GOVERNMENT OF WESTERN AUSTRALIA

Sustainable Health Review

# Public Submission Cover Sheet

**Please complete this sheet and submit with any attachments to the Sustainable Health Review Secretariat**

| **Your Personal Details**<br>***This information will be used only for contacting you in relation to this submission*** | |
|---|---|
| **Title** | Mr ☐ Miss ☐ Mrs ☐ Ms ☐ Dr ☐ Other ☒ Clinical Professor |
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**Publication of Submissions**

***Please note all Public Submissions will be published unless otherwise selected below***

☐ I do not want my submission published

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**Submission Guidance**

**You are encouraged to address the following question:**

**In the context of the Sustainable Health Review Terms of Reference listed below, what is needed to develop a more sustainable, patient centred health system in WA?**

- Leveraging existing investment in Primary, Secondary and Tertiary healthcare, as well as new initiatives to improve patient centred service delivery, pathways and transition;
- The mix of services provided across the system, including gaps in service provision, sub-acute, step-down, community and other out-of-hospital services across WA to deliver care in the most appropriate setting and to maximise health outcomes and value to the public;
- Ways to encourage and drive digital innovation, the use of new technology, research and data to support patient centred care and improved performance;
- Opportunities to drive partnerships across sectors and all levels of government to reduce duplication and to deliver integrated and coordinated care;
- Ways to drive improvements in safety and quality for patients, value and financial sustainability, including cost drivers, allocative and technical efficiencies;
- The key enablers of new efficiencies and change, including, research, productivity, teaching and training, culture, leadership development, procurement and improved performance monitoring;
- Any further opportunities concerning patient centred service delivery and the sustainability of the WA health system.

**Submissions Response Field**

***Please type your response into the field below. Alternatively you may provide your submissions as a separate attachment (Suggested Maximum 5 pages).***

**Attachments:**

1. Reducing the Reliance on Tertiary Care of Patients with Chronic Conditions (Body)
2. Appendix A: Cystic Fibrosis Model of Care 2013 (pdf)
3. Appendix B: Integrated eHealth Solution for Cystic Fibrosis Patients.(docx)
4. Appendix C: CFWA Letter of Support

## Reducing the Reliance on Tertiary Care of Patients with Chronic Conditions

This submission demonstrates how the implementation of a model that effectively transfers aspects of care for a complex chronic disorder (cystic fibrosis) from a tertiary hospital to community services can benefit both consumers and the health service. The successful implementation of a decentralised care model can reduce the burden of care on families and individuals with CF, improve sustainability and will also provide opportunities to reallocate resources within HDWA to meet unmet needs.

**Background**

The implementation of a decentralised or distributed model of care is the first recommendation of the Cystic Fibrosis Model of Care published by the Health Department of WA (HDWA) in 2008 and revised in 2013 (Appendix A) (1). Cystic fibrosis (CF) is the commonest life shortening genetic disease commencing in childhood. Cystic fibrosis is an autosomal recessive condition that affects the lungs and the respiratory tract, digestive system, sweat glands, and male reproductive organs. There are approximately 4000 children and adults with the condition in Australia. Approximately 1/24 individuals are carriers of a CF gene mutation and between 130 and 150 children with CF are diagnosed by newborn screening each year in this country. There are more than 400 patients with CF in Western Australia. A recent study indicated that reducing the burden of care associated with the management of cystic fibrosis was the first priority of consumers (2).

The most important cause of morbidity and mortality is progressive lung disease that commences early in infancy (3, 4). However, in adults the complexity of disease management is increased by comorbities such as diabetes, immune complex diseases such as arthritis, emergence of resistant pulmonary infections and new pathogens.

Expert care by multidisciplinary teams in specialist tertiary centres has greatly improved median survival so that death during childhood is now unusual and for the first time, the number of adult survivors exceeds children with CF in many centres in Australia including Perth (5).

The implementation of "comprehensive care models" in specialist centres (6) has been the single most important factor contributing to improved survival. ***The holistic care provided by such teams has resulted in a model of care almost entirely located within tertiary institutions regardless of severity of disease.*** An analysis by the Centre for Health Economics Research and Evaluation, (University of Technology, Sydney) has described the increased use of health resources with age and severity of disease (7). The proportion of individuals with more severe disease increases with age and this pattern imposes an increasing burden of care particularly on adult respiratory centres. Most importantly however, it can be anticipated that as care improves and new therapies become available, ***the absolute numbers of individuals at any age with milder disease will also increase. Even now, over 50% of young adults have relatively mild disease (defined by lung function) and most adults with CF are employed (67%) and many have undertaken tertiary education (37%) (7).*** Therefore, for many children (and their families) and adults with CF, the focus of care in tertiary institutions often imposes burdens that materially affect quality of life i.e., time lost from school/work in order to access some services that could potentially be provided closer to home (8, 9). At the same time resources are currently being stretched in tertiary centres caring for increasing numbers of adults, many with complex and severe disease (10, 11). Therefore, it is imperative to reform and ***re-focus the delivery of care, such that primary, secondary and tertiary services are better co-ordinated to provide appropriate care for individuals with CF and families, across the continuum of care*** (12) without sacrificing the quality of management that has transformed the lives of individuals born with CF. This approach is consistent with the current national health reform agenda (12).

While the care of patients with CF and complex/severe disease requires specialised skills in dedicated centres, evidence suggests that a substantial proportion of routine care could be undertaken capably and efficiently at the community level (13-16). Since the care of patients with CF has evolved solely in tertiary centres, the successful transition to a distributed care model will serve as an exemplar for cordination of tertiary and community services for other, complex, chronic diseases. The transition aims to reduce dependancy on state-funded, tertiary resources and achieve better outcomes for patients and families. This will be accomplished by improving access to a broad range of necessary services close to home underpinned by a comprehensive electronic patient record and communication platform accessible by all providers.

**Aims of the model**

- Decentralised care for cystic fibrosis

**Core enablers**

- Partnership between
  - Not-for profit, community providers (St John's Ambulance/Apollo Health)
  - Consumer organisation (CFWA)
  - Consumer-focussed, innovative tertiary respiratory service (CAHS)
- Proven CF ICT solution (Smart Health – Appendix B)
- Experienced education and training providers (CFWA, CAHS)
- Established community-based complex care providers (CFWA, CAHS)

**Deliverables**

- Consumers
  - Care close to home
  - Convenient access to multidisciplinary services
  - Access to services not limited by tertiary hospital funding
  - Integrated, holistic primary and specialist care
  - Reduced burden (time, sick-leave/carer-leave, transportation costs)
- Health system
  - Reducued wait-lists
  - Reduced cost of CF care
  - Multidisciplinary resources redeployed to areas of unmet need
  - Improved safety and quality
  - Prototype solution – can be generalised i.e., adult CF services, other programs

**Initial program implementation**

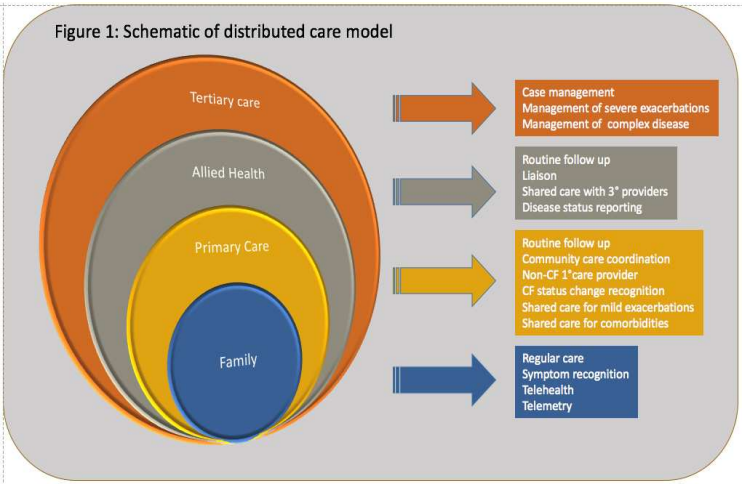
The initiating service will be established at sites of an Apollo Health practice. These multidisciplinary facilities are situated in Joondalup, Cannington, Cockburn and Armadale and are within easy reach of the families of 40 children with CF. Although initially confined to children with CF who have relatively mild disease, the program would ultimately also include adults. The program will be devloped and delivered by:

- Tertiary CF Centres
  - Case management
  - Clinical pathways
  - Management of individuals with complex/severe disease
  - Assessment of educational needs and the supervision of staff training
  - Spirometry training and quality control
- Apollo Health
  - Routine care of individuals with mild/moderate disease
  - On-site physiotherapy, nutrition, psychology, imaging, pathology and pharmacy services
  - Walk-in urgent care
  - Supervised care for mild pulmonary exacerbations
  - Liaison with community care providers
- Cystic Fibrosis WA
  - Consumer engagement/communication
  - Delivery of training packages
  - Delivery of home care for mild/moderate exacerbations
  - Contribution to hospital in the home services

**Proposed Model**

The model is consistent with the CF Model of Care developed by service providers, consumers, health economists, community development experts and health adminstrators (17). The platform translates local research into practice and provides a platform for further health systems research by groups committed to improving the health of children and adults with chronic conditions. Researchers based at the Telethon Kids Institute, other Medical Research Institutes and Universities can easily partner with clinical service providers through the WA Health Translation Network and will provide academic leadership for the evaluation processes.

Figure 1: Schematic of distributed care model




**Established enablers**

- Stakeholder support (Appendix B)
- Experience co-ordinating and delivering complex services to regional centres
- Well-defined population
- Existing training packages for GPs and allied health professionals
- Not-for-profit primary care partner
- Medicare GP extended care plan provisions
- Supportive State and Commonwealth health environments

**Essential additional requirements**

- Community capacity building
  - Specific and tailored training for GPs, allied health and nursing practitioners
  - Practice-based spirometry
- Electronic patient record and ICT solution (see–Appendix C)
- Clinical pathway development
- Stakeholder communication strategy
- Evaluation framework development, implementation and reporting

**Indicative costs**

| | Year one | Year two |
|---|---|---|
| GP training | 61000 | 31000 |
| Allied health training | 18000 | 5000 |
| Spirometry training | 5000 | |
| ICT | 291000 | 132000 |
| Communication strategy | 5000 | |
| Evaluation | | 20000 |
| Total | $382 000 | $188 000 |

Based upon 4 Apollo health centres and 20 days intensive training for 8 GPs and 12 allied health staff.

**Anticipated benefits for HDWA**

- **$100 000-$120 000 per year** of activity based funding available for redstribution (40 patients @ 2 routine consults/year in community rather than tertiary hospital)
- Improved safety
- Improved efficiencies

**Anticipated benefits for consumers**

- Reduced burden of care
- Improved access to services

**Sustainability**

- Ultimately, the provision of a broad-based community service for CF has the potential to reduce overall demand on tertiary services by 20-30% (320-480 consults/year).
- For every patient consult in the community, there is a tertiary ABF benefit of about $1300
- The ICT costs associated with additional community providers is $100/practice (4 practitioners)
- Additional training costs are approximately $3-4000 per practitioner.
- Reduced demand within the paediatric program for CF services will allow filling of unmet need for non-CF complex respiratory care.
- The model provides for growing demand due to improved survival of adults by transition of adolescent patients from paediatric care to adult care largely as a continuation within the established distributed care framework. The tertiary institution would assume responsibility for case management from the paediatric centre and additional tertiary care escalated only with increasing disease severity/complexity.
- The communication and ICT solution will improve quality and safety of care and will reduce costs due to medical errors and inefficiencies.
- The model, if successful, will be relevant to other chronic disorders with a high demand on tertiary care thus improving overall system sustainability.

**Integration of healthcare innovations**

The distributed care model will provide a platform to test innovations in healthcare and health technology that will support self-management and decentralised care strategies. Examples include:

- Smartphone technology
    - Facilitated telehealth (available)
    - Telemetry e.g., pulmonary function testing (available)
    - Integrated communication packages (in-development)
        - Provider appoinments (available)
        - Reminders (available)
        - Symptom diaries (available)
        - Pharmacy order tracking (available)
        - Health summaries (available)
    - Self-management support apps (available)
- Point-of-care/personal biomarker detection for disease monitoring (in development at Telethon Kids Institute

**Timeline**

The program will be implemented and evaluated in children over the course of 2 years (Figure 2). Once established the costs for HDWA to retain the program will be negligible and limited to to further capacity building offset by ABF savings. The continuation/expansion

of the program will be dependant upon consumer acceptability and objective, disease-specific outcomes.

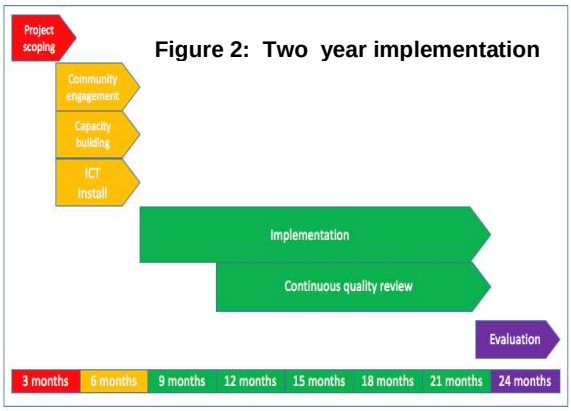



**Figure 2: Two year implementation**

**Summary**

This submission demonstrates the relaively low investment required to achieve a transformational reform of service delivery. The proposed model can deliver improved access to services for patients and families close to home, has the potential to significantly reduce demand on tertiary services and provides opportunities to reallocate resources within HDWA to address areas of unmet need.

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Government of Western Australia
Department of Health

Appendix A

# WA Cystic Fibrosis Model of Care

## Respiratory Health Network

May 2013

Text
Text

Delivering a Healthy WA

**Suggested Citation**

Department of Health, Western Australia. WA Cystic Fibrosis Model of Care. Perth: Health Networks Branch, Department of Health, Western Australia; 2013.

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## Acknowledgements

The following people are acknowledged for contributing to developing and reviewing the WA Cystic Fibrosis Model of Care (2013):

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# Executive Summary

Cystic Fibrosis (CF) is an inherited multi-system chronic disease that affects the respiratory tract and lungs, digestive system, sweat glands and reproductive organs. CF is a common genetic disease affecting individuals throughout their lifetime. The most important cause of mortality and morbidity is progressive lung disease that begins in early infancy. In 2011 there were 309 people in Western Australia (WA) with CF known to the Australian Cystic Fibrosis Data Registry.

There is no cure for CF; however expert care by multidisciplinary teams has greatly improved life expectancy. 2010 was the first year there were more adults than children living with CF in WA. As survival for the person with CF improves, so does the health, social and economic burden for the individual and the community. This trend highlights the need to plan and deliver health services which address the lifestyle needs of both adults and children, including transitional care from childhood to adulthood, ideally providing care close to home which addresses multiple health domains.

