

Respiratory Health Network

Cystic Fibrosis Model of Care

Prepared by the
Respiratory Health Network
Working Party
4 December 2007

Health Networks Branch
Working Together to Create a Healthy WA





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CYSTIC FIBROSIS MODEL OF CARE

EXECUTIVE SUMMARY

Cystic fibrosis (CF) is a hereditary disease that affects the lungs and the respiratory tract, digestive system, sweat glands, and male reproductive organs. CF affects children, adolescents and mature adults, with approximately ten babies per year diagnosed with CF in Western Australia. There is no cure for CF, however, symptoms of the disease can be treated. Proper care and treatment can greatly improve the lifespan and quality of life of a person with this condition.

This model of care was developed from the work undertaken by the Respiratory Health Network, Cystic Fibrosis Working Party, in consultation with key stakeholders groups, consumers and carers.

This model of care sets out an holistic patient-centred approach to the provision of services for people with Cystic Fibrosis (CF), with a focus on:

- transition issues across the continuum of care and disease progression
- an increase in consumer participation
- a shift from an acute hospital-based approach to a wellness approach
- services delivered where possible in community settings by a multi-disciplinary team.

The WA Cystic Fibrosis Collaborating Clinical Service (WACFCCS) will be established to support the delivery of the model of care for CF. It will provide a centre of excellence comprising tertiary paediatric and adult hospitals. The WACFCCS will focus upon:

- leadership and management systems for CF
- the processes and protocols required to deliver services across hospital and community providers

The WACFCCS will be a centralised service with inpatient services delivered within the existing arrangements at Princess Margaret Hospital for Children (PMH), Sir Charles Gairdner Hospital (SCGH) and lung transplantation at Royal Perth Hospital (RPH). It is envisaged that alternatives to inpatient services such as Hospital in the Home (HITH) and ambulatory care services will be expanded.

In the future adults will access inpatient services at SCGH, Fiona Stanley Hospital, (FSH) Joondalup Health Centre, (JHC) and in regional centres that have developed shared care models with the WACFCCS. There is also capacity for limited paediatric inpatient services at FSH.

The multi-disciplinary teams (MDT) will coordinate and where appropriate, deliver, community-based services.

The role of Cystic Fibrosis WA (CFWA) as an advocate and service provider is strengthened through the integrated service model and establishment of the WACFCCS.

Increased General Practitioner involvement in the delivery of primary care and active participation in the multi-disciplinary team will be promoted and supported by the WACFCCS, community service providers and General Practice organisations. The role of GPs will be integral to the delivery of integrated services and care management plans for people with CF in rural areas.

The increase in the adult population with CF due to improved longevity and survival has placed additional demand on services that cannot be met within the existing resources.



This is most evident in the current provision of adult CF services. The establishment of the WACFCCS and workforce strategies to address the additional resource requirements and training of clinical staff in CF are recommended. This includes the need for additional resources for the adult service in the immediate future.

The integrated multi-disciplinary approach to service delivery requires the development of an Information Communication Technology (ICT) database to store and manage patient data and care plans. This will ensure the MDT and consumers have access to accurate information to manage their health care. In addition, Telehealth will be promoted and utilised to support health service providers in rural areas and improve access to specialist support for people with CF residing in country areas.



1. METHODOLOGY

The process of developing this model of care has had a high level of involvement and participation by clinicians, key stakeholder groups, consumers and carers. The initial draft for the model of care for Cystic Fibrosis was developed by a small Working Party, made up of key stakeholders with expertise and knowledge of Cystic Fibrosis and a commitment to improving the quality and delivery of services across the continuum of care

The draft was then distributed widely to clinical staff, health professionals, health service providers, consumers and carers and the Respiratory Health Network Working Party and Advisory Group for input and comment.

Two community focus groups were also held for people with Cystic Fibrosis and their carers to contribute to the development of the model of care. The Senior Development Officer, Health Networks Branch and the Respiratory Health Network consumer representative facilitated these forums.

A workshop with key stakeholders, including consumers and carers, was then held on 3 August 2007. This workshop focused on three components of the model of care: transition; adult services; and community services. The workshop was also informed by the outcomes of the community forums held in July 2007.

The Respiratory Health Network Executive Advisory Group is accountable for the final model of care.



2. OVERVIEW OF CF IN WA

Definition

Cystic fibrosis is a genetically inherited condition that affects children, adolescents and mature adults. Whilst cystic fibrosis is a multi-system disorder, the major cause of morbidity and mortality is lung disease.

Prevalence information

In WA, the birth incidence of Cystic Fibrosis (CF) was 1:2152, based on newborn screening since 2000. This covers a range of definitions for CF, including a raised trypsin level, detection of gene mutations and an elevated sweat chloride (sweat test) but not taking into account those adults with late presenting symptoms. Overall, the Australian data demonstrate an incidence of CF in the community around 1:3001.

The textbook prevalence is usually around 1:2500, which is clearly dependent on the definition of CF (incorporating descriptors such as presence of elevated trypsin, detection of delta F508 mutation or other gene mutation, etc) and the ethnic origins of the population (for instance, in the United States, the incidence of cystic fibrosis is highest in Caucasians of northern European ancestry (1 in 2,000 live births) and lowest in African/African American (1 in 17,000 live births), Native Americans, and people of Asian ancestry). The disease occurs equally in both sexes (Source: Genomics Branch, Department of Health WA Population Data)

There are approximately 300 Western Australian families that include people with Cystic Fibrosis. Approximately 10 babies per year are diagnosed with CF in Western Australia through the newborn screening program (Figures based on data from PMH and the National Cystic Fibrosis Register).

Table 1. 2007 age distribution of CF paediatric patients in Western Australia (PMH data)

Age group	N (%)
0- 6 years	64 (39)
7- 12 years	41 (25)
13- 18 years	60 (36)

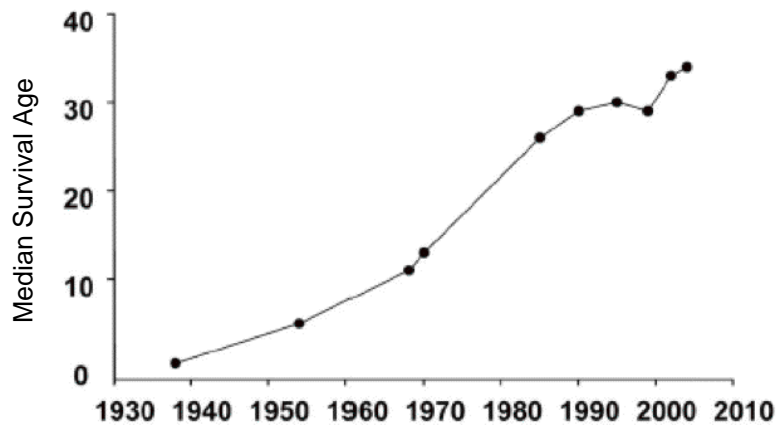
Table 2. Projected population expansion in the paediatric CF population in Western Australia 2007 - 2022 (PMH data)

Year	Number
2007	168
2012	185
2017	202
2022	223

It is estimated that the overall paediatric CF population will increase by 1.5 times in the next 15 years (excluding interstate/country transfers).



Graph 1. Median survival by year for patients with CF



The median survival for patients with CF has steadily increased since the 1930s despite the lack of any CF-specific therapies having been developed. Improvements have however, been made due to the development of centres of excellence in tertiary institutions, multidisciplinary care, antibiotics and improved nutrition. Therefore, with a shift in emphasis from tertiary to regional and community care the gains from centralised care must not be forfeited and are addressed in the proposed model of care for CF.

