EDITION 2 2019

RED

In This Issue

Getting older with CF, $20m CF research funding, 65 Roses Day and much more!

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RED CEO MESSAGE

NIGEL BARKER, CEO

I am always surprised at the huge range of issues to highlight when I sit down to write my regular column for RED magazine.

In just the space of a few months we seem to have so much to talk about to meet our members' unquenchable thirst for knowledge and news.

Just last month Prof Barry Clements announced $20m in funding from the Medical Futures Research Fund to develop a compound which breaks down biofilms in the lungs of patients with CF and thus enhances the efficacy of existing antibiotics by orders of magnitude. He's also invented a special spacer for use with nebulisers. This spacer will ensure better delivery and less waste of the nebulised drugs and mitigate any perceived risks (albeit negligible) around passive nebulisation for care givers. Prof Clements hopes to bring the device to market sooner rather than later.

If you'd like to hear more about Prof Clements progress, see a special article in this edition of RED.

In the meantime, the Hollywood blockbuster 'Five Feet Apart' was released, which served to raise awareness of this most common of rare diseases. Members of CFWA have responded positively to this and Danielle Mercer from Mandurah used the opportunity to not only raise awareness, but also raised over $5,000 for CFWA through a film night fundraiser.

We are also now racing toward finalising the 13th Australasian CF Conference with guest speakers attending from around the world. We still have financial subsidies to allow parents and carers to attend from both metro and country areas. Apply now! These subsidies represent a significant financial saving and gives you the opportunity to network with other parents and care givers.

Talking about money, in this edition of RED we look at some of the unique financial challenges faced by people growing old with CF and yes, this will also be a major topic of discussion at the Conference with a highly-specialised expert from the Public Trustee.

Mental health is a big feature in 2019 and we are very proud of our CF Talk videos which focus on the needs of young adults. If you haven't seen them yet, please check out the latest in a series of six videos funded through Lotterywest.

By the time you read this, we will be well on our way to our biggest 65 Roses Day ever: Friday 24 May. You too can get involved (if you are not already one of the hundreds of volunteers wrapping or selling 15,000 roses). Help us paint the state with roses and raise funds for CFWA, from Broome to Albany and everywhere in between. Finally, time is running out to secure your tickets to the social event of the year: the Capel Vale Conquer Cystic Fibrosis Ball! I have mine and I am looking forward to a night of fun and fundraising as together, we strive ever closer to our vision of Lives Unaffected by CF.

DEADLINE FOR THE NEXT ISSUE
If you would like to contribute to our next edition for 2019, please contact us before 11 July.

DO WE HAVE YOUR CORRECT DETAILS?
If not, please let us know so we can keep you informed

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ON THE COVER
Cindy shares her thoughts on ageing with CF. Read her story on page 7

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Strange Bedfellows With a Triple-A Rating

At Cystic Fibrosis Australia (CFA), the approach is tried and tested. The three pillars of our advocacy are ...

- Access to Medicine
- Attention to Mental Health
- Assistance with the burden of the disease.

These are the benchmarks against which we judge decisions and we are confident that our guideposts have helped us reach our goals – and as always, with even greater aspirations on the horizon.

CFA's efforts to offset the burden of cystic fibrosis (CF) as a disease have brought us into a positive partnership with the PERX app. The developers of PERX have a background in behavioural economics and a determination to help Australians dealing with chronic disease.

The phone app manages and encourages active participation in tasks, exercise and medical regimes in a procedural way, helping patients to form strong habits and stay on top of complex routines. The PERX app is now a virtual PA for over 300 members of the Australian cystic fibrosis community – we are its biggest single customer group.

CFA's collaboration with PERX is important and ongoing. We provide prizes and incentives to our users on the PERX platform, while PERX provides feedback from CF users and continues to evolve and update its software to better cater to our unique needs.

The numbers continue to climb on all fronts. More users, more giveaways, more medication adherence and more user satisfaction result in the creation of solid health habits. In fact, 84% of CF users report that PERX has been highly useful in helping them manage their medication and our adherence rates have escalated from 71% to 78%.

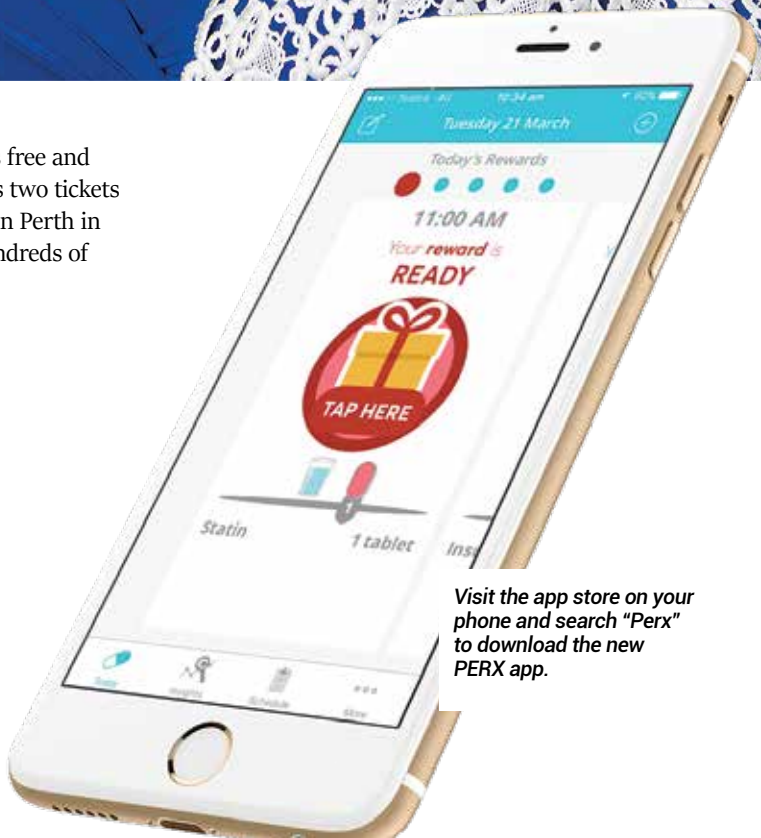
This month PERX is launching an activities section in the app with new flexible parameters to include suggestions from CF PERXs users. Activities, appointments, physio and exercise can all now win you great prizes.

When you think of CFA you don't automatically imagine iPhones and tech start-ups and Silicon Valley, but in order to stand on our Three Pillars we have had to think creatively and forge new partnerships.

Perhaps good advocacy makes strange bedfellows. And delivers Triple-A results!

Sign up now for PERX. It's free and this month's major prize is two tickets to the CF Lay Conference in Perth in August. There are also hundreds of vouchers to win.

Nettie Burke
CEO
Cystic Fibrosis Australia



Visit the app store on your phone and search "Perx" to download the new PERX app.

perx health

$20m in CF Research Funding!

The Medical Research Commercialisation Fund's Biomedical Translation Fund (MRCF BTF) announced on 15 March the launch of a new biotech company, Respirion Pharmaceuticals Pty Ltd (Respirion).

Respirion is developing a novel antibiotic therapy for the treatment of cystic fibrosis (CF), combining an inhaled version of an approved drug that is normally used to capture and dispose of excess metals in the bloodstream with the standard of care antibiotic in a novel formulation and device.

MRCF BTF has committed up to $20 million in stages in collaboration with the Telethon Kids Institute, where the technology was incubated and the spin-off created, and the Western Australian Department of Health where the product was tested. An initial trial using the technology saw an average 16 per cent improvement in the lung function of patients, demonstrating the exciting potential of the treatment to improve CF patient's quality and duration of life.

The MRCF BTF is a Federal Government-backed investment fund aimed at boosting Australia's late-stage biotech sector. This is the MRCF BTF's first investment in Western Australia.

Most of the recent attention in CF drug development has focused on addressing the genetic abnormality which causes the proteins that regulate the proper flow of water and chloride in and out of cells lining the lungs to be defective. While these new drugs have significantly improved outcomes for some CF patients with specific mutations, persistent antibiotic-resistant infections and declining lung function remain a problem for all patients.

Respirion's therapy is a combination of the antibiotic tobramycin with an adjuvant which helps break down the biofilms that bacteria erect to protect themselves in the lung. In a recent clinical study, the combination not only significantly improved the killing power of the antibiotic, but also improved the patient's overall lung function. Patients using the combination saw a 400 times greater reduction in bacterial load as well as an average 16 per cent improvement in lung function, compared to 5 per cent improvement for those patients using tobramycin alone. This demonstrates the broad potential of Respirion's therapy and represents a powerful new tool to potentially not only improve treatment of infection, but also a CF patient's quality and duration of life.

TELETHON KIDS INSTITUTE
Discover. Prevent. Cure.

The MRCF has committed up to AU$20m to Respirion in stages, to support larger clinical studies that will take place under FDA regulations in the US and Australia. The MRCF's investment is designed to validate the commercial and clinical potential for the therapy, kick start the initial studies and also potentially secure additional financial support for Respirion's development program.

