PEER REVIEW REPORT

CENTRO REGIONALE VENETO PER LA FIBROSI CISTICA
OSPEDALE CIVILE MAGGIORE, AZIENDA OSPEDALIERA VERONA
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Peer review panel

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Executive Summary and Key Recommendations

1. The CF service in Verona cares for 810 patients with Cystic Fibrosis, 472 adults and 338 children. Service is primarily delivered at Ospedale Civile Maggiore**,** however, two satellite clinics located within 100km (Ospedale Treviso and Ospedale Roverto) which operate under the direction of the Verona clinic. One hundred and sixty three patients use this shared care arrangement. The **peer review panel was impressed by the professionalism, commitment and patient-focused nature of this service**. The standard of care delivered is of a high standard.

2. This level of service contributes to the good outcomes achieved by the Verona CF team. The peer review panel was shown data from a high quality local database which **showed excellent outcomes which reflect this very high level of service delivery**.

3. Infection control policy. Clinic staff recognize the importance of an effective and proactive infection control policy. The review teamed did note several areas where significant improvements can be made; A). There are **too few hand hygiene facilities** both on an inpatient and outpatient basis. This deficit needs to be prioritized. B). **Alternative cohorting practices for ward patients need to be explored**. The practice of grouping the most vulnerable (transplant patients) with those patients with the most transmissible and resistant organisms (*B. cenocepacia* > other Bcc > MRSA) on Ward A poses significant risks to patients. Furthermore, post transplant patient numbers continue to increase and exposure risk will continue to accumulate. C). Consideration of multiple separate smaller physiotherapy/exercise rooms to maximize patient benefit and minimize cleaning time and risk for infection transmission. D). **Re-education of staff and patients on the role of masks** as merely an adjunct to hand hygiene and not as a primary means of infection control. Patients need to be cautioned about the proper type and use of masks.

4. Over the course of many years managing an evolving CF cohort, the CF team has developed an expertise in managing the many problems relevant to an aging CF population without specific training in adult medicine. Areas of evolving expertise included; CF related diabetes, dynamic bone disease and transplantation. Apart from the ongoing service provided by the CF team, **other specialties needed for a comprehensive adult CF service are also available in Verona.** The team was confident that those providing this service understood the needs of those with CF and they had every confidence in their care in these respects.

5. The CF service was a stand-alone facility or a "hospital within a hospital". There was little interaction between other disciplines which are important to support CF care including; pulmonary medicine, endocrinology, psychiatry, infectious disease, and medical microbiology. The review panel would **encourage more frequent consultations and opportunities for collaboration with other disciplines.**

6. The peer review panel were impressed with the positive developments in clinical trials research which have occurred in the last two years. At the time of the peer review, there

were nine active or recently completed clinical trial projects involving new therapeutics. and five large collaborative studies.

7. Problems unique to such a diversified clinic population were recognized and highlighted by staff. While both the historical roots of the CF service and the training of the medical staff lie in pediatrics, the patient cohort has evolved over time (as a result of successful management) to be dominated by adults. Accordingly, problems which may be more common in adult patients and therefore unfamiliar to pediatric practitioners are expected to complicate management including; drug and alcohol abuse, depression and mood disorders, sleep disturbance, sexual health, bone disease, renal impairment, etc. In the current team structure, none of the physicians are trained in pulmonology. The team would be wise to diversify to **include individuals with training in adult internal medicine and pulmonary medicine.** Furthermore, the team should consider means of increasing interaction with other health care professionals practicing in areas where there might be overlap with CF needs (infectious disease, endocrinology, medical microbiology, pulmonary medicine) so as to stay abreast of new techniques and technologies.

8. Problems unique to such a large clinic were also recognized by clinic staff. With the progressive change in patient demographics from pediatric patients to adult patients, cohorting has become as issue. Indeed the needs of these two populations are increasingly divergent. While efforts have been made to separate these two populations with "ward B" being designated for pediatrics, and new pediatric outpatient facilities being developed this problem is anticipated to only get worse with time. Whereas the medical staffs have their roots in pediatrics, other health care providers including nursing, physiotherapy, and dietetics were primarily trained in adult medicine. Indeed, the perception was that many nurses were uncomfortable providing care to children (including line placements and phlebotomy) and in particular neonates with CF. The review panel thought that the continued local emphasis on all CF services being provided under one roof was overemphasized. Given the proximity of the pediatric care building, it would be possible for specific members of the same CF team to provide ongoing inpatient care to pediatric patients under the umbrella of a pediatric hospital.

