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MEDIA RELEASE

Cystic Fibrosis Australia

Australian Researchers Take on Deadly Bacteria in the Fight Against Cystic Fibrosis

Two of Australia's leading cystic fibrosis clinicians from The Prince Charles Hospital and Mater Adult Hospital (both in Brisbane), announced two new research studies are underway aimed at improving the diagnosis and treatment of Pseudomonas bacteria - the leading cause of death among patients suffering cystic fibrosis.

Every four days a child is born with cystic fibrosis, a serious illness that mainly affects the lungs and digestive system. From diagnosis (often at birth) patients need constant medical care to survive including up to 5 hours of physiotherapy and up to 40 tablets daily.

For cystic fibrosis sufferers, a small cold can develop into a life threatening lung infection most commonly caused by the Pseudomonas bacteria. If the infection is severe, patients can undergo weeks of care in hospital to fight the bacteria that can lead to death.

These repeated chest infections damage the lungs meaning the older patients become, the harder it is to breathe. Without a cure for cystic fibrosis or the ability fight the Pseudomonas through accurately identifying the most appropriate antibiotic to treat it; in the long term, the only chance patients have of surviving is to have a lung transplant. Sadly, this is not always an option or a solution with many sufferers of cystic fibrosis not living into their teens and many not surviving past their 30th birthday.

Associate Professor Scott Bell, Director of the Adult Cystic Fibrosis Centre at The Prince Charles Hospital in Brisbane, is the chief investigator in the only national research study to tackle the Pseudomonas bacteria.

The study, the largest of its kind to be conducted in Australia involving all major cystic fibrosis clinical and research centres, is coordinated by The Prince Charles Hospital Brisbane, Royal Children's Hospital, Brisbane, Royal Prince Alfred Hospital, Sydney and the Royal Children's Hospital in Melbourne. This is a collaborative study conducted by a multidisciplinary team of researchers including cystic fibrosis clinicians, infection disease clinicians, microbiologists and scientists.

Although it is common in the environment (particularly near water and moist environs) it is rare to find the Pseudomonas infection in the majority of people. For cystic fibrosis patients, the Pseudomonas bacteria is the most common form of infection and is extremely difficult to treat. Even with powerful antibiotics the infection can and does result in death.

The study of the acquisition of Pseudomonas infection from environmental sources in young people with cystic fibrosis examines the bacteria, its natural environment and the environment of patients with cystic fibrosis. This includes environmental studies and molecular fingerprinting to make comparisons between patients and environments where Pseudomonas is present.

"By collecting and studying samples from all cystic fibrosis patients in the majority of cystic fibrosis centres, our aim is to gain a better understanding of Pseudomonas in cystic fibrosis patients so we can treat it more effectively and give patients a better quality of life through minimising infections," said Associate Professor Scott Bell.

Dr David Serisier Director Adult Cystic Fibrosis Unit at the Mater Adult Hospital, Brisbane, heads a multi disciplinary team taking on Pseudomonas bacteria by developing a new sputum sensitivity test, and comparing it with the existing method of culture and sensitivity testing to better identify the most appropriate and effective antibiotics to treat the infection.

"Currently, there is no definitive sputum culture test that accurately identifies the best antibiotics to treat this bug in the cystic fibrosis lung. We need to accurately determine the most effective antibiotic to kill the bacteria and our aim is to conduct a robust evaluation of the existing scientific sputum culture test enabling us to create a new method of testing that predicts the response to antibiotics more accurately in cystic fibrosis patients.

This means that patients with cystic fibrosis will be able to remain as well as possible enabling them to enjoy life away from hospital – this is our overall goal," Dr Serisier said.

65 ROSES DAY for CYSTIC FIBROSIS

“Please Help Us Find A Cure & Give Kids A Chance At Life”



Friday May 29 is 65 Roses Day - the annual fundraising and awareness campaign for Cystic Fibrosis in Australia to generate awareness of the illness, raise vital funds to help people living with cystic fibrosis and to fund critical research projects in the quest to find a cure.

Cystic Fibrosis Australia's CEO Terry Stewart urged all Australians to support the 65 Roses Day Appeal that will help to fund vital research projects like these.

“There are children and young adults in every state and territory of Australia who suffer this debilitating illness. There is no cure. Only with the help of the community, can we ensure every Australian living with cystic fibrosis can have the very best care until we can find the cure.”