For more information about this exciting announcement visit **www.bit.ly/TKI-19**

Let's Paint WA with Roses this May

We need your help this 65 Roses Day, Friday 24 May, to paint WA with roses for people living with cystic fibrosis

May is cystic fibrosis (CF) awareness month and Friday 24 May is our national day of CF awareness. Every year, on 65 Roses Day, we paint Western Australia (WA) with beautiful, fresh roses to raise funds and awareness for people living with CF.

We need you to help us spread 65 Roses Day across the state by taking roses to sell in your workplace, school or local community.

Funds raised from 65 Roses Day will support our vital service programs and research funding for children and adults living with CF in WA.

How You Can Get Involved

Take 65 Roses to Your Workplace or School

Your workplace or school is the perfect place to help raise awareness and funds for CF this 65 Roses Day.

Pre-order your roses on our website (no pre-payment required) and we'll deliver them to you on 65 Roses Day. Sell them for $5 a rose and deposit the funds raised to us after the event. It's that easy!

If you can't sell roses in your community, you can still support CF with a beautiful florist-arranged bouquet, delivered to a friend or loved one on 65 Roses Day, Friday 24 May.

Order your roses today at **www.cfwa.org.au/65roses** or get in touch with the team at **events@cfwa.org.au** or **08 6457 7333.**

Gift a Bouquet

Treat someone special (or yourself!) to a blooming bunch of florist-arranged roses on 65 Roses Day, 24 May 2019.

Every bunch and bouquet will make someone's day and help support our vital support services and CF research funding.

Posies are available for just $35 delivered. Order today at **www.cfwa.org.au/65roses**

Volunteer

To paint WA with beautiful roses this 65 Roses Day, we need an army of volunteers to help us!

Join our volunteer team and lend a hand for a couple of hours on Thursday 23 May as a delivery driver for our roses orders, or Friday 24 May to sell them on the streets of Perth and surrounding suburbs.

To see all our available volunteer opportunities and to join the team, visit **www.cfwa.org.au/65roses**

Thank you for helping paint WA with roses

Ageing Well with Cindy

Cindy is a highly-motivated lady with cystic fibrosis who shares with us some insights around what ageing well means for her.

When you were growing up, what sort of career options did you consider?

Bacteriology and working in labs because I was intrigued and wanted to know more about it, however, I decided that because I have cystic fibrosis (CF), it wasn't for me. At 13 I started cooking, going into hospitality and serving customers. I began to see how people's lives were better for the experience, it's not just about food or coffee, it's about relationships.

Why did you end up choosing to become a chef?

Hospitality and food provide flexible working options. I could do lunch or dinner; shorter shifts I can have a nap in between or even do my treatment in between. I did my apprenticeship in Broome. I worked on boats out of Broome and enjoyed the caring aspect, living with people and making sure that they had healthy eating options. After that I worked in mining; it was a closed kitchen with no relationship to customers which I didn't enjoy as much. So, after that I worked in a café in Augusta as a chef. I had a good mentor and learnt how to run the business; that was six years ago.

You now own a thriving café in the South West. What led you to make the decision to become a small business owner and what were some of the things that you did to plan for this?

Following this I decided to get my own café, Café Boranup in Augusta; it's done really well. I employ people from different backgrounds, people who add to the business. It's a kind of organic philosophy where staff are encouraged to have input and develop different ideas. Recently, I was approached by the Tourism Association to also run their cafés, so I'm now managing four cafés and employing 30-40 people depending on the season. I train others to cook so that they are able to duplicate the same product, always aiming for a good product and experience.

What big learning tips have you gained along the way?

- Change is a good thing
- Don't underestimate anyone
- Everyone has something to give
- Not everything or everyone is related to a monetary value

Has having your own business helped you have a better work-life balance and how do you manage this?

No. I work more, but I have flexibility and can take time off as I have people working for me. I also provide regular support to staff when I'm off and lots of training to make sure they are OK without me. I can do other things that are less taxing like trying new recipes at home.

What would you suggest to people considering running their own business?

- Take all the advice you can
- Do lots of research
- Start small and have a plan
- Don't quit your day job
- Don't start in debt
- Don't be afraid of failure and take calculated risks
- Do some small courses, e.g. Small Business Advisory offer training
- Look at YouTube and podcasts; they have great information, particularly around managing stress
- Know your own strengths and work on your weaknesses, e.g. develop your organisation skills and get a diary
- Book time out with friends and self

A-Z Guide to CFWA Services

A comprehensive list of services CFWA can offer to our WA community. We're here to help

Adult Counselling

Face to face or telephone counselling available to members 16+ years.

Advocacy

- Individual assistance, e.g. letters of support
- WA community issues, e.g. hospital WiFi
- National policy and awareness campaigns e.g. access to new drugs

Clinical Support and Advice (Nurse, Physiotherapist and Social Worker)

- General advice
- Treatment routines
- Nebuliser assistance
- Motivation

Education and Community Awareness

- School – teachers, school nurses and students
- Community – community groups, workplaces, individual and extended family
- Health Professional Education to metro and regional areas
- Community awareness to the general population, e.g. 65 Roses campaign

Equipment

- Nebulisers and exercise equipment available to members.
- Equipment education and advice

Financial Support

- Patient Support Subsidy
- Adult Regional Travel Subsidy
- Telethon Children's Regional Travel Subsidy
- Telethon Sibling and Offspring Camp Subsidy
- Adult PT, Gym and Activity Subsidy
- 2019 Australian CF Conference Subsidies
- Foodbank vouchers available
- Christmas hampers available for families

Funding for Research

We help fund research to advance treatment and understanding of cystic fibrosis.

Home Care Worker Service

An in-home service providing support with airway clearance, exercise, respite and light home duties.

Hospital in the Home

Hospital in the Home (HiTH) is a PCH-based program which is supported by CFWA. **Hospital Support**

- Weekly support visits to inpatients at both PCH and SCGH.
- Visits to Fiona Stanley Hospital as needed

Member Support Events

- Ladies' High Tea
- Parents' Retreat
- Parents and Carers Dinner
- Sibling and Offspring Camp

Newly Diagnosed

- Baby gift including newborn essentials and some useful resources
- Support to new families as directed by PCH

Personal Trainer

Personal training may be available in your area for CF members aged 16+.

Regional Support – Outreach*

- School and community education sessions including day care, community, workplace and extended families
- Health professional education including hospital staff and GPs
- Physiotherapy assistance, advice and education
- Nursing consultations, information and advice
- Counselling and social work support
- Networking opportunities and support of members
- Home care worker support in Bunbury and Busselton areas
- Skype sessions to provide ongoing support as required
- Hospital visits and support when in Perth
- Outreach dinners
- Regional travel subsidies

*Thanks to the generous support of Telethon

Resources

We have a vast array of both publications and on online resources available, including but not limited to:

CFSmart- Online resources for the education community **www.cfsmart.org.au**

Rozee- Rozee magazine for children 8 - 14 years

CF Talk - Videos and online magazines for young people, parents and carers

CF Fit - Exercise resources to assist with exercising at home or in the gym

CF Food - Nutritional resources to assist with age appropriate dietary information

CF Fact - A range of fact sheets

RED Magazine - Quarterly magazine providing details about upcoming events and topical information.

All online resources can be found at **www.cfwa.org.au and www.cfsmart.org**

Seminars

Regional sessions and workshops highlighting new research and care for the CF community.

Transition Support

Assistance with transition from Perth Children's Hospital to Sir Charles Gairdner Hospital

Transplant Support

Both pre- and post-transplant support available including:

- Home care worker and general support including cleaning
- Counselling
- Publications and online resources

Please visit our website **www.cfwa.org.au** for more information about the services we offer. Our programs and resources evolve and change as needs arise and we are always happy to discuss how we can better help you. Please call and speak to Kathryn or any of her services team on **08 6457 7333** if you would like further assistance.

Wellbeing and Ageing

Ageing for all of us can, at times, be challenging, however, for people with CF there can be extra issues to consider. In this article, we explore areas of loss, ways to manage, and hopefully thrive.

According to the National Health Survey 2017-18, 47.3% of Australians report at least one prominent chronic health condition. Chronic, by definition, is something deemed "not curable". Chronic illnesses, including CF, can also be very different for each individual and a person born with CF today will have a very different medical and psycho-social experience to someone born 20 years ago.

Regardless of how an illness manifests, the first diagnosis, and consequent changes to health, requires an adjustment to how we perceive ourselves and our relationships. Initially, there may be feelings of grief and loss: denial, sadness, anger, and hopefully, acceptance and a positive adjustment to our new situation. There is new research advising that grief is not a linear process; it's up and down and we can swing from being happy, sad and irritable, balancing a myriad of emotions. These feelings when experiencing a loss are "normal", not requiring pathologising (requiring treatment, intervention or drugs) or closure. For most of us we can learn to live with grief, balancing an array of emotions whilst still moving forward in a positive way.

People with CF and their families often experience uncertainty and worry about what the future progression of the disease might look like. Research has shown tolerance for uncertainty varies for different people and at different times of our lives dependent upon our life experiences, values and individual perceptions. People with a high tolerance for uncertainty are more likely to cope better with anxiety and worry, procrastinate less, be less avoidant and feel more confident to manage CF treatment and other issues.