9. In terms of areas which could perhaps benefit from some improvement, the main area consistently identified by the review panel was staffing. While differences in medical practice can account for some of the differences observed between the Verona clinic and UK CF Trust guidelines, there are a few areas where very large deficits are noted. For the number of patients and the increasing severity of their CF, **there is a serious shortfall in available clinic staff.** In particular areas which recruitment should strongly be pursued include; dietetics, clinical pharmacy and psychosocial support. Both nursing and physiotherapy services also displayed significant signs of being critically understaffed.

10. Nutritional outcomes in Verona were noted to be significantly lower than from published cohorts. In addition to the median clinic **BMI being ~ 1 point below comparator populations,** there were significantly increased numbers of individuals with

advanced nutritional deficiencies (BMI <18 kg/m_2). The minimal use of nutritional supplements and absence enteral feeding in those with advanced nutritional failure differ markedly from other patterns of practice.

11. Outpatient clinic space is currently functioning at or beyond capacity for a clinic population of 800. More clinic time, or expansion of existing clinics to accommodate more patients will be required

12. Development of a patient annual review visit and a resultant comprehensive report is recommended. Annual reviews provide for the opportunity for patients and care providers to reflect on the course over the last year and to focus on trends as opposed to the problems of any individual visit. A multi-disciplinary team comprising members of dietetics, nursing, physiotherapy, pharmacy services, social work and a physician should be involved with each assessment.

12. In a very limited assessment of patients, it was apparent that the high level of service delivery resulted in a high level of patient satisfaction. Patients felt they received the highest level of care. A few complaints were registered about the high level of turn over in the nursing staff and limited physiotherapy. An anonymous survey conducted in 2006 identified areas including psychological resources that were under resourced. A similar "user assessment survey" should be repeated.

Detailed findings and Observations

1 Background Information

1.1 History of Service

Dr. Carlo Castellani gave the review panel a brief history of the CF service in Verona. The service was developed in the 1970's as part of the pediatric ward and has since grown to its current state. Patients have been identified through various neonatal screening programs first implemented in the 1960's. In addition, each year several patients including adults are diagnosed on the basis of clinical grounds. In total, 6 to 30 new patients are diagnosed with CF each year. While the patient population is primarily composed of those individuals with CF who live in areas around Verona, approximately 30% of patients are from distant regions who continue to travel to Verona for their CF care on the basis of a prior relationship and confidence in their care. While the bulk of service continues to be provided at Ospedale Civile Maggiore**,** in recent years, two satellite clinics (Ospedale Treviso and Ospedale Roverto) were developed and operate independently under the general direction of the Verona clinic. Physicians and physiotherapists at each satellite clinic have received prior training in CF at Ospedale Civile Maggiore.

1.2 Service Aims

The CF team strived to provide the best care possible to all adults and children with CF, to enable them to live as full lives as possible with the least interference from their disease.

1.3 Strategic Context

CF patients are managed independently at the Centro Regionale Veneto per la Fibrosi Cistica which is located within Ospedale Civile Maggiore. Funding for the CF service comes directly to the hospital from the federal government. Staff of the Centro Regionale Veneto per la Fibrosi Cistica operate independently within the context of the larger hospital administration. Several positions are funded externally through funds raised by charitable organizations. Standards of care are met and delivery is guided using locally developed protocols for common problems relevant to CF.

2 Overview of Clinical Care

2.1 Outcomes

The outcome for surviving patients in the Regionale Veneto per la Fibrosi Cistica are very good. Mortality rates have fallen over time. In the early 1990's the annual mortality rate was 1.5-2.0%, this had dropped to less than one percent by 2006 and continues to decline. In this time, median age of death has increased from twenty-one in 1990 to twenty-eight in 2009. In particular, patient survival to transplantation has improved. In the last five years, 67 patients have been referred for transplantation and 39 were successfully transplanted.