Support 65 Roses Day by purchasing wristbands, pins, pens and other merchandise from participating LJ Hooker offices, Big W, Cheap as Chips and Ted's Camera Stores or by donating online at www.65rosesday.org.au or calling 1800 635 008.

Interview and photo opportunities include:

- Associate Professor Scott Bell, the Director of Adult Cystic Fibrosis Centre, Brisbane
- Dr David Serisier who heads the Adult Cystic Fibrosis Service at the Mater Adult Hospital, Brisbane
- Mr Terry Stewart – CEO – Cystic Fibrosis Australia
- Cystic fibrosis families from every region in all States and Territories across Australia
- CF State Organisation CEO's

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JOURNALIST NOTES

Medical Research Study by Associate Professor Scott Bell MBBS, MD, FRACP

“Clonal *Pseudomonas aeruginosa* infection in Australian patients with CF”

“Multilocus sequence typing of *Pseudomonas aeruginosa* from different niches”

- *the affects of Pseudomonas on cystic fibrosis patients*
 - *the clinical impact of Pseudomonas infection in cystic fibrosis patients*
 - *the spread of infection among and to cystic fibrosis patients*
 - *develop informed policies for infection control in the treatment of patients*
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Associate Professor Scott Bell MBBS, MD, FRACP

Thoracic Physician

Director, Thoracic Medicine

The Prince Charles Hospital

Rode Road, Chermside, Q, 4032, Australia

Background

Director of Adult Cystic Fibrosis Centre TPCH 1998 onwards

Director, Thoracic Medicine TPCH 2009 onwards

Co-Chair, Special Interest Group for CF, TSANZ, 2004-2008

Co-Chair, Centre Directors Group for CF, 2004-2008

Medical Advisor, CFQ 2000 onwards

- Associate Professor Scott Bell of the Prince Charles Hospital in Brisbane is a 2009 Australian Cystic Fibrosis Research Trust funding recipient for the project: “Multilocus sequence typing of *Pseudomonas aeruginosa* from different niches.”
- Associate Professor Scott Bell is the Chief Investigator
- The Research Team include: A/Prof. S. Bell, Mr T.J. Kidd, Prof. K. Grimwood, A/Prof. C.E. Wainwright, Prof. P.B. Rainey, Dr M. Hargreaves, A/Prof. M.D. Nissen.
- A/Professor Bell’s activities include:
 - Member of the 2009 Organising Committee for the Biennial Cystic Fibrosis Conference to be held in Brisbane in August/Sept 2009
 - Co-Chaired the Committee for the development of “The Standards of CF Care Guidelines”, released in 2008 for Australian CF Clinics and families.
 - Member of the Australian CF Data Registry Committee.
 - Active Member of the CF Centre Directors Meetings
 - CF Centre Director of the Adult CF Centre at Prince Charles Hospital

THE RESEARCH

“Clonal *Pseudomonas aeruginosa* infection in Australian patients with CF”

“Multilocus sequence typing of *Pseudomonas aeruginosa* from different niches”

Principle Areas of Study

- 1) A National research study to examine the most common bacteria infection and its effects on cystic fibrosis patients:
 - Studying all cystic fibrosis patients who are affected by *Pseudomonas* and its relatedness between patients, by collecting samples from all cystic fibrosis patients in the majority of cystic fibrosis centres throughout Australia.
- 2) Studies of the acquisition of *Pseudomonas* infection from environmental sources in young people with CF:
 - The study examines the bacteria, its natural environment and the environment of patients with CF.
 - This includes environmental studies and molecular fingerprinting to make comparisons between patients and environments where *Pseudomonas* is present.

Research Project – Key Elements

- Currently funded by multiple groups including NHMRC, CF Research Trust, TPCF Foundation, RCH Foundation, Rotary Australia
- This is the largest study surrounding CF patients to be conducted in Australia
- It is a collaborative study across all major CF clinical and research centres throughout Australia and coordinated by: The Prince Charles Hospital Brisbane, Royal Children’s Hospital, Brisbane, Royal Prince Alfred Hospital, Sydney, Royal Children’s Hospital, Melbourne.
- The research is conducted by a multidisciplinary team of researchers including CF clinicians, Infection disease clinicians, Microbiologists and Scientists.