Tolerating Uncertainty

Understanding what thoughts set off our stress response will help us to better manage our feelings and behaviour. Some useful questions you could ask yourself:

- **Is our thought fact or fiction?** We have loads of thoughts that can be unnecessarily upsetting and may not be facts, e.g. thoughts like "I can't cope" may make us feel emotionally down.
- **What is the evidence against this thought?** When we are feeling unwell, physically or emotionally, it's easy to remember all the other times we felt horrible, imagining the worst and feeling like "we can't cope". These thoughts can be challenged; we can remind ourselves of all the other times that we have coped and managed through hard times.

Practice Tolerating Uncertainty

Changing our behaviours can also lead to changing our thoughts and feelings around uncertainty. An example of this is avoidant behaviour which is often used to try and evade anxiety, however, the more we try to avoid anxiety-provoking situations the more we are affected. Changing behaviour can be difficult so it's important to start small and build confidence.

Most people thrive on certainty and can cope with a lot more when they know what the outcome is going to be, even if it is bad news. Waiting periods can be particularly bad if we are feeling uncertain or unwell. We know that many people feel relieved when they are given a diagnosis, even if the news is not so good as they feel they can then manage and deal with whatever they need to deal with (Ubel, 2006).

"Illness does not dictate a specific response; however, it does require us to make a reassessment of our identity"

Giving Up and Demoralisation

For some people there is a feeling of relentless struggle and hopelessness in trying to maintain recommended treatments and health, resulting in significant loss of meaning and motivation. Demoralisation is seen as different to depression and may not be identified and treated, however, is considered to be "the giving/given up complex" describing a state of discouragement and feeling unable to cope (Engel, 1967, in Kissane et al, 2001).

Demoralisation is seen as a pathway to decreased physical health, particularly if someone is no longer motivated to maintain recommended treatments. Seligman also discusses "learned helplessness" whereby people surrender their power, feeling they have no control over their health or their environment.

Feelings of loss and control and demoralisation are also experienced by carers. Boss (2011) advises that the best antidote is human connection, support, understanding, compassion and empathy.

Learnt Optimism

Martin Seligman has done a lot of successful work with trauma survivors, particularly war veterans. He argues that we can all learn to be optimistic. This doesn't diminish our experiences of loss; however, it helps us to feel optimistic and confident in our coping strategies (see references for short videos on learnt optimism).

Balance Feelings of Grief and Loss with Optimism

People with chronic health conditions are affected in every aspect of their being: physical, emotional, social, financial and spiritual; it may feel that everything that is important to us is threatened. Illness has variable responses requiring us to make a re-assessment of our identity and our perceptions of self and relationships.

Hope, Meaning and Spirituality

Who am I really? Why am I here? What purpose does my life serve?

Greater significance is given to the health values of a "spiritual life". Studies have shown that religion and faith can help to promote good health and ward off disease, offering social supports and a philosophy that all things have a purpose. For some, it's not religion but the quality of personal relationships, love of nature and a desire to balance our inner needs with the rest of the world.

"We are at our best when we dedicate time to something bigger than ourselves. This might be religious faith, community work, family, politics, a charity, a professional or creative goal" – Martin Seligman

Family Meetings – Enhancement of Family

Open, positive communication is key; however, many families struggle to positively articulate feelings of grief and loss. Sometimes seeking professional assistance to safely discuss feelings offers a safe space and new strategies of coping.

Engagement with Treating Team and Care Plan

The relationship should feel collaborative to build confidence and self esteem in managing treatment. Taking notes at appointments, asking questions and being honest about limitations or inability to manage is important. The treatment team will make decisions on the information that you give them and also help you to manage your treatment load. The hospital social workers are also really good at working with individuals and families in developing plans and setting goals; you can request an appointment if you are struggling.

Connection and Supportive Relationships

People who have supportive relationships feel happier and have increased ability to manage physical and emotional issues. Healthy relationships take time, commitment, respect, trust, understanding, honesty and open communication. Seeking help to build self-awareness and improve our relationships can be very positive.

The unique role of online support groups, e.g. CF Talk for Adults, and CFWA events for carers, help to build connection, acceptance and tolerance for our own situation.

Summary

There is no health without mental health. Be kind to yourself; having CF or caring for someone with CF can be challenging. Feelings of grief and loss are normal, however, there is also lots of support available. If you would like to discuss support options, please contact Kathryn **servicesmanager@cfwa.org.au**

References and further information

- Tolerating Uncertainty **www.anxietycanada.com/articles/intolerance-of-uncertainty**
- Learned Optimism – Martin Seligman **https://youtu.be/6tN9wMbdqZU**
- Identity **www.psychologytoday.com/au/blog/science-choice/201412/basics-identity**
- Healthy relationships **www.healthdirect.gov.au/building-healthy-relationships**
- **www.ncbi.nlm.nih.gov/pubmed/12406115**
- **www.abs.gov.au/ausstats/abs@.nsf/mf/4364.0.55.001**
- Beck, A.T., Kovacs, M., Weisman, A.D. (1975). Hopelessness and suicidal behavior. Journal of the American Medical Association, 1975, 234, 1146-1149.
- Boss, P. (2011). Loving someone who has dementia: how to find hope while coping with stress and grief. San Francisco, CA: Jossey-Bass.

Managing Health and Ageing

Advances achieved over the years has transformed Cystic Fibrosis (CF) from a childhood disease to a long-term manageable chronic condition with approximately 50% of the Australian CF population living well into adulthood. As people with CF are living longer, they are facing new challenges and issues compared to those they may have faced in their earlier years.

As people with CF get older, their burden of care increases and they may find they are taking more medications and requiring more treatments. This may impact their everyday life; managing CF while coping with work, relationships and family can prove to be additionally demanding. Below, we explore some of the medical issues that ageing can bring and how they can be managed.

Nutrition

The link between poor nutritional status and worsening lung function highlights the importance of eating well for people with CF. This can be a struggle for many despite the intake of a high-fat diet and dietary supplements. During these times, extra nutrition may be recommended for night feeds via a naso-gastric tube or PEG (percutaneous endoscopic gastrostomy).

Cystic Fibrosis-Related Diabetes

With increasing age there is an increase in the incidence of CF-related diabetes (CFRD). CFRD may be triggered by an exacerbation of CF, medication, or it could be a gradual onset. People with CFRD need to continue with the recommended healthy fat, high-energy diet. Insulin is the treatment of choice for people with CFRD.

Gastrointestinal Complications

A number of studies show that as CF patients age, they are at a higher risk of developing benign (non-cancerous) polyps on the mucous membrane of the large intestine and are more likely than other types of large intestinal polyps to become cancerous. This has prompted screening for people with CF to have colonoscopy screening by age forty.

People with CF are also at an increased risk of developing secondary cancers. Some CF centres are now recommending additional screening.

Bone Disease

Porous bones – low bone mineral density (BMD) – is a common complication in adolescent and adult people with CF as their health declines. The most common risk factors include: recurrent respiratory infections, poor nutrition, lack of weight bearing exercise, delayed puberty, steroid treatments, vitamin D, calcium and vitamin K deficiencies. BMD, also known as osteopenia, can lead to osteoporosis where bones become weak and brittle and more prone to fractures.

Risk factors should be minimised, and dietary intake of calcium and vitamin D should be optimised. CF researchers recommend bone densitometry scans (dual-energy x-ray absorptiometry tests – DXA scans) everyone to five years, depending on the results (ECFS best practice guidelines: the 2018 revision).

Respiratory Complications

People with CF may develop a variety of complications as they age. Although they may occur infrequently, they are more common with advancing disease severity.

Pneumothorax

A spontaneous pneumothorax is an abnormal accumulation of air in the space between the lungs and the chest cavity (called the pleural space) and can lead to the partial or complete collapse of a lung (Genetics Home Reference 2019). Spontaneous pneumothorax is a complication in people with CF and occurs more frequently with age, in those with more advanced disease as well as in those who test positive for certain infections. Treatment is by the insertion of intercostal tube drainage and pain control. For recurrent pneumothorax some people may require surgical intervention.

Haemoptysis

Haemoptysis is a medical term for coughing up blood and is a common complication in CF. It may range in severity from scant (<5 mL) to massive (>240 mL). Clear guidance should be given from the CF team as to when and who to call if haemoptysis occurs. Those with mild haemoptysis may need treatment with antibiotics. For those with moderate or massive haemoptysis, medications such as tranexamic acid may be given and bronchial artery embolization (BAE) may be performed.

Respiratory Failure

As CF lung disease progresses to the advanced stages of airway obstruction and eventual respiratory failure discussions about lung transplantation and advanced healthcare directives should already be in place.

Sinus Disease

Chronic sinusitis, with or without nasal polyps, is common and can cause significant symptoms for those with CF. Routine evaluation of sinus disease and treatment is recommended as it could be the source of lower airways disease. Treatment of CF-related sinusitis can range from antibiotics, nasal steroids, mechanical clearance with saline or antibiotic flushes, antihistamines and decongestants to surgical intervention. CFTR modulators may also prove significant in improved outcomes for those affected by sino-nasal disease.

Liver and Pancreas Complications

Those with CF who are pancreatic insufficient may have evidence of liver disease ranging in severity from mild to end-stage cirrhosis. CF-related liver disease (CFLD) usually presents before the age of twenty years and can lead to complications

such as portal hypertension. CFLD can also lead to liver failure.