Table 3. Australian Cystic Fibrosis Data Registry -Australian data register statistics - 31 December 2004

	Male	Female	Total
0-4 years	186	173	359
5-9 years	183	180	363
10-14 years	208	194	402
15-19 years	198	162	360
20-24 years	160	130	290
25-29 years	95	95	190
30-39 years	151	103	254
40-49 years	42	34	76
50-59 years	10	2	12
60 plus	0	6	6
Total population	1233	1079	2312

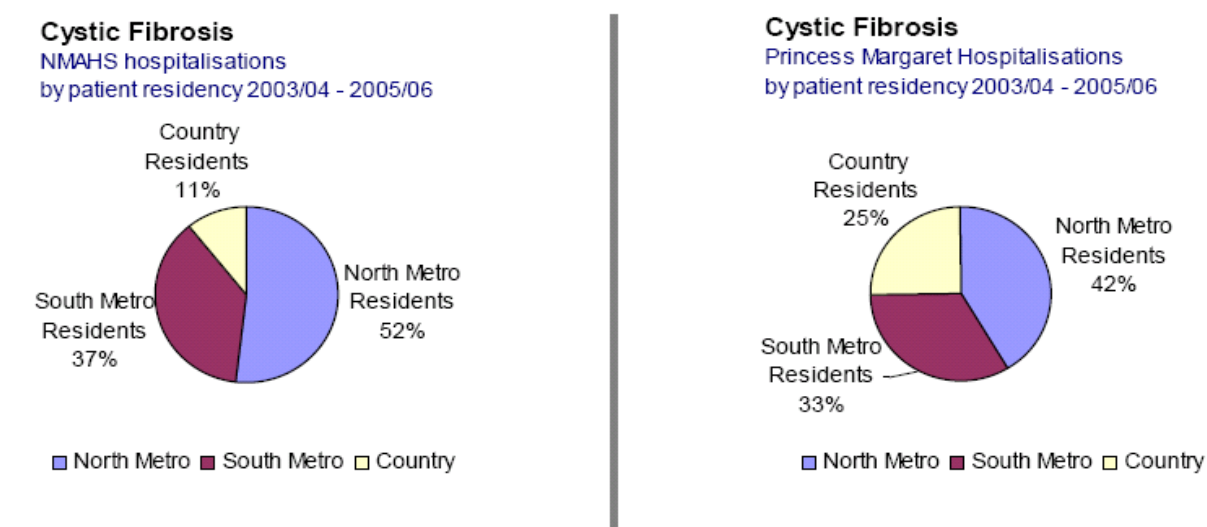


Current Hospitalisation Rates

The inflow/outflow pie chart shows that for the 3 years 2003-2006, adults with CF were admitted to the North Metropolitan Area Health Service (NMAHS). Sir Charles Gairdner Hospital (SCGH) is currently the adult tertiary centre. The graph below shows that 52% of hospitalisations were people who are resident in the north metropolitan area, 37% from the south metropolitan area and 11% from the country.

The inflow/outflow pie chart below on the right shows that for the 3 years 2003-2006, at Princess Margaret Hospital 42% of hospitalisations were children who are resident in the north metropolitan area, 33% from the south metropolitan area and 25% from the country.

Graph 2. Inflow Outflow Analysis by patient residency 2002/2006



Source: WA Morbidity Data System

Table 4. Hospital Separations with a Principal or Additional Diagnosis of Cystic Fibrosis 2003/04 - 2005/06

Place of Residence by Health Region	Area Health Service			
	North Metropolitan Area Health Service		Princess Margaret Hospital	
North Metropolitan	212	52%	248	42%
South Metropolitan	153	37%	201	33%
WA Country	44	11%	152	25%
Total	409	100%	601	100%

Source: WA Hospital Morbidity Data System



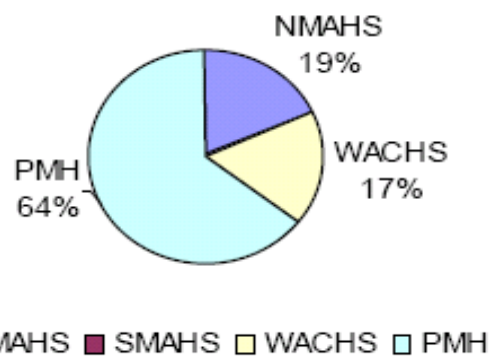
Hospitalisation of WA Country residents by Area Health Service

This graph shows the hospitalisation of country residents by area health service over 3 years, 2003-2006. Only 17% of current residents (the majority of whom are adults) are able to access hospital services in their place of residence. The remainder are either hospitalised at SCGH or in the case of children, at PMH.

Graph 3. Hospitalisation of WA Country residents by Area Health Service

Cystic Fibrosis

Hospitalisations of WA country residents
by Area Health Service/Hospital 2003/04 - 2005/06



Source: WA Hospital Morbidity Data System

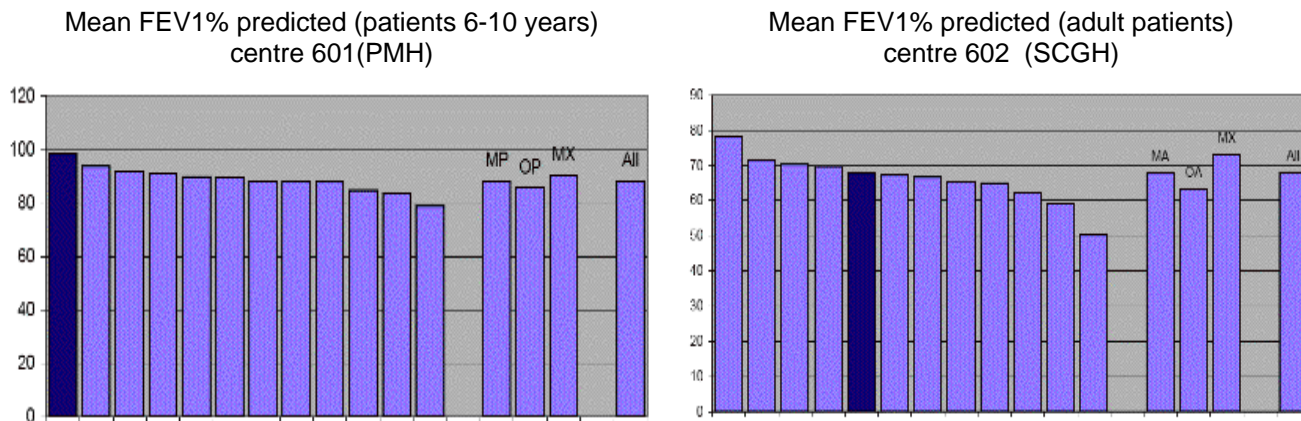
Table 5. Hospitalisation of WA Country residents by Area Health Service

Hospital Separations by Health Region	Place of residence WA Country Health Region	
	North Metropolitan Area Health Service	44
South Metropolitan Area Health Service	0	0
WA Country Health Service	40	17%
Princess Margaret Hospital	152	64%
Total hospital separations	236	100%

Source: WA Hospital Morbidity Data System



Graph 4. CF Paediatric Vs CF Adult Outcomes in Western Australia



Source Australian CF Data Registry 2007

Lung function is the single-most important outcome that determines morbidity and mortality in CF. Children in WA have the best outcomes nationally and internationally. On the other hand, outcomes in adults are below the optimal level of those of several other Australian centres although still very good by international standards. An obvious explanation for the difference in rank of the paediatric and adult outcomes is the chronic under-resourcing of transition/transfer and of adult services in WA. There is a clear disparity between the resources and services available to children with CF and their families as compared to adults with CF.



3. THE CURRENT SERVICE PROVISION

3.1 Newborns, Children's and Adolescents Service

The PMH Department of Respiratory Medicine has lead responsibility for paediatric CF care for all newborns and children. The model has evolved in the context of a multi-system disease requiring a life-long holistic approach to care, and recognition that the best health outcomes for children with CF are observed if care is conducted at a specialist centre with the resources and expertise to deliver a dedicated CF service. The quality of health of children and adolescents with CF is highly dependent on the expertise of specialist staff to ensure continuous and individualised care. The services provided by the PMH Respiratory Specialist Clinic is inclusive of inpatient and ambulatory care services, provision of all pharmaceutical supplies, community and domiciliary care. The PMH Department of Respiratory Medicine is responsible for the development and implementation of paediatric policies in Western Australia specific to respiratory disease.

There are currently 170 children with CF attending the PMH respiratory specialist clinic, with approximately 10 newly diagnosed children with CF each year. Adolescent patients aged 13-18 years constitute 36% of the total paediatric population. The number of indigenous patients is small, n=3, reflecting that CF is primarily a recessively inherited disease of Caucasian populations.