Key Objectives of the Study

- To improve treatments for cystic fibrosis patients and give them a better quality of life through minimising infections
- To gain a better understanding of:
 - the affects of *Pseudomonas* on cystic fibrosis patients
 - the clinical impact of *Pseudomonas* infection in cystic fibrosis patients
 - the spread of infection among and to cystic fibrosis patients
 - develop informed policies for infection control in the treatment of patients

Background Information on *Pseudomonas*

- *Pseudomonas* is common in the environment (particularly in or near water and moist environs).
- Can cause infection in patients with compromised immune systems, following burns, skin infections and in various forms of lung disease.
- Lung infection of the *Pseudomonas* ^{bacteria} is the leading cause of death among patients suffering from cystic fibrosis.
- When established in people with CF is difficult to eradicate and requires powerful antibiotics to control.

Medical Research Study by Dr David Serisier MBBS, DM, FRACP

“Direct sputum sensitivity testing in cystic fibrosis – a prospective evaluation”

- to improve treatments and the quality of life of cystic fibrosis patients by developing a test that will enable clinicians to easily identify the best antibiotics to prescribe
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Dr David Serisier MBBS, DM, FRACP

Consultant Chest Physician

Director Physician Training

Director Adult Cystic Fibrosis Unit

Director Adult Respiratory Medicine

Mater Health Services

- Dr David Serisier heads the Adult Cystic Fibrosis Service at the Mater Adult Hospital, Brisbane
- He is the Director of The Department of Respiratory Medicine and Director of Physician Training at the Mater.
- He is a clinical Associate Professor in the School of Medicine at the University of Qld
- Dr Serisier has recently completed his term as a board member and Vice-President of Cystic Fibrosis Australia.
- He is also Honorary Treasurer of the Thoracic Society of Australia and New Zealand.
- After finishing Thoracic Medicine training in Brisbane, he spent several years undertaking postgraduate research training through the University of Southampton, UK.
- Dr Serisier has a number of clinical and research interests which include improving the assessment and antimicrobial management of *Pseudomonas aeruginosa* infection in cystic fibrosis lung disease.

The Adult Cystic Fibrosis Unit at the Mater Adult Hospital, Brisbane – Background

- The Adult Cystic Fibrosis Unit at the Mater Adult Hospital, Brisbane is a multi disciplinary team comprising dedicated chest physicians, a care manager/coordinator, physiotherapists, a dietician, a social worker and nursing staff
- The unit does not have dedicated government funding for these staff, who therefore commit to providing this service out of goodwill and a commitment to their group of cystic fibrosis patients
- The aim of the unit is to help cystic fibrosis patients to remain as well as possible enabling them to enjoy life away from hospital.

THE RESEARCH

“Direct sputum sensitivity testing in cystic fibrosis – a prospective evaluation”

Principle Areas of Study

- Robust evaluation of the existing scientific sputum culture test
- Identify how testing for Pseudomonas bacteria can be improved leading to more direct therapies that minimise infection

Key Points

- Pseudomonas is a principle cause of respiratory infection in cystic fibrosis patients
- Pseudomonas infection in the lung is rarely found in people without cystic fibrosis
- Why Pseudomonas has such an affinity with the cystic fibrosis lung is not well understood
- Over 80% of adult cystic fibrosis patients carry the Pseudomonas bug in their lungs continually
- Treatment of the Pseudomonas organism is difficult because it develops protective mechanisms to avoid being killed in the cystic fibrosis airway
- Furthermore, there is no definitive sputum culture test that accurately identifies the best antibiotics to treat this bug in the cystic fibrosis lung – antibiotic selection currently usually relies on a ‘best guess’ approach
- Current treatments include:
 - Regular inhaled antibiotic therapies to control infection
 - IV antibiotics given in hospital when patients become more unwell
 - at least two types of antibiotics are prescribed in attempts to kill the bug

Key Objectives of the Study

- To evaluate a new sputum sensitivity test and compare it with the existing method of culture and sensitivity testing
- This research will also assess whether the new method predicts response to antibiotics more accurately in cystic fibrosis patients with Pseudomonas chest infections
- The ultimate aim of this research is to improve treatments and the quality of life of cystic fibrosis patients by developing a test that will enable clinicians to easily identify the best antibiotics to prescribe

Background Information on Pseudomonas Bacteria

1. Lung infection of the Pseudomonas bacteria is the leading cause of death among patients suffering from cystic fibrosis.
2. How often would a cystic fibrosis patient have a Pseudomonas infection?
 - a. Once it develops it is usually a permanent infection within the lungs of CF patients
3. How often would they need to be hospitalised and for how long?
 - a. This varies a lot between patients; the average might be 3 to 4 times a year, usually for 10 to 14 days
4. What is Pseudomonas bacteria and its effects on cystic fibrosis patients?
 - a. Pseudomonas is a bacteria that is present ‘ubiquitously’ in the environment but only rarely causes significant infection in humans.
 - b. It is a bug that tends to cause infections only in certain situations, particularly (and most notably) within the lungs of patients with cystic fibrosis.
 - c. Once the infection becomes established in the CF lung, the infection is rarely eradicated and will usually become a lifelong infection.
 - d. Occasionally, other patients with chronic lung diseases will develop infection in the lungs with Pseudomonas, for example some patients with bronchiectasis and occasionally subjects with emphysema.