Early identification of advancing CFLD allows for potential monitoring and for treatment of complications. Patients with portal hypertension should be referred to a gastroenterologist for screening and management. This may include immunisation for hepatitis A and B, avoidance of NSAIDs and alcohol and monitoring of liver function. Dosing of some CFTR modulator and corrector drugs may need adjustment as these may contribute to liver impairment.

For those with advanced stage liver disease, a pathway for appropriate referral to a liver transplant program should be in place.

Pancreatitis

Pancreatitis is a less common complication but can be troublesome for CF individuals with pancreatic sufficiency. Recurrent pancreatitis can result in the transition to pancreatic insufficiency. Recently developed CFTR modifying drugs may play a role in the treatment of pancreatitis.

Nephrolithiasis

Nephrolithiasis refers to kidney stones and this is common complication in people with CF. Increased fluids to maintain high urine output combined with a low -oxalate and high-calcium diet is appropriate management for these patients. Limiting oxalate-rich foods may help reduce the risk of forming new stones. Foods to avoid which are high in oxalate are nuts, rhubarb, beets, All Bran®, buckwheat flour and sesame seeds.

Referral to a specialist nephrologist and/or urologist may be indicated for complicated nephrolithiasis.

Arthropathy

Arthropathy is a term for any disease of the joints. Joint pain in CF is a common symptom in CF, however, arthropathy is poorly understood. Treatment is generally with pain relief medications and anti-inflammatory agents.

Fertility and Pregnancy

98% of men with CF are infertile because the vas deferens does not develop properly or is absent. Sperm analysis should be available for those interested in knowing their status. Access to counselling regarding fertility options is advised. A specialised form of IVF called intracytoplasmic sperm injection (ICS), where sperm is collected and injected into a partner's egg, is an assisted reproductive technique that may be a suitable option.

In women, puberty may be delayed, however, most girls with CF have normal menstrual cycles. Amenorrhoea – the absence of periods – may occur in women of reproductive age particularly if they have a low BMI and a reduced percentage of body fat. Most women who have CF can fall pregnant without difficulty, however, fertility problems in CF are addressed in the same way as for other women. It is recommended that women with CF who are considering falling pregnant consult with their CF team.

Women with CF are always considered a high-risk pregnancy because of the potential complications and should always be managed by an experienced obstetrician.

Transplantation and End-of-Life Issues

An established therapy for end-stage lung and liver disease in people with CF is transplantation. In some cases, transplant is not a suitable option or does not occur for various reasons. Outcomes for people with CF undergoing lung transplantation have continued to improve with 10-year survival rates approaching 50% (ECFS best practice guidelines 2018 revision).

Barriers to referral for transplant assessment remain and regular and detailed communication with the transplant services is vital.

Psychosocial Support

For people with CF, their family's life can be emotionally and physically challenging. The ability to deal with everyday life, study, work and unexpected life events are influenced by health decline and the increased burden of care. The need for annual mental health screening and good psychosocial care is well documented. Referral and access to appropriate social work and psychiatric support is recommended.

Any secondary diagnosis can understandably cause anxiety and frustration and patients will require added support and education. Managing another chronic illness or complication adds to the burden of care. Psychological and counselling support is paramount to ensure best outcomes.

If you are concerned about any of the issues raised in this article, please discuss them with you specialist CF team or please feel free to contact Cystic Fibrosis WA to explore community support options that may be available for you on **servicesmanger@cfwa.org.au** or **08 6457 7333**

Resources:

For additional information please see our Factsheets

www.cfwa.org.au/what-we-offer/resources/#fact-sheets

References:

ECFS BEST PRACTICE GUIDELINES: THE 2018 REVISION **www.cysticfibrosisjournal.com/article/S1569-1993(18)30029-8/fulltext**

GROWING OLDER WITH CYSTIC FIBROSIS **www.cysticfibrosis.org.uk/life-with-cystic-fibrosis/growing-old**

PNEUMOTHORAX IN CYSTIC FIBROSIS: **www.ncbi.nlm.nih.gov/pmc/articles/PMC4203988**

PNEUMOTHORAX AND HAEMOPTYSIS CLINICAL CARE GUIDELINES **www.cff.org/Care/Clinical-Care-Guidelines/Respiratory-Clinical-Care-Guidelines/Pneumothorax-and-Hemoptysis-Clinical-Care-Guidelines**

Liz's Volunteering Role

Many of you would remember Liz as the Clinical Nurse Consultant at Princess Margaret Hospital (PMH).
She has just celebrated 12 month's retirement; however, she can't sit still.
In this article, she discusses volunteering at CFWA

Why Volunteering?

I worked with people who have CF and their families for over 30 years. I thoroughly enjoyed it, even the challenges, and especially the feeling that I could make a real difference.

Volunteering at CFWA seemed like an obvious choice. I'm also working on a small project at the Telethon Kids Institute.

What is Your Role at CFWA?

Working with young people and their families in transition from Perth Children's Hospital (PCH) to the adult hospital. The ease of transition will be dependent on many factors such as current health and lifestyle. For some families and young people with CF, they have little or no difficulty in transitioning. However, for others, transition is a much more difficult time.

If your child is due to move to the adult hospital, Liz is still around and happy to catch up. Please call CFWA for further information.

Advanced Care Planning in Cystic Fibrosis

Advanced Care Planning in cystic fibrosis (CF) is particularly important because of the unique aspects of the disease. People with CF experience a slow decline in lung function and complications may occur over a long period of time. Planning should reflect and respect each person's individual journey and decisions.

Despite advances in treatment and increased longevity, people with CF will experience declining health. The major factors associated with deteriorating health are age, chronic lung infections, bacterial colonisations and the added burden of associated CF comorbidities.

Those with severe disease have complex needs and may experience accelerated decline and increased symptoms such as pain, breathlessness, cough, lack of sleep and anxiety and depression. The majority of these people will have discussions about lung transplant, and many will be referred for assessment and accepted onto the wait list. These patients will need both active CF treatments to prevent further deterioration and holistic supportive care. However, for a small proportion of people with CF, lung transplant may not be an option, or their transplanted organs may fail.

All people with CF who are approaching the severe stage of their disease require honest, open communication with all members of their CF team and other specialist providers. This is essential for the person to make informed decisions and allow for effective goal setting during the remainder of their life. Variable disease progression can mean that end of life can be difficult to predict and providing optimal care may be challenging due to this variation in disease. Advanced care planning discusses what needs to occur during periods when the person is relatively well and before a reoccurring life-threatening health crisis.

So, what is Advanced care planning and who should be involved? Advanced care planning is an ongoing discussion between a patient and their health care team, family and carers which reflects their values, beliefs, treatment and care options. The focus is on the patient's wishes for their future treatment and care should they no longer be able to make or communicate their decisions.

Preferably, Advanced care planning will result in a formal, written advanced health directive to help ensure the patient's preferences are respected at a time when they are unable to make decisions for themselves or cannot communicate. It is only used when the patient loses the capacity to make decisions and express their preferences. The lack of a written document, which is signed and witnessed, will likely give rise to genuine and reasonable doubt about validity and current applicability. An advanced health directive is a form recognised by law under the Guardianship and Administration Act 1990.

To make an advanced care directive you must be:

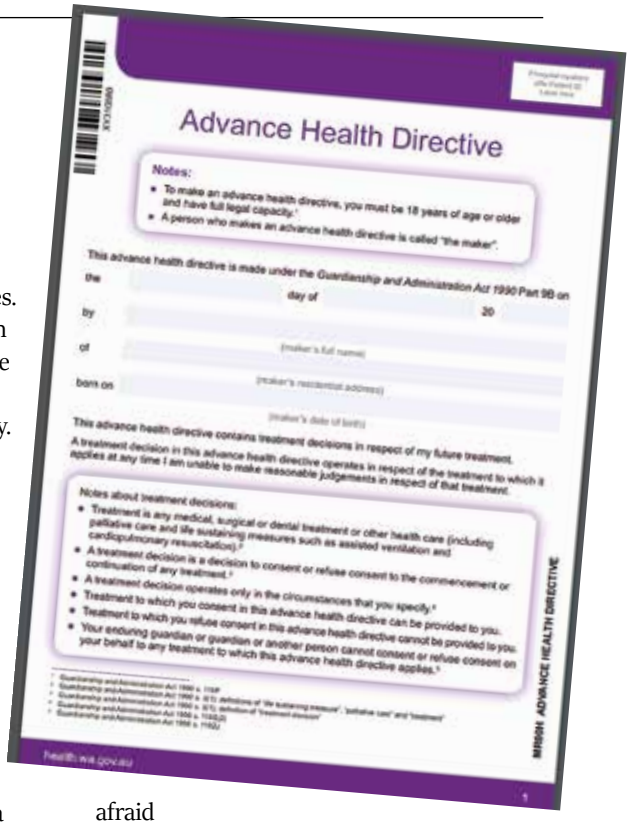
- 18 years or older
- Have full legal capacity

An advanced health directive will be strengthened if the person appoints a substitute decision-maker who has a clear understanding of their preferences and is willing to be a strong advocate for them.