Detailed pulmonary outcomes were not available to the review team. Historical cohort data show significant improvements over time. For example, the proportion of individuals with mild CF airway disease (FEV_1 >70) has increased from 41% in 1990 to 58% in 2009. Individuals categorized as having either moderate (FEV_1 50-70%) or moderate-severe (30-50%) have fallen from 27% to 20% and 21% to 17% respectively. Patients with severe airway disease with FEV_1 <30% has fallen from 10 to 5%. Chronic infection with *P. aeruginosa* has fallen over time with levels falling from 58% (one positive airway culture) to 54% from 2006-2008. The rates of *P. aeruginosa* chronic colonization among children less than twelve years of age remain less then 20%. Rates of *Burkholderia cepacia complex* colonization remains stable at <5%. *MRSA* colonization/infection rates range from 12-15%.

The peer review committee asked for detailed data with respect to BMI on all adult patients. Of the data provided for 464 adult patients, median BMI is 20.8. This value is lower than that observed in other clinics in the US (21.4)[2], UK (21.6)[4] and Canada (22.1)[1]. BMI was shown to improve with increasing age as with other datasets. In particular, 10.3% of the adult clinic population had a BMI in the less than 18 kg/m^2 category and 36% < 20 kg/m^2. **These values are markedly lower then values reported from other CF clinics[1,2,4].**

2.2 Staffing

There are significant differences that exist between the number of staff that are in the employ of the Centro Regionale Veneto per la Fibrosi Cistica and those targets set forth in the UK CF Trust Guidelines[3]. While health care delivery models are very different between the two countries and direct comparison is impossible **the contrast is striking particularly in regards to the areas of dietitics, pharmacy, and psychosocial support** (Table 1).

Table 1: Projected staffing requirements for Centro Regionale Veneto per la Fibrosi Cistica as determined by the UK CF Trust Guidelines

Team Member	Existing Verona Clinic Staff	UK Trust Recommendations					Review Panel Reccommended Team Size
		Per 50 patients	Full Care n=647	Shared Care ** n=163	Total for population of n=810	Total deficit (% Required)	
Physicians * **	7 + 1 (non-clinical) =8	1.3	16.9	2.0	18.9	10.9 (42)	10 -Adult Internist -Adult Pulmonologist
CF Nurse Specialists	-	1.0	13	1.5	14.5	14.5 (0)	N/A
Inpatient Nurses ** **	18	-	-	-	-	-	26 (18 trained / 8 untrained)
Outpatient Nurses ** **	2	-	-	-	-	-	4 trained 2 untrained
CF Physiotherapist	5	1.0	10	1.5	12.5	7.5 (48)	12
CF Dietitian	1.0	0.4	5.2	0.6	5.8	4.8 (17)	4
Social Worker	0.5	0.4	5.2	0.6	5.8	5.3 (9)	NAC
Psychologist	1	0.4	5.2	0.6	5.8	4.8 (17)	NAC
Secretary	3.5	1.0	13	1.5	14.5	11 (24)	5
Data Managers	1.0	0.1	1.3	0.15	1.45	0.45 (69)	3
Pharmacist	0	0.3	3.9	0.45	4.35	4.35 (0)	3

*As per the standards set by the UK CF Trust, UK CF Trust Standards of Care Guidelines May 2001, page 13 for provision of care to each group of 50 patients[3].

This is widely acknowledged to overestimate required team size when patient populations exceed 250.
**Patients in shared care require 50% the services of those in full care.
***Physician equivalents = UK standards/50 patients 0.7 consultants + 0.6 Staff Grades (non-certified specialist) = Rounded to 1.3, eliminating the trainee role
**** Ward nurse size estimate based on UK standards of practice for general medical wards

The peer review panel felt strongly that the most notable deficiency in terms of CF care delivery was in terms of CF dietetics. Although there is one full time CF dietitian, she is single-handed, recently appointed to the position and there is no cover for when she is absent. Furthermore, her position is externally funded through the Italian CF foundation rather than the hospital and is renewed on a yearly basis. This is unacceptable for a service of this size.

The absence of pharmacy support was also notable. Incorporation of CF specialist pharmacists has been universally adopted in other countries owing to the increasing complexity of patients, and potential for drug-drug interactions with multiple drug regimens. Furthermore, a pharmacist has the potential for to result in significant cost savings owing to drug bundling and optimizing delivery. It is evident that the savings that can be accomplished by clinical pharmacists can quickly surpass the cost of their own salary making this a cost neutral or beneficial team member.

The review panel also noted a low level of psychosocial support available for CF patients. We did not have the opportunity to meet with either the psychologist or the social worker associated with the clinic.

