Reversing the Vortex

Chris Kvam

HOW DO WE CREATE RESILIANCE IN OUR KIDS?

1. Positive self-image
2. Healthy goal setting
3. Exercise
4. Support – friends, family & pastoral care
5. Seeing the good

1. Positive self-image

When a CF kid looks in the mirror, what do they see?
Do they see a runner? A surfer?
Do they see a fantastic soccer player or a ballerina?

Do our kids see a child with CF or do they see an image of health and beauty? How we treat them, the language we use and the context we create (both in the clinic and in the home) all contribute enormously to how kids perceive themselves.

As a child moves through childhood towards adolescence and onto adulthood they are not a patient, they have become an expert on their own body and how CF affects it.

Doctors may be experts in CF
but parents are experts in their CF children

Likewise when we think about parents we must remember that the engaged parent of a child with a chronic disease soon becomes an expert in their child's CF.They notice little things others would miss. They know when a chest sounds different or breathing is more shallow or quicker or more tiring. They can tell when things are not quite right because they spend their lives with this child.

The way we treat parents also impacts whether parents see themselves as a true partner in the care of their child. The historical doctor-patient relationship assumes that it is the doctor who tells the patient what is wrong and what to do and they are expected to comply. In the care of a child with a

chronic disease, parents and kids collaborate with their team about their care. This is about listening and learning from everyone who is playing a role in the care of that child.

Give your child limited choices but hear their opinion…

Children should be given a say in their own care in an age appropriate way. They should be encouraged from an early age to participate and take responsibility in the decision making of their care but keep choices small and limited.

For example . . .

Would you like to do your physio when you first get up or after breakfast?

Would you like to get your enzymes yourself or would you like me to get them for you?

1b. How do we develop good self-image and expert-making?

We need to consider whether parents are addressing their needs when it comes to mental health, physical activity and nutritional needs. It comes as no surprise that children's health, reflects that of their parents.

The wellbeing of a child is inexplicably tied to both the wellbeing and ability of their parents to cope, physically and emotionally.

2. Healthy Goal Setting

As caregivers (both parents and care team) we can sometimes over focus on the *how* and forget to explore the *why*.

Why I should do my treatments matters more to the child than *how* I'm supposed to do them. We need to find things that matter to them.

There's a big difference between compliance and adherence.

Compliance is doing what others tell me to do. Adherence is doing the positive things that I know I should do and for kids, there's a big difference. Believe it or not for many CF people (especially teenagers), being a good CF patient is not motivating. Nor is being healthy or improving my life

motivating either because at that stage of life, our maturity dictates that we still don't fully comprehend or accept the consequences of our actions.

Passion drives adherence

What we need to do is find the causal link to passion. Adherence for the sake of adherence is not a sustainable goal. We need to find a passion that engages the child and link their treatment to this passion. Make adherence worth it.

Believe it or not, fear is not motivating. In fact, by doing the treatment those fears are not actually resolved. Health-related fears (or threats) continue after the treatment because we know that CF is a lifetime challenge. So we need to find a positive (short term) way to link them to a child's passion.

Lets figure out what's important to each child

If we can help a child figure out what's important, we can help them take the steps to move towards it. Remember in a child's eye, anything is possible and that's where sports can be great motivators because we can link them directly to treatments.

Adherence is a daily task so let's make goals that we're working towards as we do the meds, the diet, get enough rest etc. And remember, it shouldn't take a life threatening disease to make us realize our time is limited. This perspective can do wonders for family too.

3. Exercise is critical

How can families create momentum around exercise as part of their daily lives? What would family life look like if exercise was a key focus of the whole family?

Idea starter > Create regular family activities (it could be a walk after dinner or a family run every Sunday

- Set regular family activities
- Join regular family fun runs (even if some members walk)
- Set fitness goals
- Each individual competes with his or her self
- Try new activities
- Make holidays active even if its only walking or hiking or swimming

The role of exercise is critically important for people with CF

We know that building fitness over time is hard. It's hard for everyone not just CF people. Everybody needs a plan – both the CF child and the family.