Since 2000, the WA Newborn Screening Program screens all newborns for CF according to current best practice. The screening for CF is by measurement of Immuno Reactive Trypsin (IRT) in blood from the Guthrie card followed by mutation analysis and sweat test. Cystic fibrosis is diagnosed in children born before 2000 that have not been screened at birth, by mutation analysis and/or sweat test.

From diagnosis, people with CF undergo daily treatment with an array of medications, physiotherapy and nutritional supplementation to maintain health. This has considerable impact on their own quality of life and that of their carers.

The family of newborns and children diagnosed with CF are contacted by the Respiratory Nurse Specialist to advise of the test results. A meeting with the CF consultant and respiratory health nurse is then arranged. This is followed by a 3 day hospital-based education program with the multi-disciplinary health care team (MDT) including the CF medical consultant, gastroenterologist, nurse, social worker, dietician and physiotherapist. The MDT is sourced from the respective departments within PMH. This ongoing assessment and treatment phase based at PMH continues on a weekly basis until the family and child are confident with the ongoing management and treatment. Visits to PMH are then reduced to fortnightly, then monthly. On average all children with CF are assessed and monitored on a quarterly basis at outpatient clinics held at PMH. The outpatient clinics have on average 900 attendances per annum.

Cystic Fibrosis WA (CFWA) is a member based non-government organisation that advocates on behalf of people with CF and provides a range of community and home based services including social work, nursing and physiotherapy. They also have support groups for people with CF, their families and carers. The CF service staff provides information about CFWA and encourage families to contact the service. CFWA is funded by WA Health for community-based nursing, physiotherapy and social work services.

CFWA services, when required, are coordinated and referred to by the PMH Respiratory Specialist Clinic. These services are only available in the metropolitan area.



Children and families in rural and remote areas have limited access to outreach clinics coordinated from the PMH Respiratory Specialist Clinic, however, they still have to attend clinics at PMH. Outreach clinics are currently held in Bunbury, Kalgoorlie, Port Hedland and Karratha.

The process for transition from adolescence to adulthood from PMH to SCGH begins during adolescence and the transfer occurs between the ages of 16 to 21 years. Despite recent changes, the transition and transfer process does not have a clearly articulated pathway. This is recognised as a limitation in the current service provision.

3.2 Adult Services

The Department of Respiratory Medicine at SCGH, provides the WA specialist adult service for approximately 130 young adults. The demand for this service has grown significantly in recent years due to the improved longevity and survival of people with CF. However, there has been no increase in health resources to support this growth. Despite the dedication of the staff the quality and level of service does not meet best practice.

A Respiratory Physician, CF nurse practitioner and physiotherapist provide outpatient clinics with additional support from junior staff and allied health professionals within the hospital. The service is significantly under resourced by benchmarks recommended by the draft Australian CF Guidelines 2007 and the Clinical Guidelines for Cystic Fibrosis care, Cystic Fibrosis Trust, the British Paediatric Association and the British Thoracic Society and the European Cystic Fibrosis Society.

Planned and acute admissions are made to the SCGH Respiratory ward. Staff and consumers have reported frequent delays in planned admissions, which occur due to bed pressure for “acute admissions”. This service is only provided at SCGH. Thirty seven percent (37%) of hospital separations from SCGH are for residents from the SMAHS region. Country residents are required to travel to Perth for treatment. There are no outreach clinics provided in country locations. Specialist CF community services are provided predominantly by CFWA.

While there are regular outpatient clinics and a programmed annual review of adults with CF, the infrastructure and support available limits the follow up of patients. There is a “drop in clinic” for patients with acute exacerbations and limited Hospital In The Home (HITH) service provided by CFWA for IV antibiotics. This service is only available in the metropolitan area.

‘We have had no help at all in trying to access reproductive and fertility services. It has been very stressful and we are still unsure about where to go and what is available. It just feels impossible at times.’ (Partner of woman with CF)

“CFWA need to explain better what is available - it is a bit wishy washy and hard to get. The hassle of figuring out what you are entitled to all seems to hard when you are feeling sick.” (Young adult with CF)



3.3 Transplantation Service

The Royal Perth Hospital, Lung Transplant Unit provides the transplantation service for all WA CF patients. At present there is limited access to microbiological pathology services in comparison to other transplant centres. This issue is beyond the scope of this model of care. The tertiary quaternary service to be provided at FSH includes transplantation services and the required support services such as microbiology.



4. PROPOSED MODEL OF CARE ACROSS THE LIFESPAN: NEWBORNS, CHILDREN AND ADOLESCENTS; ADULTS; TRANSPLANTATION; AND END OF LIFE/PALLIATION

4.1 Rationale (Quality of Care)

The development of the CF model of care included a review of the evidence-base for the clinical management and delivery of CF services, a gap analysis and community/stakeholder consultation. This information and knowledge has been used to inform the development of the model of care.

The model builds on the quality and capacity of the existing services and identifies and addresses areas for improvement. The new model of care can be implemented through a phased approach over time. In the initial phase the emphasis will be on the reorientation and reconfiguration of existing organisational structures and service delivery to be more patient centred, responsive and accessible.

Improved longevity and survival attributed to improved medical, pharmaceutical and therapeutic interventions has resulted in an increase in the adult CF population. This growing population impacts on the capacity of the current paediatric service to deliver adolescent and transition to adult services as the demand for adult services exceeds the current resources. The current organisational structure and specialist staffing levels cannot meet the demand resulting in variability in the quality of, and access to, outpatient and hospital-based services at SCGH.

The implementation of the model of care will result in evidence based best practice quality of care. To achieve this, additional human resources and improved access to community- based care and hospital facilities will be necessary.

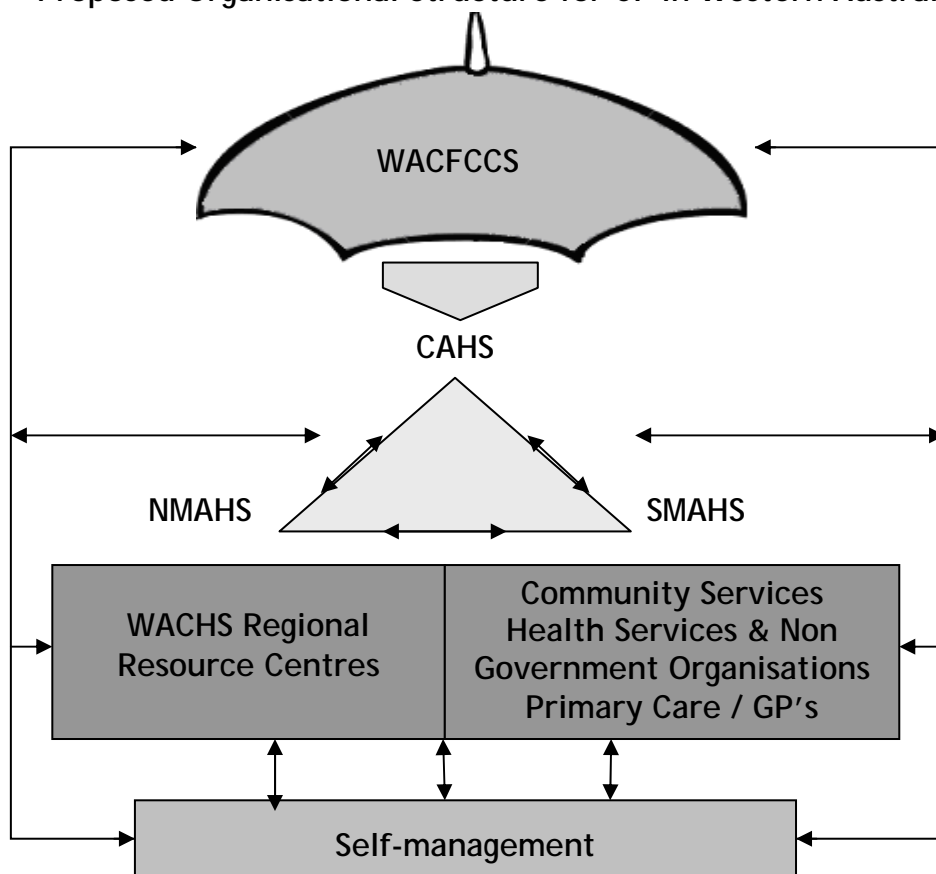
The leadership and organisational structure to support the WACFCCS proposes the appointment of a Director, dedicated staff, professional development of clinical staff in hospital and community settings, and referral pathways to relevant medical specialists and support staff.