The team worked to a number of agreed standards which were shown to the peer review panel. These included overarching standards with appropriate evidence and indicated clearly the responsible parties for delivering and implementing these standards. Quality dimensions and performance indicators were also laid down and provided a clear and comprehensive framework for the care of these patients.

2.3 Outpatient Care
Each week there are five outpatient medical clinics which take place each morning. Friday clinics are reserved for drug resistant pathogens with three MRSA clinics each month and one *B. cepacia complex* clinic. Transplant recipients are not seen specifically in a transplant clinic. Patients are shown directly to their clinic room and wait for their multidisciplinary team members to visit them, so there is limited waiting in communal areas. In each medical clinic, there are two sittings of four patients, so eight patients are seen at each clinic occasion, ie: 40 each week. *This number of outpatient clinics would seem on the small side to serve a population of 810 patients (Seeing 40 patients per week x 50 evaluable weeks (secondary to holiday) = 2000 sittings/ year available). At present there are 647 full time patients with aims to be seen 4x/year and 163 shared care patients with aims to be seen 2x/year x 50 weeks = 2914 sittings required.* **An apparent deficit of ~30% exists in the current number of clinic spots available for patient review**.

Furthermore, this would not account for those individuals with advanced disease that require assessment as frequently as weekly. As populations continue to increase this deficit would be expected to further increase. Expanding the number of clinic appointment slots into the afternoon will be required. Patients can be seen independent of clinic if there are acute changes such as symptoms of pulmonary exacerbation.

Clinics are physician led as opposed to multi-disciplinary in nature. Each patient receives lung function testing and is then reviewed by a physician. If there are issues relevant to the physiotherapists, dietician, social workers, or nursing staff arrangements can then be made for these services to be sought. Follow-up appointments can be made with each discipline should they be required. There are no nurse or physiotherapist led clinics. There are no multi-disciplinary debriefs at the end of each clinic. **There should be consideration of increasing the multidisciplinary input at out-patient visits.**

In the event a patient requires contact with the CF team outside of their regular appointments, the Verona team has developed a telephone triage system. CF nurses have distinct times of the day when they are available to answer patient's questions, provide advice and arrange urgent appointments should patient be unwell.

2.4 Annual Reviews

Annual reviews are not yet practiced in Verona. The team members had discussed these issues and have recently set forth with plans to introduce this. The structure of the planned annual reviews was not developed as of the time of the peer review process. The peer review team would encourage the clinic to proceed with annual review as this type of visit serves to enhance regular clinic visits as it focuses on the whole picture as opposed to the individual problems of each visit. **The peer review panel would encourage that Annual review be performed by a multi-disciplinary team allowing the opportunity for each discipline to re-educate and evaluate each patient**. Such Annual reviews also serve the opportunity to discuss disease progression with the patient and prospects for the future.

2.5 Inpatient Care Provision

There are twenty beds dedicated inpatient beds that exist on the 2nd floor of Ospedale Civile Maggiore, Azienda Ospedaliera Verona, exclusively for individuals with CF. Inpatient beds are divided into three separate wards. Patient rooms are all extremely well equipped. Importantly, all rooms are for a single patient use. However, each room has two beds, one for the patient and one for the care provider/support person. The review team was told that most patients are accompanied during hospitalizations by a support person. Rooms are very well suited for patient comfort, each with a television, DVD and even a safe for personal effects. Each room has its own exercise bicycle, however, these may be stored in the common hall.

Wards are separated – and allow the opportunity for patient cohorting. "Ward A" has seven beds and focuses on complex patients such as those with drug resistant or highly transmissible organisms (*MRSA, Burkholderia cepacia complex*), and those both awaiting and post transplantation. Each room has an *en suite*. **The review team was concerned**

that this model of cohorting complex patients placed the most vulnerable patients (advanced respiratory disease pre-transplant and post-transplant individuals) at high risk of acquiring highly transmissible pathogens. "Ward B" has eight beds and is reserved exclusively for pediatric patients. Not all rooms have their own *en suites*, however, renovations and expansions are planned. Work on new inpatient rooms is also about to begin. "Ward C" is part of the older structure and lacks *en suites*. It is used for those patients who do not fall into either Ward A, or B. The perception of the staff was that waiting times for hospitalization are quite low and patients are generally seen and admitted within one day of calling unwell.

