A partnership of care: An approach to the management of adults with cystic fibrosis

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Abstract

Several factors occurring more or less simultaneously have led to a major change in the management of adults with cystic fibrosis at The Prince Charles Hospital, Brisbane. Less than 50 years ago, 80 per cent of infants born with cystic fibrosis did not survive the first two years of life. In Australia today, the median survival is around 30 years of age, with many surviving into their fourth decade (Landau & Ryan 1991, p 4). Recent developments in the management and treatment of cystic fibrosis provide an exciting and promising foundation for further development and a changing focus in cystic fibrosis management.

Increased longevity has significant implications for the management of adults with cystic fibrosis. Until recently, most people with cystic fibrosis were managed within the paediatric setting, with the expectation that life expectancy would not exceed adolescence. In reality, there has been a dramatic increase in the number transferring from children's hospitals to adult facilities, and a subsequent need to provide optimal care to a group of patients coming to terms with their own perceptions of being adolescents with a life-shortening condition.

From an organisational perspective, the increased demand for resources has generated the need to consider alternative methods of providing care. Such options include home

intravenous therapy, day admission, alternative accommodation and education of staff in remote health centres. Home intravenous therapy has proven to be a popular alternative to hospital treatment and has significant implications for reducing lengths of stay and occupied bed-days.

Within the current focus on best practice, developing strategies for providing optimal health care in the hospital setting are paramount. A multidisciplinary team focus based on a 'partnership of care' philosophy underpinned by best practice principles describes the approach adopted by the cystic fibrosis team at The Prince Charles Hospital that has led to successful outcomes in meeting challenges inherent in the care of young adults with cystic fibrosis.

Introduction

Cystic fibrosis is the most common genetic disorder found in white Australian children and adolescents. One child in every 2500 live births is affected by the disorder and approximately 1 person in every 25 is a carrier of the cystic fibrosis gene. (Porteous & Dorin 1990, p 46). In Australia, the National Cystic Fibrosis Data Register has not been successful in providing accurate statistics to date, but this is being addressed.

Cystic fibrosis is an autosomal recessive disease. Autosomal means that the gene is carried on one of the 22 non-sex-determining chromosomes; the gene is carried on the long arm of chromosome seven. Recessive means that the effects of the gene defect, being homozygous, are only evident when present in two doses, that is, when the gene is inherited from both parents.

In cystic fibrosis there is a defective gene that prevents the normal formation of a protein called the Cystic Fibrosis Transmembrane Regulator (CFTR). Normal CFTR regulates a channel that moves chloride and water across the membrane of cells that are found in different parts of the body, particularly in the lungs and pancreas. When the CFTR is absent or not functioning properly, there is abnormal movement of chloride and water across the cell membrane in the mucous-secreting (exocrine) glands (Koch & Hoiby 1993).

The result is a lack of water (dehydration) in the airways and the digestive tract causing thick, sticky mucous. In the lungs, the abnormally thick mucous blocks the breathing passages (bronchi). When secretions do not move normally they become infected, resulting in chronic cough, shortness of breath, increased sputum production and chest infection. In the pancreas, the thick secretions block the flow of pancreatic digestive enzymes which are necessary for normal food digestion. When these essential enzymes cannot reach the intestine, fats and

protein are poorly digested and absorbed. This is manifested as bulky, foul-smelling fatty stools called steatorrhoea. In spite of a normal appetite, weight gain may be reduced (Chitravanshi 1989, p 2).

Although puberty is often delayed in cystic fibrosis patients, sexual maturation is normal. Fertility is believed to be reduced in females with cystic fibrosis due to increased viscosity of cervical mucous which may prevent sperm trekking along the female reproductive tract (Chitravanshi 1989, p 24). However, reduced fertility has not been evident amongst the population of young adult females with cystic fibrosis attending The Prince Charles Hospital. There have been five unplanned pregnancies within the last three years, that is, 8.5 per cent of the female cystic fibrosis population at The Prince Charles Hospital.

In 99 per cent of males with cystic fibrosis, congenital bilateral absence of the vas deferens results in infertility. In some cases, investigation into infertility in the general male population has led to the discovery that the patient has a mild, previously undetected, form of cystic fibrosis. Recent developments in this area include sperm retrieval by a technique known as microscopic epididymal sperm aspiration, used in conjunction with in vitro fertilisation (*International Association of Cystic Fibrosis Adults Newsletter* 1993, p 5).

Complications that are relatively common in cystic fibrosis patients include:

- pneumothorax
- bronchiectasis
- gastro-oesophageal reflux
- peptic ulceration
- diabetes mellitus.

The Prince Charles Hospital cystic fibrosis team has identified epilepsy as a factor in a small group of adolescent cystic fibrosis patients, with evidence of epilepsy in five cystic fibrosis patients. However, to the author's knowledge, no other documented evidence of this has been reported to date.