The North and South Metropolitan Area Health Service planners have been consulted and informed of the needs of CF consumers in their planning for Fiona Stanley Hospital (FSH) and the general hospitals in the metropolitan area.

The model of care builds on current outreach to country areas and the development of shared care models in metropolitan locations and WA Country Health Service (WACHS) hospitals to improve access to services supported by appropriately trained staff.

This model of care will improve consumer involvement and participation in decision making and self management practice. Consumer views and opinion have informed the development of the model of care with an emphasis being placed on communication, integration and coordination across the continuum of care through reconfiguration and investment in community-based services.

Figure 1. Proposed Organisational Structure for CF in Western Australia



Legend

WACFCCS	WA Cystic Fibrosis Collaborating Clinical Service
CAHS	Children & Adolescent Health Service
NMAHS	North Metropolitan Area Health Service
SMAHS	South Metropolitan Area Health Service
WACHS	WA Country Health Service
GP	General Practitioner



4.2 Overview of model of care for CF

The CF model of care is a state-wide service with a centre of excellence comprising of tertiary paediatric and adult hospitals with appropriate management structures to support this model. This service will be known as the Western Australian Cystic Fibrosis Collaborating Clinical Service (WACFCCS). The model recognises the complex medical and psychosocial needs of patients and their families and aims to provide world-class, evidence-based care to patients of all ages and disease severity. The model emphasises the need for equity of access to care for all ages from birth and skilled management of transitions across the lifespan from paediatric to adult services through to end of life and palliation services.

The WACFCCS will co-ordinate skilled, holistic, patient focused inpatient and ambulatory care. Where possible there will be a shift in emphasis from hospital-based to a primary care focus, and ambulatory care services will be delivered in community settings and in people's homes. There will be a focus on transition across the developmental age groups linked to lifestyle factors and disease progression, which recognises and is sensitive to the physical and psychosocial challenges of daily life for people with CF.

The WACFCCS multidisciplinary team (MDT) is a group of health care professionals specialising in the care of children and adults with CF and their families/carers. The core MDT comprises of respiratory physicians, clinical nurse consultants, social workers, psychologists, dieticians, physiotherapists, hospital-based teachers, pharmacists and occupational therapists. This core MDT will also access other specialist medical and social support services as required, such as geneticists, General Practice and community physiotherapy. These services will be provided by a range of agencies including government, non-government and private agencies.

The model focuses on the consumer and their families as actively participating and involved in their care and quality of life. This consumer-focused service will include self-management programs, timely access to consumer education, and information about the services and options available across developmental life stages and disease progression. There will be shared care management plans with consumers partnering with health service providers in decision making processes that impact on the health and wellbeing of the person with CF and their families. The model of care provides CFWA with an opportunity to refine its role as an advocacy and service provider for the CF community.

4.2.1 Skilled Workforce

"It would be great if clinical staff in outer metropolitan and rural areas could be upskilled in CF issues such as the flushing of "ports". Currently there are no staff at the regional hospital, where I live, trained specifically to do this with CF patients. This would save on travel and time going to Perth." (Country CF patient)

The CF model of care will require a skilled and sustainable workforce that meets the service needs of the growing CF population. This will include:

- The WACFCCS to have a core group of skilled and expert clinical staff including medical specialists, allied health and support staff.
- Shared/joint appointments to ensure quality of care across all sites.
- Telehealth and support for WACHS staff and General Practice.
- Increased community-based allied health staff.



- Specialist community and ambulatory services within metropolitan and country regions.
- Education and training of health professionals in the delivery of shared CF health care in non-tertiary centres and the community.

4.2.2 Infrastructure and ICT

Central to improvements in CF care and delivery of community-based care is the development of a web-based CF database that can be accessed remotely (in the community in the metropolitan area or regionally). This database would hold clinical data (blood test results and spirometry etc.) that would be updated as results are made available. Issues of confidentiality and privacy will need to be paramount in the development of this system.

A web-based facility utilised by paediatric, adolescent and adult care providers in the hospitals, community and regionally will improve community-based care and transition to adult care. It is particularly suitable for CF consumers given the age group and their familiarity with IT. For instance much greater use could be made of automatic SMS generation to advise or remind of appointment times, meetings, medications and so on. This system would facilitate audit and development of KPIs.

Other communication strategies to facilitate communication across the continuum of care including discharge summaries and the development of integrated care plans will be developed in consultation with primary care providers and GPs.

4.2.3 Audit and Monitoring of CF Care

There is a need to develop and maintain the WA CF data registry so that it can be used to report State data to the National CF Registry. The proposed web based CF database (see above) supports regular audit and monitoring of health outcomes for people with CF.

4.2.4 Research

The PMH paediatric service has a strong track record in clinical and basic science research. The WACFCCS will maintain a culture of research as well as regular audit across paediatric and adult services. This will ensure not only that the best treatment is available for consumers, but it will also support the recruitment and retention of high quality, motivated staff.

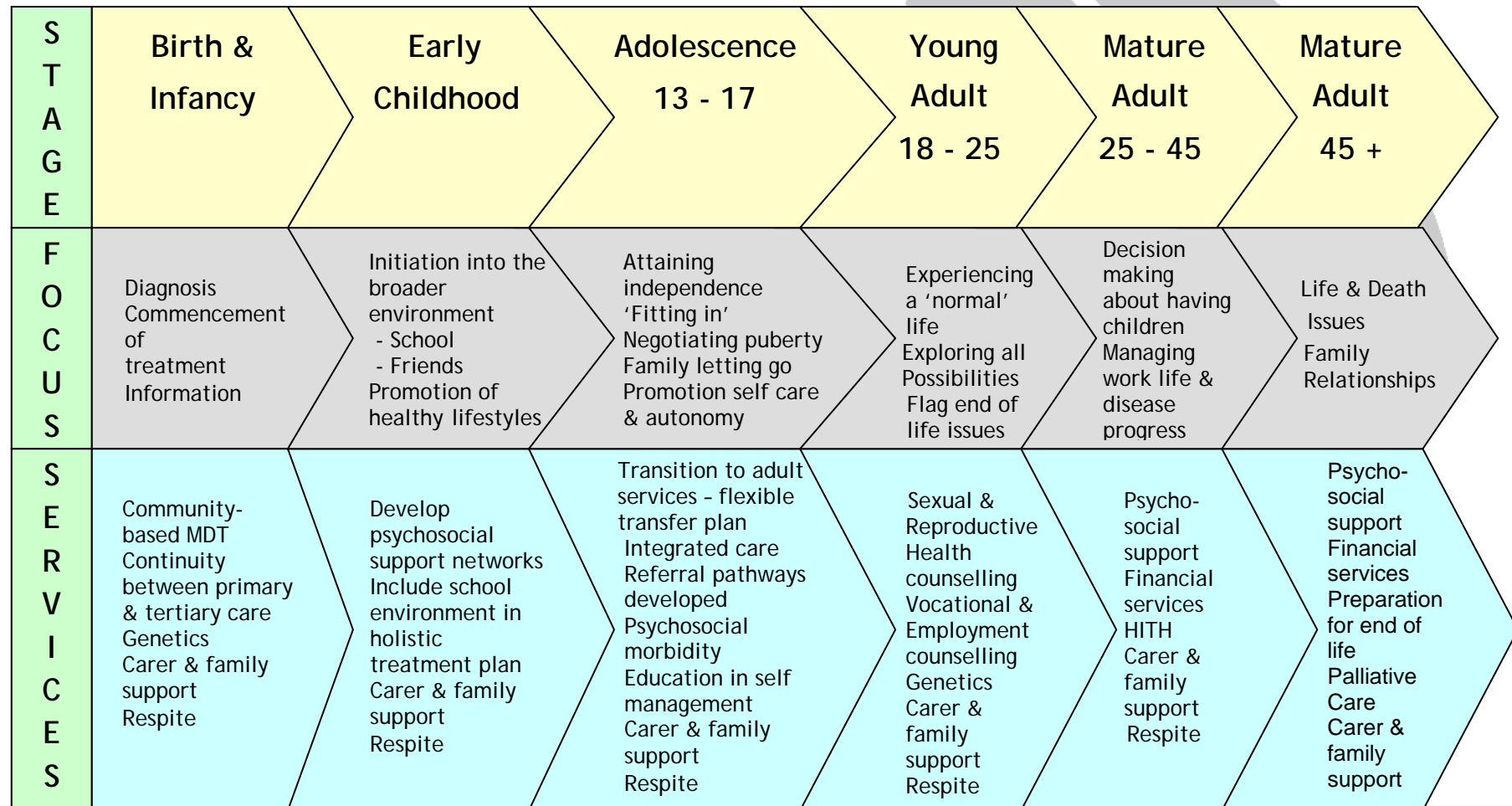
Clinical trials to improve the evidence base for the treatment and management of CF are limited as randomised control trials are difficult given the nature and progress of the disease and require the development of clinical trial networks nationally and internationally.