How can we get families re-centred around physically activity?

Physical fitness allows us to move, to live. What's more, when we're used to doing exercise we start to notice changes in our bodies. Maybe our knee feels a little tighter today or our lower back is starting to feel sore. One of the most important aspects of self-CF care (even for kids) is noticing the deviations from their normal state of health.

If they become more attune to how their body feels through exercise, they will also become more attune to changes in their body and thus, notice changes when their body starts to become unwell.

Regular exercise (and being mentally engaged in exercise) is important on many levels.

4. Support – Look in the right places

Good support from friends and family is crucial for CF kids (and CF adults) and their families.

Many CF families may look to social media for support. In times like these it can be tempting to surf facebook or chatrooms looking for others who have CF or are caring for children with CF. It's important to remember that healthy CF people are often underrepresented in social media. Probably because they are out there in life doing things! The landscape can often be one of helplessness and hopelessness.

The best thing we can do is encourage self-preservation by limiting the amount of social media exploring that we do (or being careful about the sites we go to).

5. Seeing the Good

Life is unfair. How we accept this reality and resolve it can determine how our lives turn out. Yes life is unfair but so what? This line of reasoning doesn't get us very far.

It's helpful to remember that everyone has suffering, it comes in all shapes and sizes. Living with CF does not necessarily mean a life of suffering, there will be challenges and difficulties, but there will also be joy and happiness and contentment.

Being able to see the good will determine whether this is true for you or not. Teach your children to see the good in situations and in people, this is a terrific trait and strength to have whether you have CF or not.

"My daily routine is not FIGHTING CF"

We need to be mindful about the sort of language we use when we speak about CF. If we talk of "fighting CF", what are we really saying? That our child's whole life is a fight? If they fight well, can they win? What if they get sick? Have they lost?

For a long-term chronic illness, this may not be particularly helpful. Even the idea of constantly "fighting " something can feel exhausting. We're not "fighting CF, we're living with it" . . as mindfully and peacefully as we can.

So let's remind ourselves of the 5 things that create resilience

1. Positive self-image
2. Healthy goal setting
3. Exercise
4. Support – friends, family & pastoral care
5. Seeing the good

Now let's re-phrase them to be more actively positive

1. I am not a sick person
2. Anything is possible
3. Exercise is critical
4. CF is a team sport
5. A life with CF is a path to passion and empathy. We have much to be thankful for.

Physical Fitness | Being Fully Alive

Scott Russell, PT, DPT, CGS, GCS,

Scott Russell tells us (and we know this to be true as adults) that the psychology of it all matters when it comes to physical fitness.

The headspace you're in (when it comes to physical exercise) matters.

Being physically fit also helps a CF person to recover after an exacerbation. Not just because your body is fit to start off with (which also helps) but because you're used to working hard. You can relate to the feeling of pushing your body, of getting fit again and you don't shy away from the hard work required to get there.

Pulmonary function does not predict physical fitness

Aerobic fitness is made up of:

1. Pulmonary function
2. Muscle mass
3. Mechanical ventilation

Each contributes around a third to overall physical fitness so its important to remember that even if FEV1 is down, its not the primary determinant, only one factor.

If a CF person is struggling with the mechanical ventilation (finding it hard to breath out deeply), pursed lip breathing can sometimes help.

The key factors impacting fitness are:

1. Ventilation mechanics
2. Malnutrition
3. Chronic inflammation
4. Muscle strength / mass
5. Heart & Lung health
6. Vascular adaption to inflammation

Exercise also decreases the inflammatory state

It's not just that exercise is good for your fitness and overall health, it can also improve the immune response and decrease inflammation in the body.

High intensity training is a terrific way to increase fitness

Circuit training and other high intensity interval training can be a really great way to increase overall fitness regardless of your starting fitness level. Even with end-stage lung disease, research suggests that high intensity interval training can be a great way to train as CF people can work out at high intensity for short periods of time and rest at a lower level of intensity.

What about muscle mass?

There can be a muscle mass difference with CF people but not necessarily a strength difference.

How can we maintain good bone mass with CF?