In recent years, improved treatment modalities, particularly antibiotic therapies, physiotherapy techniques and dietetic management, have led to an increased life span for people with cystic fibrosis. Ten years ago in Australia the average life span for a person with cystic fibrosis was 22 years; today the average life span has increased to approximately 30 years.

Where these patients/clients were often previously 'cared for' in the paediatric setting for their entire life span, today they are transferring to the adult hospital environment in increasing numbers. At The Prince Charles Hospital alone, there

are approximately 123 adults with cystic fibrosis. This increasing population has led to a major rethink about the goals of managing a large group with a chronic, life-shortening condition, who often have special needs related to adolescence and the impact of a fundamental change in their own perspectives on life. Management goals are centred around minimising the progression of the disease and methods include:

- antibiotic therapy
- physiotherapy/exercise
- optimising nutrition.

The first step in refocusing the care of adults with cystic fibrosis was to form a multidisciplinary team with a strong commitment to improving the quality of life for adults with cystic fibrosis. The team, led by a respiratory physician, comprises nursing, allied health and psychiatric staff, and community representation. Community representation is provided through a liaison coordinator from the Cystic Fibrosis Association. This has forged a strong and effective link between the hospital and the association. Integrating the psychiatrist into the team, in a highly visible role, has broadened the boundary of acceptance within the cystic fibrosis population and has had particular significance when dealing with issues such as transplantation or depressive illness that may occur with chronic illness.

The team meets weekly to discuss individual cases and provide a team approach to problem-solving. The format of the meeting is structured around four broad issues: medical, social, psychological and educational/employment matters. Every fourth week the meeting focuses on management and planning strategies to enhance quality of care. The group is well directed and interactive and has not experienced any adverse group dynamics. This is largely due to the strong leadership of the physician.

With the growing population, one of the early problems was facilitating a smooth transition for the increasing number of young adults being transferred from the paediatric setting to an adult facility. The Prince Charles Hospital, in collaboration with the Brisbane Royal Children's Hospital, has set up a structured approach to the transition process, as shown in Table 1.

Table 1: Transition process

Phase 1	Six-month meet with staff from Royal Children's Hospital
Phase 11	Meet potential patients at Royal Children's Hospital outpatient department
Phase III	'Checkout Charlies' – biannual information sessions at The Prince Charles Hospital, which include a tour of the hospital facilities.
Phase IV	First outpatient appointment at The Prince Charles Hospital.
Phase V	(under consideration) Twenty-four-hour admission for assessment. This may reduce some of the anxiety associated with a first admission during an exacerbation and would also expose the patient to the full cystic fibrosis team.

Checkout Charlies sessions are usually well attended and considered a great success in reducing some of the anxiety associated with transferring to an adult facility for the first time.

The multidisciplinary team is on hand to meet the young people with cystic fibrosis, answer any queries and provide information. The sessions include a tour of the Adult Cystic Fibrosis Unit facilities. An information booklet prepared by the team is distributed at these sessions and on first admission.

To assist in the transfer process, the Cystic Fibrosis Unit adopted a 'cute frog' (CF) theme. The idea behind this theme is that when young people transfer for the first time they are greeted with a cute frog of their own to look after. This is usually a small toy frog that adorns their bedside locker. The frog theme is carried throughout the unit, with ornamental frogs of all shapes and sizes decorating the area. The information booklet also carries the theme, pictorially and by almost subliminal messages that appear throughout the booklet. For example, statements such as 'going ahead in leaps and bounds' and 'keeping one jump ahead' accompany some of the information. The feedback on this theme approach has been very positive and encouraging.

When young adults with cystic fibrosis are admitted, their management is centred around a 'partnership of care' philosophy. This involves a measure of responsibility in the treatment of acute exacerbations and maintenance care. Decisions in treatment and management are discussed with the patient and are managed around both the health needs of the patient and the need to normalise their lifestyle. For example, some of the patients are able to continue part-time employment during their hospitalisation.

From a nursing perspective, the management goals are slightly broader, namely, to:

- optimise health – to facilitate a return to or improvement on previous state of wellness
- promote autonomy
- facilitate activities of daily living
- provide psychosocial support
- provide/promote health education.

As well as recognising the critical aspects of individual rights and patient autonomy, this approach endeavoured to overcome some of the early problems that the team experienced in encouraging the behaviour modification necessary to achieve optimal outcomes in health.

To provide a best practice framework on which to base our nursing care in order to achieve optimal outcomes, the nursing staff in the unit reorganised into four teams or focus groups:

- Quality Activity – producing process improvement
- Research – based on information and analysis
- Education – leading to empowerment
- Clinical Practice – guided by policies and procedures.