4.2.5 Patient Journey

The patient journey below is intended to highlight some of the key issues and service requirements across the developmental life stages and CF disease progression. The issues and services do not always fit neatly into age groups and the arrows are to represent the transitional times and flow through of care.



Figure 2. Consumer Journey





4.3 Newborns and Children

The Newborns and Children's service will retain the current best practice achieved through the newborn screening program and current guidelines and protocols. The enhancements and improvements to the service will be achieved through a consumer focussed service model with emphasis on better linkages and integration, communication, role delineation and resource allocation. There is also a greater focus on active participation and inclusion in the decision-making processes by consumers and their families.

The key components of the model of care for newborns and children are:

- Health Focus
- Community-based services
- ICT
- Shared Care
- Pharmacy

One of my biggest issues is having to go to Perth to collect medication at the hospital pharmacy - the travel time, waiting time, opening hours. It would make life a lot easier if smaller hospitals on the outskirts of Perth could dispense scripts from PMH/SCGH doctors." (Mother of a 12 year old)

Health focus - A wellness model of proactive care rather than an acute, reactive model.

Community-based services

Community and home based service delivery aimed at reducing the burden of care for families, the promotion of developmentally appropriate self-directed care, self management and minimising disruption to education and social functioning of the individual and their family.

Information Communication Technology (ICT) - Community care (home IV treatment, home physio treatment, and home health-checks) will be linked with hospital care via a web-based central CF database common to hospital sites and the community. Community nurses and physios will link with this database using laptops / Spirocards and wireless Internet access. Pathwest will automatically upload pathology results.

Shared Care - Shared care policies and protocols will be developed with non tertiary hospitals in outer metropolitan and regional areas and CFWA. This will result in care closer to home and minimise the burden of care on patients and families arising from travel and accommodation needs and social, educational and employment disruption.

Pharmacy - Implementation of a decentralised pharmacy service with prescription dispensing available at local community and hospital pharmacies. Where appropriate three month prescriptions should be routinely available and linked to consumer clinic reviews. In future electronic scripts generated by the web-based CF database will be used.



4.4 Transition From Paediatric/Adolescent Service to Adult Services

"It is hard to let go as they grow up, I know. The first time I was told that I could not attend the doctor's appointment with my daughter when she started going to "Charlies" I was really upset and couldn't understand why. I had always attended the appointments and she was happy for me to be there. There should be more information available. The idea of a formal transition process is a great idea. Maybe there should be one for parents." (Mother of young adult with CF)

There is increasing recognition in health literature of the importance of managing this process in a holistic and sensitive manner that meets the complex needs and demands of the consumer and their family/carers. The transition process from the CF paediatric/adolescent service to adult service is critical to the ongoing management and long term health outcome for people with CF.

This section defines adolescence and recommends a 3 stage process of:

- Transition
- Transfer
- Post-transfer young adult focussed care

Adolescence

Adolescence spans the period 12 - 24 years of age and may be defined as "the developmental period between childhood and adulthood - beginning with the changes associated with puberty and culminating in the acquisition of adult roles and responsibilities".

Adolescence may also be considered as "a developmental period during which young people must negotiate fundamental psychosocial tasks in their development towards maturity and independence". (*"Understanding adolescence"* www.caah.chw.edu.au/resources/gp-section1.)

Transition is defined as "the process or preparation for final transfer from paediatric to adult care systems; this transfer must be understood, ultimately, as success in aiding chronically ill children to start a productive life and achieve social integration as adults" (Schidlow 2002).

The WACFCCS team are trained and skilled in working with children and adolescence and have an understanding and knowledge of the transition process. The transition pathway is clearly articulated and consumers and families are aware, informed and included in the process.

A Planned Transition Program, initiated and implemented in early adolescence (estimated age 12 years based on available models) in partnership with patients and their family/carer will be developed for each patient comprising of the following components:

- Annual surveillance of adolescent development (MDT, adolescent specialist)
 - Promotion of self care and autonomy
 - Early detection of psychosocial morbidity secondary to the burden of chronic disease
 - Preparation for adult life ("transition check list")
- Education and vocational support (MDT, school teacher, community organisations as appropriate)
 - Promotion of healthy lifestyles



- Understanding of CF and navigating the health care system
- Maximising educational and vocational opportunities
- Counselling and psychosocial support (Social workers, psychologist, CNC)
 - Sexual health and fertility
 - Psychosocial morbidity
 - Facilitating access to health care and community support networks
- Development of a flexible transfer plan (MDT and community organisations as appropriate)
 - In partnership with the consumer, family/carer, MDT staff, community services,
 - CFWA and WACFCCS adult services staff.

Planned Transfer Process

A coordinated transfer process established between WACFCCS paediatric team and adult team, community services and CFWA as appropriate.

- Transfer planning meetings (twice-yearly) between WACFCCS paediatric and adult CF teams including community agencies as appropriate to develop a coordinated transfer and management plan. This will include:
 - Joint transition/transfer outpatient clinic
 - Introduction to adult CF MDT
 - Medical summary transfer
 - Detailed medical summary for adult services prior to transfer
 - Psychosocial support
 - Generic and CF specific
 - Consumer information/education and directory of services
 - Individually tailored adult mentoring and support services
 - Reproductive health

The adolescent/ young adult service (18 - 24 years) will be delivered at the WACFCCS adult service based at SCGH. The service will take into account the significant developmental and social factors facing young adults with a chronic illness. The key components of the ambulatory services to be delivered from youth friendly facilities with flexible booking and attendance times will be:

- **Surveillance and monitoring**
 - Autonomy and ability to self care -MDT
 - Understanding of personal health needs - MDT
 - Equitable access to health care resources - (CFWA, GP, social work, CNC)
 - Detection of psychosocial morbidity (GP, psychology, social work, respiratory physicians)
 - Promotion of healthy lifestyles (GP, CFWA, Quit, MDT)
 - Vocation and employment opportunities (Social worker, CFWA, Centre-link)
- **Sexual and reproductive health counselling**
 - Fertility counselling
 - Assisted reproduction technology



It is proposed that the services be delivered from youth friendly facilities in the hospital and community. Where clinics are required to be based at the hospital there will be a dedicated adolescent/young adult clinic with flexible booking and attendance times and an appropriate waiting area separate from adult acute areas. A dedicated young adults unit within the hospital adult acute respiratory ward is recommended, as are alternatives to hospital for treatment such as Hospital in the Home (HITH), medi hotels and community-based care.

4.5 Adult Services

The delivery of a quality driven best practice adult service will only be achieved through significant re-orientation and re-engineering of the current service model. This will be achieved through a consumer focussed service model with emphasis on better linkages and integration, communication, role delineation and resource allocation. There is also a recognised need for a greater focus on active participation and inclusion in the decision making processes by consumers and their families/carers. Additionally, there is a need for additional human resources to deliver the adult services.

