EDITION 3 2016

RED

Cystic Fibrosis WA
40th Anniversary

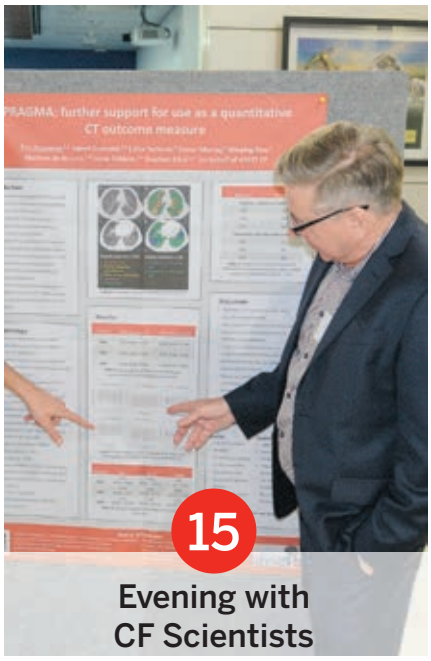
In This Issue

Nutrition

Contents

5 Roses Blossom for CF

8 Nutrition in CF



15 Evening with CF Scientists

22 CF Related Diabetes

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Features

RED

CEO Message

THE THEME OF THIS EDITION OF RED MAGAZINE IS NUTRITION.

NIGEL BARKER, CEO

Many parents and people living with cystic fibrosis (CF) face a constant challenge of creating attractive, high calorie, CF-friendly meals. It's a complex area which looks at the very different nutritional needs of people living with CF, helping teachers and peers understand why they have a different diet and the need for enzymes which can cause some anxiety.

In addition, as the CF population ages, we are seeing an increase in CF-related diabetes and this too has an impact on diet.

Help is at hand however, with the release of the latest CF Food resources sponsored by Newman's Own Foundation and the soon-to-be released Vertex Circle of Care sponsored CF Cooking videos and online resources.

The CF Cooking resources will be an international resource which is the product of a close collaboration between CFWA and Sir Charles Gairdner Hospital's Dietitian Paul O'Neill, due for release in October 2016. It will include six short videos (filming is underway) and supporting online recipes and will be launched at the Vertex Circle of Care Colloquium in the USA at the end of October.

In this edition we also look back on the hugely successful 2016 65 Roses month. This featured the Golf Day at Lake Karrinyup and a very special evening with CF Scientists at the Telethon Kids Institute where CF Scientists briefed 100 parents and members on the very latest, and in some cases yet to be published, research on CF.

There were also numerous community fundraising events and the sale of almost 10,000 roses from WAFEX on May 27th from Western Australia. This year we were also excited to see a growth in the number of regional sales from Esperance to Broome and all points in between. Thank you to Virgin Airways, Tail Lift Transport, Southern Regional Transport, Freightlines, Kalbarri Carriers, and Western Independent Foods for making this possible.

We also congratulate and introduce Clara Mok, the latest recipient of one of CFWA's PhD Top Up Scholarships. This scholarship known as the CFWA Golf Classic PhD Top Up Scholarship has been funded from the proceeds of the George Jones Family Foundation CF Golf Classic.

Finally, we are looking forward to the up-coming Capel Vale Red Tie Dinner Dance hosted by Fremantle Sailing Club Power Section in August and the Community Newspapers Halloween Fun Run in October.

Thank you for all your support.

DEADLINE FOR THE NEXT ISSUE
If you would like to contribute to our winter issue, please contact us before 19 September 2016

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
Jordyn Drayton. Read Jordyn's story on page 07.

CFA Update

I was recently in Edinburgh, Scotland for the *European Conference on Rare Diseases & Orphan Products* and I was yet again inspired by the gritty resolve and determination of people living with such a wide range of afflictions. The conference organisers engineered a very interesting mixture of clinical sessions and patient advisory and advocacy groups with a focus on a Better Future for Patients.

The Scottish Minister for Mental Health opened the Conference stating, 'it's not rare to have a rare disease as there are over 7,000 rare diseases identified.' She continued her advocacy for patients by stating that to improve patient outcomes and experience we should be asking not what is the matter **with** you, but rather what matters to you.

Alastair Kent, Chairman of Rare Diseases UK, was the standout speaker stating emphatically that rare diseases are a significant cause of human suffering and we should not have to justify the need for care or resources. Alastair believes that the advocates' job is to hold the health systems accountable ...hold their feet to the fire so they don't shy away from the big decisions.

Alastair said that the message was clear that things have changed. 2,500 years ago diagnosis was done by looking at the stars and today we look at the movement of molecules. In the past patients were the subjects. Patients are now the partners and soon they will be the agenda setters and the drivers of care.

An overarching theme of the conference was the power of the people and the communication between patients. Social media has enhanced this by giving patients a voice, a connection, a mutual support network and awareness of global breakthroughs.

In Australia, we are using social media and mutual support groups very effectively so let's maintain our own levels of gritty resolve and determination to continue our advocacy for access to the best treatments for all people with cystic fibrosis.

Nettie Burke,
CEO CFA

Cystic Fibrosis & Reproductive Choices Seminar

An information day for people with CF, their partners and parents of children with CF

Date: Friday 21 October
Time: 9am to 2.30pm
Venue: The Institute for Respiratory Health, Harry Perkins Research Institute, QEII campus

We are hoping the sessions will be streamed live for those who are interested but unable to attend.

For queries please contact
Jan Howie: nurseeducator@cysticfibrosiswa.org
or Natalie Amos: education@cysticfibrosiswa.org
or phone 08 9346 7333
More details about the agenda coming soon.

Roses Blossom for CF Around WA

THANKS TO WAFEX AND THE HUNDREDS OF SUPPORTERS WHO HELPED US SELL 9,700 ROSES ALL OVER THE STATE ON 65 ROSES DAY, OUR BIGGEST AND BEST YET!

It's hard to believe that in just a few short years, 65 Roses Day has grown so much, from 2,000 roses in 2012 to an incredible 9,700 in 2016. The sale of these roses combined with other fundraisers and donations in May raised an incredible $50,000!

This phenomenal result wouldn't have been possible without the support from WAFEX, who supplied us with all of the beautiful roses, the freight companies who transported them to regional locations, South Metropolitan TAFE who organised a small army of volunteers, and of course the organisations and individuals who lent a hand wrapping and selling all 9,700 roses.

It was particularly exciting to see such strong regional support this year with champions in Broome, Shark Bay, Geraldton, Northam, Margaret River,

THE BEAUTIFUL FRESH ROSES WERE SUPPLIED BY OUR GENEROUS SPONSOR WAFEX.

OUR JUNIOR AMBASSADOR, WILLOW, JOINED US IN PERTH CBD ON 65 ROSES DAY TO HELP US SELL ALMOST 10,000 ROSES.

OVER 100 VOLUNTEERS LENT A HAND TO PREPARE ALL 9,700 ROSES FOR SALE.

STEVE MILLS AND BASIL ZEMPILAS SPREADING THE WORD AT 6PR.

Albany and Esperance all selling roses and hosting fundraising activities. We look forward to expanding this further in the future with the help of our regional members.

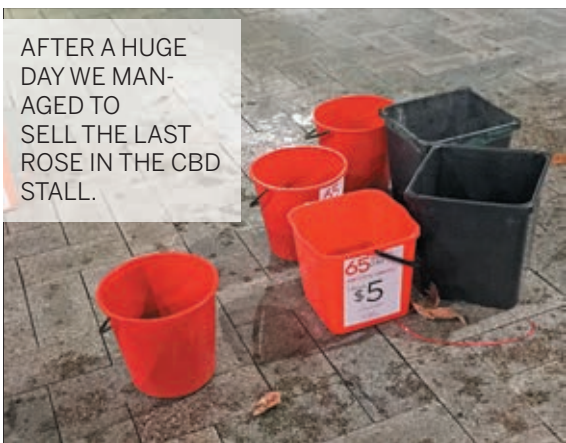
South Metropolitan TAFE events students also played a huge role by managing the 12 shopping centre stalls around Perth, allowing us to focus on the big CBD stall. It was a really big help for us and we hope the students got a lot out of it too.

As always, we are looking forward to even bigger and better things for 65 Roses Day next year. If you have any ideas for ways to improve 65 Roses Day, either in the metro or a regional area, please get in touch on 08 9346 7333 or at events@cysticfibrosiswa.org.

WAFEX GAVE US SOME "LIMITED EDITION" RAINBOW ROSES THAT WERE A BIG HIT IN OUR CBD STALL.

THANKS TO THE SUPPORT OF LOCAL FREIGHT COMPANIES, WE WERE ABLE TO DELIVER ROSES ALL OVER THE STATE, FROM BROOME TO ESPERANCE.

THE RAINBOW ROSES SUPPLIED BY WAFEX CREATED A LOT OF BUZZ AROUND THE CBD STALL



AFTER A HUGE DAY WE MANAGED TO SELL THE LAST ROSE IN THE CBD STALL.

Cooking with Jordyn

JORDYN AND HER FAMILY DESCRIBE THEMSELVES AS "FOODIES", SO WE WANTED TO HEAR MORE ABOUT HOW SHE'S DEVELOPED SUCH A GOOD RELATIONSHIP WITH FOOD. JORDYN WILL ALSO BE FEATURING IN OUR UPCOMING "CF COOKING" SHORT FILMS.

Tell us about where you grew up?

I was born in Narrogin and grew up on a hobby farm 5km out of town. The space was good; bikes, buggies and learning to drive when I was fairly young. There were also chooks, sheep, two dogs and two cats. We moved into town when I got older so that we could be closer to friends, school and all our various sporting activities. I play netball in the same team as my mum and my sister so have always kept really active.

Have you always had a good appetite and weight?

I have always struggled to put on weight, however, have always really enjoyed food. Mum and I do all the cooking, we often cook together. We hold special events for the extended family, making a nice menu, doing up the living room, it's really fun and brings everyone together. I have the same food as the rest of the family, just add a few extras, e.g. cream, butter, oils, cheese and extra ice-cream.

Do you have any strategies that help when you don't feel like eating, or if you feel you need to put on more weight?

Sometimes I don't feel like eating, I just feel full of sputum. That's when my Mum makes me eat. It just takes longer and I break it up over a longer period. I also cook things in big batches and freeze some stuff for those times that I don't feel like cooking.

When did you start becoming seriously interested in preparing food?