In an emergency, the doctors will ultimately make the medical decisions but will take the person's wishes into account and refer to their advanced care directive and/or talk to the person's substitute decision-maker and family. If the advanced care directive is not immediately available, life-supporting measures may be commenced until the treating clinicians discuss expressed wishes with the substitute decision-maker and family.

If the person does not have a directive in place and in the event of serious illness the doctors will make treatment decisions based on the best interests of the person. This may include treatments that the person may not want.

Talking about end-of-life can be difficult for most people and starting the conversation can be the hardest part. You shouldn't be afraid to raise the subject with someone in your team that you trust.


Advance Health Directive

Notes:
- To make an advance health directive, you must be 18 years of age or older and have full legal capacity.
- A person who makes an advance health directive is called "the maker".

This advance health directive is made under the Guardianship and Administration Act 1990 Part 9B on
the day of 20
by
of
born on

This advance health directive contains treatment decisions in respect of my future treatment.
A treatment decision in this advance health directive operates in respect of the treatment to which it applies at any time I am unable to make reasonable judgements in respect of that treatment.

Notes about treatment decisions:
- Treatment is any medical, surgical or dental treatment or other health care (including palliative care and life sustaining measures such as assisted ventilation and cardiopulmonary resuscitation).
- A treatment decision is a decision to consent or refuse consent to the commencement or continuation of any treatment.
- A treatment decision operates only in the circumstances that you specify.
- Treatment to which you consent in this advance health directive cannot be provided to you.
- Treatment to which you refuse consent in this advance health directive cannot be provided to you.
- Your enduring guardian or guardian or another person cannot consent or refuse consent on your behalf to any treatment to which this advance health directive applies.

health.wa.gov.au
MR5004 ADVANCE HEALTH DIRECTIVE

For more information visit Advanced Care Planning Australia: **www.advancecareplanning.org.au**

To make an advanced health directive you may obtain a form by:

1. Downloading and printing the form: **www.health.wa.gov.au/advancecareplanning**
2. Email: **acp@health.wa.gov.au**

Alternatively, you can speak to someone from WA Cancer and Palliative Care Network (WACPCN) of the Department of Health on **08 9222 2300.**

References:

www.advancecareplanning.org.au/resources/advance-care-planning-for-your-state-territory/wa

www.advancecareplanning.org.au/for-health-and-care-workers

MONEY SMART

The Australian Securities & Investments Commission (ASIC) Money Smart provide some great resources, including online budgeting plans and apps for tracking spending. This is what they recommend to enhance your financial intelligence and save for your future.

1. Create a budget – free online budget planner
2. Track your spending – free easy to use app
3. Pay off your debts – free credit card calculator
4. Start a savings plan –free savings goal calculator
5. Build your super – use the retirement planner
6. Take charge of your home loan – free mortgage calculator or mortgage switching calculator
7. Protect the important things – insurance information
8. Invest wisely – free investment toolkit
9. Financial advice – free financial advice toolkit
10. Extra help if needed – links for financial counselling, legal advice and urgent money help

For more information visit the Money Smart website.
www.moneysmart.gov.au/managing-your-money/get-your-money-on-track

ASIC's MONEYSMART
Financial guidance you can trust

CF Research News

The aim of CF Research News is to bridge the gap between people with cystic fibrosis (CF) and the researchers investigating CF, providing access to patients, parents, relatives, friends and caregivers to all scientific work published in the Journal of Cystic Fibrosis (JCF). The following article was published in October 2018.

FAILURE TO CONCEIVE IN WOMEN WITH CF IS ASSOCIATED WITH PANCREATIC INSUFFICIENCY AND ADVANCING AGE

Authors: Michal Shteinberg, Adi Ben Lulu, Damian G Downey, Zeev Blumenfeld, Christine Rousset-Jablonski, Marie Percival, Alessandra Colombo, Nili Stein, Galit Livnat, Michal Gur, Lea Bentur, Huda Mussaffi, Hannah Blau, Ifat Sarouk, Adi Dagan, Eitan Kerem, Micha Aviram, Elie Picard, Stefano Aliberti, Antonio Álvarez, Javier Perez Miranda, Eva Polverino, Isabelle Durieu, J. Stuart Elborn, Malena Cohen- Cymberknoh

What was your research question?

In men with CF, infertility (reduced ability to naturally have children) is very common due to the effect CFTR mutations can have on male reproductive organs. Women with CF are expected to have normal fertility. However, infertility is frequently found to be a problem in women with CF. Our research questions were: 1. How common is infertility in women with CF? and 2. Are there subgroups of women with increased risk of infertility?

Why is this important?

As people with CF now reach adult age, starting a family is one of the life goals people with CF may try to achieve. Infertility in women is now being successfully treated with assisted reproductive technology- such as hormonal therapy, intrauterine insemination and IVF (in vitro fertilization). If it is recognized that certain women with CF have a higher risk of being infertile, they may be referred to assisted reproduction facilities sooner.

What did you do?

In this study, 11 CF centres from 5 countries participated. Data were collected on 605 adult women with CF. We defined infertility as inability to become pregnant after 12 months of trying, and subfertility as pregnancy subsequently achieved with treatment. We also collected data on CF mutations, pancreatic insufficient status (having to take pancreatic enzyme replacement), diagnosis of diabetes, lung function, nutritional status, exacerbations in the year prior to attempts of pregnancy, and presence or type of bacteria in sputum.

What did you find?

Out of 605 women, 241 attempted pregnancy. Of these, 84 (35%) had subfertility or infertility. On average, females who attempted pregnancy were older than women who did not but were also healthier than women who did not. Women with subfertility or infertility were significantly older than women with normal fertility (subfertility was 3.9 times higher for women over 31 years), with the average age for subfertile women being 34 compared to 28 years for fertile women). Woman with subfertility or infertility were more likely to be pancreatic insufficient (subfertility 1.9 times higher in pancreatic insufficient women); 42% of women who were pancreatic insufficient were subfertile compared to only 29% of women who were pancreatic sufficient.

What does this mean and reasons for caution?

Overall, women with CF are at increased risk of subfertility: 35% is higher than expected based upon 15% subfertility in the general population. Women who are pancreatic insufficient (and need to take pancreatic enzyme replacement) should be informed that their risk of having difficulty conceiving is higher, and they should be referred early to assisted reproductive technology, especially when at an advancing age. Some caveats of this study is that data were collected from past files, therefore concerted attempts to conceive may have been missed in some cases. Also, other factors which may have been difficult to determine from past files (such as mutations and how well diabetes was controlled) may also be important in determining risk of infertility.

What's next?

In this study we did not assess the reasons for infertility, this may be determined in future studies. It would be interesting to assess whether drugs that affect the function of the CF protein (CFTR modulators such as Kalydeco and Orkambi) improve infertility in women with CF.

Original manuscript citation in PubMed:

https://www.ncbi.nlm.nih.gov/pubmed/?term=FAILURE+TO+CONCEIVE+IN+WOMEN+WITH+CF

Albany Outreach Dinner


Albany 65 roses day 2018


Albany Outreach Education

Albany Outreach

Regional outreach trips are a highlight for our team and recently Sharon and Marnie visited Albany to provide health professional education and catch up with local members.

Cystic fibrosis (CF) education was provided to nurses at the Albany Hospital with a special guest appearance from a local CF mum. The session was of great value to the staff, receiving a personal insight into a family's journey with CF. They had an opportunity to ask questions, gaining greater knowledge and empathy into the daily challenges faced and the impact of living so far away from a tertiary centre.

Our member dinner, held at the White Star Hotel, was an evening full of chatter and laughs as members reunited and caught up on the latest news. Let the countdown begin until the next Albany get-together!

Also attending the dinner were the Albany volunteer team who annually run a 65 Roses Day wrapping and selling day in their local community. They brainstormed ideas and made plans for their awareness and fundraising efforts for 2019.

Cystic Fibrosis WA (CFWA) is soon to launch a new online platform for health professionals which will allow for easy access to current CF information and related resources, CF research articles and updates on the latest CF treatments. Our team was keen to get this news out to local GPs and visited a few GP practices, providing them with CF resources and letting them know about our future plans.

If you live regionally, CFWA is able to travel to your regional centre. We offer diverse services which can include school and community education, health professional education, physio and nursing advice and support, counselling and social work support, regional subsidies and carer support events.

For further information please contact us on **08 6457 7333** or visit **www.cfwa.org.au**

Royal Children's Hospital Cystic Fibrosis Team Visit Perth Children's Hospital

The cystic fibrosis team from Royal Children's Hospital Melbourne visited Perth Children's Hospital (PCH) recently to understand how the cystic fibrosis (CF) team from PCH achieve the highest outcomes for CF patients in Australia.

Congratulations to the team from PCH on a fantastic demonstration of collaboration!

Helping to Achieve the Vision

Rachael Hosking, mother of a son with cystic fibrosis and a CFWA board member, shares some life experiences and why she is leaving a gift in her will to CFWA.

My son, James, was diagnosed with cystic fibrosis (CF) just before Christmas and his 4th birthday.

At that time, all my husband and I knew was that Telethon raised money to help kids with CF and that, therefore, it was really bad that our beautiful boy had CF.

James was born a few months before the introduction of newborn screening, so we had a tough few years when he was little, with repeated severe chest infections, hospitalisations and constant coughing.

