Brisbane CF Conference 2009
CFWA Services Presentation

Cystic Fibrosis

Overview

1. Pre-Conference Service Support Day
2. Lay Program
3. Physiotherapy Perspective
4. Nurse’s Perspective
5. Conclusion

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1. Preconference Service Support Day 28/8/09

The Services Teams from Queensland, Adelaide, NSW, WA and Victoria met to discuss service delivery.

CFWA has significant differences to its team structure, funding and service delivery:

- CFWA Home Care Workers contracted via the Health Department
- No other State receives Government funding at such significant levels as CFWA to specifically fund HCW services.
- NSW contract workers via HACC – problems with referral, continuity of care and appropriateness' of care.
- Victoria focuses on equipment rather than personal support. They receive substantial grants that pay for equipment as requested. New policy one piece of equipment each year.

1. Preconference Service Support Day 28/8/09

- Queensland have significant issues re supportive community services. They are unable to get HACC to work with their families
- South Australia only have 2.8 FTE. So only offer grants at this stage

Proposal: CFWA to do research paper outlining HCW Services, (including the history) families' satisfaction and hospital beds saved. To be presented as a model of care at the 2011 CF Conference in Melbourne.

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1. Preconference Service Support Day 28/8/09

- **CFWA Multi disciplinary team**
- CFWA are unique in that we have 4 distinct roles (Including HCW;s and Project Officer) within services plus marketing and administration.
- This has allowed us specific services at a high level eg home nursing, physio, education packages and social work. Distinct professions also smooth relationships with hospital services because we talk a common language and have something that they want eg HITH, boredom busters, equipment etc.

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- Hospital relationships were sited by many associations as problematic due in part to confidentiality agreements and historical issues. Some associations are NOT allowed to visit patients on wards.
- Most services only comprise of social work type roles.
- Qld attend clinics and give out chips etc and speak with parents
- at these clinics. They also have a take away program where they deliver a take away lunch once a week and a massage program where massages are given on the ward.
- Other than this they are not allowed on the ward and are not involved in multi disciplinary team meetings. The team advised that the relationship has been very changeable.
- People are automatically members of CFQ

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1. Preconference Service Support Day 28/8/09

- The CEO has wanted all people with CF to be contacted whether this is wanted or not as they don’t use membership forms.
- Victoria does not attend clinics and are not allowed on the hospital wards, however sites a reasonable relationship with the hospitals. Main role being in supplying equipment which they get a significant grant for.
- NSW is involved in clinics and is able to go on the wards. Relationships are reasonably positive.
- SA has no capacity.

1. Preconference Service Support Day 28/8/09

- **<u>Country clinics</u>**
- Qld has offices in Brisbane, Cairns & the Sunshine Coast. 800 people with CF. CF hospital teams are located in these places.
- NSW also has two branches – one in Sydney & one in Hunter Valley which also has a hospital CF team
- Victoria only association in Melbourne & CF hospital teams in Melbourne. No outreach clinics.
- These centres have similar outreach programs to CFWA.

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1. Preconference Service Support Day 28/8/09

- **Sibling Camp**
- All States no longer run camps, but have re programmed to run several sibling day outs in various locations. Events have included horse riding and surfing.
- Issues were that it attracted the same children each year 20-25 and many children were accessing no services. Through making it a day event in different locations more children are receiving services.
- Victoria ran no sibling events in 2008.

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- **Accommodation**
- Brisbane owns a house which they manage and use not only for country families but for events such as bereavement day.
- Victoria has a Compassionate support program which can pay for accommodation etc. eg for when country patients access transplant services.
- NSW has an accommodation program which can pay for 3 nights accommodation.

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- **Mum’s weekends**
- Not favoured by associations any more. Same participants, too high a cost.
- Associations have gone more for pamper/massage days. They advised of better turn out and access to more people.
- **Dads events**
- Difficulty sited at engaging dads. One association did a bush bash which apparently worked well. Another did a day at the football.
- **Physical Activity Subsidy**
- Qld has a subsidy of $150 per year for each individual with CF.
- SA supplies $500 for physiotherapy programs eg. exercise equipment

1. Preconference Service Support Day 28/8/09

- **Financial Assistance**
- Victoria has $300 per person a year to assistant with taxi vouchers, petrol etc.
- **Partners evenings & Grandparents Day**
- Victoria runs information/get togethers for partners & grandparents.
- **Recommendations:**
- National project eg. cookbook
- Regular contact re resources eg physiotherapy handbook (Victoria)
- Annual meetings of 2 days to work on National issues and build community services profile.
- Reasonable relationship with hospitals including confidentiality and liaison

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Other items

- CFV: have CF rose collection Jane Edmundson, have CF rose collection stand at the Better Homes and Gardens expo?