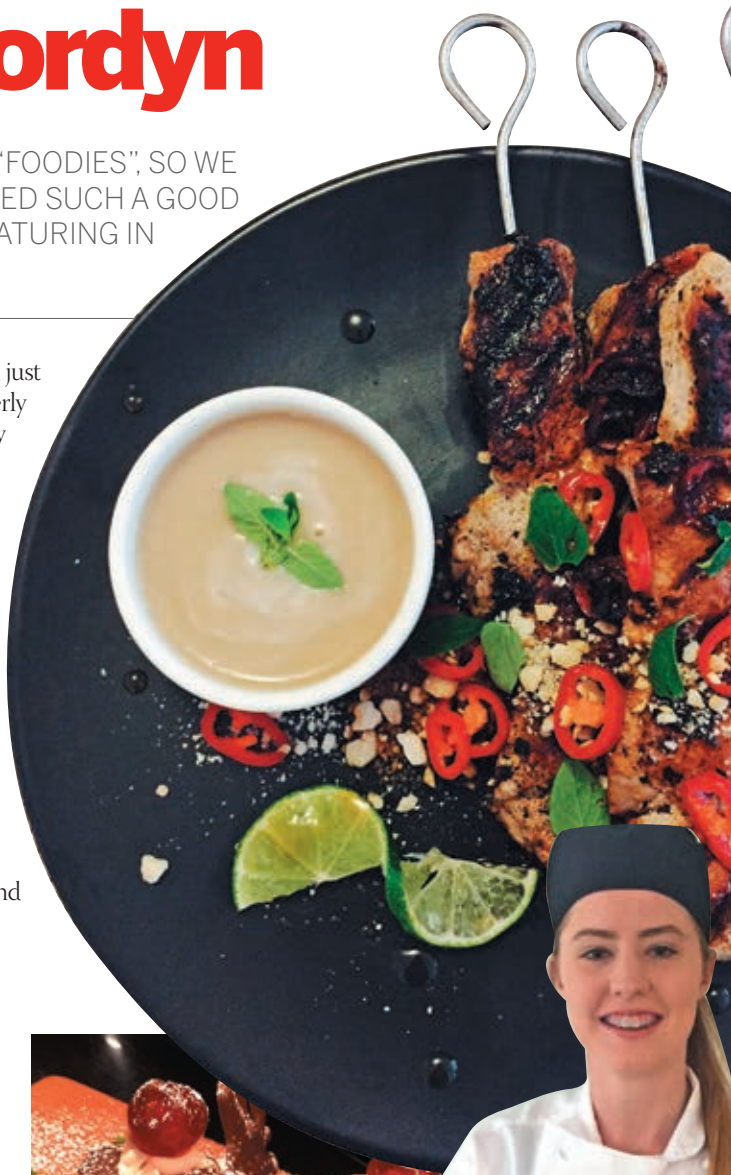
Mum always got me to cook since I was really young and when I was in high school I started cooking for the family twice a week. I started a chef apprenticeship earlier this year which was really great, but the hours were really crazy, sometimes 12 hour days, finishing at midnight. I started to lose a lot of weight because I just didn't have time to eat properly and look after myself. I really enjoyed it and it didn't really feel like a job because I loved the cooking, but it's currently on hold and I will look at something a little less strenuous.

What would you like to do in the future?

Definitely something to do with cooking, I will be looking back in Narrogin to see if I can find something and perhaps move back to Perth later in the year.

You are also featuring in one of the "CF Cooking" shows how do you think this will benefit people?

I think it will be a great resource for people to get ideas that are easy and tasty and to gain a little more knowledge about healthy CF food. Hopefully people will enjoy food as much as me, it brings people together.



Nutrition in Cystic Fibrosis

THERE IS A LOT OF NUTRITION INFORMATION IN THE COMMUNITY, BUT UNFORTUNATELY THERE IS ALSO A LOT OF MISINFORMATION. SO WHAT IS GOOD NUTRITION IN CF?

We know that nutrition is important for everybody's health, but nutrition is paramount in CF. However, the dietary recommendations between a regular and a CF diet seem opposite. Why is this so, and how did we get to a "high fat and high salt diet"?

CF diet history

In 1988 a group in Canada showed that children with CF on a high fat diet lived an average of 9 years longer compared to those in Boston who were restricting fat in their diet. Those having a high fat diet grew taller, were heavier and lived longer. If you think that at that time some people only lived into their teens or twenties, nine years was a huge difference. Especially when the only difference between the groups was a high fat diet! The results revolutionised CF dietary guidelines internationally toward a recommendation for a diet unrestricted in fat.

The importance of achieving a good body weight and growth in CF is now well known, because it is so closely related to health and lung function. Unfortunately, underweight models may make a magazine cover, or get to strut the catwalk, but in cystic fibrosis such a low body weight is simply malnutrition and is associated with a lower lung function, (by the way, the girl on the catwalk is malnourished also).

People with CF also have increased salt needs from increased salt losses. If we combine these two we have a need for a high fat and high salt diet. But who said that a 'low fat diet' was healthy?

More diet history

Another famous study from the 1940s was called the 'Seven Countries Study.' Over 15 years they recorded the cause of death and lifestyle factors in healthy men from seven different countries. The leading causes of death were heart disease, cancer and stroke, which is similar to today. Those with heart disease also had high cholesterol. The researchers observed that the death rates were related positively to how much dietary energy came from saturated fat, but not from monounsaturated fat (olive oil), or polyunsaturated fatty acids (fish), or from proteins, carbohydrates, and alcohol. They noted that the populations with a diet high in olive oil had very low death rates from heart disease.

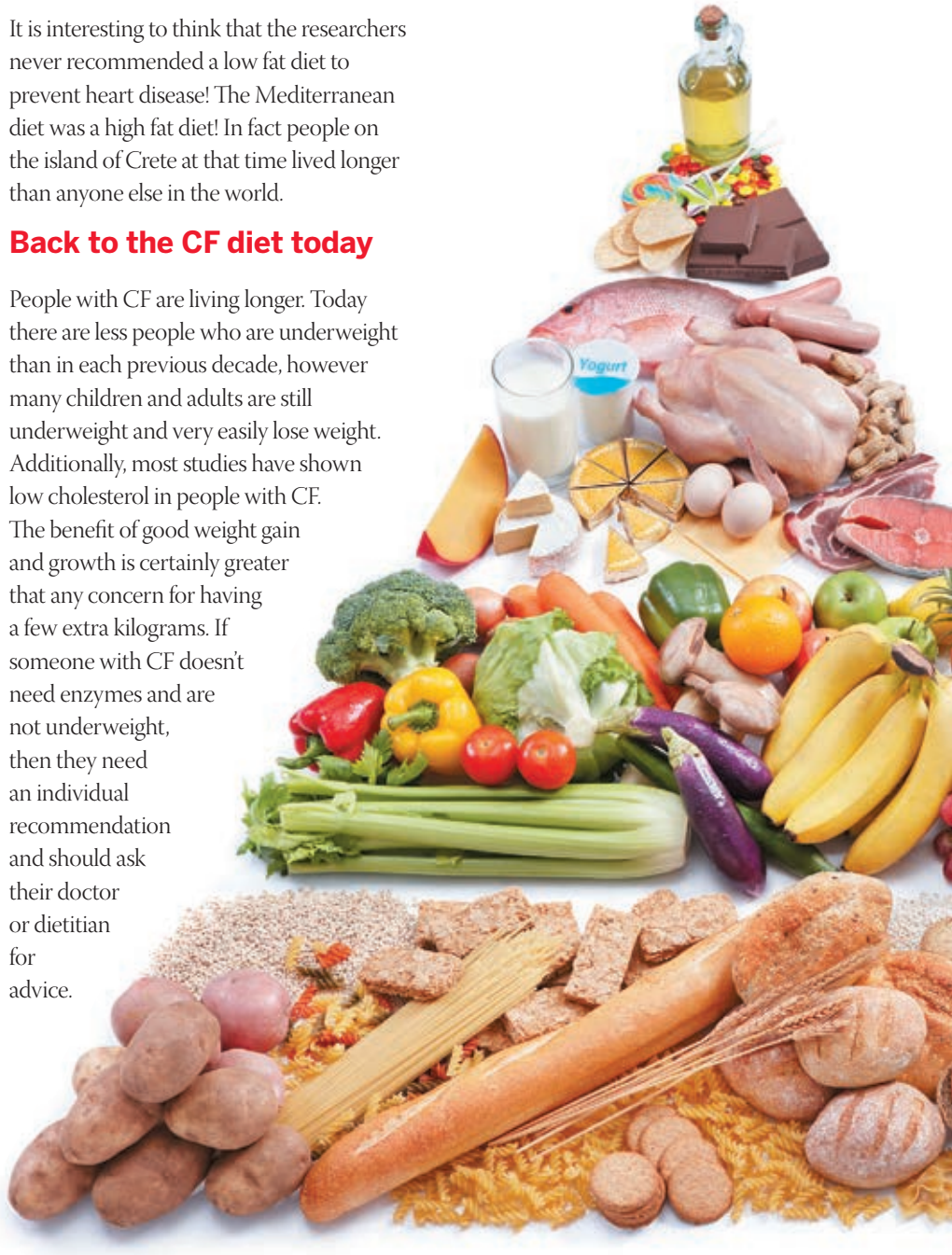
It is interesting to think that the researchers never recommended a low fat diet to prevent heart disease! The Mediterranean diet was a high fat diet! In fact people on the island of Crete at that time lived longer than anyone else in the world.

Back to the CF diet today

People with CF are living longer. Today there are less people who are underweight than in each previous decade, however many children and adults are still underweight and very easily lose weight. Additionally, most studies have shown low cholesterol in people with CF. The benefit of good weight gain and growth is certainly greater that any concern for having a few extra kilograms. If someone with CF doesn't need enzymes and are not underweight, then they need an individual recommendation and should ask their doctor or dietitian for advice.

It doesn't matter what you do, or what you dream to do one day, good nutrition can help you do it better! If you are into sport, or music, or want to have a family, or study or just need more energy, optimal nutrition is going to help you do what you want to do better.

The CF diet is healthy for people with CF. Enjoy a high fat high salt diet.



Enzymes and Cystic Fibrosis

NUTRITIONAL MANAGEMENT IS AN IMPORTANT PART OF CF CARE. GOOD NUTRITION IN CF IS ASSOCIATED WITH BETTER LUNG FUNCTION AND SURVIVAL. PANCREATIC ENZYME REPLACEMENT THERAPY IS DIFFERENT FOR EVERYONE AND SOMETIMES IT'S TRICKY TO GET THE RIGHT BALANCE.

The increased energy requirements for people with CF are a result of increased work of breathing, repeated bouts of infection and inflammation, as well as malabsorption. Malabsorption results in the loss of fat and fat-soluble vitamins in bowel motions which can lead to malnutrition and poor growth. This makes good nutrition a challenge for people with CF.

Digestion and the pancreas

In a normal situation the pancreas produces digestive juices (enzymes) that reach the intestine through small tubes called ducts. Enzymes break down the food so that it can be absorbed into our blood and transported to our organs, like muscle. Our body has different enzymes to break down different parts of the food we eat, including fat, protein and carbohydrate.

In summary the functions of the pancreas is are:

- Produce enzymes that help break down the food we eat
- Control the amount of sugar in our blood by producing insulin

How does CF affect my pancreas?

In approximately 85% of people with CF, the pancreas and/or the duct is blocked with mucus, preventing normal food digestion.

This is referred to as pancreatic insufficiency. This situation prevents the enzymes from working on the food in the intestine, and means those who are pancreatic insufficient will require enzyme supplements to assist with the digestion of food. Without enzyme supplements, poor nutrient absorption leads to poor growth, vitamin deficiency and weight loss.

Enzymes

People with CF who are pancreatic insufficient need to take enzymes (granules or capsules) to absorb important nutrients, mainly fat and fat-soluble vitamins, but also protein and carbohydrates.

In children the dose of enzymes will change as they grow and as their diet changes. Check regularly with the dietitian to review enzyme doses, including babies who are breast fed. It is important to manage enzyme replacement well so your child grows normally.

The most commonly used enzyme supplement in Australia for babies, children and adults with CF is Creon® which comes in microsphere form for babies or as capsules. Panzytrat® is also available and works well for some individuals.