Since diagnosis, James and the rest of our family have been on a journey, learning how to adjust to life with CF. The challenges will be known by any family affected by this horrible disease: getting the physio and nebuliser done before school; packing a high fat high salt lunch; not forgetting the enzymes and all the other pills; making swimming lessons twice a week; keeping away from people with colds; finding a parking spot at PMH for clinic; dealing with the boredom of time in hospital; and, of course, coughing, coughing, coughing.

My amazing son, James, has patiently waited for hours at clinic visits, overcome his needle phobia and proclivity to faint when doing blood tests, put up with countless spirometry and other tests, taken thousands of pills a handful at a time, swam so much for physio that he made the interschool swim squad, endured muscle pains from lack of salt and nearly passed out running a relay for lack of oxygen. James is now 19 and in his second year at university and is learning to manage his own health in the adult world of Charlies [Sir Charles Gairdner Hospital].

The reality is that CF affects not only my beautiful son's physical health but also his mental health. He faces many challenges that most people his age don't, and really struggles to stay motivated and positive at times.

As a CF parent, I am determined to make a difference. I joined CFWA's board after my dear friends and Charlie's respiratory team nurses, Siobhan Dormer and Tara Hannon, put the idea to Sue Morey, who was then a CFWA board member. Siobhan and Tara thought my background, parent of a person with CF and lawyer, were a good combination. As a board member, I have a role in making a difference to people living with CF. I see the scope and impact of the services CFWA provides to people living with CF. I know that CFWA makes a difference to the lives of people with CF and families like mine. I know that CFWA is working towards delivering on its vision of 'lives unaffected by CF'. I know that we have to keep persevering until a cure is found.

In 2017, I embarked on another journey. I was diagnosed with metastatic breast cancer. I thought I had hurt my neck gardening. In fact, my vertebrae were disintegrating due to the cancer.

I read somewhere that cancer is a word, not a sentence, and everything in life is a choice. My choice is to live the rest of my life living, not dying. Whilst my health has now recovered, having cancer has caused me to think both about what I want to accomplish in the rest of my life and what legacy I want to leave. How do I want to be remembered?

There are so many things on my bucket list it fills a foolscap page: everything from learning French to swimming with the whale sharks. Another of those things is to encourage a philanthropic spirit in my children by encouraging them to be kind and generous and to help others. To this end, I am leaving a gift in my will to CFWA as an example to them and to make a difference in the future.

I know that the team at CFWA will use my future gift prudently and apply it to help people with CF and other families like mine. I know that my gift will help us move closer to a world where lives, like that of my son James, are unaffected by CF.

CFWA thanks Rachael for sharing her commitment to the vision of 'Lives unaffected by CF'. If you would like discuss leaving a gift in your will, please contact Karen De Lore at **bequests@cfwa.org.au** or **08 6457 7333.**

Little Steps, Big Achievements

The HBF Run will be back in May and some of our smallest team members will be making their debut run for cystic fibrosis!

Norah, who has cystic fibrosis, and her brother, Cole, have participated in the run for the last two years as part of the team 'For Norah'. In the past they have been along for the ride in their pram. This year, they have told their mum and dad, Nyssa and Cody, to park the pram because they want to take their own steps for CF.

Nyssa, Cody, Norah and Cole have a huge team of dedicated supporters who join them every year, wearing red CF shirts in amongst the sea of blue to raise as much awareness as they can. You may have even seen them in the HBF Run TV ads recently!

But they are not the only runners who are making CF their reason to run this year. We have 15 other runners who are raising funds and awareness for Cystic Fibrosis WA.

We wish all our teams a safe and happy run and can't wait to share how much of a difference their efforts will make in our community after the big day!

If you are running and would like to help by raising awareness and funds for CF, please contact Marnie on **events@cfwa.org.au** or **08 6457 7333** to find out how.

Annual Ladies' High Tea

Female carers (and their +1) are invited to join us for an afternoon of high tea, champagne and lots of laughter as we get creative and paint a rose to celebrate 65 Roses Day!

A great opportunity to meet up with other ladies in a fun and relaxed environment.

When: Saturday 4 May 2019
2.45pm – 6pm
Where: The Niche, 11 Aberdare Rd
(cnr Hospital Ave), Nedlands
Cost: $25 per person
RSVP: http://tinyurl.com/h3qlrxn

Guest Speakers: Bianca, one of the stars from our CF Talk videos, will be coming along to say a few words and answer any questions.
Cabernet and Canvas: A blank canvas. A guided painting. A great afternoon out!
www.cabernetandcanvas.com.au

Event also includes raffles and door prizes.
For more information, contact Paula at **services@cfwa.org.au** or **08 6457 7333.**

Superannuation, Insurance and Centrelink

Security of finances provide options to adjusting work-life balance and in some cases, stopping work and focussing on health.

Superannuation

Superannuation, or super, is money paid into a retirement fund by your employer. There are many different types of funds, each a little different, offering slightly different fee structures, plans and options. For most people your employer has to pay an amount equal to 9.5% of your gross salary into your super fund on top of your normal salary. These funds accumulate over your working life.

Keeping Track and Lost Super

If you register with the Australian Taxation Office's myGov you can track all super accounts and consolidate them into the one account. This will save unnecessary fees and make it easier to track. From February 2019, super accounts which have not received a contribution for 16 months and have balances below $6,000 will be transferred to the Australian Taxation Office (ATO) who will attempt to auto-consolidate the amounts into your active account, if you have one. If it cannot be auto-consolidated, the balance will remain with the ATO until it can be validly claimed, through your myGov account.

Self-employed and Super

If you have super from previous employment you need to check to see if you can continue with contributions, otherwise you can choose another super fund. Super gets preferential tax treatment in that you pay less tax on your earnings if they go into your super. It's also a really great way to save! Speaking to a tax consultant or an accountant could also be useful.

Early Access to Super

Most people are unable to access their super until retirement from the workforce, however, if you can demonstrate specific medical conditions or severe financial hardship, it's possible to get early access.

Access Due to Temporary Incapacity

If you are temporarily unable to work or need to drop down your hours due to a physical or mental health condition, super can be released in regular payments over the time you are unable to work.

Super Insurance Policies

Some super has automatic insurance policies in the advent of death or disability. These types of insurance are often cheaper than similar cover outside of super. Basic cover can usually be increased, decreased or cancelled. Your super fund will have more detailed information on their website.

- Income protection policy (IPP) pays you an income stream for a specified period if you are unable to work due to serious illness or disability.
- Total and permanent disability (TPD) a benefit available to you when you can demonstrate serious disability or ill health whereby it's unlikely that you'll be able to work again.
- Total and temporary disability (TTD) a benefit available to you in the advent of temporary disability where you are unable to work and are in financial difficulty.
- Death cover or life insurance is a benefit that your beneficiaries will receive in the advent of your death. Most super funds offer life insurance for their members.

Superannuation Co-Contribution

A scheme where you can add extra money into your super for low income earners. The ATO will co-contribute $0.50 for every dollar you contribute up to a maximum of $500 per year. You must be earning less than $46,920.00. For self-employed, or part-time workers, this could be a good option.

Superannuation Top-Up

You need to arrange with your employer to take an agreed-upon amount – e.g.

$20 -$40 per fortnight – out of your after-tax salary and put this into your super fund, or you can do it as a lump sum. These extra contributions add up over your working career. Total payments into your super fund (both employer contributions and personal top-ups) are capped each year; refer to the ATO for current caps.

Centrelink benefits for people 16 years of age and older:

- Disability Support Pension is available for those unable to work due to illness or disability
- Sickness Allowance is a temporary payment available if you need to take time off work due to hospitalisation or temporary illness. Full-time students are also eligible if they are unable to study due to illness or injury
- Mobility Allowance is when you are unable

to use public transport without assistance or due to illness. This is not means tested so you are able to participate in work or studies

- Pensioner Education Supplement is a supplement is based on your study load and your specific benefit
- Youth Allowance is for 16-24-year old's looking for full-time work, studying full-time, doing an apprenticeship or living independently
- Austudy is for full-time students over 25 in an approved course or Australian apprenticeship program
- Carer's Allowance is a fortnightly payment if you give daily care to someone who has a disability or a serious illness
- Continence Aids Payment Scheme is a yearly payment to cover some of the cost of products that help you to manage incontinence

References and further information

Superannuation

www.ato.gov.au/Individuals

www.industrysuper.com/understand-super/super-changes

http://guides.dss.gov.au/guide-social-security-law/4/13/1/20

www.my.gov.au

www.moneysmart.gov.au/superannuation-and-retirement/how-super-works

www.etax.com.au/superannuation-co-contribution

Centrelink

www.humanservices.gov.au/ individuals/services/centrelink/disability-support-pension

www.humanservices.gov.au/individuals/services/centrelink/sickness-allowance

www.humanservices.gov.au/individuals/services/centrelink/mobility-allowance

www.humanservices.gov.au/individuals/services/centrelink/pensioner-education-supplement

https://www.humanservices.gov.au/individuals/services/centrelink/austudy

www.humanservices.gov.au/individuals/services/centrelink/pensioner-education-supplement

www.humanservices.gov.au/individuals/subjects/caring-someone-illness-or-disability

www.humanservices.gov.au/individuals/

The information provided in this article is general in nature and does not constitute financial advice. Before making any decision, we recommend you consult a financial planner to consider your individual situation.