Each ward is supervised by an individual physician (changing weekly, unless overnight coverage disrupts the schedule), a nurse and a physiotherapist. This team structure seemed to work very effectively for providing high level patient care and ensuring continuity of care. The team meets daily to discuss on-going issues and to round on the patients. Attending physicians rotate to provide 24 hour inpatient care. The review team noted that the high level of seniority (consultant physicians) for on call responsibilities was unique among other models of CF care. On call facilities are limited to a bed with a privacy curtain within the physiotherapy department.

2.6 Weekly Routine

The weekly routine for the CF team includes a regular multi-disciplinary team meeting on Mondays lasting for 1.5 hours. Here all inpatient matters, patient's receiving IV antibiotics at home and urgent outpatient matters are discussed with the entire team. The nurses and physiotherapists stated that they felt comfortable with their role in the team meeting and freely contributed to discussions. It was not apparent if information is available in a spreadsheet format for each meeting to provide structure and keep a record of the plans from the meeting. It was not readily apparent if a hierarchical structure existed to deal with disagreements in planned management. Encouragingly, satellite centers have access to these meetings through Skype.

There is also a "Pasta Tuesday" which serves as a professional development day. Team members gather to discuss current controversies in treatment delivery and diagnostic modalities. Working groups to develop consensus protocols are often convened within this context which aids in the delivery of patient care.

Outpatient care is delivered is delivered on floor 3, along with the offices of all personnel. Outpatient clinics occur daily each morning and patients are segregated according to sputum microbiology (ie MRSA, Bcc). There are no independent nurse run clinics.

2.7 Infection Control

Infection control was emphasized by clinic staff. For inpatients, all are instructed to follow a pre-defined set of guidelines on ward behavior. This information is documented in a booklet which is given to each patient. The review panel was impressed with the comprehensive nature of this booklet. All patients have single rooms. Patients were instructed on hand hygiene before and after leaving their room. In hallways patients would wear surgical masks. In common areas such as the kitchen patients were not

allowed to enter – but rather support personnel would heat all meals, etc. **There was, however, a notable absence of alcoholic hand foams/sprays for hand sanitization and sinks in common areas**. We were told there were issues at the hospital administrative levels regarding the appropriateness of implementation of alcohol hand hygiene. As far as the review panel was concerned, **alcoholic hand hygiene is absolutely appropriate for non-soiled hands for hand hygiene for staff and patients alike**.

While patient exercise is possible within individual's rooms (with exercise bikes) to limit possibility of transmission, a common gym also exists. This room is well stocked with multiple weight benches and free weights (3), exercise bikes (4) and treadmills (2), however, can be used by a single patient at a time as a result of risk of cross infection. Furthermore, with the proximity of all the exercise equipment, the reviewers were concerned that devices not used could also be contaminated by aerosols generated during exercise requiring extensive equipment cleaning. The storage of all this equipment together seemed a waste of valuable space. **The clinic should consider the construction of several, smaller physical therapy studios such that multiple patients can exercise at the same time using the already available equipment, with a minimum of cleaning required.**

While patient segregation and cohorting was practiced, we have some concerns regarding its implementation. "Ward A" cohorts all complex patients including those with Bcc, MRSA, those awaiting transplantation, and those post transplant hospitalized with complications. This practice puts extremely vulnerable post transplantation patients at risk of acquiring aggressive pathogens. Furthermore, patients with MRSA and *B. cepacia complex* other than *B. cenocepaicia* are cohorted with more aggressive and transmissible *B. cenocepacia*. Rooms were apparently constructed with the idea of viral vectors for gene therapy becoming common place in the 1990's. The review panel thought that this designation provided a false sense of reassurance regarding the capacity of these rooms to limit pathogen transmission. These rooms are ventilated merely by opening windows – no true air exchange/or HEPA filters exists within these rooms. Furthermore, no rooms capable of containing airborne transmissible pathogens exists within the entire complex. While the current cohort does not include high numbers of these transmissible pathogens, recent outbreaks in other large centers continue to highlight the importance of forward planning. **Alternate cohorting practices need to be explored. The practice of grouping the most vulnerable (transplant patients) with those patients with the most transmissible and resistant organisms poses future risks to patients.** Furthermore, post transplant patient numbers continue to increase and exposure risk will continue to accumulate.