Enzyme dose

The dose required will vary from person to person, as people with CF vary in their degree of pancreatic insufficiency. The Australian Guidelines recommend a dose for enzymes as a range between 4,000 - 5,000 units of lipase per gram of fat. This range is between a very low dose and a very high dose, as can be seen on the right of the table below:

Enzyme Strength (units)	Mid range dose (fat gram)	Range (fat grams)
10,000	8	From 2 - 20
25,000	20	From 6 - 50

Although you may need to adjust your dose, it is recommended not to increase the dose without first discussing this with your doctor or dietitian for guidance. This is partly because of the need to avoid exceeding the 'safe upper limit' This limit was introduced after very high doses of enzymes used in children in the 1980s was suspected to be involved with damage to the bowel called fibrosing colonopathy. As a result, the 'safe upper limit' was introduced and since that time the problem has been avoided. The safe upper limit is 10,000 units per kilogram of body weight per day. Ask your dietitian if you are exceeding this amount.

Enzyme doses need to be monitored regularly and adjusted based on the different amounts of food and fluid consumed. Capsules and beads should not be crushed or chewed. Beads should be mixed with an acidic food (e.g. fruit puree, fruit gel, jam or tomato sauce) until capsules can be swallowed.

How do you know if there are digestion/ malabsorption problems?

It is important to learn the warning signs that may indicate that you or your child has problems digesting food.

Changes that you may notice or should monitor:

- Tummy pain
- Discomfort
- Excessive smelly wind
- Greasy, fatty poos that are sometimes difficult to flush
- Diarrhoea and/or constipation
- Hunger despite eating lots of food
- Poor weight gain, poor growth

Your dietitian and doctor will work together with you to work out the best plan to improve you or your child's absorption. This will help ensure optimal nutrition and weight gain.

Your dietitian will also be able to provide advice on the type of food to eat for a high energy diet that will promote normal nutrition, growth and weight gain.

Getting the balance right

Enzymes need to be taken before, or during eating and with nearly all food and fluids. They are not necessary for foods that contain mainly simple sugars. Some examples include:

- All fruit: fresh, dried or canned
- Non-starchy salad vegetables
- Lollies, jellies, sorbet, Roll-ups
- Juice, cordial, soft drinks, electrolyte replacement drinks, water

Remember that while enzymes are dosed according to how much fat is in food, enzymes work on fat, carbohydrates and protein. Therefore, they are still needed with foods that are lower in fat such as rice cakes, lean meat, baked beans and pasta. Sometimes enzymes may need to be spread out if it takes longer than half an hour to eat your meal e.g. when out to dinner or if your child regularly takes longer to finish their meal.

Talk to your CF dietitian if you have any digestion problems.

Reference

Pancreatic enzyme replacement therapy and Cystic Fibrosis

https://www.health.qld.gov.au/trition/resources/paeds_pert.pdf

Member Profile: Mitch Messer

MITCH HAS BEEN AN ICONIC AND VERY PUBLIC FACE OF CYSTIC FIBROSIS FOR MANY YEARS.

If you don't know him, Mitch worked at Cystic Fibrosis WA (CFWA) for approximately 18 years, commencing as an Education Officer, then Fundraiser, and becoming the Executive Director of CFWA. Following this he became the President of Cystic Fibrosis Australia (CFA) for about 6 years and then President of Cystic Fibrosis Worldwide (CFWW) for another 6 years. He is now Vice President of CFA, taking a slight back step concentrating his activities on his position as CEO of CLAN WA, a fantastic community-based organisation that offers FREE training workshops and in home support for carers.

The focus of this interview is not so much about Mitch's achievements, however, on a more personal level about taking enzymes and doing Intravenous Therapy IVs in a public space such as a restaurant.

Were you always open about telling people that you had CF?

Yes, always, it never occurred to me not to tell somebody. I grew up in a country town and kind of assumed everybody knew. CF was unknown then and I was never told that it was a bad thing.

Was there ever a time in your life that you were more self-conscious about having CF, such as through the teen years?

Yes. when I was a teen I was short and scrawny, my growth spurt didn't happen until after high school. My teeth were also stained black through tetracycline antibiotics as a young child. Princess Margaret Hospital (PMH) used to take photos for lectures. The teeth were the hardest thing; I used to get picked on and teased and stopped smiling so people wouldn't see my teeth. As an adult I had all my teeth crowned by the dental clinic so that they looked normal.

Has there been a time in your life that you felt discrimination about having CF?

Not because of CF. People pick on you for normal things. I have never not done anything because of CF.

CF doesn't seem to get in the way of you leading a full and interesting life. You seem comfortable with doing IVs in public such as restaurants etc, were you always comfortable to do this?

CF doesn't get in my way. I do things I can do, not things I can't. I push the limits and pull back when needed. Consultancy was good, the hours worked to suit. Now I have a constant workload which I'm learning to manage. I'm also lucky as I've always had supportive employers, it's a lot of give and take and I've had flexible workplaces. I think if you try to hide things it's more noticeable. I just go and do things like take insulin, Creon etc. as if it's just normal. I do things in plain sight and just treat it as it's supposed to be. I do IVs in public sometimes just because I don't want to miss out on anything, particularly long meetings, I just have to. If you make it part of your life it therefore seems normal.

Have you ever had any issues from other people about doing treatments such as IVs or taking enzymes in public?

Never had issues from people. Some people ask "what's that"? They're generally very supportive.

Not all people are comfortable about taking enzymes in public or doing IVs in public. Would you be able to offer some advice on how this could become easier? Or do you think this is simply an individual choice?

People make choices, IVs are a hard choice but thankfully I'm happy to be the centre of attention. With enzymes, I don't try to hide them, people don't really care and they stop noticing, like it's just normal.

Maintaining a good healthy weight is integral for good lung health. Do you have any tips on eating well when not feeling so good?

I like food that is easy and when unwell I like comfort food, e.g. stews, mashed potato. I eat lots of different things when I go out such as Thai, Japanese and then top up with extra CF food.

My parents were always very open, CF is not something you can control. I think it's important to make the best of whatever you have. If you hide something then it's as though you're ashamed of it. I have always disclosed up front with employment. There were always "age milestones"; I kind of ignored these things, and I never expected not to do the next thing. I never took on the statistics they aren't people, just numbers, I always assumed that I would be a survivor and perhaps had some denial about the implications of CF.

Managing Medical Milestones

RESEARCH INDICATES THAT ALTHOUGH CYSTIC FIBROSIS HAS A HIGH TREATMENT AND PSYCHOLOGICAL BURDEN, MOST PEOPLE ARE VERY RESILIENT, HAVE A GOOD QUALITY OF LIFE AND ARE ABLE TO DEVELOP BENEFITS FROM THEIR CF-RELATED EXPERIENCES. THIS ARTICLE DISCUSSES SOME OF THE DEVELOPMENTAL STRUGGLES AND SOME USEFUL STRATEGIES TO NAVIGATE THROUGH THEM.

Newborn

The complexity of CF and its treatment often means that it becomes a family diagnosis; it affects the whole family and how the family manage this diagnosis will impact on treatment and health outcomes.

Many parents often experience sadness and guilt due to the hereditary component of CF. If unresolved, parental distress can have negative impacts on attachment. Elevated levels of anxiety are frequently accompanied by hypervigilance and parents may misinterpret normal newborn behaviours as CF symptoms.

Confirmation of diagnosis can be a relief as CF is then a "manageable disease", however, many parents have fluctuating moments of psychological distress often termed "chronic sorrow or chronic grief." This psychological distress can be ongoing and unresolved, easily triggered by medical interventions, hospitalisation or even the expectations of treatment. Feelings of anxiety at challenging moments are normal, and most of the time, manageable, however when they persist, can have significant impacts on both the child and parent's mental health.

Strategies

Developmentally appropriate routines are essential around general daily activities that incorporate CF-specific treatment tasks. This will promote a positive relationship between the parent and child and increase the likelihood of better adherence later on.

CFWA can provide home care worker support to assist with good routines. Other useful links:

- **http://raisingchildren.net.au/behaviour/newborns_behaviour.html**
- **http://www.ngala.com.au/**
- **https://www.carerswa.asn.au/**
- RED magazine, Ed 2/2016, The Challenge of Setting up a Daily Routine **www.cysticfibrosis.org.au/wa/redmag/**
- **http://www.blackswanhealth.com.au/services/ataps/**

Early childhood

Language and cognitive development help the child to process and communicate their personal experience of CF. Children start to make assessments about what they can control in their environment, however for the child with CF, there can be a number of stressful situations that they can't control e.g. medical treatments and interventions. Extreme behavioural reactions could occur at this age in an attempt to avoid or escape particular procedures. Such situations could result in trauma or aversions for future medical procedures if not handled well.

These children are learning about behavioural/emotional self-regulation whilst taking on CF-related tasks. Parents try to develop strategies to enhance compliance, whilst children continue to test "limits". Conflict is not uncommon with parents trying various strategies; sometimes either a more authoritative approach in an attempt to get their child to do essential health related treatments or a more permissive/over protective approach, only doing minimal treatments or choosing the battles.

Mealtime behavioural issues are noted as being more problematic than in non CF families, with reportedly less than a quarter of children reaching recommended dietary requirements. Nutritional intake has direct correlations to lung function and overall health and can be an ongoing battle throughout the life span. Children with CF may demonstrate behaviours that interfere with eating such as delaying meals by talking, leaving the table, crying and whinging. To counteract this behaviour, parents may use increased coercion, commands, and physical prompts, sometimes feeling that they have little control over their child's eating.

Strategies

Behaviour modification can alter future maladaptive issues around food. Behavioural techniques include the praising of desired eating, ignoring noneating behaviours, setting time limits for meals, and the use of goal setting and reinforcement of goal setting. Useful supports:

- **CF food http://www.cysticfibrosis.org.au/wa/cffood**
- **www.clanwa.com.au/**
- Speak with PMH treatment team, dietitian and or psychologist

School age

Peer relationships become increasingly important at this stage. For some children, going to school can bring about a second diagnostic period where they realise at a deeper level the impact of having CF and being different to their peers. There can be a high cost in appearing to be "normal" through minimising the more visible aspects of CF. Keeping CF a secret can have a negative impact on the ability to maintain a supportive peer network. Children at this age can develop anxiety and/or depressive symptoms, particularly if parental distress is not managed.

Strategies

Age-appropriate and accurate information about CF is important to assist the child to understand their illness, the relationship between CF and their daily treatments and medical procedures. Coping strategies about CF-related experiences are also very important e.g. distraction and pain management if undergoing a medical procedure.

- **Procedural Anxiety fact sheet coming soon http://www.cysticfibrosis.org.au/wa/fact_sheets**
- **http://www.cysticfibrosis.org.au/wa/brochures (Kids magazines)**

- **https://www.dcp.wa.gov.au/CrisisAndEmergency/Pages/Familyandparentinghelplines.aspx**

Adolescence

Adolescence is typically a challenging phase with proportional time spent between peers and family; peers often becoming more influential. For some teens, CF symptoms such as cough and fatigue can worsen, especially for females. This effectively means more reliance on family and at times, hospital, rather than with friends where they would more often rather be.

On the up side, there are some positive associations with having CF such as increased social skills, assertiveness and a higher degree of negotiation skills due to regular discussions with adults about health treatment and the need to manage treatment.

Several barriers to adherence have been identified such as a lack of personal knowledge about CF and their treatment plan, disagreement with treatment plans, complexity of treatment, psychological issues (child/parent or both), "forgetting", time management, skills to manage routines and low level of connection to disease symptoms. Research indicates that adherence to treatment plans could be as low as 50%, particularly if the teen or their parent is struggling with anxiety or depression.

Strategies

Teens need to start owning their treatment and setting individual goal plans with their treatment team, home care worker and/or parent. Research has shown that teens who believe that their treatments are necessary are more motivated to do them. Self-management generally increases at this age, however, supervision by an adult still correlates to better adherence. Motivational interviewing or health coaching can be a useful strategy to help set some health goals and break down barriers to treatment. CFWA can assist with this.