Volunteer Spotlight

We are excited to introduce our newest volunteer, Rachel Rogers. Rachel is a student in her final year of health promotion studies at Curtin University as well as a mum of a child with CF.

With this degree, Rachel is interested in making meaningful changes and seeing better health outcomes through advocacy and promoting safe and healthy communities.

Rachel has had an interesting working career, starting out with an outdoor adventure company. She spent eight years teaching children at camp how to do exciting activities like canoeing and abseiling, eventually moving into first aid training and managerial roles. Choosing to travel eventually brought Rachel to Perth where she has made her new home.

As part of Rachel's studies, she had to choose an organisation to do practical work experiences outside of the university – she chose Cystic Fibrosis WA (CFWA)! Although her practicum doesn't start until next semester, Rachel wanted to give back to CFWA by volunteering, for the support she feels her family has received over the years. Rachel hopes that during her volunteer work she is able to gain a better understanding of this support at an organisation level, as well as gain another perspective of the research, education, social, and clinical support, and lived experiences within the CF community. CFWA will benefit from a consumer's perspective in the enhancing of current and future resources.

Rachel's passion and interest in the health of communities and populations has already driven her to create change in her own community, we are excited to see what the future will bring. Welcome to the team Rachel!

Insurance Broking Peak Body, NIBA, Continues to Support CFWA

WA Divisional Committee Chair Ross Bethell recently spoke with Nigel Barker about the incredible 17-year NIBA partnership

The annual National Insurance Brokers Association (NIBA) gala luncheon provides an opportunity for members to network and catch up on changes within the industry. For Ross Bethell, Chair of NIBA's WA Divisional Committee, the luncheon also provides a strong connection to the community.

"Since 2002, the National Insurance Brokers Association (NIBA) WA has proudly supported Cystic Fibrosis WA (CFWA) and the comprehensive services they provide to people affected by the disease in Western Australia.

Thanks to our generous members, last year during our annual gala luncheon, we were delighted to raise money for CFWA during a live and silent auction.

We look forward to continuing our partnership with CFWA and thank them for the tremendous work they do to improve the lives of those affected by cystic fibrosis."

For CFWA the incredible generosity of NIBA – representing some 320 firms and more than 3,500 individual intermediaries – has raised more than $50,000 since 2002 through its annual luncheon. In addition, many member organisations extend support throughout the year for CFWA's vital services.

Since 1982, NIBA has been a driving force for change in the Australian insurance broking industry. It has supported financial services reforms, encouraged higher educational standards for insurance brokers, and introduced a strong, independently administered and monitored Insurance Brokers Code of Practice.

NIBA strives to represent the interests of insurance brokers, on behalf of their clients (policyholders), with integrity and in a manner that is respected, credible and relevant.

NIBA sets and promotes quality standards for insurance broking in Australia, enhances and promotes the professionalism and standing of brokers, and community confidence in brokers.

In addition to fundraising, peak bodies like NIBA provide an important role in increasing awareness for CF and CFWA in the community.

If you belong to a peak body or service group that would like to help CFWA drive greater awareness and support vital services, please contact Karen De Lore at **marketing@cfwa.org.au** or **08 6457 7333**.

NIBA
National Insurance Brokers Association

THE 13TH AUSTRALASIAN CF CONFERENCE MEETS PARENTS RETREAT!

With special thanks to Telethon, Lotterywest and CFWA,
we're combining Parents Retreat and the CF Conference to offer a weekend at Crown!
Our conference package for both metro and regional parents and carers provides a hefty financial contribution toward registration, carers dinner and Crown accommodation.
The conference is for you – Parents and Carers. How we can support your weekend.

Single Package	Cost	CFWA Subsidy	You Pay $
Registration*	$270	$150	$120
Registration* & Dinner	$335	$200	$135
Registration* & Dinner & Accommodation*	$535	$385	$150

Couple Package	Cost	CFWA Subsidy	You Pay $
Registration*	$540	$300	$240
Registration* & Dinner	$670	$400	$270
Registration* & Dinner & Accommodation*	$870	$570	$300

* Based on early bird registration fee available until 30 April 2019 **Based on 1-night stay at Crown Promenade

The Conference

- The Conference is packed with interesting and practical sessions that focus on different aspects of living and caring for someone with cystic fibrosis (CF)
- With over 15 different sessions running concurrently, you can choose which sessions you would like to learn more about
- At least 10 international experts, as well as presentations from other parents and people with CF
- The conference presents the latest advances in CF research, care and drug development as well as new ideas and visions for the future!

Parents Retreat

- The weekend provides a wonderful opportunity to meet and connect with other parents and carers from WA and interstate.
- Catch up over dinner to chat about the new things you've learnt during the day
- Enjoy a luxury overnight stay at Crown and some well-deserved rest and relaxation

An opportunity not to be missed as the Conference heads west!
Sat and Sun 3 - 4 August 2019

To register visit **www.cysticfibrosis.org.au/acfc**
Contact Paula for further information about the subsidy on **services@cfwa.org.au** or **08 6457 7333**

lotterywest supported

CELEBRATING 50 YEARS telethon 7

CYSTIC FIBROSIS Western Australia

Alicija, Danielle and Rebecca (left to right)

Danielle and her team of supporters

'Five Feet Apart' Movie

You may have heard about, or seen, the new film 'Five Feet Apart', which follows the story of young adults living with cystic fibrosis (CF).

Discussing the film

While this film acts as a great opportunity for raising awareness of CF and the risks of cross-infection in the broader community, we understand that it may raise concerns for some people.

Our team have put together a list of discussion questions and helpful resources which cover some of the main themes portrayed in the film. They can be found on our website at **www.cfwa.org.au/news/5feetapart.**

If you would like to talk to someone, or would like more information on any of the topics mentioned on the website, please contact our team at **servicesmanager@cfwa.org.au** or **08 6457 7333.**

"Five Feet Apart" Mandurah Movie Night

Danielle, CFWA member and mum to a child with CF, held a Five Feet Apart movie awareness and fundraising event. It was a hit with the local Mandurah community, selling out in 24 hours and attracting 100+ attendees to watch the new film and learn more about CF. Through ticket sales and raffles that Danielle and her team of supportive volunteers sold before the show, over $5,000 was raised for the cystic fibrosis community.

A massive thank you to Danielle and her supporters Alicija Lawler, Jon Randell, Rebecca Wreford, Reading Cinemas Mandurah, Jenny Doy and Mercer Harries First National Real Estate for putting together this fantastic event.

Early lung disease in infants and children with CF: Looking to the future

Sarath Ranganathan, Graham Hall, Peter Sly, Stephen Stick, and Tonia Douglas undertook the Concise Clinical Review on behalf of Australian Respiratory Early Surveillance Team for Cystic Fibrosis (AREST-CF).

Very early treatment of children with cystic fibrosis is key if we are to help them live longer and have a better quality of life, a clinical review has found.

The Review by senior Australian paediatric CF clinicians has established lung damage occurs very early in life, often without symptoms or signs. Because of this, the specialists now believe early childhood is a critical period for intervention to delay or stop the start of irreversible damage.

As well as future research strategies, doctors are also considering immediate changes to clinical practice based on the Review. They believe it's not feasible to await the results of future studies before intervening. Logic and experience, they believe, mean certain initiatives can be started to improve care of infants and children with CF, even if direct evidence is not yet available.

As a result, parents possibly face potentially increased surveillance and treatment of their healthy-looking children. Parents in this situation may benefit from psychosocial support, the Review says.

Because doctors now know that lung disease develops very early in life, the Review makes the case that they are therefore obliged to provide better CF education and support for parents and families after diagnosis.

Doctors examined a decade of research findings about early lung disease in children with CF to set up research and

therapy directions for the future. They found irreversible, progressive lung disease develops early in life in CF because of a genetic defect that leads to dysfunction in the CF transmembrane regulator (CFTR), causing mucus in various organs to become thick and sticky.

Lung inflammation, infection and structural lung damage are common in infants and preschool children, and the Review found clinical approaches so far had failed to stop most children with CF having established bronchiectasis (permanent lung scarring) by the age of five.

Future studies will be designed to alter early disease progression to stop lung damage before it becomes irreversible. Areas of focus include treating lung inflammation, minimising lung function decline and preventing infection using environmental strategies.

The exact mechanisms linking the basic CF defect to organ damage (including irreversible damage to the lungs) are unclear, but data from intensive CF early surveillance programs, such as those developed by the Australian Respiratory Early Surveillance Program for Cystic Fibrosis, have provided important insights into the biological mechanisms and natural history of lung disease in early life.

Review findings at a glance

Pulmonary Inflammation

- Lung inflammation in infancy and the preschool years is associated with worse nutritional status, undesirable organisms in the lower respiratory airways, bronchiectasis, and lung function abnormalities.
- Infection is a key contributor in inflammation. Its increase with age during the preschool years appears to be relatively independent of current or previous infection.
- Normal lung defence mechanisms are overwhelmed by inflammation and infection, creating the potential for lung structure weakening.
- Research is examining the role of thick, sticky mucus in the cause of inflammation. The challenge is to identify and trial potential therapies for reducing inflammation and infection as a way to prevent future lung disease.