This was so that we could coordinate and critically evaluate our activities.

For example, the Quality Activity team develops audits and surveys and is involved in organising trials and evaluations of new products. The Research team is involved in various research studies and provides the unit with the latest journal articles, among other activities. The Education team is involved in Home IV therapy education in collaboration with the Home IV coordinator, organises weekly in-services, updates the patient information booklets, and so on. The Clinical Practice team has been involved in developing relevant policies, updates the procedure manual and provides the resource people for procedures such as care of venous access devices and peripherally-inserted central catheters.

The appraisal system (performance, planning and review) has been linked into our focus teams, with the expectation that each nursing staff member in the unit will develop at least one performance, planning and review objective that is directly related to their team.

When several young adults with cystic fibrosis are admitted simultaneously, a peer group culture often predominates, with the group sometimes forming into a 'pack' that, on occasion, can lead to adverse behaviour. For example, some of the initial problems were group decisions to avoid physiotherapy treatments and even to 'hit the town' for the evening. Open discussion, group counselling sessions and expectations that have been formalised into guidelines within the partnership of care philosophy have helped to overcome some of these initial problems.

With the increasing population has come an increased incidence of *Pseudomonas cepacia*, a particularly virulent organism that can have serious implications on health outcomes for this group of patients (Govan et al. 1992; Isles et al. 1992). Approximately 10 per cent of the adult cystic fibrosis population (n = 9) have had sputum cultures showing evidence of *Ps. cepacia*. The team has now recognised the need to 'stagger' routine admissions to reduce the chances of cross-infection. In addition, patients with *Ps. cepacia* are admitted to another area of the hospital where single room facilities are available. Separate outpatient clinics are now held for the population with *Ps. cepacia*.

Staggering admission dates for those requiring maintenance care, or 'tune-ups' as they are referred to within the cystic fibrosis population, has also reduced some of the problems that have been more often associated with adolescence than with chronic illness.

All the CF team members have experienced a sharp learning curve in the last few years, with several challenges being met. These challenges have included dealing with the death of young adults, fertility and pregnancy issues, non-compliance with treatment, and extremes of behaviour including drug and alcohol abuse. Another issue is lung transplantation which is a major decision and often a crisis point for young adults with cystic fibrosis. As these challenges have arisen, the team has developed strategies for dealing with each issue on an individual basis and with all the skills and experience available from within the broad scope of the multidisciplinary team. These strategies have included:

- having team conferences
- using primary nursing in appropriate situations
- encouraging family involvement (without handing control back)
- accommodating work commitments
- creating a supportive environment
- resourcing other health professionals, for example, family planning staff, genetic counsellors, psychologists and obstetricians.

In terms of successful outcomes, it has not been possible to collect objective data to date. There are too many confounding factors. For example, weight gain and improved respiratory function depend very much on patient compliance with treatment options. Our admission rate and length of stay will reduce as our home intravenous therapy program gains momentum, but is presently confounded by ever-increasing numbers.

Our success has been measured in terms of providing a supportive environment and seeing young people, who previously adopted risk-taking behaviour because they could see no future beyond their teens, become motivated to seek fulfilling and productive lives.

Given the diversity of psychosocial issues, not least the transfer to an adult facility that affects the management of young adults with cystic fibrosis, the multidisciplinary team approach has enabled a comprehensive plan of care to be established for each individual patient and has provided for a rapid response to crises and challenges. Underpinned by the partnership of care philosophy, the team approach has certainly 'gone ahead in leaps and bounds' over the last few years and expects to 'leap' ahead in the management of adults with cystic fibrosis.

In order to deliver quality health care, ongoing improvement is necessary. By using a team approach and seeking methods of optimising health care, adopting appropriate strategies to meet challenges and aiming to evaluate outcomes, the principles of best practice can be fulfilled.

References

Chitravanshi S 1989, 'An information manual on cystic fibrosis for patients and parents', Organon Australia.

Govan JRW & Nelson JW 1992, 'Microbiology of lung infection in cystic fibrosis', *British Medical Bulletin*, vol 48, no 4, pp 912–30.

International Association of Cystic Fibrosis Adults Newsletter 1993, September, Issue 34, p 5.

Isles A et al. 1992, '*Pseudomonas cepacia* – more than a harmless commensal?' *The Lancet*, vol 339, June 6.

Koch C & Hoiby N 1993, 'Pathogenesis of cystic fibrosis', *The Lancet,* April 24, vol 341, pp 1065–74.

Landau L & Ryan G 1991, 'Requirements for adequate level of care for those with cystic fibrosis', *Thoracic Society News,* September, p 4.

Porteous D & Dorin J 1990, 'Cloning the cystic fibrosis: Implications for diagnosis and treatment', *Thorax*, 45, pp 46–55.