The WA CF Model of Care (MoC) addresses the issues of:

- Disease progression and transition across the continuum of care
- Partnerships between the person with CF, their family/carer, community based heath care providers and Western Australian Cystic Fibrosis Care Service (WACFCS) multidisciplinary teams
- A shift from tertiary centred management of the person with CF to a multidisciplinary community centred approach
- Offering the person with CF and their families an opportunity for effective self management using a wellness approach instead of an acute hospital-based approach.

The WA CF MoC describes the WACFCS; a model of service delivery driven by partnerships between the consumer and their family/carer, paediatric and adult tertiary hospital services and other specialist and community-based services.

The aim of the WACFCS is to provide lifetime care for the person with CF across the continuum of care; addressing the biological and psychosocial needs of patients with CF and their families/carers including end of life and palliation services. The WACFCS will provide leadership and the WACFCS Service Director will facilitate the paediatric (Princess Margaret Hospital [PMH]) and adult (Sir Charles Gairdner Hospital [SCGH]) based services and the large component of community based service delivery.

WACFCS tertiary centre activities may include:

- Delivering excellence in clinical practice, teaching and research
- Building workforce capacity among multidisciplinary professionals
- Addressing the needs of consumers with complex conditions or who require services only available in tertiary hospital settings
- Clinical governance: developing clinical pathways, referral pathways, policies and practice standards for CF.

The WACFCS consumer-centred service delivery pathway, consistent with the Cystic Fibrosis Standards of Care, highlights the need for:

- Specialist care, usually provided at a tertiary facility
- Clinical care, usually provided in a community-based facility
- Provision of multidisciplinary health services
- Access to lung transplant services
- Self management support using a wellness approach.

The Model builds on the knowledge and capacity underpinning the current services for people with CF and evidence-based best practice as documented in the WA Clinical Services Framework 2010-2020.

The key recommendations of the Model are to:

- Establish linkages between CF tertiary hospital services and community-based organisations (e.g. general practices, Medicare Locals, GP Super Clinics) to facilitate the transition of health services from tertiary services to the community in a partnership model. An evaluation of consumer-centred, clinical and economic outcomes should be undertaken to judge the safety and effectiveness of this model of service delivery in WA.
- Develop clinical governance frameworks utilising evidence-based guidelines and protocols to facilitate safe and effective tertiary-led delivery of care in community settings.
- Develop referral pathways to guide the delivery of tertiary-led care in the community sector, with a focus on:
  - Transition from paediatric and adolescent services to adult services
  - Transplantation services
  - Interdisciplinary care
  - Palliative care.
- Delivery of care in the community sector may be facilitated through implementation of a CF-specific or chronic lung disease liaison team to provide a link between tertiary CF centres and community-based centres.
- Where possible, decentralise pharmacy and dispensing of medication to community and local hospital pharmacies using a pharmacy network.
- Promote 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. UUSmart Health Solutions may be an appropriate platform in this context. Both patients and health professionals should have access to the stored data.
  - Provide consumers with access to reliable and valid information about CF and self-management, including access to innovative tools such as smart-phone ‘apps’.
- Continue to build workforce capacity (clinical skills and knowledge) across the sector, especially in primary care – in collaboration with CF centres, CFWA, and Universities. Professional development opportunities should be offered within an interprofessional framework that acknowledges the critical role of co-care and self-management.

- Promote a culture of research and evaluation by:
  - Undertaking research at a state level, including research that examines safety, efficacy and efficiency of alternative models of health service delivery
  - Participating in national and international research, particularly in clinical trials.
  - Continuing to contribute data to the National CF Registry.
- Undertake a review of CF centres in WA to identify activity over the last five years which aligns with the 2007 Model of Care and explore opportunities to facilitate implementation of the 2013 Model of Care recommendations.

# 1. Introduction

Cystic Fibrosis (CF) is an inherited, progressive, multi-system disease of the secretory glands affecting the respiratory tract, digestive system, sweat glands and reproductive organs. A defective gene causes CF, resulting in sticky, thick mucous production, causing chronic infections, inflammation and obstruction of the lungs and difficulty in digesting food and nutrients [4,5].

CF is an incurable chronic disease affecting both males and females through infancy, childhood, adolescence and adulthood. In 2011 the national median age of death for a person with CF was twenty seven years, which was the same in 2010. Nationally, fifteen (60%) of the twenty five CF deaths reported in 2011 were attributed to respiratory complications from CF [6].

Given the multi-system and complex nature of CF, optimal outcomes may be achieved when health services are delivered by a multidisciplinary team, focusing on prevention of airway obstruction and infection, physiotherapy, enzymes, vitamins, medications and dietary regimes [7]. Advances in research, symptom management and the implementation of multidisciplinary teams are improving the quality of life and longevity of people with CF [8].

Although declining pulmonary function is typical in CF, the rate of decline is highly variable. Life expectancy for a person with CF has increased because of improved treatment, such that a baby born with CF today could expect to live into their forties or fifties. It is projected that within the next decade most people with CF living in developed countries will be adults rather than children [9]. In 2010 the average age of a person with CF in Western Australia was 18.6 years, with 50.9 percent of people being over 18 years of age, the first year where there were more adults living with CF than children [10]. This trend highlights the need to plan and deliver health services which address the lifestyle needs of both adults and children, including transitional care from childhood to adulthood, and ideally providing care close to home which addresses multiple health domains.

This updated Model of Care, informed by evidence and expert opinion, will guide planners and health service providers in delivering best-practice care to Western Australians with CF. In particular the Model will focus on the systemic structures and strategies to improve service delivery in the context of the State's health priorities, resource availability, organisational capability, operational factors, contemporary policy and local/community environments.

# 2. Methods

The 2007 Cystic Fibrosis Model of Care [11] was developed by a small Working Party, made up of key stakeholders with expertise and knowledge of CF. A review of the evidence-base for the clinical management and delivery of CF services, a gap analysis and community/stakeholder consultation were undertaken to inform the development of the 2007 Model of Care. In 2012 the Working Party was re-convened to update the 2007 Model of Care. The 2013 Cystic Fibrosis Model of Care underwent an extensive consultation process involving consumers, carers, health professionals, non-government organisations and other key stakeholders. The update aimed to ensure it aligned with contemporary evidence, current epidemiology of CF, and opportunities offered by the Australian health reform agenda to improve health service delivery to consumers with CF. A particular focus was placed on improved service integration with primary care.

To ensure a consistent approach to CF management and treatment at a state and national level in Australia, the revised 2013 Cystic Fibrosis Model of Care is informed by the following key documents:

- Cystic Fibrosis Standards of Care, Australia 2008 [12]
- National Chronic Disease Strategy, 2006, National Health Priority Action Council, Australian Government Department of Health and Ageing [13]
- WA Health Promotion Strategic Framework 2012-2016, Chronic Disease Prevention Directorate, Department of Health [14]
- National Tobacco Strategy 2004–2009, Ministerial Council on Drug Strategy [15]
- Framework for the Treatment of Nicotine Addiction, Respiratory Health Network, Department of Health WA [16]
- WA Chronic Health Conditions Framework 2011-2016, Health Networks Branch, Department of Health WA [3]
- WA Chronic Conditions Self-Management Strategic Framework 2011-2015, Health Networks, Department of Health WA [17]
- WA Clinical Services Framework 2010-2020 [18]
- WA Primary Health Care Strategy, Health Networks Branch, Department of Health WA [19]
- WA Chronic Lung Conditions Model of Care, Respiratory Health Network, Department of Health WA [2]
- Liverpool Care Pathway [20]
- Palliative Care Model of Care [21]

# 3. What has happened since the 2007 Cystic Fibrosis Model of Care?

There have been a number of positive system improvements since the publication of the 2007 Model of Care for Cystic Fibrosis. These are summarised below.

**Health Services – Adult CF Centre at Sir Charles Gairdner Hospital (SCGH)**

- Workforce (2007 Recommendation 8):
  - Employment of an additional Adult CF physician.
  - Employment of a 0.5 full time equivalent (FTE) CF dietician.
  - A CF physiotherapist visited CF centres in Europe and the United Kingdom for professional development.
  - Members of the Adult CF Team visited Johns Hopkins CF Centre in Baltimore, Maryland (USA) to consolidate knowledge in centre-led CF care.
  - Service changes at the Adult CF Centre at SCGH include:
    - The establishment of segregated clinics.
    - CF nurse consultant transitioned to CF nurse practitioner.
    - Establishment of joint clinics with a specialist in CF-related diabetes.
    - Use of telehealth consultations for country patients.
    - Clinical CF research projects with CF clinical trials conducted through the Lung Institute of WA (LIWA), WA Lung Research, and Linear Clinical Research Ltd.
- Improvements in transition links between SCGH and PMH through the transition program in partnership with Cystic Fibrosis WA (CFWA) (booklets on the process of transition are available at PMH and SCGH). (2007 Recommendation 5).
- Australian CF Data Registry – up to date data input from SCGH to inform the WA Adult CF Centre annual report generated each year. (2007 Recommendation 10).
- Booklet on CF and Fertility is now available at SCGH. (2007 Recommendation 5).
- Seven CF patients received lung transplants at Royal Perth Hospital (RPH) in 2011, reinforcing the strong link between the RPH lung transplant team and the SCGH Adult CF Team. (2007 Recommendation 4).

**Health Services – Paediatric CF Centre at Princess Margaret Hospital (PMH)**

- Outreach services (2007 Recommendation 6):
  - Extended to include Broome
  - Doubling of outreach clinics to Karratha, Port Hedland, Bunbury, and Geraldton
  - Increased use of telehealth for consultations with rural-based clients and families.
- Clinical services planning:
  - Engagement with South Metropolitan Health Service (SMHS) planners regarding service delivery at Fiona Stanley Hospital (FSH) and development of a partnership framework for service delivery in the community. (2007 Recommendation 3).
- Transition services framework (2007 Recommendation 5):
  - Significant update of the transition framework for paediatric patients of PMH with CF to the adult CF centre at SCGH [22,23].

## Cystic Fibrosis WA (CFWA)^

CFWA has supported and been involved with a number of projects supporting the 2007 CF Model of Care (MoC) which include (2007 Recommendations 4 & 5):

- The Regional Respiratory Training Program (RRP) has completed three years of training of regional nurses and physiotherapists. In 2012, numbers reached 33, far exceeding the original target of 20.
- CFWA employ home care workers to deliver airway clearance, exercise programs, respite and home duties for people with cystic fibrosis.
- Training of home care workers has included; mental health first aid, general first aid, life coaching and a boundaries workshop.
- Life Coaching was offered to adults with CF and their parents from early 2012 to assist with identifying goals and developing strategies to achieve them.
- In late 2012 a personal trainer program commenced in the north western suburbs for adults with CF assessed as likely benefiting from participating in a personal exercise program.
- Cleaning services are offered to those people with CF who require extra support and who may not necessarily need a home care worker.
- Education expositions were held in 2010 and 2011. In 2012 a series of seminars were offered; the most successful being a fertility workshop.

- Ongoing programs are provided to reach all ages and stages of people with CF including: newly diagnosed program; transition program; transplant support program; sibling support program, and regional outreach program.
- Successful grant applications have enabled CFWA to provide medical, physiotherapy and exercise equipment for adults and children with CF.
- CFWA have initiated a national approach to education to streamline and standardise CF-related education throughout Australia.
- Dietary requirements and infection control are being addressed by CFWA through their “CF friendly schools” approach.
- Demand on services has resulted in expansion of physiotherapy services, development of new roles including a services manager and a recreation and health promotions officer, and employment of a nurse educator to deliver education to allied health professionals and general practitioners.
- CFWA have committed over quarter of a million dollars in 2012 including $30K to provide seed funding to help establish the first adult CF research capability at SCGH.
- PhD top-up scholarships have been provided.

^ CFWA is a member based non-government organisation that advocates on behalf of people with cystic fibrosis and the CF community, including carers, and extended family. CFWA provides a range of community and home-based services including social work, nursing and physiotherapy, and support groups for people with CF, their families and carers.

**New clinical research: The Role of the Receptor for Advanced Glycation End Products (RAGE) in CF Related Airway Inflammation & CF Related Diabetes**

The risk of developing CF related diabetes (CFRD) increases with age and this can be associated with increased respiratory disease and morbidity. The causal relationship between diabetes and worsening lung function is unknown.

This relationship will be investigated via a research project assessing whether increased amounts of a specific type of molecule (RAGE) and ligands (commonly seen in diabetes and have been found in CF airway fluid) are implicated in the deterioration in lung function that is seen in individuals with CFRD.

We propose that CFRD may be associated with increased ligands and that these lead to activation of the RAGE. The RAGE activation then leads to increased lung inflammation and worsening lung function

Healthy subjects and subjects with CF, CFRD and diabetes will be assessed. Other important clinical research being conducted at LIWA and SCGH is summarised in Appendix 1.

# 4. 2013 Cystic Fibrosis Model of Care

## 4.1 What is the Cystic Fibrosis Model of Care?

The Model of Care for Cystic Fibrosis describes the WA Cystic Fibrosis Care Service (WACFCS); a model of service delivery driven by partnerships between the consumer and their family, paediatric and adult tertiary hospital services and other specialist and community-based services (including General Practice, nursing and physiotherapy services located in community settings).

Importantly, to enable sharing of health information and drive safety and quality WACFCS requires:

- Information and Communication Technology (ICT) systems
- A skilled workforce
- The capacity to offer opportunities for effective self-management by consumers and their families
- Resources to develop a culture of clinical research to drive optimal safety, quality and service efficiency.

By working towards a partnership-based Model of Service Delivery, informed by evidence and service priorities, Western Australians living with CF will receive integrated and consumer-centred care, with more services delivered closer to their home. That is, the *right care*, at the *right time*, by the *right team*, in the *right place*.

The Model of Care aligns with the WA Chronic Lung Condition Model of Care [2], which describes the common core service components of the existing respiratory Models of Care, and thus sits hierarchically above the Asthma, Chronic Obstructive Pulmonary Disease and CF Models of Care.

All of these WA Respiratory Health Network Models of Care are underpinned by four guiding principles outlined by the WA Chronic Health Conditions Framework 2011-2016 [3]:

- Integration and service coordination
- Interdisciplinary care planning and case management
- Evidence-based and consumer-centred care

### 4.2 Addressing geographic distribution of people with CF in WA

Figures 1 and 2 illustrate the geographic distribution of adults and children with CF who reside in the Perth metropolitan area and rural WA, respectively (correct at August 2012). The figures indicate that many people with CF live a considerable distance from the tertiary centres which provide care to children (PMH) and adults (SCGH) with CF. For example, Figure 1 illustrates a relatively larger proportion of people with CF living in the south-eastern (Bentley and Armadale) and northern metropolitan (Wanneroo and Joondalup) corridors. This situation underlines the need to deliver care closer to where people live. This is achievable by shifting the focus of service delivery from tertiary centres to primary care facilities, as clinically appropriate. Further, the number of people living in the community with CF is increasing, including those with complex diseases and those with less complex diseases (refer to section 6 for more detailed information). Therefore, for a sizable proportion of people with CF, particularly those with less complex disease states, health services could feasibly be delivered in community-based settings by a skilled workforce that has access to tertiary hospital support, such as through liaison services or Telehealth. Indeed, preliminary evidence is emerging to substantiate the possible benefits of this model of service delivery[24-26].

