapy)
FAFRM, FACHPM, MMed (Pain Management)

Dr Sam Steele studied as a Physiotherapist at The University of South Australia and subsequently worked in Australia and the United Kingdom. He went onto study Medicine at The University of Wollongong. He has completed fellowships in Rehabilitation Medicine and Palliative Medicine. He completed his training in several major hospitals in New South Wales including, Nepean Hospital, Liverpool Hospital, Camden and Campbelltown Hospital, Braeside Hospital, Blacktown Hospital and Prince of Wales Hospital.

Dr Steele has completed a Masters of Medicine in Pain Management through the University of Sydney.

He currently works at South Western Local Health District and Sydney Adventist Hospital in Palliative Care. He is the Clinical Lead for Cancer Rehabilitation at Sydney Adventist Hospital.

He is an active member of the Australia and New Zealand Society of Palliative Medicine (ANZSPM), Rehabilitation Medicine Society Australia and New Zealand (RMSANZ) and The International Society of Physical and Rehabilitation Medicine (ISPRM). He is a member of the Palliative Care Clinical Studies Collaborative (PaCCSC).

Dr Steele has a special interest in cancer pain, cancer rehabilitation and symptom management associated with chronic disease.

Specialist Expertise:

- Cancer Rehabilitation
- Pain & Symptom Management
- Palliative Care

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The San achieves highest ranking in NSW for outpatient cancer services again

For the sixth year in a row, the Integrated Cancer Centre at Sydney Adventist Hospital (the San) topped the State in the annual Outpatient Cancer Clinics Survey, showing that our commitment to whole-person care for patients with cancer is making a difference to their lives.

Each year, patients' experience of outpatient cancer care throughout NSW is independently surveyed by the Bureau of Health Information (BHI) in partnership with the Cancer Institute NSW. 8,280 patients from 43 facilities – public and private – were surveyed.

The purpose of the survey is to gain an understanding of patients' experience of multiple aspects of their cancer care. Patients were asked to evaluate their experience including: access to services, care planning, facilities, coordination of care, complications, follow-up and support.

In the report released on 25th October, 2023, the San's Integrated Cancer Centre (ICC) led the State by achieving the highest ranking with 12 measures being significantly higher than the NSW results – this was the highest number of any facility in the survey. Some of these measures included: overall experience of care received; well-organised care; polite and courteous staff; comfort and cleanliness of facilities; and how well the healthcare team worked together.

Professor Gavin Marx, Clinical Director of the Integrated Cancer Service at the San, expressed, "This is fantastic and humbling feedback from our patients. Earning the top state ranking consistently year after year is a clear demonstration of our entire team's unwavering dedication to providing the best possible care for those undergoing cancer treatment. It's a privilege to collaborate with such an exceptional team whose primary focus and driving force is to enhance the quality of care for our patients."

"While we are very proud of these results," said Prof Marx, "we are always striving to improve patient experience during cancer treatment. While it is great to know patients experience excellent cancer services here, we won't rest on these results – each year we continue to look for ways that we can improve our services."

The San's Integrated Cancer Centre operates in partnership with Icon Cancer Centre Wahroonga, which offers radiation therapy treatment for all cancer types. Icon's Clinical Director of Radiation Oncology, Dr Andrew Fong, says the results of the BHI report is affirmation "we're on the right track".

"The multidisciplinary team approach to cancer treatment – all in the one location – offers convenience for patients, and a concentration of expert clinicians and first-rate facilities," said Dr Fong. "It is a privilege to care for our patients and their families impacted by cancer. And it's good to know that the measures we take every day to improve the lives of those we care for is making a positive difference – despite a very difficult time in their lives."

Many factors contribute to Australia's high rates of cancer, and the San has long been committed to not only the early diagnosis and cutting-edge treatment of disease, but also prevention and research.

"The San's purpose and mission is whole-person health," said CEO Brett Goods. "And perhaps in no other area is this more important than in the area of cancer. We are very grateful to our superb team at the Integrated Cancer Centre."

"Receiving this recognition for the hospital's cancer services just spurs us on to deliver even better outcomes for our patients and their families. Alongside the ICC, our Cancer Support Services – as well as research – all contribute towards efforts to treat disease and promote survivorship and wellbeing for our community."

About Sydney Adventist Hospital

Sydney Adventist Hospital, fondly known as 'the San', is operated by Adventist HealthCare Limited. It has been a leading healthcare provider for 120 years (since opening in 1903) and has grown to become NSW's largest not-for-profit private hospital with more than 700 beds. The San is the biggest employer in the Hornsby Ku-ring-gai region.

www.sah.org.au

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