Patient focussed services - emphasis on informed self management of care and active participation in decision making on all management and treatment plans by consumers and, where appropriate, families.

Access to service provision - the WACFCCS adult service will operate from SCGH with a satellite adult service to be provided in the South Metropolitan Area Health Service (eventually Fiona Stanley Hospital). There will be dedicated staff responsible for the provision of services to adults with CF supported by WACFCCS staff.

"There is too much red tape and delays when I need to get into hospital, like being advised to go through Emergency when the consultant has requested an admission.

My experience is there is a lack of understanding of many hospital staff about CF treatment. I hear staff comment that CF patients are coming in for a holiday and treat the hospital like a hotel. This is unfair as we are trying to maintain our health to prevent the illness progressing and keep our jobs at the same time. Trying to work all day and then crash when back in hospital does not mean we are lazy or do not deserve a bed." (Young adult with CF)

Protocols and clinical pathways - for the treatment and management will be developed and implemented based on best practice evidence based guidelines. These will build on current practice.

Referral pathways - for the clinical management of the multi-system disease associated with CF are developed and implemented. This includes but is not limited to endocrinology, gastroenterology and hepatology, psychiatry, rheumatology, reproductive health, lung transplantation and interventional radiology.

Improved access to community-based/ambulatory settings for care - alternatives to hospital such as self management or monitoring by health staff for treatment of acute exacerbations eg: HITH, medi-hotel style accommodation within the hospital setting. Integration with GPs and primary health providers will improve access to



general health care and where appropriate management of CF health issues, reducing the reliance on the tertiary outpatient service.

I don't like going to hospital, I would do anything to stay away. What I would go to is a community one stop shop "Tune Up Centre". (Young adult with CF)

4.6 Transplantation Services

The transplantation service will continue to be provided at Royal Perth Hospital with a move to FSH in the future.

Transition - to transplantation and liaison with the transplant team will be facilitated by joint appointments of the WA CF Centre staff and transplant unit staff where possible.

Continuity of care - WA CF Centre will promote continuity of care pre and post transplantation through referral pathways and protocols.

As stated previously, the tertiary quaternary service to be provided at FSH includes transplantation services and the provision of the required support services such as microbiology.

4.7 End of life care and palliative care

The declining health of people with CF should be managed in a holistic and sensitive manner to meet their complex needs and those of their families. An optimal end of life care model includes engagement whilst the person with CF is well.

Palliative care aims to improve the quality of life of patients and their families and includes the provision of services to meet medical, practical, emotional, spiritual and psychological needs of the person and their family.

Maddocks (2003) states "the benefits of palliative care are not limited to the final days and weeks of dying. Palliative care is relevant to managing symptoms in many clinical situations and can contribute to care in decisions early in the course of any eventually fatal illness".

In the case of CF it is recognised there can be tension between providing active treatment in the hope of transplantation versus maximum comfort for declining health. This can also raise conflicting expectations and demands for the person with CF, their families and the CF team.

Recent research "A Palliative Care Model for Progressive, Non-Malignant Disease: Cystic Fibrosis" undertaken by Wilson and Braithwaite 2007, presented at the 2007 National Palliative Care conference recommends five key components that should be addressed in the delivery of end of life and palliative care services for people with CF and their families. The five key components are:

- Information
- Communication
- Psychological Support
- Family Inclusion
- Timing



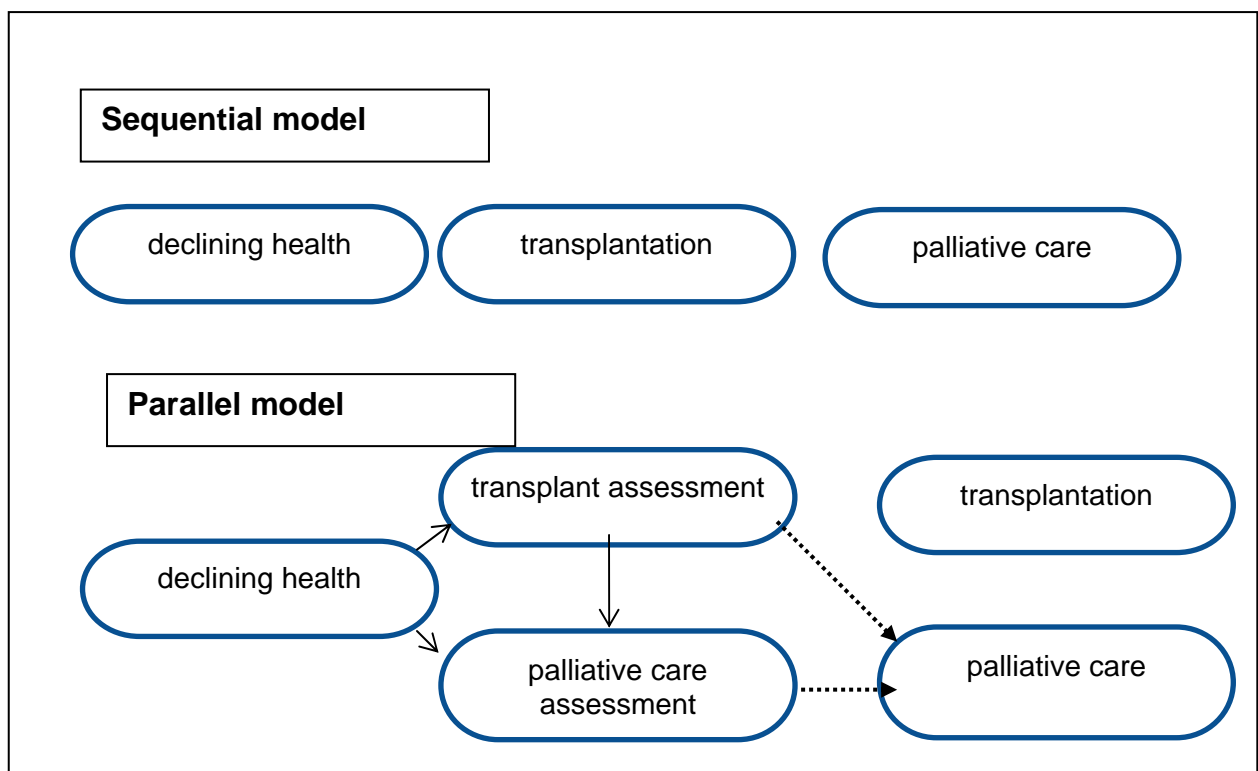
The provision of timely and sensitive information/education for consumers and family about end of life issues and palliative care is critical to discussions about transplantation. Written **information** will be developed that encourages discussion of end of life issues.

Consumers and families prefer open and honest **communication** about end of life issues, including having information available about what to expect and the options for care. A care plan will be developed by the multi-disciplinary team, the person with CF and their family/carers to facilitate communication about end of life management options and plans. The palliative care plan should identify the range of **psychological and psychosocial support** services appropriate to the needs of the individual, family and carers.

Family inclusion, for end of life planning, and **access to support services** for families of people with CF is a crucial aspect of the palliative care plan. It has been reported that in some instances families are unprepared for their loved one's death. Families are often confronted with life threatening health crises where the person with CF has survived, or where there has been good health and then sudden decline or unexpected progression of the disease.

A parallel, rather than sequential, model to end of life care in CF is the preferred model since **timing** is important to the discussion of end of life issues when there is declining health and referral to the transplant team.

Figure 3. The sequential and parallel models of end of life care for CF patients





4.8 Continuous Improvement

The Working Party undertook an audit of the current services provided for people with CF and identified areas for improvement to implement best practice principles and guidelines and service delivery. An extensive literature review (see appendix one) has been undertaken that sets out the levels of evidence base across the lifespan and progression of the disease. This best practice evidence base and the European, UK and draft Australian CF Guidelines are the basis of the model of care. The evidence base is rated A, B or C as set out below:

- A.** Many well designed randomised controlled trials directly relevant to the recommendation, yielding a consistent pattern of findings.
- B.** Some evidence from randomised controlled trials, but not optimal. More interpretation of the evidence was needed. For example, there were not many randomised controlled trials, their results were not consistent, and were not directly relevant to the recommendation. They may not have been directly relevant because, for example, the study population was different.
- C.** No randomised controlled trials but the issue is important enough to merit a recommendation that is based on published evidence and expert opinion of the authors and reviewers.