Typical adolescent autonomy is developed by being able to take a few risks and learn the consequences. This has bigger implications for a teen with CF and parents often need to stay in a supervisory role much longer than teens without CF, but need to shift their role into a more collaborative one where their teen can start managing one small aspect at a time. This may be tied to natural consequences of more freedom in other areas. Useful resources for teens:

- **http://www.blackswanhealth.com.au/services/ataps/**
- **http://headspace.org.au/headspace-centres/ (including regional)**
- **http://youthfocus.com.au/**
- **https://www.youthbeyondblue.com/**

Second diagnosis

CF-related diabetes (CFRD) and CF-related bone disease (CFRBD) are not uncommon secondary diagnoses on top of having regular CF symptoms. Getting a second diagnosis such as CFRD can be considered a medical milestone associated with worsening symptoms of CF. This can occur during late adolescence and affects many adults. A new diagnosis can trigger a lot of anxiety, particularly if CF symptoms have worsened. Some adults find that they have to take a closer look at their management strategies and perhaps cut back on some activities, such as work. If people are already managing CF well,it is generally easier to manage the requirements of a second diagnosis.

Follow these links to get more information on how to manage CFRD and bone health:

- **https://www.cysticfibrosis.org.uk/life-with-cystic-fibrosis/publications/factsheets**
- **http://cfnz.org.nz/wp-content/uploads/2015/12/FS_Bone_Health_Mar_13.pdf**

CF-related pain

CF-related pain is not uncommon for both children and adults, particularly when unwell. Pain can significantly impact quality of life and willingness to do airway clearance techniques, affecting health outcomes. Anticipatory fear affects approximately 80% of both children and adults when facing particular procedures such as insertion of intravenous therapy. The following links may help with strategies for managing pain and anxiety:

- **Procedural Anxiety fact sheet coming soon http://www.cysticfibrosis.org.au/wa/fact_sheets**
- **http://www.sickandhappy.com/im-getting-old-and-thats-good-right/**

Weight and nutrition

With each developmental stage comes a readjustment in both perspective and treatment. Dietary requirements are one significant factor that can constantly need adjustment, particularly at times of illness where higher energy requirements are necessary. Studies indicate that approximately 12-18% of people with CF have issues around food and weight including the omitting of enzymes, insulin, overusing laxatives, not complying with supplements, over exercise and food restricting. Low weight has serious implications for overall health including lung function, the ability to fight infection and bone health. Useful resources and links:

- **http://95.138.169.194/search.php?sid=8eb4382926b9a478280f4d67526a55b6 (CF Trust Forum)**
- **Seek advice from your dietician and set goals for your ideal weight**
- **CF Food – Booklets**

References

http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2874200/

http://www.hindawi.com/journals/pm/2012/134132/

http://www.sickandhappy.com/depression-sucks-and-what-you-can-do-about-it/

Didsbury, Thackray 2010, Cystic Fibrosis and Body Image

Golf Classic 2016

WE ARE PLEASED TO ANNOUNCE THAT THE GEORGE JONES FAMILY FOUNDATION GOLF CLASSIC AT THE PRESTIGIOUS LAKE KARRINYUP COUNTRY CLUB, NOW IN ITS SIXTH YEAR, WAS ANOTHER GREAT SUCCESS IN 2016.

The weather was looking patchy as the 32 teams arrived at registration, but the sun was shining by the time the players teed off, and it stayed that way throughout the day as the teams made their way around the world-renowned course. After the game, players enjoyed refreshments on the terrace before the evening's host, Steve Mills, kicked off the proceedings. Guests enjoyed a fabulous meal, paired with wines from our generous sponsor Capel Vale, and fought it out for some great auction items including a luxury Karma Resorts accommodation package.

An incredible $103,500 was raised from this year's event through team sponsorship packages, auctions, raffles and donations. This means that over $577,500 net profit has been raised for cystic fibrosis through the Golf Classic in the past six years! Thank you to all of the sponsors and donors for their ongoing support, many of whom have now been with us since the first Golf Classic in 2011.

We'd particularly like to thank our naming rights sponsor, the George Jones Family Foundation, who have been an incredible supporter over the past five years, and the volunteer committee, chaired by Bradley Coutts, who have worked tirelessly throughout each year to organise such a prestigious event.

Registrations for the 2017 will open later this year. If you'd like more information about the 2017 Golf Classic, please get in touch on 08 9346 7333 or at marketing@cysticfibrosiswa.org

George Jones Family Foundation

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WINNERS ON THE DAY

1st	Bankwest	Paul Simpson	Don Leary	Mark Hector	Glen Tilehurst
2nd	Monitor WA Pty Ltd	Dan Peardon	Dave Ritchie	Brad Downey	Mark Simpson
3rd	Alder Tapware	Tony Brankovic	Matt Ingram	Jamie Harnwell	Adrian Hall

Healthy Bones are Important

OPTIMISING HEALTH THROUGH GOOD NUTRITION IS ESSENTIAL TO DEVELOPING HEALTHY BONES. THE FOLLOWING NOTES ARE A SUMMARY FROM THE 11TH CF AUSTRALASIAN CONFERENCE 2015, ENTITLED HEALTHY BONES: MEDICAL, NUTRITIONAL AND EXERCISE INTERVENTIONS TO OPTIMISE BONE HEALTH.

What determines bone growth and strength in the general population?

- Genetics (80%)
- Exercise (15%)

What determines bone mineral density (BMD) in the CF population?

- Genetics and having Δ 508 mutation
- Inflammatory markers released in chronic lung infection reduce bone formation and reduce muscle mass which influences BMD
- Delayed puberty
- Diabetes interferes with the pathway for bone growth
- Poor diet/malnutrition
- Poor growth

How does nutrition influence bone growth?

Adequate intake of dietary calcium is needed to form calcium in bone. Dairy foods are the major contributor to calcium intake and should not be avoided. Hard cheese is lactose free so can be eaten on a lactose-free diet. If you drink lactose free milk make sure it has added calcium. A high protein intake increases calcium absorption, bone formation and BMD.

What type of exercise is best for bone development?

It has been shown that there is a strong link between regular weight bearing physical activity and increase in BMD. The intensity of exercise is important. Those with CF who participated in high impact activity (50 jumps, 3 times/week) were shown to have a significant increase in BMD. Weight bearing exercise in adolescence increases bone mass in adulthood.

Preventing bone problems, e.g. kyphosis (outward curvature of the spine, causing hunching of the back):

- 50 % of adults with CF have kyphosis and 30-50% report some type of bone pain. Better bone health by improving BMD will help reduce this.
- Be aware of your posture
- Look at your posture in the mirror. Try to keep your chin up and shoulders back. See your physiotherapist if you are concerned.
- Keep your trunk mobile; include regular stretching in your exercise program.
- Encourage children to stretch arms upwards; racquet/bat and ball games are good, e.g. tennis, T-ball, basketball etc.

Reference: 2015 Munns, C; King, S & Middleton, A. Healthy Bones: Medical, nutritional and exercise interventions to optimise bone health. 11th Australasian Cystic Fibrosis Conference.

An Evening with CF Scientists

ON THE 4 MAY 2016, CYSTIC FIBROSIS SCIENTISTS FROM THE TELETHON KIDS AND THE HARRY PERKINS INSTITUTES PRESENTED THEIR LATEST RESEARCH RESULTS.

Presentations were made by Anna Tai, Jamie Wood, Tim Rosenow, (who also presented on behalf of Shannon Simpson), and Cindy Branch-Smith.

Genetic variation within a single bacteria strain Pseudomonas aeruginosa, found in Australian patients with cystic fibrosis

Anna explained that 50% of Australian patients with cystic fibrosis (CF) were infected with the bacteria, Pseudomonas aeruginosa. The two most common strains AUST-01 and AUST-02 were only ever found in CF lungs, and not in patients without CF suggesting that AUST-01 and AUST-02 had evolved specifically to survive in CF lungs. For this to happen it was necessary for cross-infection to spread these strains within the Australian CF population.

1. The study showed genetic changes in the AUST-02 strain were associated with the development and spread of a new multi-drug resistant AUST-02 substrain (M3L7) within an adult CF centre in Queensland.
2. This strain was highly resistant to most antibiotics compared with other Pseudomonas strains.
3. This highlighted the importance of paying careful attention to cross-infection precautions to restrict the spread of new multi-drug resistant Pseudomonas strains within a CF clinic.
4. Based on Anna Tai's findings, a new screening process was developed which enabled quick and cost effective testing for the busy CF clinics.
5. This resulted in a new purpose-built CF ward with 14 single room facilities atThe Prince Charles adult CF centre in 2014.

The new Perth Children's Hospital due to open this year in Perth will incorporate the very best cross infection precautions available to help prevent spread of multi-drug resistant Pseudomonas strains within a CF clinic.

Telehealth for adults with cystic fibrosis in rural and remote Western Australia

Work undertaken by Jamie Wood, Siobhain Mulrennan, Kylie Hill,Nola Cecins, Sue Morey and Sue Jenkins seeks to address the challenges of delivering services to CF patients living in rural and remote WA using Telehealth.

Jamie Wood, is the Physiotherapist at Sir Charles Gairdner Hospital (SCGH) the WA state adult CF centre, explained that 15% of CF adults live in regional and remote areas of WA but only 22% of them attended four or more CF clinics per annum.

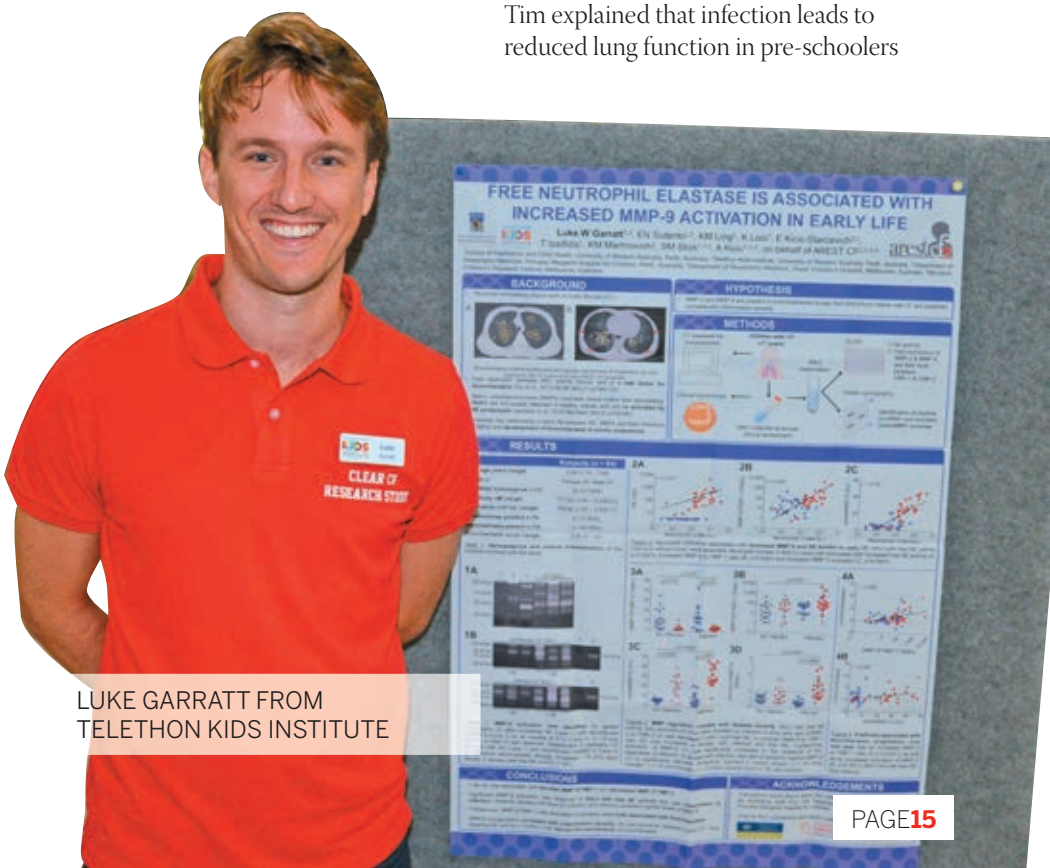
Telehealth allowed these patients to receive services without having to travel to Perth. Most participants preferred this and increases in antibiotic use and hospital admissions highlighted the increased detection of exacerbations and provision of appropriate treatment. A significant number of participants gained weight, and there was an improvement in the vitality domain of the Cystic Fibrosis Questionnaire Revised.