> '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)

The service delivery characteristics may necessarily vary between community-based sites, reflecting diversity in operational requirements between sites and community needs. Transition between tertiary hospital and community-based providers may be most efficiently facilitated through CF liaison teams or chronic lung conditions (CLC) teams [27], in partnership with Cystic Fibrosis WA (CFWA). These teams would be responsible for creating referral and clinical-support links between tertiary centres and community-based providers, as well as linking patients and their families with community-based services, in co-operation with CFWA. The operational attributes of the liaison teams would be determined by local requirements, particularly the volume of patients with CF in a given area. For example, CF-specific liaison team may be used in areas where a high volume of patients with CF live, while in lower volume areas, liaison may be facilitated by teams with generic chronic lung disease expertise. Table 1 outlines the potential role of a CF or chronic lung disease team (or person). An initial key responsibility of a liaison service would be to establish referral pathways across the sector which may ultimately be implemented in partnership with other organisations, such as CFWA and Medicare Locals.

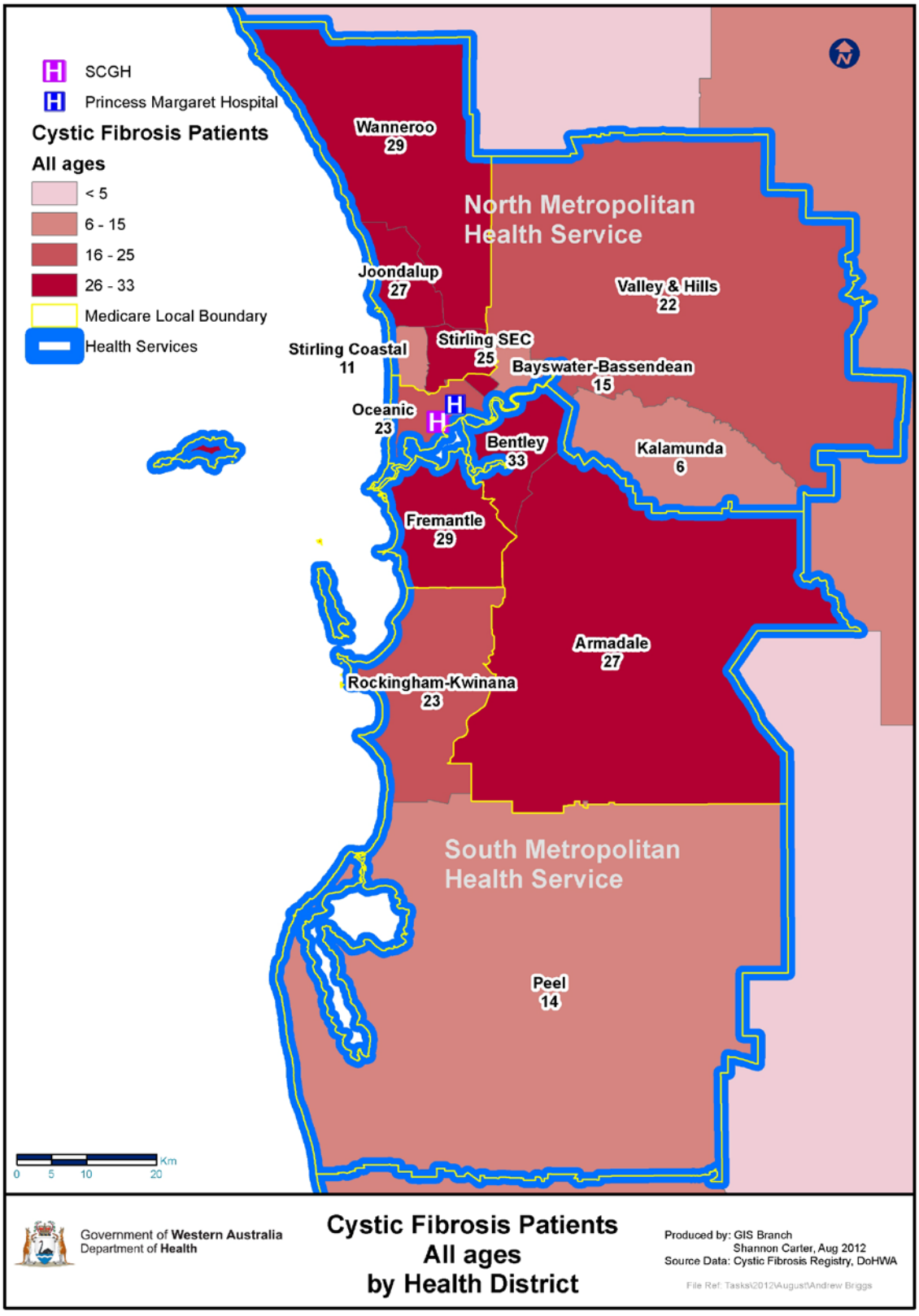



**Figure 1:** **Geographic distribution of adults and children in WA with CF in metropolitan Perth,** relative to the locations of tertiary centres and WA Health Service and Medicare Local boundaries (data correct at August 2012; source: Cystic Fibrosis Registry).

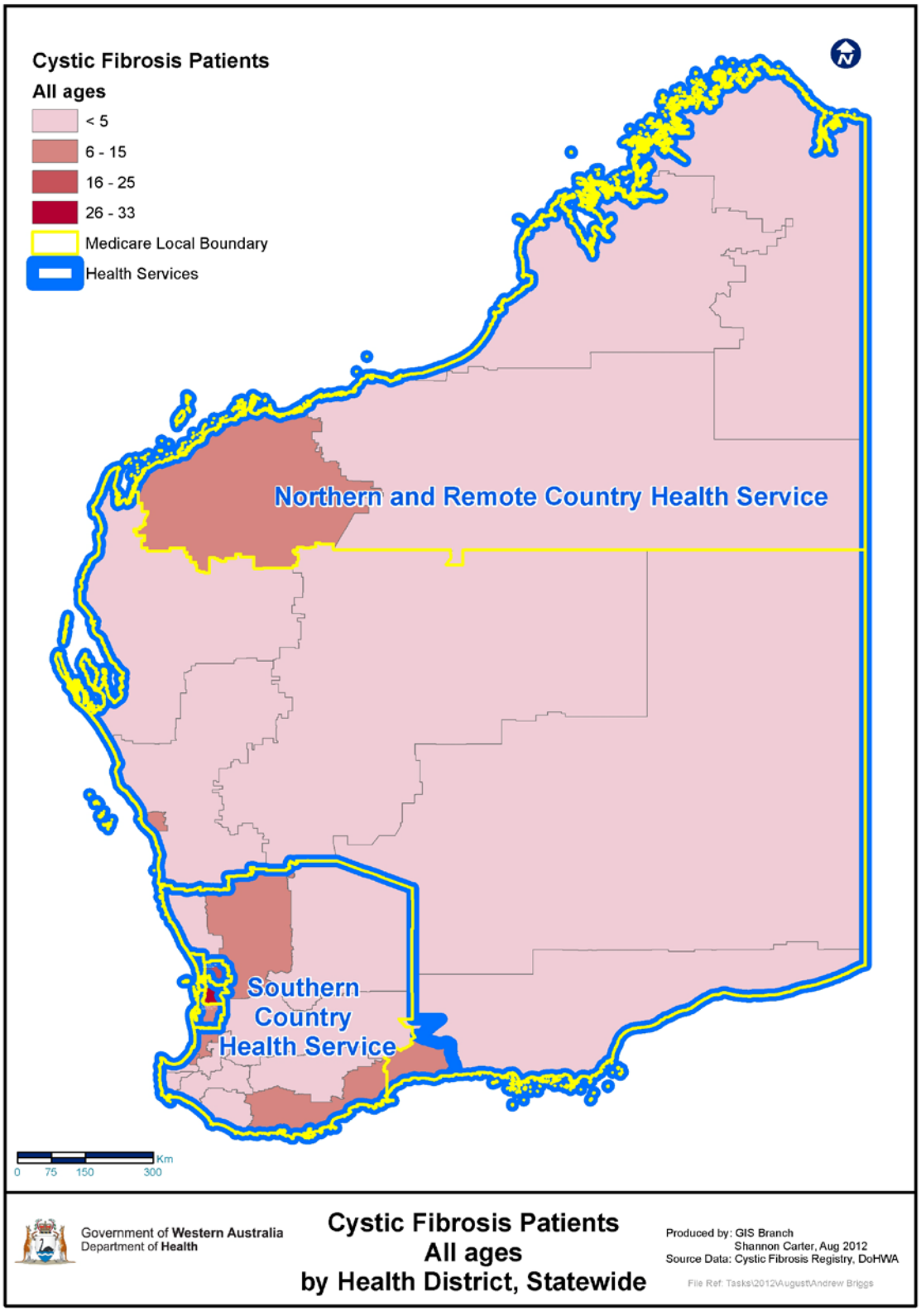



**Figure 2: Geographic distribution of adults and children in WA with CF in regional WA,** relative to WA Country Health Service and Medicare Local boundaries (data correct at August 2012; source: Cystic Fibrosis Registry).

### 4.3 A model for service delivery: The Western Australian Cystic Fibrosis Care Service (WACFCS)

In order to provide health services across the continuum of care to people with CF, the WACFCS is underpinned by a partnership model, where public (State and Commonwealth), private, and non-government service providers work collaboratively to provide care across the metropolitan and rural areas, consistent with the WA Primary Care Strategy [19] (Figure 3). The WACFCS incorporates paediatric and adult tertiary hospital services, other specialist services, and community-based care (including General Practice and other health services located in community settings). Integration between services is facilitated by liaison services (either CF-specific liaison or liaison services for chronic lung disease). Working in partnership enables the right care to be delivered in the right place and is supported by current funding directions from the Commonwealth Government, particularly for the primary care sector.

The WACFCS represents a consortium of service providers and organisations which can provide care to consumers with CF through tertiary hospital services and community-based services across the State (Figure 3). Service enablers are described in section 4.6.

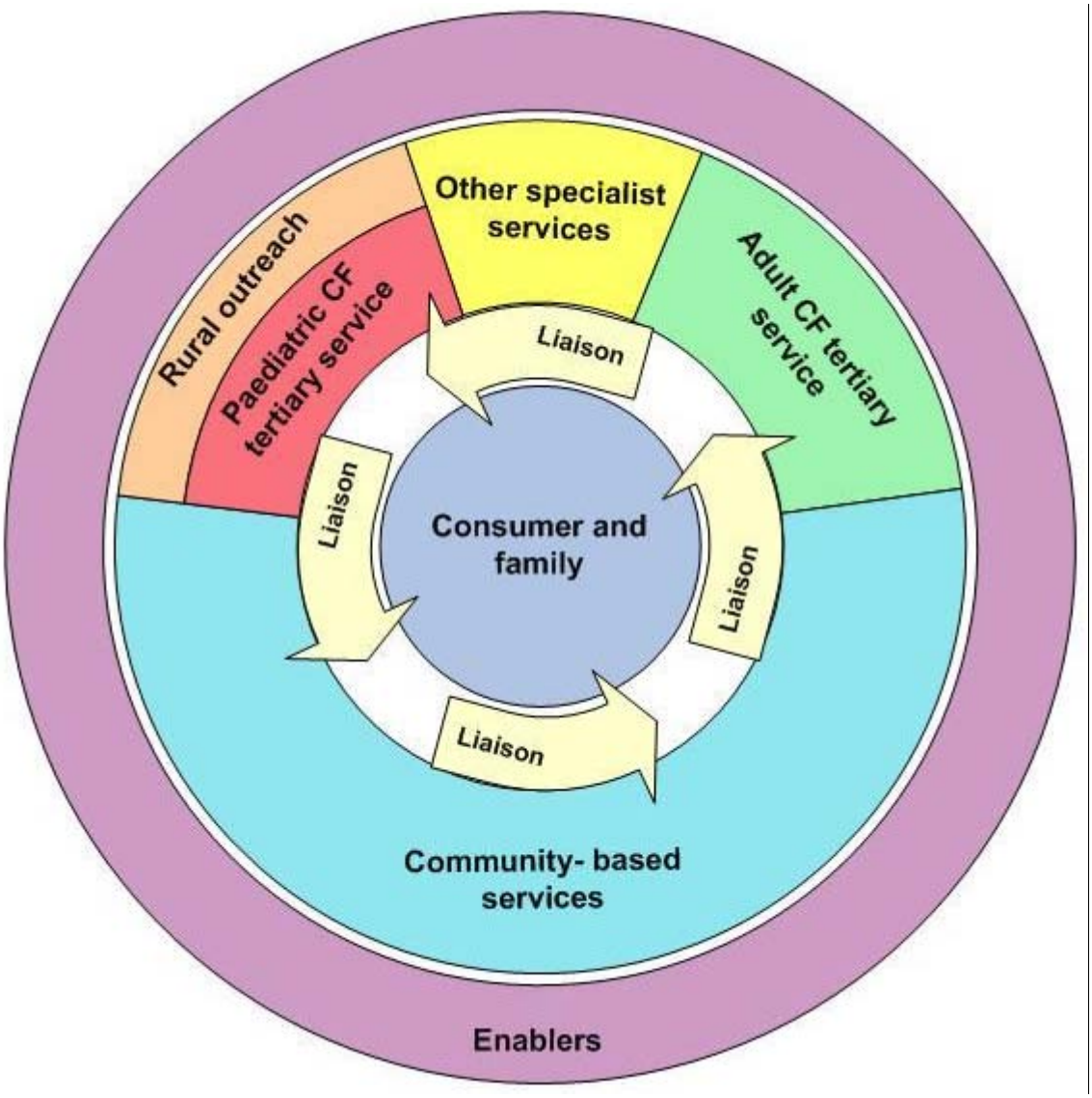



**Figure 3: Schematic of the WA Cystic Fibrosis Care Service (WACFCS).**

Table 1 outlines the partners of the WACFCS, their components and possible responsibilities, acknowledging that specific roles will be informed by local operational requirements. The premise underlying the WACFCS is that leadership in a range of areas, facilitated by a Service Director, is provided by the paediatric (PMH) and adult (SCGH) tertiary hospital services, while a large component of service delivery may occur in the community. Therefore, the WACFCS adopts a tertiary-led, but community-delivered health service model.