Patients have been taught the importance of infection prevention practices in outpatient environment. All individual rooms for outpatient assessments also exist within the facility. While patients wear masks immediately when entering common areas, they congregate in common waiting areas. No or limited opportunities exist for hand hygiene in the outpatient clinics as well as entry into the buildings. **This deficit needs to be prioritized. Basic hand hygiene is the most important step in minimizing pathogen transmission**.

This is applicable to both patient and clinic staff alike. Clinic segregation does exist with separate clinics for MRSA and *B. cepacia* complex being operated on Wednesdays.

Patient masks are routinely employed as a standard of care. This practice is not unique to the Verona clinic as it has been adopted by a number of centers. **The peer review team was concerned, however, that staff and patients may be falsely reassured by mask use as illustrated by lack of facilities for hand hygiene for patients and staff alike.** Furthermore, as an infection prevention strategy for respiratory pathogens hand hygiene remains the most crucial step with masks serving to reduce risks only marginally further. Patients were also observed to be wearing masks inappropriately (surgical masks upside down, or with the nose not covered). One patient was observed to be wearing a N95 mask as well. These masks are; exceedingly expensive, need to be fit checked, and have the potential to cause hypercarbia - even in health care workers when worn for more then twenty minutes – let alone individuals with advanced airway disease. **Re-education of staff and patients on the role of masks as merely an adjunct to hand hygiene and not as a primary means of infection control is required. Patients need to be cautioned about the proper type and use of masks.**

2.8 Prescribed Chronic Disease Management Therapies

Inhaled antibacterial therapies for chronic maintenance of lung health are used by the Verona clinic primarily in those individuals chronically infected with *P. aeruginosa*. Tobramycin solution for inhalation (either TOBI or Bramitob) are used as first line therapy. Colistin is used less frequently. At time of preparation the review team did not have data available regarding drug use stratified by *P. aerugionsa* infection or severity of lung disease. In total, 42% (344/810) patients received inhaled antibacterial therapies. This compares to data from the UK and US in individuals chronically infected with *P. aeruginosa* of 88.6% and 67.4%, respectively. Direct comparison accordingly, cannot be made. DNase use was reported to be used in 220 individuals (27.1%). In the UK DNase is used by 37.2% of all patients and in the US DNase is used by 76.1% of patients >6 chronically infected with *P. aeruginosa*. Chronic macrolide were utilized in 60.7% (492/810) of patients (all patients). This compares favorably to the UK (60.4%) and US (65.6%) in patients chronically infected with *P. aeruginosa*.

2.9 Home Intravenous Antibiotic Programme

Home delivery of parenteral antibiotic therapy is available to patients attending the CF clinic in Verona. Patients (and care givers) have been taught safe practice for drawing up medications and the self administration of these therapies. Competencies are evaluated by nursing staff and patients have to be certified prior to home IV therapy being available to patients. The review panel did not get the opportunity to explore the frequency with which home parenteral therapy was employed relative to delivery in hospital.

2.10 Transition Arrangements

As one team (within the larger CF team) provides care to pediatric patients formal transition of care does not occur as it might between separate pediatric and adult facilities. All patients are commonly discussed during the weekly MDM such that all staff are aware of active issues. We did not have the opportunity to discuss with the clinic staff

how patients transition between "ward B" and (either A or C), nor how teenagers are encouraged to take greater responsibility for their chronic disease management. We did not get to explore the role in which psychology or social work team members played in the transition from adolescence to adult hood. Home visits by clinic psychosocial staff are not practiced as a routine.

2.11 Microbiological Services

The microbiology service is delivered by the local hospital and a single microbiologist is responsible for CF sputum studies. We did not have the opportunity to visit with the microbiologist, nor review the specific protocols used in the analysis of sputum composition. We did note, however, that quantitative sputum analysis was being performed. The relationship between the CF team and the microbiologist is for the most part limited to reporting of sputum pathogens. There is limited consultation that occurs. The review team felt that a closer relationship with the consultant microbiologist might enable opportunities for clinical insight and advice with respect to treatment. There are no local prospective studies to examine for clonality of pathogens to establish if cross infection is occurring. A research study is underway with collaborators in Milan looking for the presence of clonal *P.aeruginosa*..