Clearly, Telehealth is here to stay and provides an important weapon in the arsenal for delivering world class services to the rural and remote population of WA.

The Effect of early infection on lung function in infants with cystic fibrosis

Tim Rosenow presented a paper on behalf of Shannon Simpson which looked at the effect of early infection on lung function in infants with CF.

Tim explained that infection leads to reduced lung function in pre-schoolers



LUKE GARRATT FROM TELETHON KIDS INSTITUTE

and school-aged children, but the impact of infection on infants was not as well understood.

Tim and his colleagues tracked the lung function of 108 infants over the first two years of life. The lung function test they used was the multiple breath washout (MBW) test. MBW assesses how evenly inhaled air mixes throughout the lungs by determining the lung clearance index (LCI). A higher LCI means that ventilation is more variable throughout the lung, indicating worse lung disease. Infection was detected using bronchoalveolar lavage.

The results of this study showed that those who had an infection detected had a higher (worse) LCI than those that did not. In addition, once an infection was detected, the rate of lung function decline accelerated, even if there was no infection at the subsequent visit. H. influenzae had the largest effect on lung function.

This study shows that early infection with respiratory pathogens has a significant effect on lung function. It highlights the need for close monitoring of pulmonary infection and early intervention in infants with CF who otherwise may seem well.

PRAGMA-CF: A sensitive clinical trial outcome for young children with CF

AREST CF and in particular the work of Tim Rosenow has been pioneering in the development of highly sensitive detection techniques to identify early CF-related lung disease in infants.

Tim and his collaborators developed the Perth-Rotterdam Annotated Grid Morphometric Analysis for CF (PRAGMA-CF), a system of measuring the extent of lung disease in young children(aged below six years) with CF. This method involves overlaying a grid over the CT images of the lungs and allocating each grid square a category based on the presence of various lung abnormalities. From this, the extent of lung disease (as a percentage of total lung volume) is calculated.

TIM ROSENOW EXPLAINING HIS RESEARCH.

GERARD DONALDSON, PROF. STEPHEN STICK, CHRISTINE DONALDSON AND NIGEL BARKER.

With this method, they were able to detect lung disease affecting less than 1% of the lung and its progression from ages one to three.

The results from these studies have been published in the "Blue Journal," the highest rated respiratory medical journal in the world.

Infants and young children stand to benefit the most from the many new medications and therapies becoming available, with the aim to prevent structural lung damage. Without a reliable outcome measure, clinical trials in this age group had not been possible.

Supporting young people in school

The final presentation was made by Cindy Branch-Smith who reported her findings regarding supporting young people living with CF, particularly in school.

Cindy showed that young people living with chronic conditions often miss significant opportunities at school to develop socially and emotionally.

The research team is currently co-developing, with young people with CF and their families, a paper-based version of an app that addresses their social and emotional health needs and engagement with school. Future funding will enable this paper prototype to be developed into an online resource and app, and test its effectiveness. Based on previous research, this will both increase their wellbeing and result in substantial academic gains.

You can view the full lectures on You Tube by searching for "An Evening with CF Scientists 2016". Full lay summaries may be viewed at **www.tinyurl.com/hcw2xnt**

CFfood

Thanks to generous funding from Newman's Own Foundation, we are proud to announce that we have developed a new set of resources under the title CFfood. The resources provide information about the dietary requirements and how to maintain good nutrition for people with CF from babies to adults.

The components of CFfood include:

- Nutrition and Cystic Fibrosis: A Guide for Feeding Babies (Aged 0 to 2 years)
- Nutrition and Cystic Fibrosis: A Guide for Feeding Children (Aged 3 to 12 years)
- Food for Young People with Cystic Fibrosis (Aged 13 to 17 years)
- Nutrition and Cystic Fibrosis: A guide for Adults (Aged 18+)
- A cookbook: " High Energy Recipes for Cystic Fibrosis" for toddlers through to adults.
- Upon admission to hospital, a Fat Booster Hospital Bag, filled with a range of nutritional, high calorie goodies, is also provided for children and adults with CF.

The resources developed for CFfood are available online at www.cysticfibrosis.org.au/wa/brochures or in hard copy. Contact CFWA for more details if you would like a hard copy of one of the booklets.

CF Cooking is Coming

CYSTIC FIBROSIS WA IS EXCITED TO ANNOUNCE THAT WE ARE PARTNERING WITH OUR WA CF ADULT DIETITIAN, PAUL O'NEILL, TO PRODUCE A SERIES OF COOKING DEMONSTRATIONS WHICH WILL BE PROFESSIONALLY FILMED AND MADE AVAILABLE ONLINE. THIS WILL BE AN INNOVATIVE HEALTH RESOURCE AND IS POSSIBLY ONE OF THE FIRST OF ITS KIND IN THE WORLD.

Paul has been the adult CF dietitian at Charlie's for eight years. He has worked as a chef in another life and is excited to be working on some delicious recipes for this project.

He says, "We know that nutrition is important for people living with CF, yet the 'CF diet' seems opposite to the healthy diet for the non-CF population". This can often be a dilemma because when we eat food, it is often a shared experience. For example, at work, in the playground, at the family table, or meals with friends can produce what seems like a clash between different health recommendations. CF Cooking has been designed to help people with CF cook a healthy CF diet and to negotiate some of these difficulties.

There will be a series of six videos that will be specific to CF food and nutrition. The project is currently in the planning and development stage, but filming will start soon and will be completed by November this year.

The videos will be placed online so that anyone with a smartphone, tablet or PC can access them wherever there is internet coverage. The recipes and other resources will also be available in a downloadable format. So keep your eyes out for the launch of 'CF Cooking' later this year.

CFcooking
cystic fibrosis healthy cooking

Tips for Managing Toddlers Who are Fussy Eaters

MEAL TIMES CAN BE A CHALLENGE FOR ANY PARENT, BUT EVEN MORE SO WHEN THEIR CHILD HAS CYSTIC FIBROSIS (CF). HERE ARE A FEW SUGGESTIONS THAT MAY MAKE MEAL TIMES MORE ENJOYABLE.

As a parent, it's your job to decide what foods are offered, when foods are offered and where foods are eaten, but your toddler can decide how much to eat and which foods to eat.

- Offer a range of colourful foods on the plate and allow your child to choose what he or she wants to eat
- Allowing your child to help prepare some of the meals could encourage their interest in food
- Encourage self-feeding and exploration of food from an early age
- Offer alternative foods from each food group
- Encourage your child to feed independently and to sit down while eating
- By the evening, toddlers can be very tired, so small meals of easy to eat foods can be more beneficial for your cranky toddler; sometimes if things are too tricky a nutritious milkshake may be the way to go
- Avoid using sweet food as a bribe and keep them out of sight until savoury foods have been eaten at meal times

Be patient. It's been reported that kids will need to try new foods anywhere from 7 to 15 times before acquiring a taste for it. That's a lot.

Toilet Training Tips!

TOILET TRAINING CAN SEEM VERY DAUNTING FOR PARENTS BUT WHEN YOU ADD CYSTIC FIBROSIS (CF) INTO THE MIX, PARENTS CAN FEEL OVERWHELMED AT THE THOUGHT OF THE WHOLE PROCESS. WITH THE ADDED FOOD AND FLUID INTAKE AND POSSIBLE GUT ISSUES, WHICH EQUALS INCREASED TRIPS TO THE BATHROOM, YOU COULD BE WONDERING; ARE WE EVER GOING TO BE ABLE TO TOILET TRAIN OUR CHILD?

I am very pleased to tell you that many parents have been pleasantly surprised at how easy it ended up being! Below are some tips to assist you.

When should toilet training begin?

Toilet training should begin when your child shows clear signs that he or she is ready. There is no right age and the process should be treated as just another step in their development. The average age is 3 years for daytime continence. Most children show signs of bowel and bladder awareness between 18 months to 2 years but with children with CF and different stool consistency, this may be tricky to navigate!

If the family environment is experiencing a significant change, such as an arrival of a new baby, moving house or your child is starting daycare, try and delay commencing toilet training until things settle.

Every child is unique!

Signs of readiness

Signs that your child may be ready for toilet training include the following:

- Your child's nappy is staying dry for longer periods (about 2 hours at a time)
- Your child is aware that they have had a bowel motion in their nappy
- Your child shows signs that they are uncomfortable when their nappy is wet or dirty

- Your child has an interest in the toilet and tries to copy parents or older children
- Your child can label body parts
- Your child has a bowel motion at a similar time/times each day
- Your child can sit still for short periods of time
- Your child can follow simple instructions

Getting started

Be prepared for the routine of toilet training because initially it might be toilet timing. In the beginning it is best to spend some time at home whilst working on this. It is often suggested to parents to work on the morning first and build on that success. Children are generally less tired and more enthusiastic in the morning!

Prepare the environment

Is the toilet close by to play and living areas? Is there ample light as some children are fearful if the toilet is dark? Decide whether you will be using a potty or the toilet. If you are using the toilet, you will need to purchase a small step which can be found at any baby shop or department store.

- Reading a book together on toilet training can be a helpful reference for your child and will help to reduce the pressure on them.
- Ensure your child is wearing clothes that are easy to remove. Being able to navigate and remove clothing quickly is essential.
- Show your child how to use the toilet or potty.
- Explain to your child, in simple words, what they need to do. How much paper to use and how to wipe themselves, how to flush the toilet and wash their hands each time.
- Purchase 'grown up' undies and lots of them. Plan a special and exciting shopping trip together!
- If your child has a usual time for bowel movements (such as after a meal) you can take your child to the toilet at that time of day.
- Always try to remain positive and calm about toilet training. Your child is not ready if you are constantly having a battle to get them onto the toilet. If this is the case, stop everything and try again in a few months.
- Coach, praise and encourage every small step. Don't wait until they can do the whole task properly before praising them. Saying things like, "Did you know you could do that?" Or "Well done for trying to get to your potty" (even if they are a bit late). Focus on the positive and ignore the negative.
- Remember to encourage and practice good hygiene as part of toilet training. Remind your child to wash, dry and moisturise their hands each time they use the toilet. This should be completed whether successful or not!

Other tips

Staying dry at night may take some extra time. Complete night time control may not occur until your child is 4 or 5 years old, or even older. If your child is 5 or older and does not stay dry at night, you may want to discuss this with your child's medical team.

Even when children are toilet trained they may have some accidents when excited or busy playing, or setbacks due to illness or emotional situations. Be patient and calm and be reassured that this is normal and expected.