Key leadership activities undertaken by the tertiary centres include, but may not be limited to:

- Delivering excellence in clinical practice, teaching and research.
- Building workforce capacity among multidisciplinary professionals (medical practitioners, nurses, clinical psychologists, physiotherapists, social workers, dieticians, teachers, occupational therapists, pharmacists).
- Addressing the needs of consumers with complex conditions or who require services only available in tertiary hospital settings (e.g. transplant services, clinical genetics, and fertility specialists).
- Clinical governance: developing clinical pathways and referral pathways, policies, and practice standards for CF, in partnership with liaison services. Specifically, protocols and clinical pathways for the treatment and management should be developed and implemented based on best practice and evidence-based guidelines. Referral pathways for the clinical management of the multi-system disease associated with CF should be developed and implemented. This includes, but is not limited to, endocrinology, gastroenterology and hepatology, psychiatry, rheumatology, reproductive health, lung transplantation and interventional radiology.

Within the WACFCS, complimentary activities and services interact to provide optimal care for, and management of, people with CF. CF care involves acute treatment relating directly to the individual's daily health needs, while CF management includes a more holistic perspective including consideration of how CF may affect life decisions such as career choices, family planning and social inclusion.

**Table 1: Summary of proposed components of the WA Cystic Fibrosis Care Service**

| Partner | Components | Responsibilities |
|---|---|---|
| Consumer with CF and their family/carer | | ▪ Engage in co-care with health professionals and organisations<br>▪ Self-management, supported by the broader care team |
| Tertiary hospital service | Adult tertiary centre (SCGH), paediatric tertiary centre (PMH) and associated workforce (WACFCS Director, respiratory physicians, nurse practitioner, clinical nurse consultants, clinical psychologists, social workers, dieticians, physiotherapists, teachers, occupational therapists, pharmacists, clinical microbiologists, research coordinators) | ▪ Health service delivery for those people who require Level 6 (tertiary) care or inpatient care<br>▪ Link with other tertiary facilities (e.g. RPH transplantation services and Fiona Stanley Hospital cardiothoracic services from 2014)<br>▪ Care planning<br>▪ Facilitating co-care<br>▪ Linking with community-based services<br>▪ Outreach services to rural and remote WA<br>▪ Workforce training across the sector<br>▪ Clinical research<br>▪ Clinical governance |
| Community-based services | Cystic Fibrosis WA, General Practitioners, Community Physiotherapy Services, Allied Health professionals, community nurses, secondary hospitals, Medicare Locals, community pharmacy, RITH, HITH, Chronic Lung Conditions teams | ▪ Integrated health service delivery in community based settings, with an emphasis on self-management through the principle of 'co-care'<br>▪ Care planning, including collaborative development of management plans for acute exacerbations<br>▪ Linking with tertiary hospital services as required<br>▪ General health promotion |
| Liaison services | Health professionals with knowledge in best-practice clinical service delivery and for people with chronic lung conditions and/or CF. Liaison personnel may work across sites and organisations (e.g. hospitals, CFWA, Medicare Locals) | ▪ Referral and clinical support systems between primary care providers and specialist centres<br>▪ Link with community based support services for people with CF and their families provided by community-based agencies such as CFWA. |

### 4.4 Key service components of the WACFCS

#### 4.4.1 Co-care

Co-care care promotes service delivery, education, and self-management support to be delivered by a range of providers, such as non-tertiary hospitals, primary care organisations (e.g. General Practice), and consumer organisations like CFWA, in partnership with consumers and a tertiary centre [26]. Consistent with recommendations in the Chronic Health Conditions Framework 2011-2016, delivery of community based care for people with CF could be facilitated through the use of GP-initiated Medicare funded schemes such as the Chronic Disease Management Plan or Better Access to Mental Health Care Plan. While the number of Medicare-funded occasions of service are currently limited under these plans, these initiatives may complement care provided through the public sector.

Individuals with CF need to be provided with an approved care plan, which should be developed and reviewed annually by CF clinicians in tertiary services as a central service. Components of the care plan should involve elements of self-management and services that can be provided by community-based services.

Health service providers, particularly those in primary care, will need to be supported with professional education in order to ensure contemporary understanding of care for CF patients, available services and referral pathways. System support is also important for facilitating delivery of care (e.g. referral pathways and clinical governance frameworks). In this context, General Practice may be supported by Medicare Locals working within the WACFCS. Consumers should be supported to partner with health service providers in decision making processes that impact on their health and wellbeing and that of their families.

Ultimately, co-care arrangements are likely to promote 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. Access to hospital pharmacies for prescription medication for CF has been difficult for many Western Australians with CF. An alternative access system is to have the medicines dispensed from the hospital and use current distribution networks to deliver for pickup from a pharmacy near the person's home.

#### 4.4.2 Newborns and children

- Screening: 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. CF is diagnosed in children born before 2000 that have not been screened at birth, by mutation analysis and/or sweat test.
- Delivery of health services: Standards of Care for CF suggest that children diagnosed with CF should be transferred to the specialist paediatric tertiary centre for initial assessment as well as parental education and genetic counselling [12]. Once a comprehensive care plan its established, community-based CF clinics may be engaged for delivering care closer to home, such as follow-up appointments, as outlined in section 4.3 and Figure 3. For newborns or children requiring specialised care and access to facilities at a level 6 institution, care should be arranged at PMH.

### 4.4.3 Transition from paediatric to adult services

Transition is defined as 'the process of preparation for final transfer from paediatric to adult care systems; this transfer must be understood, ultimately, as success in aiding chronically ill children start a productive life and achieve social integration as adults' [1].

Transition planning between paediatric and adult tertiary hospital systems, as well as transition planning to community-based services, is important. The transition process from the CF paediatric/adolescent service to adult service is critical to effective management and long term health outcomes for people with CF [28,29].

There is increasing recognition in health literature of the importance of managing the transition process in a holistic and sensitive manner that meets the complex needs and demands of the consumer and their family/carers. The Paediatric Chronic Diseases Transition Framework [30] (PCDTF) provides a guide to transition planning for children with chronic health conditions and their families. The PCDTF outlines five guiding principles to inform transitional care:

1. Planned and coordinated care
2. Readiness for transition
3. Ownership of transition by the young person
4. Shared responsibility by all involved in the transition
5. Accessibility and availability of services.

The development of an implementation plan for the PCDTF is a priority for the Child and Youth Health Network in 2013. A working party is expected to be convened to design a system wide approach to transition which will inform CF transition services across the three stages of health care transition: active preparation, active transition, and integration.

The WACFCS team should be trained and skilled in working with children and adolescents to ensure they have an understanding and knowledge of the transition process. A planned transition initiated and implemented in early adolescence (estimated age 12 years based on available models) in partnership with patients and their family/carer, should 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, facilitated through the use of a 'transition check list'.
- Education and vocational support (MDT, school teacher, community organisations and peers as appropriate):
  - Promotion of healthy lifestyles
  - Understanding of CF and navigating the health care system, including resources available in the community
  - Maximising educational and vocational opportunities.
- Counselling and psychosocial support (social workers, psychologist, clinical nurse consultant, genetic counsellors):

† MDT: multidisciplinary team

- Sexual health, fertility and pregnancy planning
- Psychosocial morbidity
- Facilitating access to health care and community support networks (e.g. peer 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, GP and other health professionals as appropriate.
  - CFWA and WACFCS adult services staff.

## Transition Planning

A coordinated approach towards transition planning is essential for optimising the health outcomes of adolescents transitioning towards adult-based tertiary hospital and community services. Figure 4 illustrates three key processes involved in this approach: planning, young adult-focused care and surveillance and monitoring.

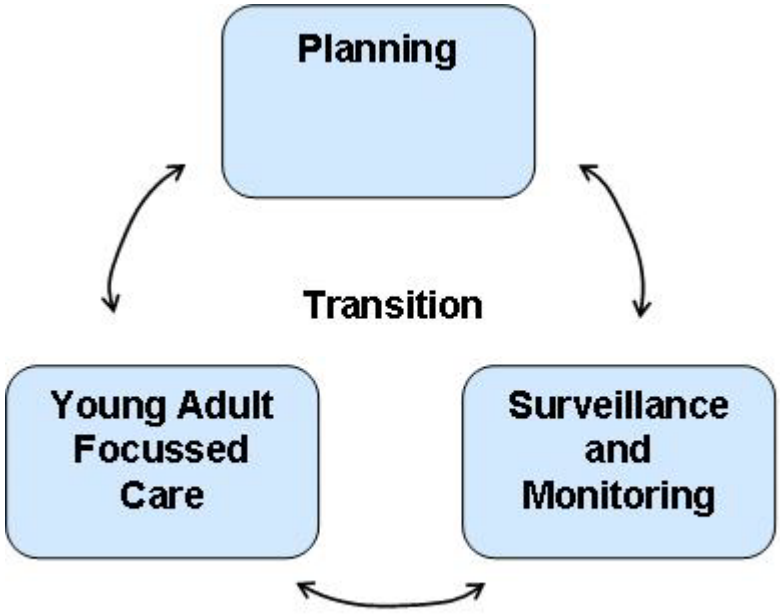



**Figure 4:** **Transition from paediatric to adult services.** This process should include a coordinated approach towards transitioning between paediatric and adult services in WA (planning); tertiary care aimed at the adolescent/young adult with CF (young-adult focussed care) and, holistic surveillance and monitoring of the disease by the patient and multidisciplinary team (MDT).

A coordinated transition process should be established between the WACFCS paediatric and adult teams, community-based services and CFWA. This should include:

- Joint transition outpatient clinic
  - Introduction to adult CF MDT
- Medical summary transition
  - Detailed medical summary prepared for the adult service prior to transition from the paediatric service

- As people become more reliant on their General Practitioner on transition to adult care, a copy of the medical summary should also be provided to the patient's General Practitioner.

- Psychosocial support including:
  - Generic and CF-specific support
  - Consumer information/education and directory of services
  - Individually tailored adult mentoring and support services
  - Reproductive health education.

For the purposes of transitioning case management for adolescents from PMH to SCGH, there is a transition meeting at PMH for the patient and their family/carer to meet the adult team. A tour of SCGH is conducted by CFWA for the patient and their family/carer. While case management is transitioned between tertiary CF centres, the delivery of care should occur, where possible, in community settings, consistent with the model of service delivery described in this Model of Care.

Booklets on the process and implications of transition are available at PMH and SCGH.

**Post-transition young adult focussed care**

At the tertiary hospital level, the adolescent/young adult CF service (18 – 24 years) is provided by the WACFCS adult service based at SCGH. The service takes into account the significant developmental and social factors facing young adults with a chronic illness. Whether services are provided from a tertiary hospital or community-based facility, the site(s) should be youth-friendly and adopt flexible booking and attendance times, and include the following key components:

**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, clinical nurse consultant)
- Detection of psychosocial co-morbidity (GP, psychology, social work, respiratory physicians)
- Promotion of healthy lifestyles (GP, CFWA, Quit, MDT)
- Vocation and employment opportunities (Occupational Therapist, Social worker, CFWA, Centre-link)
- Fertility and pregnancy planning counselling
- Assisted reproduction technology

Where hospitalisation is required, 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.

> '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)

### 4.4.4 Services for adults

- The demand for adult CF services continues to increase owing to increased longevity of adults with CF, with most Australian centres struggling to meet published standards of care. The number of Western Australians with CF is increasing, including adults with complex disease presentations and those with less complex disease. The CF Model of Care proposes a partnership between community and tertiary providers in order to deliver these standards of care. The workforce capacity needs to be reviewed in both community and tertiary settings in order to ensure that appropriate standards of care are available to all patients with CF regardless of service setting. Australian CF Standards of Care recommend specific ratios of patients to health professionals in order to deliver care of adequate safety and quality [12]. Tertiary-based CF centres have a key role in helping to build the capacity of the community-based workforce to work in a co-care model of service delivery with clinicians based at tertiary CF centres but the transition will require investment in both sectors for this to be accomplished. Furthermore, as patients grow older the focus of care will inevitably shift to include a broad range of services that are currently only available in tertiary hospitals.

As more adults with CF engage in study and work, delivery of care close to home becomes increasingly important, in order to minimise disruptions to lifestyles associated with relying on care from tertiary centres only. For example, data from the 2008 Australian Cystic Fibrosis Data Registry (ACFDR) indicate that 9.8% of people with CF had a university degree and 37.7% of people were engaged in full-time paid employment. Three years on, the 2011 ACFDR data report an increase to 14.9% and 40.1% in those aforementioned areas, respectively [6,31].

### 4.4.5 Self-management and self-management support

Self-management and self-management support represent a fundamental component of a client and family/carer-centred Model of Care. To enable effective co-care through self-management, comprehensive information should be provided to enable families to adopt positive health habits for children and adolescents with CF. Both Cystic Fibrosis WA and the tertiary centres have information resources regarding effective self-management and health promotion strategies. Adopting a family/carer-based, wellness model of care, supported by the care team, promotes proactive care rather than acute, reactive care. There is some evidence to support the use of self-management education for improving behaviour and knowledge in patients with CF and their caregivers [32].

### 4.4.6 Outreach services to rural WA

While children and families living in rural and remote areas may still need to attend appointments at tertiary centres, some may have limited access to outreach clinics coordinated from the PMH Respiratory Specialist Clinic. Outreach clinics are currently held in Broome, Bunbury, Kalgoorlie, Port Hedland and Karratha and will receive ongoing support using telehealth. The outreach service includes a doctor, nurse, physiotherapist or dietician and with an emphasis on engagement of local community based services for clients with CF.

For rural and remote adult CF patients, telehealth conferences are scheduled for Albany, Broome, Karratha and Geraldton.

### 4.4.7 Lung transplantation

The Royal Perth Hospital Lung Transplant Unit provides the transplantation service for all patients in WA with CF. Patients with CF currently comprise one quarter of indications for lung transplantation (RPH Lung Transplantation Service, 2011). At present there is limited access to microbiological pathology services in comparison to other transplant centres. Consideration should also be given to the provision of palliative care services for people with CF awaiting lung transplantation [33] (see 4.4.8). Western Australia's principal cardiothoracic service will be based out of Fiona Stanley Hospital (FSH) from 2014/15.

### 4.4.8 End of life and palliative care

The declining health of people with CF, including those people awaiting lung transplantation, should be managed in a holistic and sensitive manner to meet their complex needs and those of their families. Sensitive initiation and discussion of advance care planning for end of life care, including initiation or review of Advance Health Directives (AHD) and/or Enduring Power of Guardianships, should be provided as needed.

The provision of information and education for consumers and families about the role of palliative care is critical. Palliative care aims to improve the quality of life of patients and families by providing pain and symptom relief, and spiritual and psychosocial support [34] [35]. In the case of CF, it is recognised that there can be tension between providing active treatment in the hope of transplantation versus palliative care for declining health. This can raise conflicting expectations and demands for the person with CF, their families, and the WACFCS team.

Six key components that should be addressed in the delivery of end of life and palliative care services for people with CF and their families include[‡]:

- Information
- Open and honest communication
- Pain and symptom management
- Psychological support
- Family/carer inclusion
- Timing

Planning for palliative care services should be undertaken among those with advanced CF, their family/carer and the WACFCS team, with support from a palliative care team [33].