Bone health is important and needs bone strain to develop. This means doing some kind of bone impact activity (could be jumping or running) and it needs to be higher than typical daily activities (some kind of novel impact like running or resistance training or gymnastics or jumping).

Its worthwhile remembering that puberty represents the peak accrual period for bone density and is therefore the optimum time to engage in exercises for bone health.

What about resistance training for kids?

Recent studies suggest that weight bearing exercise does help with bone density and does affect pulmonary health.

Some specific pediatric studies that relate to resistance training in children are:

1. Pediatric Resistance Training, 2014 , A Journal on Pediatrics, Neonatology, Adolescent Medicine,
   Child and Adolescent Psychiatry, Moro T. [1], Bianco A. [2], Faigenbaum A. D. [3], Paoli A. [1]

2. Strength Training for Children and Adolescents: Benefits and Risks, Davide Barbieri and Luciana Zaccagni , University of Ferrara,

Department of Biomedical and Specialty Surgical Sciences, Ferrara, Italy

3. Benefits of combining inspiratory muscle with 'whole muscle' training in children with cystic fibrosis: a randomised controlled trial. Br J Sports Med. 2014 Oct;48(20):1513-7. doi: 10.1136/bjsports-2012-091892. Epub 2013 May 16

How can we know how hard to push ourselves? (or our kids)

If you suspect you are de-saturating, get your care team to measure your oxygen levels, work out the heart rate level at which you begin to de-saturate and keep your exercise within healthy limits.

Temporary de-saturation (during periods of high intensity exercise) for periods < 20 seconds are potentially okay, as long as your body has time to recover and rest during a lower intensity phase after that.

The CF Therapeutic Pipeline | Ushering in a great era of hope

Dr Margaret Rosenfeld, MD, MPH
Professor of Pediatrics, UW School of Medicine
Pediatric Pulmonologist, Seattle Children's Hospital

Let's remind ourselves of the overall CF process:

We start with a CFTR defective gene
Which makes our bodies defective in ion transport (salt)
Which means the depletion of surface liquid
Which makes it difficult to clear mucus from the body
Which results in mucus obstruction
Which leads to an ongoing cycle of inflammation and infection

It's also useful to clarify the various stages of clinical research trials and what they mean as we consider new therapies in the pipeline:

Lab development:
Drug testing generally starts with animals and the focus here is on examining whether toxicity presents itself during animal testing.

Phase # 1 :
This is generally 10-20 patients and it is the first study in humans to determine dosage and whether there are any side effects that will prevent the drug from moving ahead.

Phase # 2:
This can be anywhere from 20 to several hundred patients and here safety is the primary concern to be explored. There is some evaluation of effectiveness but it is not definitive.

Phase # 3:
This is where randomized placebo controlled trials are conducted. It can involve anywhere from several hundred to a thousand in a sample size and is the definitive study as to whether the drug is effective or not.

Phase # 4:
This stage is focused around safety monitoring over the long haul.

It's also helpful to remind ourselves of the two major *types* of drugs that are used to combat CF:

A. Correctors – which work to address the CFTR protein to the cell's surface eg. Lumacaftor

B. Potentiators – which increase the activity of the defective CTR at the cell surface to improve the transport of chloride. eg. Ivacaftor / Kalydeco

A much more detailed explanation can be found here:
http://www.cftr.info/about-cf/clinical-management-of-cf/cftr-modulators/potentiators-correctors-and-production-correctors/

Kalydeco [Ivacaftor] was the first major [potentiator] drug to come to market and addressed the defect for those with the G551D "gating mutation".

In this mutation the protein cell is present on the cell surface but the ion transport is reduced. In this case, Kalydeco led to sustained improvements in pulmonary function and nutritional measures - increased weight gain, decreased pseudomonas cultures and increased lung function approx. 10%.

In CF patients the small intestine can also be overly acidic. Ivacaftor also works to restore the CFTR function which also normalizes the small bowel PH. Ivacaftor also reduced the rate of fecal elastase.

Orkambi is the most important treatment that has come onto the market in the US for DF508 thus far.