You will have to adjust your routines slightly to include potty or toilet time. Some CF parents have commented that "we are now acquainted with the toilets at every restaurant and shopping centre we've been in the past few months".

A message for parents approaching toilet training:

"Remember that soon this is something they will deal with on their own and helping getting them there is the least we can do!"

References

http://www.ngala.com.au/files/files/375_PRG_22_Toileting_Tips.pdf

http://www.stanfordchildrens.org/en/topic/default?id=toilet-training-90-P02300

http://www.continencevictoria.org.au/one-step-at-a-time-toilet-tips/

http://raisingchildren.net.au/articles/toilet_training.html

http://sisterfibrosis.com/potty-training-cf-edition/

CYSTIC FIBROSIS
Western Australia

Toilet Training Tips and Starting School with CF

Monday **1 August** 2016

5.30pm – 8.30pm at
Cystic Fibrosis WA (CFWA)
The Niche Building

11 Aberdare Road,Nedlands

AGENDA:

5.30 - 6.30 pm: Toilet training tips for toddlers with CF
6.30 - 7.00 pm: Chat with other parents
7.00 - 8.30 pm: Starting school with CF

You can attend one session or both sessions if you wish. A variety of CFSmart education resources and toilet training handouts will be available. Drinks and nibbles will also be provided.

Please RSVP as to which sessions you will attend by
2pm Monday 1st August to Natalie Amos
email: education@cysticfibrosiswa.org or phone: 08 9346 7333

Taking Enzymes in High School

IF YOU ARE A TEEN WHO FINDS IT EMBARRASSING TAKING YOUR ENZYMES AT SCHOOL, KEEP READING. WE HAVE A FEW TIPS ON HOW TO BE DISCREET WHILE YOU TAKE THEM....

The peer pressure to conform to what other teens are wearing and doing can be a minefield, so doing something different like taking medication at school, can make things even more tricky.

Being different and being treated differently

The thing is when you have something like cystic fibrosis (CF) it does make you different. There's no getting around that. The choice you have is whether to see this as a bad thing, a good thing or just one thing about you, when there is so much more. You have some power over how people treat you. If someone is not treating you well, you can ask them not to. If someone is treating you well you can reinforce that by thanking them or encouraging that response. It might take a bit of time but eventually people will realise that taking medication is just what you do and it's not a big deal.

If there are students making a big deal about you taking your medication, it's worth speaking to a supportive teacher or the school nurse about it and also with your parents. Getting someone else in to the school to provide education to the teachers and students about what CF is and how it can affect a person who has it, can be useful. Teachers and students who understand more about CF are better equipped to provide you with more support and understanding and peers can learn about a condition that they could potentially be a carrier for themselves. Cystic Fibrosis WA (CFWA) can provide education sessions about CF for teachers and students in high school.

How do I explain what the enzymes are for?

Take a few minutes to think up a quick explanation for your enzymes so that if someone asks, you can easily think of what to say. This can be your 'go to' phrase for all occasions if someone asks. Your explanation can be very simple such as: "I need these tablets so I can digest my food".

There will also be other students in your school who will be taking medication for different reasons and they could be embarrassed about it too, so sometimes it is good to remember you are not alone.

Tips for disguising taking your enzymes

- Store enzymes in a mint or lolly container. (Make sure the date of the enzymes that you put into the container is checked – if they are out of date they won't work as well. Write the date in a black marker on the bottom of the container).
- Take the enzymes while heading to the drink fountain. Walk away from the group of friends you are with, have your enzymes in your pocket and quickly put them in your mouth before taking a drink.
- Ask a teacher if there is somewhere private where you can take your enzymes on a regular basis.
- Test the waters by taking your enzymes in front of someone you know well and trust, first. Their positive reaction might help build your confidence.
- Distract your audience. Ask another person a question so the group is looking at them or start a new topic of conversation so people become involved in that. Or, you could have your enzymes when everyone's attention is naturally focused elsewhere.
- If you are very worried, maybe you can take your tablets just before you eat, in the bathroom, at your locker or ask your teachers if you can take them in the classroom once the students have gone.
- If you forget to take your enzymes at the start of the meal, take them during your meal, or you can still take them at the end, as long as it's within 5 -10 minutes of eating. Better late than never!

Someone you trust like your parents, close friends, school nurse, teachers or your CF dietician may be able to suggest other ways to help you feel more comfortable taking your medication.

Other useful resources

- Look up "How to be a teenager with a chronic illness" by Charles Michael Duke, on YouTube. He has some interesting advice explained in a very humourous way.
- CF explained for teachers: **https://cfsmart.org/teachershs/**
- CFWA can post you a High School Teacher booklet or arrange to visit your school to speak to teachers and/or students about CF. Contact Natalie Amos at **education@cysticfibrosiswa.org or 08 9346 7333**
- For more information about CF nutrition we have booklets available in hardcopy "Food for Young People with Cystic Fibrosis" or online at **http://www.cysticfibrosis.org.au/wa/brochures**

Have You Seen our Nutrition Fact Sheets?

CFWA HAS BEEN BUSY DEVELOPING A NUMBER OF FACT SHEETS TO PROVIDE UP-TO-DATE INFORMATION TO GUIDE YOU IN THE MANAGEMENT OF CF. THE FOLLOWING IS A SUMMARY OF TWO OF THE NUTRITION FACT SHEETS WITH A LINK BELOW TO MORE DETAILED INFORMATION.

Salt and fluid replacement

CF causes changes in many parts of the body including the sweat glands, resulting in increased salt loss when sweating. The Salt and Fluid Replacement Fact Sheet discusses the impacts of increased salt loss on people with CF and how this can be managed. Some key points covered include the main causes of salt loss, symptoms of resulting dehydration and prevention and management strategies.

Percutaneous endoscopic gastrostomy (PEG)

One of the most important goals of CF care is maintaining optimal nutrition. This can be very difficult for some people who are unable to reach their goal weight with regular meals, snacks and supplements. In these cases weight gain can be assisted by placement of a PEG feeding tube. The PEG fact sheet discusses how to care for your PEG feeding tube and troubleshooting any problems that may arise.

For more information on Salt and Fluid Replacement or PEGs or to access any other facts sheets follow the linkhttp://www.cysticfibrosis.org.au/all/fact_sheets.

Cystic Fibrosis-Related Diabetes (CFRD) and You

CF-RELATED DIABETES IS CAUSED BY DAMAGE TO THE PANCREAS OVER A PERIOD OF TIME. IT IS COMMON FOR PEOPLE WITH CF TO BE DIAGNOSED WITH CFRD – IT OCCURS IN AROUND 20% OF ADOLESCENTS AND 40 - 60% OF ADULTS WITH CF. CFRD SHARES FEATURES WITH OTHER TYPES OF DIABETES BUT IS A DISTINCTLY DIFFERENT CONDITION.

To understand CFRD, you need to understand a bit about the pancreas, glucose and insulin. The pancreas is an organ that has two main functions. The first is to make and secrete a hormone called insulin and the second is to make and secrete digestive enzymes and juices. In CFRD, the pancreas' ability to produce enough effective insulin is affected.

Insulin is a hormone that helps to control blood sugar levels, which can also be referred to as blood glucose levels. Glucose is a type of sugar that is used by cells in our bodies for energy. This is our body's main source of fuel. It is essential for life. Without glucose, our cells can't operate and carry out all their different functions.

We get most of our glucose from carbohydrates. The carbohydrates we eat are digested in our stomach and broken down into small particles - mostly glucose - and then transported around the body by the blood to the cells where energy is needed.

However, glucose can't get into the cells without insulin. Think of it like a lock and key system. The cells are 'locked' and in order for the glucose to get into the cells, insulin is required to act like a 'key,' allowing the glucose in.

If glucose cannot enter our cells two things happen:

1. The cells do not receive the energy they need to work properly
2. Blood sugar levels rise

CFRD is different to other types of diabetes

- Type 1 diabetes is an autoimmune condition in which the body attacks the cells in the pancreas (beta cells) which produce insulin. Type 1 diabetes accounts for around 9% of the diabetes population. It is usually (but not always) diagnosed in children and young adults and is managed with insulin.
- Type 2 diabetes occurs when the body becomes resistant to, and /doesn't produce enough, insulin. Type 2 diabetes is not always preventable as development may be linked to family history, however, it may be managed with diet and exercise alone. Some people will need to use medication as well as lifestyle changes to manage it. Type 2 diabetes used to be thought of as a disease only affecting adults, however, it is becoming increasingly common in younger people.
- CFRD is different to other types of diabetes and the symptoms may vary from person to person. It may be

triggered by an exacerbation of CF and treatment for infection or may be a gradual onset. It is often worse during CF exacerbations, as inflammation and infection make the cells of the body less sensitive to insulin and medications used to treat exacerbation can increase blood glucose levels, as can pregnancy and some other medications.

The major risks for uncontrolled CFRD come from the body not being able to transport enough glucose to the cells for them to function optimally. The concerns seen in uncontrolled CFRD include weight loss, loss of protein stores in the body and poor health outcomes such as decline in lung function.

What are the symptoms of CFRD and high blood glucose levels (hyperglycaemia)?

- Lack of energy
- A decrease in lung function
- Weight loss
- Increased hunger
- Excessive thirst
- Increased urine output

Does everybody who has CFRD have symptoms like this?

No. Often people are diagnosed with CFRD during a routine blood test. Some symptoms of CFRD may also be very similar to other CF-related symptoms. This is why people with CF are regularly screened for CFRD.

Why is it important to screen for CFRD?

If the symptoms of CFRD are left untreated, your CF symptoms can become worse. This can lead to:

- Increased thickness of mucus which becomes more difficult to clear
- Reduced ability to fight infection
- Loss of muscle
- Loss of weight

Increased blood glucose levels over time can lead to other diabetes-related complications, such as damage to small blood vessels, impaired kidney function and vision impairment.

Screening and diagnosis of CFRD

The gold standard for CFRD testing is the Oral Glucose Tolerance Test (OGTT). Ideally this should be done annually. Other tests are used to check for Type 1 and Type 2 diabetes but can sometimes miss people with CFRD. The OGTT is more sensitive and the best choice for screening in CFRD.

Managing CFRD

As patients with CFRD are insulin insufficient (don't produce enough insulin), taking insulin is the treatment for CFRD. This helps people achieve 'glycaemic control' which means they have the right amount of glucose in their blood.

In other types of diabetes, losing weight and changes to the diet can play a big part in diabetes management, but a diagnosis of CFRD does not change the existing recommendations for nutrition in CF. Weight loss is not part of the management plan, fats are not restricted and a high energy; high protein diet is still recommended. It is important that carbohydrates are consumed throughout the day. You should discuss this in more detail with your CF dietitian.

Diabetic Ketoacidosis or DKA, is a serious complication of diabetes which can occur in other types of diabetes, but it is extremely uncommon in CFRD.

What are the symptoms of hypoglycaemia?

- Symptoms of hypoglycaemia can vary from person to person and early signs may include:

Feeling shaky and weak

- Hunger
- Sweating
- Lightheadedness, dizziness
- Headache
- Pins and needles around the mouth
- Mood change

How do you manage a hypo?

- The first signs of a "hypo" can be hunger and you may start to feel shaky. This can lead to sweating and possibly fainting.
- It is very important that you understand how to treat low glucose levels, particularly if you are taking insulin.
- If you think you might be experiencing a "hypo" you need to eat 15g of fast acting carbohydrates. This is equivalent to 3 big jelly beans, 3 teaspoons of sugar or honey or 50ml of lemonade
- It is very important that you DO NOT drive if your BGLs are below 5 mmols.