5.0 KEY RECOMMENDATIONS

Recommendation 1:

The Respiratory Health Network Cystic Fibrosis Taskforce model of care for Cystic Fibrosis in WA Health is endorsed.

Recommendation 2:

That Area Health Services develop and implement the WACFCCS organisational structure to deliver the model of care for CF in WA Health.

Recommendation 3:

An Implementation Plan to support the allocation of additional resources to meet the identified gaps and needs in service delivery for adults with CF is developed.

Recommendation 4:

Referral pathways across the service transitions to ensure a holistic multi-disciplinary approach to service delivery are developed and implemented. These should include:

- Paediatric/adolescent to adult services.
- Adult services to end stage/palliation.
- Inter-disciplinary - multi-disciplinary and medical specialists
- Discharge planning - hospital to community and GPs
- Community services to hospital services
- Hospital in the Home (HITH)

Recommendation 5:

That a “roadmap” of transition across the developmental age groups for people with CF and their families is developed including:

- Information about treatment, therapy and service providers across the lifespan and disease progression
- Education materials and information about key issues eg. Genetic counselling, transfer from paediatric to adult services, end of life issues, financial assistance
- A service directory for people with CF and their families
- Consumer focus on self management and peer support (people’s stories)

Recommendation 6:

That there is service enhancement through the development of evidence based guidelines and protocols for the appropriate management of CF including:

- Establishment of shared care models with metropolitan health services and WACHS
- Decentralisation of pharmacy and dispensing of medication to community and local hospital pharmacies



Recommendation 7:

That there is the development of **ICT solutions** including:

- A web based CF database that can store patient data and be used via laptop/spirocards and wireless internet access in hospital and community settings
- Develop electronic scripts generated by the web based CF database

Recommendation 8:

That **workforce strategies** are investigated and developed that include:

- The establishment and appointment of a Director of the WACFCCS
- Recruitment of medical, nursing and allied health staff to be upskilled and trained in the clinical management of CF
- Consideration of additional resources to meet the existing demand for adult services
- Capacity building in the primary and community care sector by training GPs and community-based health professionals in CF management
- Training and clinical support to WACHS staff and general metropolitan hospital through Telehealth and access to WACFCCS staff for consultation.

Recommendation 9:

That there is ongoing **evidence based research and advocacy** into CF by:

- Undertaking research at a state level
- Participating in national and international research
- Lobbying Federal Government for changes to the Commonwealth health care card status for people with CF

Recommendation 10:

That there is the development of appropriate quality and governance systems including:

- Development and ongoing maintenance of the WA CF data registry to support and report state data to the National CF Registry.



5.1 Implementation of Recommendations

The Working Party has recognised there are different levels and phases for implementation of the recommendations. They are:

- Achievable within existing resources and service delivery model
- Require further planning and development
- Require additional human resources, funding and endorsement

5.1.1 Phase one: Achievable within existing resources and service delivery model

- Inclusion and participation of consumers and families/carers in decision making and development of care management plans
- Exploration of options to increase community services e.g. Dedicated Community Physiotherapy classes
- Develop referral pathways as set out in recommendation 3.
- Review of current consumer and family information, education and resource material
- Commence negotiations with WACHS to formalise current outreach clinics
- Commence negotiations with WACHS to improve communication, staff training and support and shared care models via existing outreach clinic service
- Establish Telehealth and telephone support services for GPs and clinical staff in metropolitan and WACHS.
- Roadmap of transition - develop project brief and seek funding for relevant agencies to undertake this project.

5.1.2 Phase Two: Require further planning and development

- Develop organisational structure for WACFCCS (Rec 2)
- Decentralisation of pharmacy and dispensing of medication to community and hospital pharmacies (Rec 5)
- Workforce strategies and professional development/training (Rec 8)
- Roadmap of Transition project (Rec 5)

5.1.3 Phase Three: Require additional human resources, funding and endorsement

- Establishment of WACFCCS and appointment of Director.
- Additional dedicated medical, nursing and allied health resources to meet the demand for services (Rec 8)
- ICT CF database (Rec 7)
- Shared care models between and across metropolitan hospitals and with WACHS (Rec 6)

6.0 EVALUATION

The CF model of care will be evaluated periodically. At each phase of implementation a variety of strategies will be utilised including:

- stakeholder surveys;
- hospital morbidity data analysis; and
- comparative clinical outcome metrics using the national CF data registry.



GLOSSARY

CF	Cystic Fibrosis
CFWA	Cystic Fibrosis Western Australia
CNC	Clinical Nurse Consultant
CQI	Continuous Quality Improvement
ENT	Ear Nose Throat
FSH	Fiona Stanley Hospital
GP	General Practitioner
HITH	Hospital in the Home
ICT	Information Communication Technology
ICU	Intensive Care Unit
IV	Intra Venous
KPI	Key Performance Indicator
LDU	Low Dependence Unit
NATA	National Accreditation Training Association
NGO	Non Government Organisation
NMAHS	North Metropolitan Area Health Service
MDT	Multi Disciplinary Team
PACS	Phosphofurin Acidic Cluster Sorting Protein
PMH	Princess Margaret Hospital
RPH	Royal Perth Hospital
SCGH	Sir Charles Gairdner Hospital
SMAHS	South Metropolitan Area Health Service
WACFCCS	Western Australian Cystic Fibrosis Collaborating Clinical Centre
WACHS	Western Australia Country Health Service



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APPENDICES

Legend for Appendix 1 - Proposed Model of Care for CF

Levels of evidence:

- A Many well designed randomised controlled trials directly relevant to the recommendation, yielding a consistent pattern of findings.
- B Some evidence from randomised controlled trials, but not optimal. More interpretation of the evidence was needed. For example, there were not many randomised controlled trials, their results were not consistent, and were not directly relevant to the recommendation. They may not have been directly relevant because, for example, the study population was different.
- C No randomised controlled trials but the issue is important enough to merit a recommendation that is based on published evidence and expert opinion of the authors and reviewers.

Note

The management guidelines for routine care, pulmonary exacerbations and self-care are to be regularly reviewed in order to take account of current evidence. From the analysis that follows it is clear that current practice falls short of best practice in a number of areas



Appendix 1: Newborn & Children's Proposed Model of Care

What	Who	Where	Institutional	Policy
Diagnosis Newborn screening (NBS) ^{AC} Late diagnosis Counselling ^{BC} Education ^B	NATA accredited lab ^C Geneticist ^C Trained counsellors ^C Multidisciplinary team ^C (MDT) Non Government Organisations (NGO)	Initial Tertiary hospital ^C Low dependency unit (LDU) Follow-up Tertiary centre ^C Home	LDU Communication strategy Patient register/database Clerical support ICT support	Admission policy Management protocol ^{ABC} Audit and Continuous Quality Improvement (CQI)
Multidiscipline care^C	Respiratory specialists Gastroenterologist Adolescent specialist Nurse consultant Junior medical staff Physio Dietician Social worker Clinical psychologist Teacher Community care workers	Tertiary centre led ^C	Weekly meeting Meeting facilities Communication strategy Patient register/database Clerical support ICT support	Governance Confidentiality Data-linkage/sharing
Routine surveillance Outpatient review ^C Imaging ^B Microbiology ^C Pulmonary function ^C Bronchoalveolar lavage (BAL) ^B Pathology Pharmacy ^C	MDT Radiologist Anaesthetist (to 4 yr) Respiratory scientist Microbiologist Histopathologist Clinical chemist Pharmacist	Tertiary hospital ^C Regional hospital	Pulmonary function laboratory ^{BC} Portable spirometry Combined CT/BAL ^B Pseudomonas typing ^{BC} Telehealth ^B Communication strategy Clerical support ICT support	Annual review Management protocol ^{ABC} Anaesthetic protocols Micro. policies/protocols Outreach funding policy Audit and CQI



Newborn & Children's Proposed Model of Care (continued)