Consumers and families expect open and honest communication about end of life issues, including having written information available about what to expect and care options. The palliative care plan should identify the range of psychological and psychosocial support services appropriate to the needs of the individual, family/carer, as well as access to support services for families of people with CF as a crucial aspect of the palliative care plan.

Families are often confronted with life threatening health crises where the person with CF has survived, or where there has been recent 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

[‡] "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

of end of life issues when there is declining health and referral to the transplant team (Figure 5). Written information should be developed that encourages discussion regarding end of life issues. The WA Palliative Care Model of Care [21], developed by the WA Cancer and Palliative Care Network, will inform the development of an appropriate palliative care pathway for people with CF and their families. The use of Advance Health Directives and enduring guardians in end-of-life planning for people with CF should be promoted. The Liverpool Care Pathway is a best practice tool that supports care of the dying patient and is being implemented in health care settings across [20].

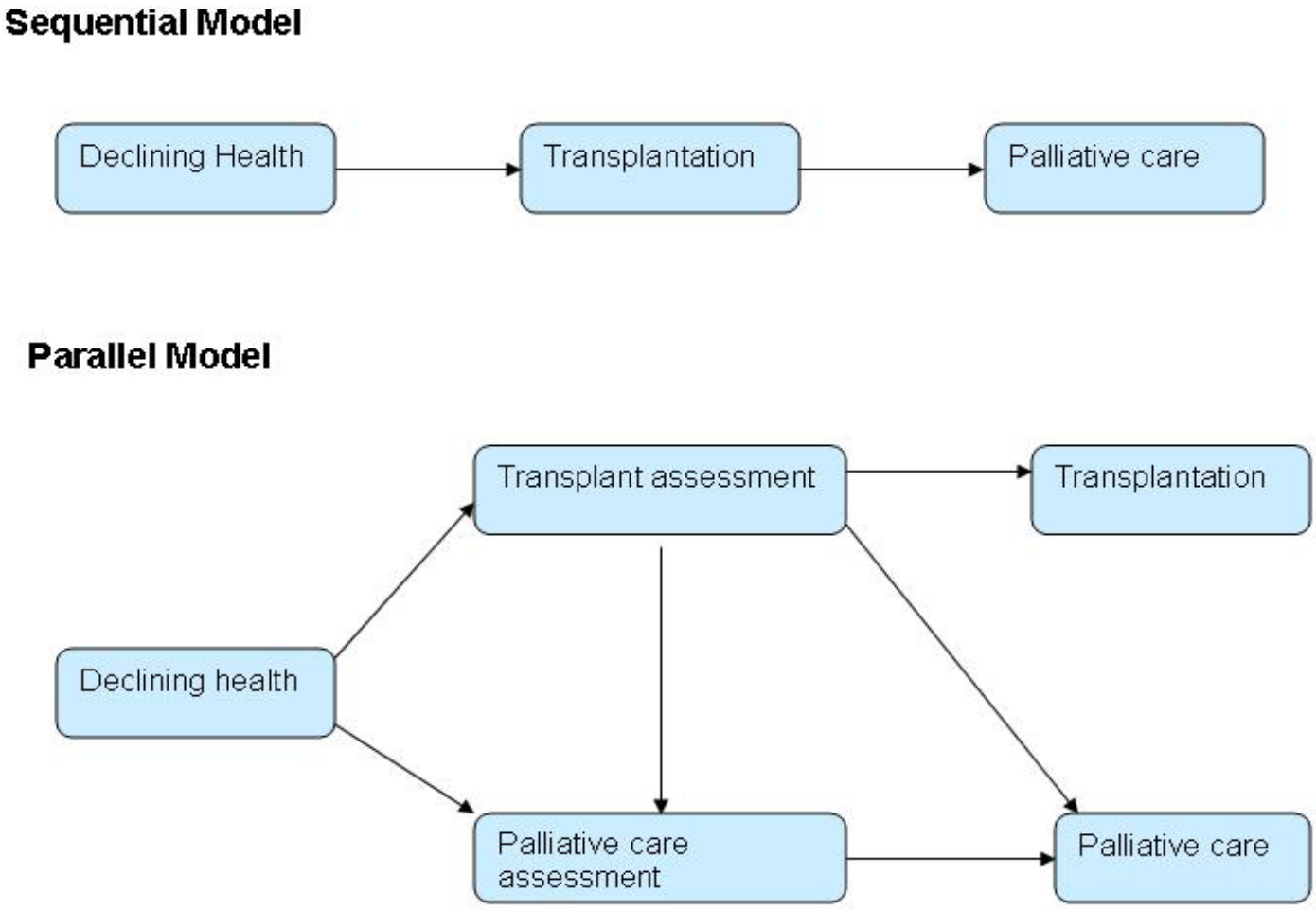
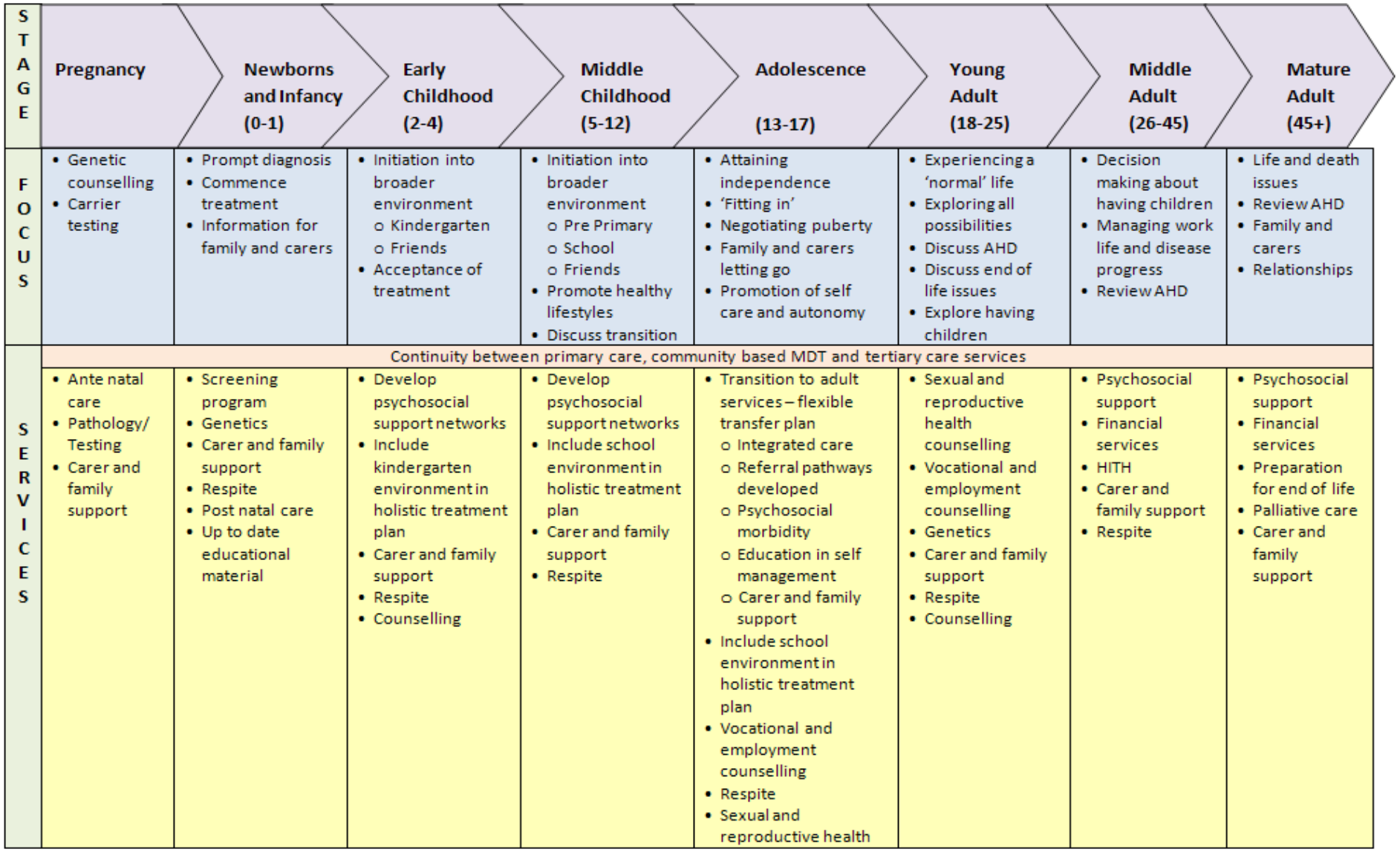



**Figure 5: Palliative care models for declining health and transplantation in people with CF.**

### 4.5 The WACFCS consumer-centred service delivery pathway

The aim of the WACFCS is to provide care across the lifespan for people with CF and across the continuum of care for the condition; addressing the biological and psychosocial needs of patients with CF and their families/carers, including end of life and palliation services. Figure 6 outlines the typical consumer pathway and key services that should be provided or facilitated by WACFCS across the typical course of the disease. A consumer pathway, consistent with the Cystic Fibrosis Standards of Care, Australia [12] highlights the need for:

- Specialist centre care, usually provided at a tertiary facility
- Clinical care, usually provided in a community-based facility
- Provision of interdisciplinary health services
- Access to lung transplant services



| STAGE | Pregnancy | Newborns and Infancy (0-1) | Early Childhood (2-4) | Middle Childhood (5-12) | Adolescence (13-17) | Young Adult (18-25) | Middle Adult (26-45) | Mature Adult (45+) |
|---|---|---|---|---|---|---|---|---|
| FOCUS | • Genetic counselling<br>• Carrier testing | • Prompt diagnosis<br>• Commence treatment<br>• Information for family and carers | • Initiation into broader environment<br>o Kindergarten<br>o Friends<br>• Acceptance of treatment | • Initiation into broader environment<br>o Pre Primary<br>o School<br>o Friends<br>• Promote healthy lifestyles<br>• Discuss transition | • Attaining independence<br>• 'Fitting in'<br>• Negotiating puberty<br>• Family and carers letting go<br>• Promotion of self care and autonomy | • Experiencing a 'normal' life<br>• Exploring all possibilities<br>• Discuss AHD<br>• Discuss end of life issues<br>• Explore having children | • Decision making about having children<br>• Managing work life and disease progress<br>• Review AHD | • Life and death issues<br>• Review AHD<br>• Family and carers<br>• Relationships |
| SERVICES | Continuity between primary care, community based MDT and tertiary care services | | | | | | | |
| | • Ante natal care<br>• Pathology/ Testing<br>• Carer and family support | • Screening program<br>• Genetics<br>• Carer and family support<br>• Respite<br>• Post natal care<br>• Up to date educational material | • Develop psychosocial support networks<br>• Include kindergarten environment in holistic treatment plan<br>• Carer and family support<br>• Respite<br>• Counselling | • Develop psychosocial support networks<br>• Include school environment in holistic treatment plan<br>• Carer and family support<br>• Respite | • Transition to adult services – flexible transfer plan<br>o Integrated care<br>o Referral pathways developed<br>o Psychosocial morbidity<br>o Education in self management<br>o Carer and family support<br>• Include school environment in holistic treatment plan<br>• Vocational and employment counselling<br>• Respite<br>• Sexual and reproductive health | • Sexual and reproductive health counselling<br>• Vocational and employment counselling<br>• Genetics<br>• Carer and family support<br>• Respite<br>• Counselling | • Psychosocial support<br>• Financial services<br>• HITH<br>• Carer and family support<br>• Respite | • Psychosocial support<br>• Financial services<br>• Preparation for end of life<br>• Palliative care<br>• Carer and family support |

**Figure 6: Pathway for consumers with CF and their families, with focus areas and health services by life course stage.**

AHD = Advance Health Directive.

### 4.6 Enablers for the WACFCS

A range of support systems are required to ensure the effective functioning of the WACFCS and the feasibility to deliver the proposed service pathway, as outlined it the WA Chronic Health Conditions Framework [36].

#### 4.6.1 Audit and monitoring through the national CF registry

Comprehensive data and data collection methods are critical to informing performance and quality. WA contributes to the national CF data registry, maintained by Cystic Fibrosis Australia (CFA). CFA maintain a data registry for CF patients in all states in Australia. Annual reports are published and contain more that ninety percent of people with CF. A committee, comprised of CF physicians from around Australia, meets regularly with the registry directors to ensure the data registry reflects useful information. For example, it was agreed that the clinics are now identified in the public report. This will assist clinics to look across Australia for best practice in managing CF and encourage quality improvement across the clinics. CFA is further developing the registry to accommodate the inclusion of quality of life data and investing in the management and development of the registry to support research, regular auditing and monitoring health outcomes for people with CF. Data specific to WA can be extracted upon request.

#### 4.6.2 ICT solutions

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 (e.g. blood test results and spirometry) that would be updated as results become available. Issues of confidentiality and privacy are paramount in the development of any such system. This system would facilitate audit and development of key performance indicators. One possible system is the Smart Health platform, already used in other states for the online sharing of electronic health records. Smart Health enables both the patient and health professional to access data. Alternatively, client-held e-records could be utilised to further empower people with CF and their families to actively participate in co-care. The national Personally Controlled Electronic Health Record (PCEHR) system may be a suitable tool to enable people with CF and their families to achieve this.

Strategies to facilitate communication across the continuum of care including discharge summaries and the development of integrated care plans should be developed in consultation with primary care providers, liaison teams and GPs.

Consumers also require ICT solutions to provide improved access to information and services, particularly those individuals who live in rural and remote WA. Access to reliable consumer-focused websites (e.g. CFWA), SMS reminders for appointments, and smart phone ‘apps’ to facilitate self-management, are examples of possible ICT strategies to encourage greater participation in co-care [37]. ICT can also provide consumers with increased social communication opportunities, improving psychosocial networks and reducing the risk of isolation.

#### 4.6.3 Research

PMH has a strong track record in clinical and basic science research related to CF. The WACFCS should foster a culture of research as well as regular audits across paediatric and adult services. This will ensure not only the best treatment is available for consumers, but will 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. For example, randomised control trials are difficult given the nature and progress of the disease and require the development of clinical trial networks nationally and internationally. Most trials are conducted on adult cohorts and none have investigated the feasibility and health benefits of preventing the consequences of lung disease in early CF. A critical mass of research in prevention of lung disease in people diagnosed with CF during their first few years of life is needed.

Health services research is also critical to evaluate consumer satisfaction and the clinical and economic outcomes of different models of service delivery. The National Health and Medical Research Council (NHMRC) Partnerships for Better Health funding scheme provides an opportunity to evaluate policy-relevant health services research and forge links between researchers and decision-makers.