Talk with your CF team about how to prevent and manage your "hypos" and always carry fast-acting carbohydrates with you.

For more information about CFRD, make an appointment to see a Diabetes Educator.

Acknowledging Sandra Wilberforce Diabetes Educator SCGH as a resource for this information

National Diabetes Week will take place from **Sunday 10 to Saturday 16 July 2016**. To mark the event, this year Diabetes WA will be shining a light on foot health and diabetes.

Diabetes can damage the nerves and blood vessels in your feet, leaving you at risk of ulcers. The good news is that you can reduce your risk by paying attention to your feet and looking after them properly. To raise awareness about importance of looking after your feet, Diabetes WA has developed a range of resources that can be downloaded for free.

Cystic Fibrosis-Related Liver Disease and Pancreatitis

MILD CYSTIC FIBROSIS-RELATED LIVER DISEASE (CFLD) IS COMMON, WHILE SEVERE CFLD AFFECTS 10% OF PEOPLE WITH CF. PANCREATITIS RARELY AFFECTS PATIENTS WHO ARE PANCREATIC INSUFFICIENT, HOWEVER, NEARLY ONE IN FIVE PATIENTS WITH CF CAN BE AFFECTED.

Cystic fibrosis-related liver disease

CFLD is a broad term which refers to a spectrum of abnormalities ranging from mild liver enzyme derangement on routine blood tests to biliary cirrhosis with portal hypertension and liver failure. Severe CFLD is the third leading cause of death in CF, after respiratory failure and transplant-related complications.

What causes CFLD?

The underlying genetic defect in CF is due to mutations in the CF transmembrane regulator (CFTR) gene which encodes the CFTR chloride channel. The CFTR protein is located on the epithelial lining of the bile duct and gall bladder. Its key function is to hydrate bile and facilitate bile flow. Abnormal CFTR function leads to thickened and sluggish secretion which blocks the bile ducts (1, 2). This leads to excessive accumulation of toxic bile salts which causes inflammation, fibrosis and cirrhosis of the portal tract inside the liver. Furthermore, the CF gut is thought to have increased permeability. This leads to the leakage of bacterial factors from the gut into the portal circulation which incites further liver inflammation and fibrosis (1).

Clinical presentations of CFLD

Mild CFLD is common in CF. Transient liver enzyme abnormality is common in children and adults with CF and generally does not cause major issues. However, severe CFLD affects 10% of patients with CF. Severe CFLD refers to biliary cirrhosis with portal hypertension and liver failure. Portal hypertension causes enlargement of the spleen, low platelets and oesophageal and gastric varices (enlarged blood vessels). Patients with portal hypertension are at risk of a significant variceal bleed. Progression of CFLD can lead to liver failure. Severe CFLD mainly affects those with severe CFTR mutations (Class I- III). Patients with the same CFTR mutations can have different disease trajectories (3). This is likely to be due to the complex interaction between the CFTR gene with other modifier genes and environmental factors in the development of CFLD. Males tend to be affected by CFLD more than females.

Other types of CFLD include neonatal cholestasis, hepatic steatosis, microgallbladder, gallstones and biliary sludge (4). Cancer of the bile duct and gall bladder are uncommon but had also been reported in CF.

Investigations

The diagnosis CFLD diagnosis is based on clinical examination, blood test and radiological imaging including abdominal ultrasonography (US), MRI or CT scans and newer techniques such as elastography. Liver biopsy is sometimes required to confirm the diagnosis.

Management of CFLD

To date, there is no effective treatment for preventing or slowing the progression of CFLD. Supportive management are important in CFLD. This includes optimising the nutritional status, avoiding alcohol and hepatotoxic medications and vaccination against hepatitis A, hepatitis B and varicella zoster virus. Patients with an enlarged spleen should consider avoiding contact sports to prevent splenic injuries. In patients with portal hypertension, regular surveillance gastroscopies are required for the early detection and banding of varices. In patients with liver failure, liver transplant is a potential option.

Ursodeoxycholic acid (URSO) improves liver enzyme abnormalities but its efficacy in CFLD is controversial. To date, there is insufficient evidence to show URSO improves long-term clinical outcomes in CFLD.

Exciting developments in CFTR modulation therapies (e.g. ivacaftor) have demonstrated substantial clinical benefits in patients with specific CFTR mutations. These benefits include improvement in respiratory symptoms, lung function and nutritional status. However, it remains unclear if CFTR modulation therapies can also prevent or halt the progression of CFLD and further studies are required.

CF-related pancreatitis

Pancreatitis almost exclusively affects patients with CF who are pancreatic sufficient and rarely affects patients who are pancreatic insufficient (5). Nearly one in five patient with CF who is pancreatic sufficient is affected by pancreatitis (5, 6). Pancreatitis is also an independent risk factor for developing pancreatic insufficiency later in life (1, 5, 6).

What causes CF-related pancreatitis?

CFTR is expressed on the epithelial lining of pancreatic duct. It regulates the acidity, fluidity and the enzyme content of the pancreatic fluid. Patients who are pancreatic sufficient have preserved pancreatic tissue which produces digestive enzymes. However, abnormal CFTR function causes abnormal thickened pancreatic fluid which blocks the pancreatic duct. This leads to the leakage of digestive enzymes into other parts of the pancreas, causing injury, inflammation, swelling, pain and subsequently scarring.

Clinical presentations of CF-related pancreatitis

Acute pancreatitis commonly present in late adolescence or early adulthood. A classic symptom is severe abdominal pain which might be associated with nausea

and vomiting. Pancreatitis can be the first presenting symptom in patients with non-classical CF.

Diagnosis

Diagnosis is confirmed on blood test which shows raised levels of pancreatic enzymes (e.g. amylase and lipase). Radiological imaging, such as US or CT of the abdomen, is sometimes required for further assessment.

Management of pancreatitis

Recognition of pancreatitis symptoms and early presentation to medical assessment is the key to facilitating the effective management of acute pancreatitis. Management is supportive which generally include gut rest, intravenous fluid and pain relief. Potential precipitating factors such as alcohol, medications and dietary factors will need to be carefully addressed. Further studies are required to assess if CFTR modulation therapies are effective in preventing CF-related pancreatitis.

Long term outcome

Recurrent pancreatitis causes progressive inflammation and destruction of the remaining pancreatic tissue. Therefore, recurrent pancreatitis is a risk factor for the development exocrine and endocrine pancreatic function in patients who are previously pancreatic sufficient. Close clinical monitoring is required to detect pancreatic insufficiency in patients with recurrent pancreatitis.

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Fundraising News

ONCE AGAIN THERE HAS BEEN AN AMAZING SHOW OF SUPPORT FOR CFWA, ESPECIALLY DURING THE MONTH OF MAY! THANK YOU FOR MAKING A REAL DIFFERENCE IN THE LIVES OF PEOPLE LIVING WITH CF! YOUR ONGOING SUPPORT MEANS A LOT TO US.

The money raised will continue to fund research initiatives and provide vital services. We couldn't do this without your help. Thank you to everyone who took part.

65 Roses at school

The school community in WA continues to amaze us by stepping up to the challenge raising funds for CF! Thank you to all the teachers, students and parents that participated.

Floreat Park raised $864.70, Endeavour Primary School also raised funds and reached a total of $388.45. Great work!

The schools in the town of Merredin got on board for 65 Roses with Merredin College raising $595.20 and St Marys Catholic College raising $650. A brilliant result all around!

Gairdner Primary School had a gold coin donation day, totalling $60.00 and Bullsbrook College raised $607.50. Thank you!

Wesley College raised $527.41, holding a sausage sizzle and gold coin donation day and Ellenbrook Christian College raised $796.00 through various fundraising activities. Such a great job!

Pickering Brook Primary School raised $205.90 with crazy hair, while Yanchep District High School with Allyssa Rowe heading up the team raised $335. Well done and thanks so much!

Emma Tander and the school community at St John Bosco College sold roses on 65 Roses Day and raised $830. Thank you to all involved!

Crack a Cure for CF!

If you have spent any length of time on social media in the last few months you have probably come across two darling sisters, Isobel and Ruby Donaldson 'Cracking a Cure for CF', doing their part to raise funds and awareness for cystic fibrosis.

The idea is Isobel and Ruby's brain child and involves cracking an egg any way you think you can, although most people have been cracking them on their head and uploading the photo or video on social media, nominating a friend to do the same and making a donation to CF.

The concept has been far reaching with participants Cracking a Cure from across the globe, including internationally recognised Professor Stephen Stick and Rugby legend Nathan Charles.

Well done Isobel and Ruby for coming up with such a creative and effective idea to raise funds and awareness. To date, the Crack a Cure challenge has raised $11,591.46 and is still rising.

If you have missed the challenge you can see it here **http://www.crackacure.com/** . Help save lives and send in your own challenge.

Community fundraisers do it again with the 65 Roses Challenge!

CFWA has had a team of faithful community fundraisers making their presence known in their local communities in the lead up to and during the month of May. Each one doing their own version of the 65 Roses Challenge. You may have spotted them selling merchandise, roses, cooking sausage sizzles, baking cakes, holding raffles and doing various other activities to raise funds to help improve the lives of people living with cystic fibrosis.

This year, the fundraising activities went from Broome to Esperance and Kalgoorlie, Northam, Albany and Margaret River. A great big thank you for all the hard work, hours given and tireless support from our exceptional fundraisers.

Thanks so much for your support, every dollar raised makes a real difference. We always love to hear the stories about your fundraising adventures! Here are just a few.

Kate and Paul Spaapen cooked up an amazing $2,598 by hosting four different dinner parties during the month of May. Thanks so much!

Kate Hillard set up her 65 Roses Challenge raising a fabulous, $2,810 and then also went on to raise an additional $580 selling roses. Thanks Kate!

Ghupie Venter and the Just Breathe team held a health and wellbeing day with face painting, sausage sizzle, community stalls and exercise classes, raising $1,258.97. Thanks so much!

Joel and Elysia Lawrence and the team from the Margaret River Masters Football Club had a night of fun and fundraising, raising a sizeable $4,000! Great effort and thanks so much!

Maggie and Katie Di Re held a quiz night for CF, raising $922 and then went on to raise and additional $644.55 selling roses. Thanks again!

Shari Douglas, Nicole Forgione and Mathew O'Brien embarked on a challenge to be remembered by signing up to do the Kokoda Trek in October. Already they have raised $1,606.60 and the figure is still rising. Good luck guys on the challenge and thanks so much for raising funds for CF!

Sally Edwards organised a fundraising challenge at her gym raising, $109.65 and Nena Johnston and her workmates raised $700 having afternoon tea. Great job and thanks so much!

Brooke Murphy held a Superhero Dance Party and got the town of Manjimup dancing the night away, raising $4,435.35! A fabulous result!

Cindy Watterson-Bolst and the team at Café Boranup held a coffee fundraiser, raising $200. Thank you!

Alex Inkster had another very busy May month participating the HBF Run for a Reason and then selling cupcakes and other goodies to reach an impressive fundraising total of $1,636.80

The Community Recycling & Collection Service raised $121.26 by recycling clothing and Michael Wood from MTM Physiotherapy raised $310 by organising football parking. What great ideas! Also Jump About Trampoline Park raised $63. Thanks so much.

Paul Fowler is another member of the CF community who has taken on a rather large challenge. Between August and October, Paul plans to ride a bike across Europe, from the Atlantic in France to the Black Sea in Romania. He is doing this to try and raise awareness and money that will go towards research for new therapies and, ultimately, a cure for cystic fibrosis.