1. Background

65 Roses Day is Cystic Fibrosis Australia's (CFA) primary awareness campaign that is used to raise funds for people living with CF, and to finance critical research projects.

Cystic Fibrosis Australia raises money for the Cystic Fibrosis Research Trust as well as support services for young people who have CF and their families.

2. Why "65 Roses Day"?

"65 Roses" is a small child's effort to pronounce his sister's condition. As he couldn't pronounce "cystic fibrosis" he called the condition "65 Roses". The red rose is the symbol for cystic fibrosis.

3. When is 65 Roses Day?

Friday 29 May - 65 Roses Day - is the national fundraising day for Cystic Fibrosis Australia. However, we are aiming to gain media coverage from mid April to the end of May, commencing with the call for Champions.

4. How does CFA raise funds?

CFA raises funds through the sale of 65 Roses Day merchandise throughout the month of may include wristbands, pens and other merchandise. Outlets include: participating L.J Hooker offices, Big W and Cheap as Chips.

CFA also calls for Champions to volunteer to sell merchandise to family, friends, workmates and neighbours.

5. What is Cystic Fibrosis?

Cystic Fibrosis (CF) is the most common recessive genetic condition affecting Australian children and young adults today. It primarily affects the lungs and digestive system by clogging them with thick, sticky mucus. Repeated chest infections can cause irreversible damage and untimely death. Mucus can also cause problems in the pancreas preventing the release of enzymes needed for the digestion of food.

6. Who is affected?

CF is an inherited condition. For a child to be born with CF both parents must be genetic carriers for CF. They do not have to have CF themselves.

7. How common is CF?

Approximately one in every 2500 babies will be born with CF with one child born every four days. Two-thirds of infants born with CF will be diagnosed in the first year of life. In all, about 3,000 adults and children are living with this disorder. There are 1 million carriers of CF in Australia.

8. How is CF diagnosed?

In Australia, all babies are screened at birth for CF. A blood spot test is applied and if this test proves to be positive a sweat test will be done to measure the amount of salt in the sweat. It is with this test that a final diagnosis is made.

9. What are the symptoms of CF?

CF affects primarily the lungs and digestive system. People with CF may have the following symptoms:

- ❑ Persistent cough, particularly with physical effort
- ❑ Some difficulty in breathing or wheezing with effort
- ❑ Tiredness, lethargy or impaired exercise ability
- ❑ Frequent visits to the toilet
- ❑ Salt loss in hot weather which may produce weakness
- ❑ Poor appetite

10. Is CF contagious?

CF is not contagious. Coughing is a frequent symptom of CF. Some people are afraid they may catch it, but CF is an inherited condition which is present at birth. You cannot “catch” CF and you cannot give it to anyone else.

11. What is the life expectancy for someone with CF?

Until quite recently CF was a genetic mystery, and most people with the disease died before they reached their teen years. Due to improved research, the average age expectancy is now around the mid 30s.

12. What has been the impact of CF research?

Over the past 10 years researchers have made great progress in understanding the genetic basis of CF, and this has led to earlier detection. Research has also led to better knowledge and management of the condition, and extended life expectancy.

13. Is there a cure for CF?

There is no cure for Cystic Fibrosis. However, with today’s improved treatments most people with CF are able to lead reasonably normal and productive lives. A great amount of time, energy and money is being directed towards finding new and improved ways of treating CF and finally finding a cure. It is hoped that the emerging field of gene therapy may soon help to correct lung problems in people with CF.

14. What treatment is currently available?

While there is currently no cure, CF sufferers undertake intense daily chest physiotherapy to move mucus in the lungs. 40 enzyme replacement tablets are taken each day to aid digestion.

15. What is needed?

Money!!! Cystic Fibrosis Australia does not receive government funding for research. The money raised through 65 Roses Day products and donations fund research that will improve treatments and hopefully find a cure for this debilitating and life threatening disorder.