- Working with Techni Pro as larger cohort than separately?
- Bereavement support
- Transplant assistance program ($1000)

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2. Lay Program

The latest in Gene Therapy studies : Eric Alton

(Professor of Gene Therapy and Respiratory Medicine, National Heart and Lung Institute, Imperial College London)

- Has been involved in developing gene therapy for CF for 15 years and coordinates the UK CF Gene Therapy Consortium- 3 UK centres involving 80 clinicians and scientists.
- Gene therapy opposite of evolution. Involves putting a gene into a virus
- Gene therapy involves the addition of a healthy, working copy of the faulty gene into the appropriate cells of the body. So instead of treating the symptoms of the disease, like most conventional medicines, gene therapy has the potential to correct the underlying cause.

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2. Lay Program

Other problems eg. transporting the liposome?

- Rubber from cap, legal issues and agreements
- Run study in 200 patients for 2 years, long intensive program
- Gene therapy for CF has been tested in humans using both viruses and liposomes. Five of the liposome trials were undertaken by members of the UK CF Gene Therapy Consortium. These early studies were concerned mainly with safety issues.
- 12+ age group best because lungs stop growing
- Lipid approved by regulators, treated 1st of 3 patients

Possible Outcomes:

- Small or variable benefit- low dose prevention of treatment
- Most get better or stop getting worse
- No benefit
- They have proved the concept that some patients can get somewhat better

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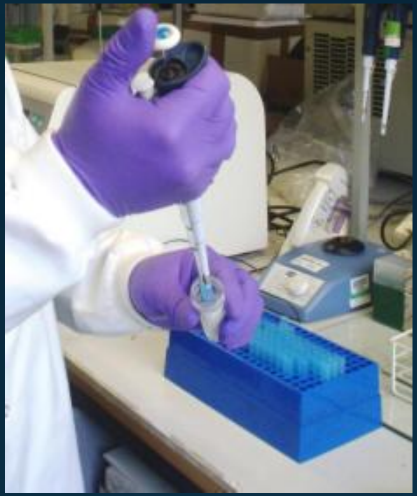
2. Lay Program

- Ready for Wave 2 of testing if Wave 1 doesn't work
- Sendai virus- able to put a gene into as virus can be repeated- Lenti virus
- Discovered that Wave 2 product 500 times better than Wave 1, tested on mice
- But Lenti virus has cousin can cause cancers
- Not ready for trials yet need safety study
- This is world's biggest trial for "getting better"
- Fighting against evolution and practical problems
- Expensive and need large team
- http://www.cfgenetherapy.org.uk/links/links1.2.htm



2. Lay Program

Clare Collins: Associate Professor in Nutrition and Dietetics

- Balance: everyone’s balance point is different
- Expectation or burden of treatment exceeds what is realistic
- Appetite and energy imbalance: extra work of breathing and fighting infections and loss of protein and fat via malabsorption
- Suggestion for adolescent at high school: No enzymes= no high fat food at school, eat it at home
- Female adolescent weight still lower than males
- School difficulties: Less time to eat, child’s weight can go down, by 12-17 even lower to 35%
- Challenges: Families let clinic be the “Bad Cop” and they be the “good cops”
- Young people have a goal- nutrition support does make a difference, vigilance will always be required to tackle the energy imbalance.

2. Lay Program

Case Study of CF patient with Borderline Personality Disorder

- Had been healthy in early years so was embarrassed and difficult transition to hospital, not used to being admitted
- Was very difficult with hospital staff, was taken off transplant list
- Developed hobbies in hospital , positive reinforcement,
- Trained ward staff in dealing with difficult patients,
- Boundary setting, appropriate behaviour, regular meetings with whole team

Case Study with CF patient waiting for lung transplant

Case Study of a person undiagnosed with CF

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2. Lay Program

Professor Alexandra Quittner

- Professor of psychology and paediatrics , University of Miami
- Research focuses on the measurement of adherence and health related quality of life in children and adolescents with chronic illnesses.
- Has developed a health-related quality of life measure for CF, that is used in international clinical trials
- Is working on a web enabled cell phone to promote adherence in teens with CF.
- Also doing a study on depression in children and adults with CF and parent caregivers.

2. Lay Program

- Conducted the largest survey on CF adults 865 in US asking questions about adult life. Average age 30, adult CFers, 95% Caucasian, 2% Hispanic
- From survey main point discovered that the CF adults are pursing typical adult activities such as working, living independently, getting married etc.
- Life expectancy in US 37.8
- ***Independent living very important step to developing as an adult- key to living unaffected by CF?***

Challenges for caregivers:

- Depression in relation to adherence, from study 30% of parents (mums mostly) depressed = worse adherence.
- High rates of depression in newly diagnosed- responsibility more on the mums, dads wanting to be more involved but mums not allowing it.