### 4.6.4 Workforce development

Excellence in service delivery is underpinned by an experienced and flexible 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.' (Adult with CF)

To engage with the community a skilled and sustainable workforce relies on:

- WACFCS having a core group of skilled and expert clinical staff including medical specialists, nursing, allied health, pharmacy and support staff;
- Sufficient numbers of skilled practitioners to provide services that meet appropriate standards of care;
- Shared/joint appointments to ensure quality of care across all health care sites;
- Telehealth and support for staff working in rural WA and General Practice;
- Increased community-based allied health staff;
- Specialist community and ambulatory services within metropolitan and rural WA; and
- Education and training of all health professionals involved in the care of people with CF in the delivery of shared CF health care in non-tertiary centres and the community.

### 4.6.5 Consumer and Family/Carer Involvement

At the centre of the WACFCS is the consumer and their family/carer. In order to provide a model of care that meets the needs and preferences of consumers, individuals with CF and their families/carers need to be involved in the planning of services, development of policy and advocating for CF issues. Structured opportunities should be provided to consumers and carers/families to comment on and identify areas within the care pathway (Figure 6) that could be improved. This patient-led evaluation will enhance understanding and usage of the services available.

# 5. Key recommendations

**Recommendation 1:**

Establish linkages between CF tertiary hospital services and community-based organisations (e.g. General Practices, Medicare Locals, GP Super Clinics, Cystic Fibrosis WA and SilverChain) to facilitate the transition of health services from tertiary services to the community in a partnership model. An evaluation of consumer-centred, clinical and economic outcomes should be undertaken to judge the safety and effectiveness of this model of service delivery in WA.

**Recommendation 2:**

Develop clinical governance frameworks utilising evidence-based guidelines and protocols to facilitate safe and effective tertiary-led delivery of care in community settings.

**Recommendation 3:**

Develop referral pathways to guide the delivery of tertiary-led care in the community sector, with a focus on:

- Transition from paediatric and adolescent services to adult services
- Provision of services to rural and remote regions
- Transplantation services
- Interdisciplinary care
- Palliative care.

Delivery of care in the community sector may be facilitated through implementation of a CF-specific or chronic lung disease liaison team to provide a link between tertiary CF centres and community-based centres.

**Recommendation 4:**

Where possible, decentralise pharmacy and dispensing of medication to community and local hospital pharmacies using a pharmacy network.

**Recommendation 5:**

Promote 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. Smart Health Solutions or the Personally Controlled Electronic Health Record (PCEHR) system may be an appropriate platform in this context. Both patients and health professionals should have access to the stored data.
- Provide consumers with access to reliable and valid information about CF and self-management, including access to innovative tools such as smart-phone 'apps'.
- Improved access to services for consumers, families/carers and health professionals in rural and remote regions.

**Recommendation 6:**

Continue to build workforce capacity (clinical skills and knowledge) across the sector, especially in primary care – in collaboration with CF centres, CFWA, and Universities. Professional development opportunities should be offered within an interprofessional framework that acknowledges the critical role of co-care and self-management.

**Recommendation 7:**

Promote a culture of research and evaluation by:

- Undertaking research at a state level, including research that examines safety, efficacy and efficiency of alternative models of health service delivery
- Participating in national and international research, particularly in clinical trials.
- Continuing to contribute data to the National CF Registry.

**Recommendation 8:**

Undertake a review of CF centres in WA to identify activity over the last five years which aligns with the 2007 Model of Care and explore opportunities to facilitate implementation of the 2013 Model of Care recommendations.

# 6. Epidemiology

## 6.1 The national picture

The Australian Cystic Fibrosis Data Registry (ACFDR) is a national registry which captures data regarding people diagnosed with CF throughout Australian CF centres and hospitals. It is estimated that data from the 2009 ACFDR has about 90% coverage within Australia, leading to some underrepresentation of patients with CF [38]. Data from the ACFDR in 2011 registered 3133 people in Australia living with CF [6]. Assuming 90% coverage, approximately 3481 people with CF currently live in Australia.

Although CF is one of Australia's most common inherited genetic conditions, the incidence of CF is expected to decline in coming years [5]. For example, a Victorian study utilising ten years of data from 1989 - 1998 estimated the incidence of CF to be approximately 1 in 2800 live births [39] whilst it has more recently been reported as 1 in 2986 (95% CI, 2735-3288) births, as calculated over 5 years of ACFDR data from 2004 to 2008 [38]. Despite a decreasing incidence, the prevalence of CF (mild and severe disease severity) is expected to remain relatively high, owing to increased life expectancy for people with CF, including people with complex and less complex CF conditions. Australian data supports the trend for increased longevity in people with CF and the need for services for people with CF in later years of life [5,8]. It is projected that the life expectancy at birth for Australians born with CF between 2003 - 2005 is 38 years [40], this aligns with international estimates of life expectancy between 34 - 47 years [8]. In contrast, in the 1970s life expectancy of individuals with CF was predicted at only 16 years of age [41].

Despite increasing life expectantly among people with CF, disease severity generally increases with age. For example, Figure 7 illustrates the proportion of patients with CF in five disease severity categories, based on forced expiratory volume ($FEV_1$), which is considered a key measure of quality and length of life for people with CF [42]. This model is based on three years of ACFDR data from 2003 - 2005 and predicts the life expectancy at birth to be 38 years of age for this cohort. At 45 years of age approximately two thirds of people with CF have died. Of the remaining cohort, approximately 10% have received a transplant, with 1%, 15%, and 8% in severity categories 1, 2 and 3, respectively[^]. Table 2 illustrates increasing lung function impairment by age and by sex for Australian people with CF. Only 8.9% of people with CF at age 30 or above have a normal range $FEV_1$ (≥90% $FEV_1$) [6].

[^] Disease severity categories: Severity 1= mild disease where $FEV_1\% \geq 70$; Severity 2= moderate disease where $30 \leq FEV_1\% < 70$; Severity 3= severe disease where $FEV_1\% < 30$; Severity 4= where a patient has received a heart and/or lung transplant; Severity 5= where a patient has died.

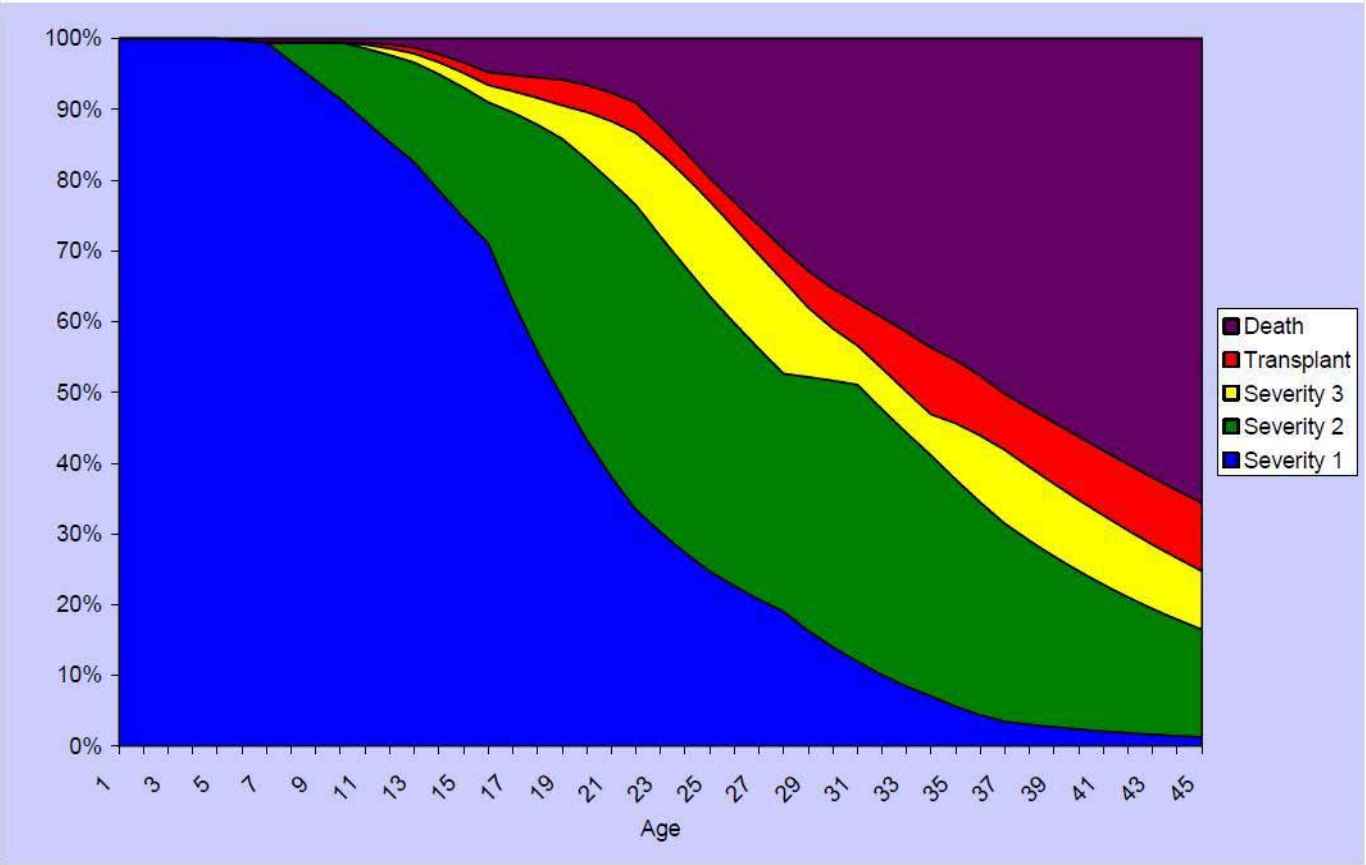



**Figure 7: Proportion of patients in each disease severity category by age.**

Severity 1= mild disease where FEV1% ≥70; Severity 2= moderate disease where 30 ≤ FEV1% <70; Severity 3= severe disease where FEV1% <30; Severity 4= where a patient has received a heart and/or lung transplant; Severity 5= where a patient has died. (Reproduced with permission from University of Technology Sydney, 2011)

**Table 2: Lung function impairment by age and by sex (Source: ACFDR 2011)**

| | Severe[1] | Moderate[2] | Mild[3] | Normal[4] | Total | Severe[1] | Moderate[2] | Mild[3] | Normal[4] | Total |
|---|---|---|---|---|---|---|---|---|---|---|
| | Number | | | | | Per cent | | | | |
| Age/sex group (years): | | | | | | | | | | |
| 6 - 11 | 1 | 40 | 133 | 331 | 505 | 0.2 | 7.9 | 26.3 | 65.5 | 100.0 |
| 12 - 17 | 13 | 70 | 184 | 201 | 468 | 2.8 | 15 | 39.3 | 42.9 | 100.0 |
| 18 - 29 | 102 | 297 | 228 | 142 | 769 | 13.3 | 38.6 | 29.6 | 18.5 | 100.0 |
| 30+ | 111 | 223 | 95 | 42 | 471 | 23.6 | 47.3 | 20.2 | 8.9 | 100.0 |
| Total measured | 227 | 630 | 640 | 716 | 2213 | 10.3 | 28.5 | 28.9 | 32.4 | 100.0 |
| Males | 140 | 325 | 344 | 371 | 1180 | 11.9 | 27.5 | 29.2 | 31.4 | 100.0 |
| Females | 87 | 305 | 296 | 345 | 1033 | 8.4 | 29.5 | 28.7 | 33.4 | 100.0 |

[1]$FEV_1$ <40%; [2]$FEV_1$ 40≥70%; [3]$FEV_1$ 70≥90; [4]$FEV_1$ >90%.

### 6.2 National health service utilisation

Based on ACFDR data from 2003-2005, the estimated direct cost of 3000 patients with CF is around $67 million to the health system (distribution: inpatient costs, 58%; pharmaceuticals, 29%; medical services, 10%; complications, 2%; and diagnostic tests 1%) [40]. Data from the 2011 ACFDR indicate that 49% of persons with CF who have been hospitalised accumulated at least 14 admitted days through the year. The mean length of stay for these patients was 22 days (median = 13), indicating that people with CF spend considerable time hospitalised [6].

### 6.3 State epidemiology

In Western Australia, 339 people were identified in the ACFDR 2011 dataset, representing 10.8% (339/3133) of the total number of people in Australia living with CF [6]. The average age of Western Australian people with CF for 2010 was similar to the ACFDR 2009 dataset – 18.8 years for males and 18.4 years for females (state: average age = 18.6 years; median age = 18.5 male vs. 17.8 female). This was lower than the Australian overall average of 19.0 years at 31 December 2010 [10]. Table 3 summarises the age and sex of ACFDR 2010 registrants both nationally and in WA. The proportion of males to females was greater both nationally and in WA, however, across age groups this trend varied. These data are similar for the 2009 ACDR dataset [43] and similar to European Union country CF registers as reviewed by Salvatore et al. [44].

**Table 3: National and WA age and sex of registrants at 31 December 2010 (collated from national ACFDR 2010 and WA ACFDR 2010 reports)**

| | National data registry | | | Western Australian data registry | | |
|---|---|---|---|---|---|---|
| **Age** | **Male** | **Female** | **Total** | **Male** | **Female** | **Total** |
| 0-4 years | 236 (15%) | 223 (16%) | 459 (15%) | 30 (17%) | 25 (16%) | 55 (16%) |
| 5-9 years | 222 (14%) | 218 (15%) | 440 (14%) | 23 (13%) | 30 (19%) | 53 (16%) |
| 10-14 years | 214 (13%) | 219 (15%) | 433 (14%) | 19 (11%) | 21 (13%) | 40 (12%) |
| 15-19 years | 212 (13%) | 187 (13%) | 399 (13%) | 22 (13%) | 13 (8%) | 35 (10%) |
| 20-24 years | 215 (13%) | 174 (12%) | 389 (13%) | 31 (18%) | 18 (11%) | 49 (15%) |
| 25-29 years | 200 (12%) | 161 (11%) | 361 (12%) | 16 (9%) | 25 (16%) | 41 (12%) |
| 30-34 years | 110 (7%) | 96 (7%) | 206 (7%) | 13 (8%) | 10 (6%) | 23 (7%) |
| 35-39 years | 76 (5%) | 79 (6%) | 155 (5%) | 2 (1%) | 11 (7%) | 13 (4%) |
| 40-44 years | 73 (4%) | 36 (3%) | 109 (4%) | 10 (6%) | 5 (3%) | 15 (4%) |
| 45+ years | 69 (4%) | 43 (3%) | 112 (4%) | 7 (4%) | 3 (2%) | 10 (3%) |
| Total population | 1627 (100%) | 1436 (100%) | 3063 (100%) | 173 (100%) | 161 (100%) | 334 (100%) |

In WA as at 31 December 2011, the proportion of adolescents and adults (18 years or older) with CF was approximately equal (n = 169 (49.9%) vs. n = 170 (50.1%), respectively). The adult proportion of people with CF is higher in WA than the national average of 48.8%. National ACFDR data from 2000 support the trend of an ageing population with CF, indicating a proportion of only 34.7% adults with CF at that time [6]. In 2008 the ACFDR registry recorded Western Australia and Victoria as the only two

states with adult CF populations >50% of the total number of people with cystic fibrosis in the state [31]. As of 2011, the proportion of adults per state has grown to include South Australia and Tasmania [6].