ORKAMBI is a combination of lumacaftor, which is designed to increase the amount of mature protein at the cell surface by targeting the processing and trafficking defect of the *F508del* CFTR protein, and ivacaftor, which is designed to enhance the function of the CFTR protein once it reaches the cell surface. ORKAMBI is taken every 12 hours - once in the morning and once in the evening.

Initial Orkambi results showed that it showed a sustained increase in FEV1 scores by 3% and had an effect on decreasing hospitalisations and the need for IV antibiotics.

Worsening of liver function, including hepatic encephalopathy, in patients with advanced liver disease has been reported in some patients with CF while receiving ORKAMBI. Cataracts have also been cited as a potential risk.

Respiratory events (e.g., chest discomfort, shortness of breath, and chest tightness) were observed more commonly in patients during initiation of ORKAMBI compared to those who received placebo.

It is important to note that correction is a multi step process and may require more than one drug to adequately perform the process.

There is another drug currently being trialled – VX661 which is also a corrector [like Lumacaftor].

Initial research shows that this drug has less drug interractions than Lumacaftor and less chest tightness.

Preliminary results suggest an increase in lung function of between 5% and 7% and a decrease of up to 7% in chloride sweat output. However Phase # 3 clinical trials are unlikely to be starting for a few years.

Dr Rosenfeld talked about a future combination being explored which would be made up of Ivacaftor , lumacaftor and a second corrector drug for DF508.

At this stage it has only been trialled on a sample of 7 patients but potentially may be another drug to come on to the market in years to come.

It's important to understand that none of these drugs is a 'cure' for DF508 but the good news is that there is a lot of work being done on this mutation and there is likely to be more results in the coming years.

The Birds and The Bees | CF and Reproductive Health

Anna Tsang, MSN, NP, CDE,
Nurse Co-ordinator / Practitioner St Michael's Hospital
Ontario, Canada

Sexual health should form a routine part of CF care.

In malesl

The CFTR protein causes poor formation of the male reproductive structures, essentially a developmental defect which can result in:

- Infertility [because of the bilateral absence of vas deferens]
- Delayed hormes / puberty [normal testosterone but delayed puberty]
- Smaller volumes of ejaculation

It is crucial to understand that infertility does not mean impotence, but rather it relates to "Azoospermia" which means that there is no sperm found in the ejaculation fluid.

So in short, CF men are able to have children but the challenge is that their sperm will not be ejaculated normally, but rather needs some help reaching its target.

Katherine Fraymen [Royal Melbourne Children's Hospital] is an expert in this area and will be speaking at the 2015 Australasian CF Conference.

What's the best way to tackle the issue of sexual and reproductive health?

12-14 yrs	Now is a good time to have an initial conversation especially as it relates to delayed puberty. It's important to emphasise here that puberty is not arrested, it is simply delayed by 12-24 months.

14-15yrs	Males should be aware of the infertility issues by this age. Body image can become negative around this age in general [and heightened by medical issues] so this is a crucial time to talk about these challenges.
17-20yrs	This is generally when Anna would offer a semen analysis if the male patient was interested and discuss in more detail, what this means for future reproductive health.

Peer support [and open discussion with parents] is crucial throughout this stage. It is absolutely crucial to keep reinforcing that puberty will be delayed only temporarily and that boys will 'catch up' with their peers.

Good nutrition, exercise and muscle building activity will also support boys during this process by providing them with a stronger and more positive body image.

In females I

It is important from 12 years it is important to start talking to girls about sexual [personal] female health with a key focus around 14-15 yrs.

Potential urinary incontinence

Leakage may occur during activities such as coughing, sneezing and laughing. Cough is the major cause of leakage in people with cystic fibrosis. Risk of UI is greater at times of increased cough such as during a chest infection. The amount of leakage may vary from a few drops to emptying the full bladder.

Teaching girls how to strengthen their pelvic floor is a crucial part of sexual health and Ana recommends introducing this idea early on to avoid urinary incontinence.

Reducing caffeine [or excluding] also improves urinary continence and should be suggested.