2. Lay Program

- Need more balance with partners in care giving for long haul
- International depression site: www.tides-cf.org/
- Funding in Germany etc for parents for counselling
- Adherence: families finding it hard to get all treatments done,
- Families need to understand why and how the treatments are helpful- lack of info and skills on administering the medication
- Lack of skills on how to manage daily regime
- Skill defects in the professionals giving advice and care
- Miscommunication between physicians and adolescents during clinic visits- don't always remember what was discussed and find the clinic stressful, difficult to admit "don't know something" and little time given to ask questions. = treatment plan

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2. Lay Program

- Noticed that if time spent with Dad doing treatment or if parent in the room, but not participating in the treatment, there was increased adherence
- Mobile phone : icare has alarms, for when to take medication, can text other adolescents, has chat site, live database and linked to pharmacy.

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2. Lay Program

Val Hall: nurse consultant at Belfast Regional CF centre

- Collaborative care vs traditional care
- Collaborative= self management, emotional management and patient defined problems
- copying clinic letters to CF patients, receiving test results via email
- Peer review of Clinic team- use report findings to get more funding etc

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2. Lay Program

Michael Boyle: Comparative study of CF Centres in North America

- Can add years of lives to patients by optimising delivery of treatments we already have
- 7 year gap of survival rate at top centres
- Common things to make a good performing centre: multidisciplinary team, well developed centre networks, ability to measure outcomes and passion for patients
- Measure, intervene remeasure

Practical steps to improve, this is what clinics with outstanding performance did:

- 1) maximise use of clinic time- preclinic meeting,
  preclinic form for patients to identify needs
- 2) review the basics at every visit- education of the basics personalised care plan stating what discussed eg
  airway clearance, nutrition, medications, diabetes etc

2. Lay Program

- 3) kind but not too nice, need blunt facts (top centres more aggressive with antibiotics – nutritional status as important as pulmonary status- aim to keep patient as normal as possible, prepared to fight for it!!
- 4) when in doubt bring patient back more frequently
- 5) involve patients in their care by regularly giving them feedback, show graphs of lung function and BMI- push for healthy patients not to be complacent
- 6) take advantage of available resources, help with projects or build on them
- www.portcf.org
- 7) teach good practices in children

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2. Lay Program

- 8) make it personal- identify where obstacle to adherence is eg embarrassment with peers, fear of affecting an up and coming event, engage family outside of clinic with Education days etc
- 9) It's all about the team- give chance for team member to shine, develop a passion for providing best possible care, family education care
- 10) Be determined to improve, pay attention to a specific area, get involved with quality improvement project, whole team

- No centre perfect at everything

To make change : focus on 3-5 details to improve outcome and pay attention to them for 2 years,

key areas to focus on

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3. Physiotherapy Perspective

Exercise and Physiotherapy

Validation of the 3 minute step test as a tool for predicting decline in 12 months. (Anne Holland)

- 15cm high step, external pacing at 30 steps per minute (metronome), continuous monitoring of oxygen saturation and heart rate
- Desaturation on 3MST is associated with decrease in lung function and increase in hospital admissions over the next 12 months
- Not useful in mild disease
- Relevance to us – quick, easy test with minimum space required, valid to use in the home for moderate to severe patients

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3. Physiotherapy Perspective

Balancing exercise and daily life for young people with CF – a trial of a novel exercise program (Allison Mandrusiak)

- RCH in Brisbane developed a FITKIT for home exercise – exercise in a bag which included instruction cards, exercise equipment and daily exercise log
- Positive outcomes were that patients maintained lung function, vs deterioration in the control group; some positive comments on the FITKIT
- Common theme amongst participants in intervention group was increased burden of care involved in completing the exercise log book
- Relevance to us – achieving a balance between exercise prescription and daily life is essential if a program is to be adhered to

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3. Physiotherapy Perspective

Effects of exercise on respiratory flow rates and mucus viscoelasticity in CF (Tiffany Dwyer)

- Exercise is proposed to increase mucus clearance; this research proposed to compare treadmill and cycle exercise (at 60% max) on airflow and mucus properties with a control group (resting breathing)
- Concluded that exercise improved ease of expectoration of mucus in adults with CF
- Relevance to us – promoting exercise in the home is very important to assist with airway clearance

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3. Physiotherapy Perspective

Physiotherapy treatment of infants with CF – a survey (Esta-Lee Tannenbaum)

- Aim to investigate the level of physiotherapy care which parents of infants undertake at home and their perceptions of competency with a 2 page, 12 question survey
- Burden of care physio places on families needs to be recognised as parents highlighted negative aspects of physio
- Physio does not always fit into family lifestyle, therefore physios need to assist families to find strategies that will ensure individualised and effective physio at home
- Relevance to us – we are part of the strategy which helps make physio more achievable at home

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3. Physiotherapy Perspective