### 6.4 Hospital utilisation in Western Australia

Table 4 illustrates for financial years 2008/09 – 2010/11, the majority of patients with CF living in the North Metropolitan Health Service (NMHS), South Metropolitan Health Service (SMHS) and WA Country Health Service (WACHS) sought hospital care from NMHS. The major tertiary centres for adult and paediatric care are SCGH (NMHS) and PMH (Child and Adolescent Health Service), respectively, which are both within the geographic area covered by NMHS. There was no trend in the rate of hospitalisation during 2008/09 – 2010/11 (see Table 5). The utilisation of NMHS for CF is also demonstrated across all age groups 0 - 6 years, 7 - 12 years, 13 - 18 years and 19 years and older (see Table 6).

**Table 4: Total separations, principal and additional diagnosis for CF, all ages, health service of hospital by patient's health service of residence, WA, 2008/09–2010/11 (Source: Epidemiology Branch, WA Department of Health; HDMS)**

| | North Metropolitan HS | South Metropolitan HS | WA Country Health Service | Total |
|---|---|---|---|---|
| HS of hospital | | | | |
| NMHS[1] | 518 | 489 | 194 | 1,201 |
| SMHS[2] | <5 | 6 | <5 | 9 |
| SCHS[3] | - | - | 32 | 32 |
| NRCHS[4] | - | - | 15 | 15 |
| Total | 521 | 495 | 241 | 1,257 |

[1]North Metropolitan Health Service; [2]South Metropolitan Health Service; [3]Southern Country Health Service; [4]Northern Remote Country Health Service

**Table 5: Total separations, principal and additional diagnosis for CF by age group, WA, 2008/09–2010/11 (Source: Source: Epidemiology Branch, WA Department of Health; HDMS)**

| | Rates per 100,000 persons | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|
| Age Group | 0-6 | 7-12 | 13-18 | 19-85+ | Crude | ASR | LCI | UCI | ASR S.E. |
| 2008/2009 | 57.0 | 17.6 | 43.1 | 9.6 | 17.4 | 17.5 | 15.8 | 19.3 | 0.9 |
| 2009/2010 | 62.3 | 36.5 | 35.8 | 11.6 | 20.2 | 20.5 | 18.6 | 22.4 | 1.0 |
| 2010/2011 | 58.1 | 25.9 | 43.8 | 11.0 | 19.1 | 19.4 | 17.6 | 21.2 | 0.9 |
| Total | 59.1 | 26.7 | 40.9 | 10.8 | 18.9 | 19.2 | 18.1 | 20.2 | 0.5 |

The rate of hospitalisation of people with CF in WA showed no trend (P = 0.17). ASR = age standardised rate per 100,000 persons; Crude = crude rate per 100,000 person years; LCI = lower 95% confidence interval; UCI = upper 95% confidence interval; ASR S.E. = age standardised rate standard error.

**Table 6: Total separations, principal and additional diagnosis for CF by age group and health service of hospital, WA, 2008/09–2010/11 (Source: 2011 ACFDR)**

| Health Service | 0–6 years | 7–12 years | 13–18 years | 19 years and older | Total |
|---|---|---|---|---|---|
| NMHS[1] | 352 | 133 | 220 | 507 | 1,212 |
| SMHS[2] | <5 | <5 | <5 | 6 | 9 |
| SCHS[3] | <5 | <5 | <5 | 22 | 32 |
| NRCHS[4] | 6 | <5 | <5 | 6 | 15 |
| Total | 364 | 138 | 225 | 541 | 1,268 |

[1]North Metropolitan Health Service; [2]South Metropolitan Health Service; [3]Southern Country Health Service; [4]Northern Remote Country Health Service

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# 8. Glossary

| Acronym | Desription |
|---|---|
| CF | Cystic Fibrosis |
| CFA | Cystic Fibrosis Australia |
| CFWA | Cystic Fibrosis Western Australia |
| CLC | Chronic lung conditions |
| CSIF | WA Chronic Respiratory Disease Service Improvement Framework |
| FEV1 | Forced expiratory volume |
| FSH | Fiona Stanley Hospital |
| GP | General Practitioner |
| HITH | Hospital in the home |
| ICT | Information Communication Technology |
| MDT | Multi Disciplinary Team |
| NMHS | North Metropolitan Health Service |
| NRCHS | Northern Remote Country Health Service |
| PMH | Princess Margaret Hospital |
| RITH | Rehabilitation in the home |
| RPH | Royal Perth Hospital |
| SCGH | Sir Charles Gairdner Hospital |
| SCHS | Southern Country Health Service |
| SMHS | South Metropolitan Health Service |
| WACFCS | WA Cystic Fibrosis Care Service |
| WACHS | WA Country Health Service |
| WA HDMS | WA Hospital Data Morbidity System |

## Appendix 1: Clinical research at the Lung Institute of Western Australia and Sir Charles Gairdner Hospital (2007 Recommendation 9):

(1) Clinical impact of clonal Pseudomonas aeruginosa in CF. An Australian study, PIs Scott Bell, Peter Bye. About 100 people from the WA Adult CF Centre have contributed microbiological and clinical data. Sue Morey managing study in WA. Ongoing.

Low Rates of Pseudomonas aeruginosa misidentification in isolates from Cystic Fibrosis Patients.

Timothy J. Kidd, Kay A. Ramsay, Honghua Hu, Peter T. P. Bye, Mark R. Elkins, Keith Grimwood, Colin Harbour, Guy B. Marks, Michael D. Nissen, Philip J. Robinson, Barbara R. Rose, Theo P. Sloots, Claire E. Wainwright, Scott C. Bell, and the ACPinCF Investigators. J Clin Microbiol 2009; 47: 1503–1509.

From 214 samples from the WA Adult centre there was one 'misidentification', equivalent to 0.5%, compared to the average 'misidentification' rate for all Australian centres of 2.3%.

(2) A Phase 3, Randomized, Double-Blind, Placebo-Controlled, Parallel-Group Study to Evaluate the Efficacy and Safety of VX-770 in Subjects with Cystic Fibrosis and the G551D Mutation. Now in follow-up open label phase. A multi-centre industry sponsored study managed by LIWA Clinical Trials Unit and Siobhain Mulrennan. Completed and published,

Ramsey BW et al. A CFTR potentiator in patients with cystic fibrosis and the G551D mutation. N Engl J Med 2011; 365:

(3) A Phase 2, Multicenter, Double-Blinded, Placebo-Controlled, Multiple-Dose Study to Evaluate Safety, Tolerability, Efficacy, Pharmacokinetics and Pharmacodynamics of VX-809 Alone and in Combination with VX-770 in Subjects with Cystic Fibrosis, Homozygous for the F508del-CFTR Mutation. A multi-centre industry sponsored study managed by LIWA Clinical Trials Unit. Ongoing.

(4) Siobhain Mulrennan. The Role of the Receptor for Advanced Glycation End products (RAGE) in CF Related Airway Inflammation & CF Related Diabetes. Completed.

(5) Jamie Wood, Physiotherapy Department, SCGH. The effect of a supervised exercise training program on glycaemic control in people with CF-related diabetes. Collaboration with Dr Joey Kaye, Dr Siobhain Mulrennan, Sue Morey, SCGH and Dr Grant Landers, Dr Kym Guelfi, Sugumaran Muniandy, UWA. Completed.

(6) Assessing Cholecalciferol Recommendations in the Adult Cystic Fibrosis Population. SCGH Adult CF MDT. Ongoing.

This document can be made available in alternative formats on request for a person with a disability.

GOVERNMENT OF WESTERN AUSTRALIA

Sustainable Health Review

# APPENDIX B

## *Integrated eHealth Solution for Cystic Fibrosis Patients*

# *Integrated eHealth Solution for Cystic Fibrosis Patients*

## *Executive Summary*

The goal of the project is to improve the accessibility, integration, quality, retrieval and use of patient information to provide a better service to patients in their local settings.

The project focuses on the care of CF patients and will deliver a model that is applicable to a range of other chronic illnesses, demonstrating the value of improved access to clinical information for chronically ill patients across the state.

This project will build upon a unique and proven CF specific eHealth application that is already installed in other states. The application solution is available for the WA program and will require only minor customisation and interfacing for each of the participating health sites and practices in WA. The core technology is a secure web based application that allows the capture and review of patient clinical information at any point of care to authorised care providers based on consent.

This project will be an exemplar for how cross-jurisdictional eHealth solutions for the chronically ill can be implemented with very low risk and at for very low cost.

The service delivery model that will be enabled by the integrated eHealth solution is a partnership between HDWA, CAHS, St Johns Ambulance/Apollo Health and CFWA. The model will be implemented initially at community-based, multidisciplinary facilities in Joondalup, Cannington, Cockburn and Armadale and can be easily extended to non-Apollo Health facilities and existing outreach centres including Bunbury, Kalgoorlie, Port Hedland, Karratha and Broome.

## *Addressing the SHR Terms of Reference*

| | |
|---|---|
| *Leveraging existing investment in Primary, Secondary and Tertiary healthcare, as well as new initiatives to improve patient centred service delivery, pathways and transition;* | This program deliver CF patient and provider centred care by leveraging existing investment, particularly ICT investment, to integrate a specialist CF clinical information system with WA Health information systems. This will result in detailed, CF-specific, patient-centric health records that will support seamless treatment for CF patients across the health sector, regardless of location. Authorised healthcare providers will have access to patient clinical details at all points of care.<br><br>This program will enable patients to be treated closer to home, with less dependence on expensive public health facilities and services. |
| *The mix of services provided across the system, including gaps in service provision, sub-acute, step-down, community and other out-of-hospital services across WA to deliver care in the most* | Steve and Andre probably have the best words for this one … |

| | |
|---|---|
| *appropriate setting and to maximise health outcomes and value to the public;* | |
| *Ways to encourage and drive digital innovation, the use of new technology, research and data to support patient centred care and improved performance;* | This program will use an innovative, cloud-based clinical information system that will be integrated with WA Health core information systems. This technology has been proven in other Australian states and is therefore low risk.<br><br>This solution provides WA Health with an opportunity to demonstrate a state-wide clinical information solution that will be a benchmark for other chronic disease and condition managed programs. It will provide clinician and research access to clinical data that is presently either unavailable (paper-based) or incomplete. |
| *Opportunities to drive partnerships across sectors and all levels of government to reduce duplication and to deliver integrated and coordinated care;* | This program will rely on collaboration between tertiary health services, private primary care services and regional healthcare services. Access to patient-centric patient clinical data across patients' points of care will reduce patient management, reduce redundant diagnostic testing and improve the efficiency of care delivery. |
| *Ways to drive improvements in safety and quality for patients, value and financial sustainability, including cost drivers, allocative and technical efficiencies;* | CF patients represent a disproportionately high cost to the health system. The opportunity to treat patients in primary care settings will reduce the care burden for hospitals and their expensive outpatient and inpatient services. |
| *The key enablers of new efficiencies and change, including, research, productivity, teaching and training, culture, leadership development, procurement and improved performance monitoring;* | The key enabler for this program is access to detailed patient clinical data at all points of care. The sheer volume of paper-based records for CF patients results in critical data being missed, tests being repeated and substantial inconvenience when delivering outreach services around the state.<br><br>The program will have major downstream benefits for research, teaching and training. It will support clinical efficiencies and enable clinical performance monitoring that is not possible with current paper-based systems. |
| *Any further opportunities concerning patient centred service delivery and the sustainability of* | The CF program will inform and provide a model for other, high cost disease programs, such as |

| | |
|---|---|
| *the WA health system.* | kidney disease, heart disease and asthma. The CF program could be readily extended to provide similar benefits for patient safety and clinician efficiency for other managed health health programs. The selected solution already provides support for chronic kidney disease that could be rolled out to health services in weeks. |

# Objectives

- Prove the usefulness of clinician access to patient-centric health records in metropolitan, rural and regional areas to provide improvements in patient care.
- Demonstrate that the health sector can work together to deliver innovative services
- Demonstrate that service charging models are appropriate for health care and are commercially sustainable
- Assess and refine the e-Health technology infrastructure components used in the project.
- Provide foundation infrastructure for additional online health management programs to be offered after this project is completed

# Cystic Fibrosis - Patient Information Management

CF patents generate enormous volumes of clinical data from their broadly-based care teams. Paediatric services will have generated as many as a dozen volumes of paper files by the time the patient reaches his or her early teens. This program will support seamless transition of paediatric CF patients into the adult CF system with complete patient histories available for the transition. Clinical data is contributed by physicians, nurses, dietitians, physiotherapists, psychologists, social workers, lung function testing, pathology and diagnostic imaging services. The data that is maintained includes an extensive list of indicators, from routine measures such as height and weight, to detailed dietary analysis, lengthy lists of procedures and conditions, physiotherapy regimes, living and social conditions, psychological assessments, lung function assessment data, airway organism analysis, and routine pathology and imaging as diagnosis tools.

These patients have complex medication regimes that must be managed carefully to avoid risk of misadventure, particularly when patients are treated outside their normal public health CF Service setting.

With the exception of in-house (hospital) diagnostic test results (pathology, diagnostic imaging and lung function), typically patient data is maintained on paper, which for constitute many volumes of information.

These files are unwieldy and inaccessible, even within a CF clinical team, resulting in care team members maintaining their own separate notes to ensure access to patient information. Information is not accessible between care team members except at periodic team meetings or by telephone calls and emails to solicit information when required. This is extremely inefficient and exacerbates the risk of misadventure for these patients.

# External Healthcare Practices

The paediatric CF program at Princess Margaret Hospital, in collaboration with the adult cystic fibrosis program at Sir Charles Gairdner Hospital, is developing an innovative healthcare model that will see CF patients treated more frequently in primary care settings, significantly improving the patient's experience, reducing the need for patient travel, reducing the burden on patients' families and reducing the burden on

tertiary healthcare facilities and CF specialists services in public hospitals.

This care model includes focused care for CF patients in specific community healthcare facilities that will collaborate and partner with Western Australia's two hospital-based CF services and CFWA.

External healthcare providers (regional emergency services, GPs and physicians) have no access to patient history except by telephone calls to the patient's home practice, and that is problematic if the patient's hospital physician or nurse is not available (out of normal working hours, staff not available, paper file cannot be located). The already high risk for these patients is further heightened in these circumstances, given than inappropriate treatment for CF patients can be fatal.

Regional Western Australia is now well served by broadband networks that have been developed by regional health alliances, as well as by the Federal Government's Broadband for Health program. These networks provide infrastructure that is well suited to improved access to clinical data by regional and rural practitioners when treating patients from metropolitan hospitals. At present, these practitioners rely on patient knowledge, phone calls and faxes to metropolitan colleagues (who may or may not be available) or simply practising without detailed knowledge of the patient's condition.