Infection

- Bacteria such as Staphylococcus aureus, Haemophilus influenzae, and Pseudomonas aeruginosa are thought to cause respiratory infection in CF, but the role of these organisms in the development and progression of structural lung disease and lung function decline remains unknown.
- The eradication of chronic P. aeruginosa (a significant pathogen in declining health and increased death) during the preschool years does not appear sufficient to prevent the development of structural lung disease or lower lung function.
- Saureus is commonly cultured shortly after diagnosis and in up to 30% of infants during the first six months of life.
- The environment may play a part in acquiring infections. Living in a regional versus a metropolitan area is a significant risk factor for the first acquisition of P. aeruginosa. It's possible P. aeruginosa infection may occur in those who already have worse lung disease or disease susceptibility. It remains unknown whether the number of a particular organism as opposed to the presence of the organism present in the airways, is important in CF lung disease.

Structural Lung Disease

- Structural lung damage caused by infection and inflammation is difficult to assess in its early stages, and until now has not been the main focus for doctors. Advances in technology have markedly improved the sensitivity with which structural lung disease can be detected.

Lung Function

- Infants who avoid lung infections during infancy experience lung growth similar to that of healthy children. It's likely that lung clearance index (LCI) may be useful to monitor structural lung disease in preschool and school-age children.

Implications for Clinical Practice

- Despite normal FEV1, almost 80 per cent of children with CF show evidence of bronchiectasis by the time they reach school age.
- New treatments will be aimed at preventing the onset and delaying the progression of bronchiectasis, inflammation and lung function decline during the preschool years. The next target for treatment arising from newborn screening for CF is to develop genuine primary prevention strategies intended to avoid the development of the disease.

Clinical Care and Future Strategies

- Change in attitude in CF clinics is needed, with attention being given to the critical role of intervention in the early years.
- The Review advocates the development of policies and guidelines for a unified proactive approach to care in apparently 'healthy' CF children without symptoms. It says the detection of lung abnormalities in early life makes a persuasive argument for intervention, even if evidence for specific treatments is scarce.
- Therapeutic options may include antibiotics for CF pathogens detected in both symptomatic and asymptomatic individuals.

While using therapies earlier and more intensively seems an appropriate response to current scientific evidence, there is the potential risk of treatment-related side effects and increasing health costs.

Currently, treatments are recommended for the majority of patients because of the inability to predict a child's risk of disease progression. Being able to target children with the greatest need for such treatments would not only save money, but would reduce the potential risk of side effects from unnecessary treatments. The Review argues identifying better predictors of severity of disease from time of diagnosis should remain a crucial aspect of further research. New, potentially disease-modifying treatments are likely to be extremely expensive and may have as-yet-unknown side effects.

If early intervention is to succeed, appropriate tools are also required to safely and effectively monitor disease progression and the effect of treatments.

The main takeaway from the Review is that maximising the benefits provided by newborn screening programs through the early introduction and implementation of new CF treatments is important if we are to improve the longevity and quality of life for the next generation of people with CF.

Flight Centre Perth Supporting Our Vision

Staff at Flight Centre Perth step on board to help.

For Grant Parsons at Flight Centre Hay Street, a conversation with Cystic Fibrosis WA's (CFWA) Karen De Lore about workplace giving became the opportunity to get his team on board with workplace giving through the Flight Centre Foundation (FCF).

Grant's wife, Sandi, lives with cystic fibrosis (CF) and Grant understands that the work carried out by CFWA makes a huge difference for individuals living with CF, and their families. With the FCF's vision of 'Building Brighter Futures', Grant could see the synergy between our organisations and put the process in motion to have CFWA nominated as a workplace giving recipient organisation.

Workplace giving provides a great opportunity to support CFWA. Many organisations incorporate volunteering and workplace giving within their community engagement program. At CFWA we welcome the opportunity to work with organisations to spread awareness and support events like 65 Roses Day through volunteering and raise vital funds for ongoing services and research.

If an organisation you, or a family member, work for has a program in place, we would love the opportunity to talk to them about how their program can help us achieve our vision. Alternatively, if you would like to know how to set up a workplace giving program in your organisation we can help.

For more information please contact Karen De Lore at **marketing@cfwa.org.au** or **08 6457 7333.**

Remember also that that the team at Flight Centre Hay Street (729 Hay St Mall) on **1300 509 397** are there to help with your travel needs. Make sure you mention that you are aware of their support for CFWA!

FLIGHT CENTRE®

Fundraising News

It's been a busy couple of months with birthday celebrations, live performances and 'Five Feet Apart' fundraisers!

Gloria and her magnificent cake

Gloria celebrating her 75th birthday in style!

Sixty+ Duo

Gloria's Celebration for CF

Cystic fibrosis (CF) grandmother Gloria celebrated her 75th birthday in style recently by bursting out of a cake at her party held with family and friends. However, Gloria's birthday celebration wasn't all about her living out a lifetime dream; she also used the opportunity to raise awareness and funds for her grandson who lives with cystic fibrosis. Gloria's very generous guests raised over $1,000 on the night which was donated to Cystic Fibrosis WA (CFWA).

Sixty+ Music Gig

Community member Jennifer Davies and her friends, Terry and Edward from the duo Sixty+, put on a music gig to raise awareness and funds for the CF community. They held their performance at The Vic in late February and had 168 guests attend to dance along to the music in support of CFWA, raising over $3,000!

Rottnest Channel Swim

The 2019 Rottnest Channel Swim landed on a beautiful sunny day in late February. This year, it included a very important team who were swimming to raise funds and awareness for CF. They swam the 19.7km stretch from Cottesloe to Rottenest and raised over $480! Thank you to Natasha O'Sullivan and her team for supporting CFWA and creating awareness in the community.

For details about upcoming events, or information about how to organise your own awareness and fundraising activity, visit our website **www.cfwa.org.au** or contact Marnie on **events@cfwa.org.au** or **08 6457 7333.**

You Can't Ignore It...

For sisters Isobel and Ruby, sticking to their daily cystic fibrosis (CF) treatment is a routine they help each other get through.

When Ruby was born and diagnosed with CF, her parents, Christine and Gerard, knew what they were dealing with. Two years earlier, Ruby's sister, Isobel, was also born with CF. With two older brothers, the routine required to manage CF in their household was already understood.

Everyone has a role, with Isobel and Ruby working together every day, helping each other through their treatment, and older brothers acting as great hospital visitors and carers when the need arises.

Isobel and Ruby, with their family, are passionate about raising awareness for CF, initiating the 2016 'Crack-a-Cure' campaign, participating in research trials, and more recently, the 2018 Orkambi® advocacy campaign. At Cystic Fibrosis WA (CFWA), we are very grateful that they are the face of the ***'You can't ignore it.... CF is for life'*** tax appeal.

In collaboration with Cystic Fibrosis Community Care and Cystic Fibrosis Queensland, the appeal supports the many services that CFWA provides to our CF community in WA and aims to educate the broader community on the relentless need for treatment and vigilance in daily CF life.

Christine reflects that she and Gerard want the same thing for all their children; 'to live a happy, healthy full life and to grow old surrounded by loved ones'. The opportunity to be part of the CFWA-funded AREST CF program, and the support received through home care, hospital and education programs, 'assist in the daily battle and help put the odds in our favour'.

Please help us to spread our 'You can't ignore it... CF is for life' message. Together we can change the future for CF families. If your family would like to participate in future awareness and fundraising campaigns, please contact Karen at **marketing@cfwa.org.au** or **08 6457 7333.**

May

1 – Evening with the CF Scientists

1 - Barbagallo Men's Health Session

4 – Ladies' High Tea

5-10 – Regional Respiratory Training Program

19 – HBF Run for a Reason

23 – 65 Roses Rose Wrapping Day

24 – 65 Roses Day

August

3-4 –Australasian CF Lay Conference

4-6 –Australasian CF Medical Conference

TBC - Perth City to Surf

September

1-30 - Serve It Up for CF in September

October

2-4 – Sibling and Offspring Camp

19 – Parents' Dinner

TBC – Telethon Weekend

November

1 – Cystic Fibrosis Golf Classic

10 – Men's Support Event

22 – Sponsors and Volunteers Evening

*Please note, dates may change if conflicts arise.

For more information and to register for these events visit **www.cfwa.org.au/get-involved/upcoming-events/**

THE CAPEL VALE

CONQUER CYSTIC FIBROSIS

GRAND BALL 2019

SATURDAY 18 MAY, 7PM – 1AM
GRAND BALLROOM, CROWN PERTH.

A FUN FILLED EVENT WITH LIVE MUSIC, SILENT AND LIVE AUCTIONS ALL RAISING FUNDS TO CONQUER CYSTIC FIBROSIS.

FOR FURTHER DETAILS: WWW.CONQUERCYSTICFIBROSIS.COM OR
EMAIL: INFO@CONQUERCYSTICFIBROSIS.COM

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Let's Paint WA With Roses

65 Roses Day Friday 24 May

www.cfwa.org.au/65roses