Challenging Physiotherapy Practice

Do nebs take longer if done in alternate side lying? NO

- Now investigating if improved deposition of drugs using this positioning
- We are often involved in assisting with nebs at home

Using PEP in babies with CF and floppy airways

- Generally not introduced until school age
- Currently looking at earlier use in CF patients
- ?possible change of practice with young children at home

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3. Physiotherapy Perspective

Reflux during physio with head down tip in 1-3yos

- Very unpredictable
- Some increased reflux, some decreased and some stayed the same
- Importance of individualised treatments with no rules or recipes

Improving lung health in frequent flyers with intensive outpatient physio

- A group of 10 children who accounted for a high % of hospital admission days were given intensive outpatient physio for 12 months
- Hospital admission days decreased from 677days in 2007 to 349 days in 2008
- Also increased lung function and improved fitness
- Possible move towards more intensive outpatient based treatment to free hospital beds

3. Physiotherapy Perspective

HiTH in Qld – general discussion

- HiTH is proposed to decrease need for admission, risk of cross infection and burden on child and family
- Research shows families take in more information in the home
- Physio services are usually provided by a physio from the hospital team
- Charity funds are often relied on to fund service
- Issue remains of how to provide an equitable service – should the focus be on an "at risk" group or equally distributed among patients?

How many on HiTH at one time?

How do we sustain funding?

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3. Physiotherapy Perspective

Overwhelming take home message

- Physiotherapy accounts for the highest burden of care for families with CF and the lowest level of adherence to therapy
- Treatment must be individualised to reduce burden of care and maximise treatment effectiveness for each patient
- We must invest more time, effort and energy into uncovering the barriers to treatment for each individual and finding ways to overcome these

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4. Nurse's Perspective

Day one

Nurses session Sunday am

- Discussion on the Google Nurses Interest group.
- Will be used for Benchmarking discussion, Policy formation.
- Name and email left with presenter to enable contact in the near future, as it is invite only group and the group is hidden from search engine.

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4. Nurse’s Perspective

Keynote address:

- Prof Alton gave summary of progress after 20 yrs from discovery of the CF gene and our present understanding.
- There are five classes dependent on were the CTFR production is halted.
- Treatment is aimed at the genetic level, airway biology level, Airway Inflammation level, Airway infection level, Clinical care. So far genetic level has not contributed much to an effect on life expectancy.
- we have vx770 and vx809 to correct sodium absorption, at airway biology level.
- PTC124 maybe repair the gene in some cases depending the protein folding stop being side stepped.

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4. Nurse’s Perspective

Keynote 2 - Claire Collins - dietician

- Resting energy expenditure higher in type 1 - 3 classes
- Recommends - Enzymes start at lower doses, have high fat diets with omega 3 for less inflammation and higher energy consumption before satiation.
- Target 100 grams fat/day Easy to match with PERT and easy to calculate.

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4. Nurse’s Perspective

Post Lunch sessions:

- Peter Sly re collaborative efforts between Perth and Melbourne - gossip - Peter Sly going to Brisbane to set up program there.
- Early eradication of Pseudomonas Aeroginosa good outcome but increased risk of Stenotrophanomas and Aspergillosis - consider use of antifungal.
- Oral abstract presentations.
- SA trialling video phone use during hospital in the home treatment. Daily sessions, can show some problems over the video although still a little loss of detail.

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4. Nurse’s Perspective

- Small country hospital assisted in better care by having one contact person, set up direct admission protocol through A&E as person looks well but needs care. - Aid continuity of care and earlier presentation for treatment.
- Moriarty - Requirement for secrecy if child in vitreo had cf.
- Finlayson - Introduce the topic of Palliative care early - ie before transplant. It allowed for patient and family to elect for palliation earlier if the situation appeared to require palliation.

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4. Nurse's Perspective

Mon am

- Optimising health - a balancing act.
- Use of exercise PEP. Consider number of tasks to complete before living normal life. Out of hours exercise clinic - Wii fit popular in UK.
- Diet stimulants discussed - Males want to bulk up Females want to be less.
- Plenary 2
- Eric Alton - Gene therapy presentation - Wave one treatment with cleaned up liposomes to get genetic material into cells to bypass protein problem but only will last one month.
- Looking to use Wave two treatment with Lenti virus.

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4. Nurse’s Perspective

Case presentations

- One case involving Encephalopathy after binge eating at show on protein due to some liver disease.
- Standards of care and Peer review - allow accreditation of CF centre with recommendation of more staffing and resources.
- Tuesday Breakfast session - Data Registry. 10 yrs old this year.
- Will be able to compare to overseas centres - and other Australian centres. 2007 report launched. 2008 report due March 2010 and 2009 report due Oct 2010.
- Session on travelling with CF - considerations - Plan early, get there safely, play safe, recover when back home.
- Endocrinology sessions

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