What	Who	Where	Institutional	Policy
Pulmonary exacerbations	MDT Anaesthetist Microbiologist Clinical chemist Pharmacist Radiologist	Tertiary hospital ^B Regional hospital Home ^B	Emergency beds Timely IV access Same day care IV access Single rooms ^C LDU Pathogen surveillance/typing ^C Regional education/training Effective home supervision ^B Communication strategy	Management protocol ^{ABC} Admission policy Microbiology policies and protocols Infection control Regional funding policy HITH Audit and CQI
Patient/carer self-management ^B	MDT NGO Patient family Teachers Sports coaches	Hospital School Home Sports clubs etc.	Education ^B Review Evaluation	Confidentiality Management guidelines ^{ABC}
Lung function ^{BC}	Respiratory scientist Community nurse Physiotherapist	Tertiary hospital Community	Pulmonary function lab Portable spirometry Education and training	Laboratory accreditation
Complications	Multidisciplinary team Transplant specialist Specialist surgeons Specialist physicians Psychiatrist Anaesthetists Radiologists Specialist nurses Occupational therapist	Tertiary hospital	Communication strategies	Referral policy



TRANSITION FROM PAEDIATRIC TO ADULT SERVICES

What	Who	Where	Institutional	Policy
Transition Outpatient surveillance c Education c Vocational support c Counselling c Psychosocial support c Transfer planning c	Multidisciplinary team ^c (MDT) NGO CFWA, QUIT Community care workers Adolescent specialist	Paediatric Tertiary hospital ^c Community/home via NGO	Weekly meeting Meeting facilities Communication strategy Patient register/database Clerical support ICT support	Annual review Management protocol ^{ABC} Audit and CQI Confidentiality
Transfer^c Transition meeting Transfer outpatient clinic Psychosocial support	MDT NGO CFWA Adult respiratory specialist Adolescent specialist	Paediatric and Adult Tertiary centre led ^c NGO	Regular joint meetings between tertiary centres Meeting facilities Communication strategy Patient register/database Clerical support ICT support	Shared management protocol Data-linkage/sharing Audit and CQI
Young adult-focussed CF care Outpatient surveillance ^c Youth friendly CF services Education c Vocational support c Counselling c Psychosocial support c	MDT Adolescent specialist NGO General practitioners Obstetrician Fertility specialists Geneticist	Tertiary hospital ^c Regional hospital General practice HITH	Ambulatory service Emergency beds Timely IV access Same day care IV access Home IV support and supervision Single rooms ^c Weekly meeting Meeting facilities Communication strategy Patient register/database Clerical support ICT support	Annual review Management protocol ^{ABC} Regional funding policy Outreach funding policy Audit and CQI



ADULT MANAGEMENT

What	Who	Where	Institutional	Policy
Routine surveillance Outpatient review (at least quarterly) ^C Sputum microbiology ^C Imaging ^B Pulmonary function ^C Pathology Pharmacy ^C Annual review ^C	Multi-disciplinary team (MDT)- see below ^C	Tertiary hospitals ^B Regional hospitals	North & South Metro Adult CF centres Proximity / linkage of tertiary centres to regional centres / community ^{BC} Pulmonary function laboratory Portable spirometry Pathogen surveillance/typing ^{BC} Telehealth Communication strategy Data-linkage Patient register/database Electronic patient record Outreach clinics ^C Exercise training ^B	Joint appointments across regions Data-linkage Management guidelines (universal) Microbiology policies and protocols Infection control policy Outreach funding policy Pharmacy funding / dispensing policy Changing demographics Privacy policy Audit and CQI KPIs



ADULT MANAGEMENT (continued)

What	Who	Where	Institutional	Policy
<p>Multidisciplinary care^c</p> <p>Care Plans including: Psychological & psychosocial support Transplant End of life care Palliative care Family & Carer support</p>	<p>Core CF team^c</p> <p>Respiratory specialists Clinical fellow Junior medical staff Nurse consultant Home care nurse Specialist Physio Specialist Dietician Social worker</p> <p>Gastroenterologist Hepatologist Microbiologist Pharmacist Endocrinologist Diabetic educator Thoracic surgeon ENT specialist Radiologist Clinical psychologist Psychiatrist Community care workers Respiratory scientist Histopathologist Fertility specialists Obstetrician / gynaecologist Rheumatologist Administration officer Data clerk</p>	<p>Tertiary hospital led^B</p>	<p>Weekly meeting Meeting facilities Communication strategy Patient register/database Electronic patient record Research Professional development^c Training^c</p>	<p>Governance Confidentiality/privacy Data-linkage Audit and CQI KPIs</p>



ADULT MANAGEMENT (continued)

What	Who	Where	Institutional	Policy
Pulmonary exacerbations^B	Core CF team ^C MDT ^C Venous access specialists Physiotherapy support Nutritional support Microbiologist Pharmacist Radiologist	Tertiary hospital ^B Regional hospital Low Dependency Unit (LDU) Home ^B	Emergency beds - within 48hr IV insertion within 48hrs Same day care IV access Single rooms ^C Low Dependency Unit (LDU) Pathogen surveillance/typing ^C Regional education/training Appropriate home supervision ^B	Management guidelines ^{ABC} Admission policy Microbiology policies and protocols Infection control policy Regional funding policy HITH supervision/reporting Audit and COI KPIs
Lung function^{BC}	Respiratory scientist Community nurse Physiotherapist	Tertiary hospital ^C Community	Pulmonary function lab Portable spirometry Education and training	Laboratory accreditation Data-linkage Infection control
Patient/carer self-management^{BC}	MDT NGO Patient family Employers	Hospital College/University Work Home	Education Review Evaluation	Confidentiality Management guidelines
Late Diagnosis Counselling^{BC} Education	NATA accredited lab ^C Geneticist ^C Trained counsellors ^C MDT ^C NGO	Initial Tertiary hospital Low dependency unit (LDU) Follow-up Tertiary hospital Home	LDU	Admission policy Management guidelines



ADULT MANAGEMENT (continued)

What	Who	Where	Institutional	Policy
Complications	Core CF team Multidisciplinary team Specialist surgeons Specialist physicians Psychiatrist Anaesthetists Radiologists Specialist nurses Occupational therapist Transplant MDT	Tertiary hospital	Communication strategies Gastrostomy service Bronchial artery embolisation Non-invasive ventilation service	Referral policy



TRANSPLANTATION

What	Who	Where	Institutional	Policy
Transplant Assessment / Active List ^C	Transplant MDT Transplant physiotherapist Transplant microbiologist Psychiatrist / psychologist Social worker	WA Lung Transplant Unit ^C Referring Centre Low Dependency Unit (LDU)	Transplant MDT meetings ^C LDU Transplant gymnasium Assessment and active list clinics Multiple combination antibacterial testing ^C Pathogen surveillance/typing ^B Education End of life & palliative care planning	Referral policy ^B Microbiology policies and protocols ^C Shared care on active list Meeting minutes
Transplantation ^B	Transplant MDT ^C Transplant surgeons Transplant physicians Transplant microbiologist Hepatologist Gastroenterologist Endocrinologist Radiologist Specialist nurses Physiotherapist Occupational therapist Social worker Psychiatrists	WA Lung Transplant Unit ^C	Non-invasive ventilation Theatres ICU Electronic patient record / database Communication	Infection control policy ^{BC} Privacy Public relations Audit / CQI KPIs



TRANSPLANTATION (continued)

What	Who	Where	Institutional	Policy
Post-transplant Care ^B	Transplant MDT ^C Transplant surgeons Transplant physicians Transplant anaesthetists / perfusionists Transplant microbiologist Hepatologist Gastroenterologist Endocrinologist Radiologist Specialist nurses Pharmacist Physiotherapist Occupational therapist Social worker Psychiatrists	WA Lung Transplant Unit ^C Home ^C Regional centres LDU	Transplant gymnasium Transplant clinics Non-invasive ventilation Theatres (bronchoscopy / lung biopsy) ICU LDU Electronic patient record / database Education Communication Research	Local transplant protocols / guidelines Specialist referral policies Microbiology policies and protocols ^{BC} Infection control policy ^{BC} Shared care on active list Audit / CQI ^C KPIs ^C



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