## Proposed Program

The proposed program will provide multi-disciplinary teams of healthcare providers at two of Perth's metropolitan hospitals, as well primary healthcare partner practices, with an integrated on-line clinical information management solution that replaces current dependencies on paper records. Clinical staff will include:

- Physicians
- CF Service administrators
- Nurses
- Dietitians
- Pharmacists
- Physiotherapists
- Lung Function specialists
- Social Workers
- Occupational Therapists
- Psychologists

This same clinical data will be available to all healthcare providers that provide patient care between CF Clinic visits and in emergencies, particularly in primary care settings and in regional hospital emergency departments, including rural physicians and nurses.

## Complementary Initiatives

CF Australia operates the CF Data Registry, which depends on extensive manual entry of patient information by CF Services around the country. The proposed integrated eHealth clinical information solution includes functionality to populate the Registry automatically from clinical records maintained by participating CF Services, which will result in significant efficiencies for staff in these centres.

## Extensibility

This submission includes the potential to extend the proposed program to other chronic disease programs in Western Australia, at very low cost and with negligible risk. The proposed eHealth solution already supports chronic kidney disease and infectious diseases, both of which lack detailed clinical information support in WA, as well as cancer programs. The proposed solution can be readily extended

to support a broad range of additional chronic disease and health management programs, based on the existing core application.

# Ability to Undertake the Program

The project consortium includes healthcare and other service organisations that will ensure the successful completion of this project:

1. Princess Margaret Hospital.
2. Sir Charles Gairdner Hospital
3. St John's Ambulance (Apollo)
4. Smart Health Solutions – unique CF eHealth solution vendor.

# Implementation Plan

The project implementation plan describes the detailed tasks to enrol practitioners and patients in the program, install and train users and to provide operation support.

During the course of the project, interfaces will be developed with existing systems where they exist and where they support automated, standards based interfaces. This process, which will be conducted in parallel with implementation of the program, will address issues such as:

- Existing data being largely paper based
- Absence of standards based health records in most practices
- Absence of standards based message interfaces, particularly in regional/rural practices

An implementation plan will be developed.

# Risk Management

A detailed risk management strategy will be provided if required.

# Governance, Monitoring and Reporting

A project steering committee, comprised of executive representatives from the stakeholder organisations, will oversight all aspects of the project.

The project management team will report to this committee on progress against milestones, progress against budget, resource management and change management issues to a schedule that will be determined during the project set up phase.

All of the project participants have existing, strong governance structures for clinical and administrative programs. Further details can be provided if required.

# Indicative Budget

| **Clinical Information System Costs (12 months)** | | |
|---|---|---|
| *Service fee (System Vendor)* | Based on number of treatment centres, users and patients ($5,000 per month) | $60,000 |

| | | |
|---|---|---|
| *Project Management (WA Health, System Vendor)* | A central focus point for the project | $50,000 |
| *Project set up (WA Health, System Vendor, other stakeholders)* | Initial training, configuration of interfaces | $50,000 |
| *Expenses* | Vendor, over two years | $20,000 |
| | **SUBTOTAL** | **$180,000** |
| **CHANGE MANAGEMENT** | | |
| *Governance (WA Health)* | Services provided by stakeholders | |
| | **SUBTOTAL** | **$0.00** |

Notes:

- The Service fee for the eHealth solution is inclusive of all customer use and support; there are no licence fees or separate maintenance and support fees.
- The business model below includes additional line items for customisation and a formal project review, which are not essential to implement the solution.

# *Sustainability*

The sustainability of the proposed program is based on a long-term business model in which subscribing to on-line health information management systems to manage patient records is extremely cost-effective compared to the manual alternatives for communicating patient information between practices (telephone tag, faxes, paper files, letters and even email).

In the proposed solution, patient clinical information is available to the practitioner (subject to patient consent) on demand, when it is needed. Studies in the UK have identified costs of up to $50 simply to locate a hospital file when an external clinician requests information. This does not include the effort to make copies, send faxes, make telephone calls or the risks that are introduced when pages are misfiled, not returned to the file or simply lost.

The development of sustainable eHealth programs is typically constrained by the capital requirement to initiate and establish the project, which is the purpose of this grant application.

The following five-year budget indicates how the cost of implementation, together with service fees, will generate efficiencies (at a modest 5% improvement to services in participating practices) that create sustainability for the program. The model assumes that:

- The patient group expands by 10% annually
- CF Patients will require 50 provider interactions a year on average (all provider types, all treatments)
- Savings of 5% of program costs may be achieved across clinic visits, regional practice visits, in-patient admissions and medications. Other savings may also be identified.

Indicated full year savings may not be achieved until year 3, with 25% and 50% savings possibly being

achieved in Years 1 and 2 respectively. The amounts in the following table have been discounted by 75% and 50% in years 1 and 2.

| Items | | | $,000'S | | | |
|---|---|---|---|---|---|---|
| | Year 1 | Year 2 | Year 3 | Year 4 | Year 5 | **Total** |
| **ADMINISTRATIVE EXPENDITURE** | | | | | | |
| Project Setup | 50 | | | | | 50 |
| Project Management | 50 | 25 | 25 | 10 | 10 | 120 |
| Customisation and Interfaces | 50 | 25 | 25 | | 10 | 110 |
| Review | 50 | | | 10 | | 60 |
| Expenses (Smart Health) | 10 | 10 | 5 | 5 | 5 | 35 |
| **Sub-total** | **210** | **60** | **55** | **25** | **25** | **375** |
| Contingency @ 10% | 21 | 6 | 6 | 3 | 3 | 39 |
| **TOTAL** | **231** | **66** | **61** | 28 | 28 | **414** |
| **OPERATIONAL COSTS** | | | | | | |
| Service fees | 60 | 66 | 74 | 81 | 89 | 370 |
| **Total Recurrent Expenditure** | **60** | **66** | **74** | **81** | **89** | **370** |
| **TOTAL EXPENDITURE** | **291** | **132** | **135** | **109** | **117** | **784** |
| | | | | | | |
| **BENEFITS** | | | | | | |
| Hospital clinic visits (2.5k @ $500 * 5%) | 16 | 35 | 76 | 83 | 92 | 302 |
| In-patient days (7.5k @ $750 * 5%) | 70 | 155 | 340 | 374 | 412 | 1,351 |
| Pharmacy ($7.5m * 5%) | 94 | 207 | 454 | 499 | 549 | 1,803 |
| **TOTAL BENEFITS** | 180 | 397 | 870 | 956 | 1,053 | 3.456 |
| **TOTAL EXPENDITURE** | **291** | **132** | **135** | **109** | **117** | **784** |
| **NET (COST)/BENEFIT** | **(111)** | **265** | **735** | **847** | **936** | **2,672** |

# *Technology*

The core technology for this project is a unique and proven CF and other chronic disease specific integration of secure web technologies and secure infrastructure that delivers patient histories (summary clinical event records) to the workstations of participating healthcare providers. It provides an industry strength security framework for access to patient information across the healthcare continuum, integrating Internet and secure identification technologies in line with existing government and industry standards.

The proposed solution is established, proven, low cost and can be implemented quickly. The proponents have a unique mix of healthcare delivery experience and access to appropriate providers and solution delivery in similar projects in the health sector. These attributes will significantly mitigate risk and minimise costs.

The solution enables general practitioners, specialists, hospitals, diagnostic services and other authorised healthcare providers to share summary patient clinical information in a simple, user friendly and secure manner.

It is the only solution of its kind in Australian that specifically supports CF. It is the only solution available that can support clinical information management for CF patients across care-settings state-wide, including private practices.

The proposed solution was designed by Australian healthcare providers and built in Australia to fulfil the business requirements of those providers. The proposed solution is proven and is ready to be deployed in projects of this scale and beyond. It is flexible, extensible, and simple to rollout to participants.

The application is readily adaptable to specific aspects of e-health, particularly chronic illness management and is ideal for the case management of cystic fibrosis. It can be expected to provide significant value in terms of improvement in care delivery and reduction in overall cost of service delivery.

# Registry Support

One of the elements of the proposed project is its support for automated reporting to the national CF Data Registry, which is maintained by CF Australia. Annual CF reporting is currently undertaken manually by public CF services, either using paper forms or a separate on-line web-based service. Completing the reporting process requires a nurse to go through each patient's paper files (typically several volumes) to find the extensive range of reporting items that must be entered. This process takes time away from patient care, is prone to transcription errors and can generate inconsistent data when a patient has attended more than one service.

This project includes the electronic generation of CF Australia reporting data from clinical records that are collected routinely when patients are treated by CF Services; no additional transcribing of data is required.

# Evaluation

## Proposed Key Performance Indicators

This project will improve the availability, integration, and quality of patient health information. The following KPI's will be used in review processes as measures of its success:

- More effective use of scarce hospital resources by treating patients in the community when hospital care is not required
- A significantly improved experience for patients and their families, who will have to visit Perth metropolitan hospital services less frequently
- More appropriate and timely clinical decision making and healthcare delivery
- Patients empowered to take an active role in making informed decisions regarding their health
- More appropriate and timely referral and feedback to support patient health care
- Use of stored information to fill the current gaps in knowledge and improve the existing evidence base
- Use of the stored information to effectively target healthcare needs

## User Assessment

Clinical users will complete a survey/questionnaire prior to commencement of the installation and training program, and again at the end of each evaluation period. This questionnaire is comparatively brief and covers a range of simple qualitative and quantitative usability issues. Analysis of the questionnaire will allow direct comparison to be made regarding the effectiveness of patient information management, with an emphasis across teams and practices, in pre and post implementation operating environments.

## Patient Assessment

CF WA will conduct a similar program with patients, including patient interviews, to assess the impact of the solution on the delivery of care on this group of participants.

## Application Use and Performance

The application maintains a complete log (event record) of all activities (for audit and review purposes). This data will be used to report on detailed application use, including measures of frequency of use, session times, numbers of records accessed and updated, patients enrolled, providers enrolled, use over

the period of the project, etc.

## *Stakeholder Review*

Finally, a formal stakeholder review will be conducted after the initial period of use, to consider the various review outputs and to formally report to the participants in the project and to external bodies followed by minor follow up reviews.

CYSTIC FIBROSIS Western Australia

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Prof. Stephen Stick
Princess Margaret Hospital
Roberts Road,
Subiaco WA 6008

Tuesday, 3 October 2017

Dear Steve,

**Distributed Service Model**

It was a pleasure to meet with you yesterday and discuss our involvement in the "distributed service model" pilot project in Joondalup.

CFWA would like to take this opportunity to offer this letter of support.

We would also like to take this opportunity to confirm where we feel we can add value as the community service provider for people living with Cystic Fibrosis in WA and as a health and allied health educator.

With regard to our training programmes, we recognise that St. John is a well-known Registered Training Organisation and we would welcome an opportunity to collaborate with them to further develop appropriate resources and particularly to adapt our Regional Respiratory Training Program (RRTP) which is now in its 8th year of delivery here in Perth.

As promised, I have attached our draft programs for the RRTP for 2018, for nurses and physiotherapists. We will be making a few small changes to these so that participants can attend either a 2-day acute training program or the fuller more comprehensive 5 day training. A small component of this may be appropriate for staff from Joondalup to attend.

We are currently developing e-learning modules for health professionals which could be used to train health and allied health professionals in the pilot. We will need to source approximately $100,000 to complete this.

We already have an e-learning platform in place delivering our CFSmart program. This would be used to deliver this program. E-Learning could also be supplemented with face to face training for small groups. Modules under development include:

***Health Professionals***

**Module 1 - What is Cystic Fibrosis?**

- Genetics
- Demographics
- Anatomy and Physiology - overview
- Overview – systems impact

**Module 2 - *Screening and Diagnosis***

- CF Carrier Screening

- CF Newborn Screening

**Module 3 - *Respiratory system***

- Anatomy and Physiology
- Treatment: - AREST CF
  - Medications
  - Airway Clearance Therapy (overview)
  - Exercise
- Complications

**Module 4 - *Infection***

- Causes
- Prevalence of Organisms
- Side effects
- Infection Prevention and Control

**Module 5 - *Airway Clearance Therapy and Exercise***
(Physiotherapists)

**Module 6 - *Gastrointestinal Tract***

- Anatomy and Physiology
- Pancreatic Insufficiency
- Treatment
- PEG
- Complications

**Module 7 - *Nutrition***

- Diet
- Healthy fats
- Supplements
- Salt
- Vitamins

**Module 8 - *CF Related Diabetes***

- Causes and statistics
- Effects
- Treatment

**Module 9 - *Co-morbidities***

- Sinusitis
- Liver
- Renal disease
- GORD
- Metabolic Bone disease
- Cardio vascular disease
- Anxiety and Depression

**Module 10 - *Mutations and Modulators***

**Module 11 - *Medications***

- Antibiotics
- Anti – inflammatory
- Anti-fungal
- PPI's
- Modulators .....

**Module 12 - *Puberty and Transition***

**Module 13 - *Fertility Male/Female***

- Anatomy and Physiology
- Pregnancy
- Assisted Reproduction Technology, IVF, Surrogacy

**Module 14 - *Psychosocial***

- Parents
- Child
- Adult
- Anxiety and Depression

**Module 15 - *Research***

- Trials

**Module 16 - *Transplant***

**Module 17 - *Living with CF, an adults journey***

- Study
- Employment
- Marriage
- Financial security
- Burden

**Module 18 - *History of CF***

- Journey – origin
- Genetic pool – Asia, Aboriginal, Iranian
- Where are we going?

**Module 19 - *Support Organisations***

- CFWA
- CFA
- PMH
- SCGH
- FSH

I have also attached a copy of our CF Smart "A Guide to Cystic Fibrosis for Health Professionals" This was developed by us with input from PMH and SCGH. Whilst it may have been a little pre-emptive on page 14 in stating that the New Children's hospital would be open in 2015, it nevertheless illustrates the quality of collateral we will be producing. This can also be downloaded from our website http://www.cysticfibrosis.org.au/media/wysiwyg/CFF045_-_Services_Brochure_Health_Online__WEB.pdf

Finally, we would like to be involved in the public focus group you have foreshadowed in Joondalup. We would be happy to promote this meeting through our communication channels, attend the meeting itself and if required, we would be happy to speak at the meeting.

Importantly, we would see this pilot as embracing both children and adult services for localised non-acute management of CF.

We believe that the "SmartHealth" electronic record system would provide better care for patients, assist in providing information for the CF data registry and streamline referral processes to our home care services in the future. In addition, we would see a role for leveraging technology with PMH, SCGH and FSH through on-line consultations with clinicians at the tertiary centres which again would reduce the impact of the disease on lifestyle and assist in cross infection control.

I trust that this will give you sufficient information to proceed and I look forward to hearing from you in the near future.

Yours sincerely

Nigel Barker

CEO