Paul was diagnosed with cystic fibrosis at 16 months, and it is generally not something he tells people about but increasingly he had the desire to do something for the cystic fibrosis community.

Paul has set himself a sizeable goal and so far has raised a remarkable $7,587.20, with funds continuing to come in. You can follow Paul's journey by going online **https://65roseswa2016.everydayhero.com/au/paulmichael**

Good luck Paul! And thank you for doing this challenge for the CF community.

Not to be overlooked are all of the hardworking fundraisers who worked heartily to raise funds for CF on 65 Roses Day, Friday 27 May 2016! We couldn't do the work we do without you! Thank you for your commitment, your effort and the time that it took to raise funds.

Thanks to Joesphine D'Alessandro $165, the team at Panizza Dental Trust $100, Cathy North $1,380, Edith White $120, Andrea Halliday $230, Elisha Mason $875, Mary Allingame $500, Community Newspaper Group $392, Feliciano Sanchez $302.50, Hayley Gillard $1,027.90, CBH Group $527.75, Val York $826.25, Donnelle Oakley $1270.50, Tammie Rafferty $1334.10, Nick Joannides$300, Price Waterhouse Coopers $250, Lisa Padua $1475, Sally Edwards $715, Herbet Smith Freehills $720, Cherie Scrivener $1,174.30, Suncorp $130, RachuelKnapinski $1,252.75, Liz Balding $239.60, Lyn Murphy $250, Fabienne Brown $1,895, Trident Broking $250, Simone Blennerhassett $100, Anne De Jagger $125

Here are their individual fundraising results:

Krystal Burton	**$2,686.50**
Elizabeth Lewis	**$2,300.00**
Adelaide Withers	**$2,167.87**
Jacquie Carter	**$1,592.00**
Angela Sims	**$1,458.00**
Zoe Docherty	**$1,354.75**
Louise O'hare	**$983.90**
Tiff Miller	**$824.98**
Natalie Aylmore	**$822.31**
Olivia D'arcy	**$608.75**
Lois Edwards	**$534.00**
Hayley Cooper	**$526.25**
Jaclyn & Haydn	**$525.00**
Kerry Bell	**$417.50**
Bethan Smillie	**$392.00**
Sophie Edelman	**$333.00**
Arnie Van Biljon	**$265.50**
Melanie Denby	**$249.50**
Alessandro Luculanoa	**$188.00**
Jacqui Robertson	**$165.75**
Pri Adilbert	**$105.00**
Perry Cunningham	**$105.00**
Jamie Greenleaf	**$105.00**
Kris Carvell	**$94.50**
Jordan Lindley	**$73.50**
Karla McKenzie	**$21.00**

and HHG Legal $250.

Loose change sums it up!

Have you ever wondered what happens to your loose change after you pop into the donation tin?

Well thanks to the following businesses, the total money from loose change since the last edition of RED came to a total sum of $1,602.80!

Many thanks to the following businesses, Scarborough 7 Day Chemist, John Hughes Hyundai, Glengarry Sports Medicine, Beacon Lighting, Harmony Dental, Parkland Mazda, Cash City, Bayswater Night & Day Guardian Pharmacy, The Angove Street Collective, Cannington Motorcycles, Walter Road Pharmacy, Electric Tattoo, Fraser Motor Cycles, Jenny Craig Morley, Ambassador Tavern, Chemart Pharmacy, Lotus Indian Groceries, Riverton Summerfield Bakery, Jetts Fitness Bassendean, Chan Brothers, Dankz Furniture, Prime Products, Nola Café, Gioivanni's Hair Studio, Xpresso Code, Parkland Mazda, Payless Shoes Victoria Park, Price Attack Victoria Park, Mount Claremont Pharmacy, McKenzies Pharmacy, Newspower, Gosnells Pharmacy, The Good Guys Rockinham and Corfield Doctors Surgery.

HBF Run For a Reason

After months of training and preparation the HBF runners took to the streets to run to raise awareness and funds for CF.

Thank you to all the runners for your time and effort spent training and running and the voice you gave to our cause, your sweat, calories and miles. And of course, thank you for the $18,899.56 funds you raised!

Thanks so much to all the fundraisers and donors who made a contribution to the grand total!

Unblocking DIOS

DIOS (DISTAL INTESTINAL OBSTRUCTION SYNDROME) IS A COMMON COMPLICATION OF CF WHICH AFFECTS APPROXIMATELY 8 - 16% OF ADULTS AND CHILDREN.

The causes of DIOS

Inflammation and infection affects the output of enzymes in the gut which may change the environment of the gut and make it more acidic and also have thicker gut mucus which can slow the time for bowel contents to pass through the system.

Risk factors for DIOS

History of previous DIOS, pancreatic insufficiency, dehydration, post-transplant, severe genotype, failure to pass the first stool at birth (meconium ileus) and gut surgery.

How common is it in Australia?

In the Australian CF population the incidence has been reported to be 14-16% in adults and 7-8% in children. Reportedly in those who have had an episode of DIOS it is likely to reoccur in 77% of cases.

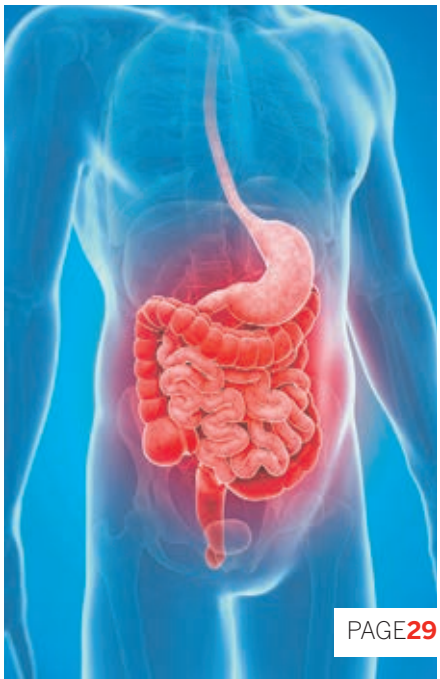
Does climate influence the occurrence of DIOS?

A study of people with CF in Brisbane and Sydney looked at when DIOS occurred and found the outside temperatures were higher in the week before the patients presented in emergency with DIOS (24°C adults & 27°C children).

Prevention of DIOS

- Regular use of polyethylene glycol based laxatives (Movicol, Golytely) osmotic laxatives that draw water into the large bowel
- Stay well hydrated, include salt supplements in the diet; pear juice is naturally high in sorbitol (a natural osmotic laxative) and may be useful
- The priority is enzyme control in relation to fat intake. Some doctors prescribe medications to control gut acidity, however. This is controversial; some also use probiotics
- Elite athletes should weigh themselves pre & post exercise to estimate fluid loss and rehydrate accordingly
- Develop an awareness of signs of dehydration: dry lips, reduced urine output, fatigue, nausea
- Educate the health professionals you come across regarding your history i.e. previous episodes of DIOS, meconium ileus

Reference: 2015 Matson, A& Ooi K. Unclogging DIOS. 11th Australasian Cystic Fibrosis Conference.



Outreach

Albany and Denmark 11-12 May

The CFWA services team travelled to Albany and Denmark in May to provide professional education to 10 nurses at Albany Hospital, support to a number of families and an education session for daycare staff. We also held a social dinner at the Earl and Spencer for families from Albany and surrounding areas.

Mandurah 23 June

Our Mandurah coffee morning was held in June and provided an opportunity for CFWA staff to catch up with local members over coffee and cake. A family support visit was also provided in the area.

Bunbury 23 June

It was great to connect with a few new and older members at the Water's Edge Café Bar & Restaurant. Our physiotherapist also had the chance to touch base with a young man to discuss physio. We are available on country trips to provide education sessions for day cares, schools, workplaces, extended families, hospital staff or community groups. Or if you would like a one to one session with the physiotherapist, community nurse or social worker/counsellor, please let us know.

CFWA Golf Classic PhD Top Up Scholarship

CLARA MOK, RECENT CFWA GOLF CLASSIC PHD TOP UP SCHOLARSHIP RECIPIENT, DISCUSSES HER PROPOSED RESEARCH PROJECT

Beginning in infancy, structural lung abnormalities are often the first sign of respiratory disease, and are the primary cause of morbidity and mortality in cystic fibrosis (CF). Chest computed tomography (CT) scans are now widely recognised as playing a critical role in monitoring the earliest changes in the lung seen in young children with CF, thanks largely to the efforts of the researchers, clinicians, patients and their families involved with AREST-CF. CT air trapping is the most common structural abnormality in early CF and appears as geographic areas of decreased density on expiratory CT scans. It is strongly associated with overall respiratory health and therefore, has the potential to be used as part of an important outcome measure in CF clinical management or clinical trials aiming to alleviate small airways disease. Despite its importance in CF, the trapped air appearance on CT has not been well characterised:what causes its appearance and how it progresses over time in relation to clinical markers of CF lung disease remains poorly understood.

Although air trapping and its progression can be assessed by CT, the requirement of ionising radiation (albeit in very small doses) limits the frequency at which it can be performed. Hence, there is a need to develop a safe, reliable and non-invasive tool to assess the extent and severity of lung disease that is appropriate for children with CF. Unlike CT, magnetic resonance imaging(MRI) does not have any known adverse biological effects (i.e. radiation-free) and therefore is suitable for serial monitoring of lung disease progression. A further potential advantage of MRI is the ability to assess the functional properties of the lung including: how evenly the lung ventilates and how effectively blood flows through different parts of the lung. This information is important because it will allow us to determine which pathology best reflects the overall extent (i.e. the cause) of trapped air on CT. However, functional MRI in the lungs has not been thoroughly investigated in paediatric CF and its sensitivity to mild disease is not yet known. Therefore, further validation of functional MRI in early CF is warranted.

The aim of my PhD is to characterisein detail the trapped air appearance on CT in early CF. Specifically, we aim to further validate functional MRI in the lungs and use this information to determine the cause of CT trapped air. We also aim to map the progression of CT air trapping in relation to clinical markers of CF lung disease. Ultimately, a better understanding of CT trapped air may lead to its use as a novel therapeutic target in early CF lung disease. Further validation of functional MRI in the lungs will provide greater insight into the mechanisms underlying early CF lung disease progression and allow us to individualise therapy based on the functional performance of the lung.

Save the Dates: 2016 Events

CYSTIC FIBROSIS WA'S 2016 CALENDAR IS JAM-PACKED, SO MAKE SURE YOU MARK YOUR DIARIES NOW. THERE ARE EVENTS FOR EVERYONE, WHETHER YOU ARE LOOKING FOR OPPORTUNITIES FOR FURTHER EDUCATION, VOLUNTEERING, FUNDRAISING OR TO COME ALONG AND MEET OTHER MEMBERS AND ENJOY SOME RESPITE

August

1 Starting School with CF Information Seminar
27 Red Tie Dinner Dance

September

9 Coffee Morning Bunbury
13-23 Deloitte Spring Rose Art Exhibition

October

6 and 7 Sibling & Offspring Camp
14 Parents' Dinner
21 CFWA Expo: Fertility and CF
23 Halloween Run: a Great Strides Event

November

5 Convicts for a Cause
6 Post-Transplant Support Day
25 Christmas and Awards Party

More information about each event will be made available closer to the time. Please note: dates may be subject to change.

Halloween Fun Run for Cystic Fibrosis

Sunday 23 October 2016
Perry Lakes Reserve Floreat

Save the date, more info coming soon...

A GREAT STRIDES event

Proudly supported by: Community newspaper group