Thrush is another challenge girls face

Ana recommends a strong probiotic during teen years as well as home made yoghurt and other fermented foods to increase good bacteria in the body.

When acute thrush persists, she recommends treatment with an anti-fungal pill and says "Girls shouldn't put up with it, it can be treated."

Female hormones also play a role in CF

There is evidence to suggest that hormones play a role in the health of CF girls during the teen years. There is thought to be a change in pulmonary status coinciding with increases in estrogen during puberty years.

A Swedish study measured the hormone structure during menstruation and concluded that it hormones may play a role. Further reading can be found here http://www.sciencedirect.com/science/article/pii/S0954611100908916

Sexual and personal health is always best delivered when it is:

- well planned
- organized
- developmentally appropriate

What's New in Infection Control | Prevention and Control

Lisa Saiman, MD, MPH
Professor of Pediatrics, Columbia University
Attending Physician & Hospital Epidemiologist New York Presbyterian Morgan Stanley Children's Hospital New York

Historically pseudomonas patients have generally cultured pseudomonas one to three times early on [intermittent cultures] and as they get older they are more likely to experience persistent cultures which eventually become mucoid.

MRSA

However in Lisa's experience, it is MRSA that is increasing in both incidence and prevalence. Whereas pseudomonas is decreasing in both incidence and prevalence.

Lisa's point of view is that patients' better young function at younger ages combined with the more aggressive treatments leading to successful eradication along with an increase in community acquisition of MRSA has led to this shifting landscape.

Historically MRSA used to be a healthcare infection, one that kids might acquired after a stay in hospital. More recently researchers have started to see an increase in community-onset MRSA [and a decrease in hospital acquired MRSA].

For those of you not familiar with MRSA, this refers to methicillin-resistant Staphylococcus aureus . A fairly nasty bacteria which can cause an infection on the skin and in the lungs. Although it is resistant to several common antibiotics, MRSA can be treated with some antibiotics, nose drops and other therapies. [CFF.org]

MRSA can spread from one person to another through casual contact or through contact with objects that have become covered with the bacteria. If MRSA is in the lungs, it can be spread in tiny drops of liquid when a person coughs, sneezes or laughs, or on objects that touch the mouth.

You an read more about MRSA here https://www.cff.org/Living-with-CF/Germs-and-Staying-Healthy/What-Are-Germs/Methicillin-Resistant-Staphylococcus-aureus-%28MRSA%29/

There is currently a study being done exploring the early eradication of MRSA via Vancomycin and also exploring topical skin treatment as the skin is an important reservoir of MRSA.

Pseudomonas

Lisa also talked about current standard practice pseudomonas in the US. At present, the current US treatment is twice a day for 28 days [inhaled Tobi] followed by more treatment if the patient continues to culture pseudomonas.

Current studies also suggest that combination inhaled Tobi and Azithromycin may also improve the rate of eradication.

Lisa's topline recommendation when it comes to pseudomonas is that all CF clinics have a standard response treatment when a patient cultures pseudomonas.

There is currently a research study underway [funded by CFF] exploring the feasibility of standardizing treatment for exacerbations [using existing therapies]. More info on the study STOP-OB-13 can be found here https://clinicaltrials.gov/ct2/show/NCT02109822

General infection control

Lisa also discussed hygiene and how we limit cross infection within clinic as well as limit infection outside of clinic.

Guidelines for limiting cross infection in clinic

The updated clinical infection control guidelines now state:

- all CF patients should wear a mask when attending clinic
- all nebs should be sterilized after each use
- lung function activity should be tightly controlled
    - mobile unit brought to the exam room at the start of the visit
    - In a negative pressure room or lab
    - In PFT lab with a HEPA filter
    - In PFT lab without a HEPA filter but allowing 30 mins to lapse between kids o

You can read more about the most recent infectious control recommendations here

https://www.cff.org/uploadedfiles/aboutCFFoundation/InfectionPreventionControlPolicy/Webcasts/How-to-Lower-the-Risk-of-Spreading-Germs-in-CF-Clinics-and-Hospitals.pdf