2.12 Additional Specialty Support

There is limited interaction with other disciplines. On site radiology facilities are available for all required nuclear and radiographic imaging studies and easy access to consultant radiologists. Totally implantable vascular devices (TIVADs) are managed by either the vascular surgery or radiology team. Bronchial artery embolisation is available for individuals with significant hemoptysis. There is also ready access to an obstetrician/gynaecologist service for females pursuing pregnancies. Thoracic surgery or pulmonary medicine are responsible for chest tube placement and maintenance in the event of a pneumothorax. There is very limited access to endocrinology/diabetologists. There are no joint clinics reported for the 140 (17.2%) patients with CF-related diabetes. Staff reported even phone consultations were difficult to arrange with consultant endocrinologists. One CF physician has had limited prior training in endocrinology and serves to assist in the management of CFRD. The review team did not get to meet those individuals involved in psychosocial support but would have been very interested in finding out about the availability of psychiatry services, and outpatient social services.

Lung transplantation is not done locally. Patients are referred typically to one of two centers depending on projected waiting list times; Padua or Bergamo. In rare instances, patients may be referred to other centers such as Palermo or Innsbruck. There is limited share care practiced following transplantation. For the most part post transplant patients follow up with their CF clinics who manage complications such as infection and episodes of rejection. There was some sense of abandonment by the transplant service in those individuals with failing grafts who were not considered candidates for re-transplantation. Liver transplantation has also been performed on several patients with end stage cirrhosis. The one patient also reported his perception that the transplant team did not communicate recent results or clinical decisions well with the local CF team.

The Centro Regionale Veneto per la Fibrosi Cistica was described as "a hospital within a hospital" by some interviewees. The review team was given the impression that there was very limited involvement of other services with CF patients (infectious disease, microbiology, hepatology, endocrinology, etc.). There was no indication that staff participated in mutli-disciplinary medical rounds with other divisions of medicine. Several physicians had stated that on occasion consultations from other specialties were sought, but lack of CF specific knowledge of the consultancy service limited the utility of the information. Certainly, this seems like a vicious circle bound to repeat itself as these individuals do not end up seeing more CF patients**. The review team would encourage the center staff to increase their exposure to other disciplines such that new techniques and technologies can be adapted to the CF population.**

2.13 Facilities

The peer review panel was shown the inpatient facilities, the outpatient facilities, the day case areas, the lung function laboratory and the administrative areas. The facilities were all adequate and appropriate for their purpose. All patient rooms were single rooms and were well equipped with televisions, DVD players, telephone and safes. All rooms also contained a second bed, available for a spouse or support person's use. The review panel was impressed by the inclusion of an exercise bike in each room. Not all rooms had *en suite* facilities. Renovations are planned with development of additional *en suites* in mind. A large gymnasium is available with multiple aerobic and weight based exercise machines available for use. Risk of infection transmission limits the use of this room to one patient at a time, despite its size and abundance of equipment. Given the proximity of equipment, the review panel would be concerned that the amount of cleaning required to properly sterilize all relevant surfaces between patient use is not done at present.

The lung function laboratory was staffed by three individuals and managed by a physician. While meeting functional requirements, the pulmonary function laboratory lacked modern equipment and were not employing any newer technologies for measuring lung function (infant PFTs, nitrogen gas lung washout studies). Furthermore, the lung function testing using water displacement is an aged device and may risk infection transmission. The addition of an adult trained pulmonologist may enable the team to explore ways and means of improving existing facilities. Sweat chloride testing and nasal potential differences are other technologies regularly performed in the laboratory.

There is a CF specific bronchoscopy suite available for routine studies. One physician has been trained in bronchoscopy and performs them with the assistance of ward personnel. In instances in which problems are anticipated an anesthesiologist may be on hand. The review panel questioned the need for a CF specific bronchoscopy suite given the low frequency with which the procedure is required in CF. Furthermore, the issue of patient safety was raised given the stand alone nature of the facility and lack of specifically trained support staff, and the ability to maintain ones bronchoscopic competencies with limited procedures.

2.14 Research

The Centro Regionale Veneto per la Fibrosi Cistica has traditionally focused on the delivery of quality clinical care. The large patient population that they serve has meant that little time has traditionally been available for clinical trial research. However, since 2007 great efforts have been made to pursue clinical trial research. Distinct clinical trial research staff have been hired using charity funds to recruit and monitor patients in clinical trials. At present fourteen studies are completed or underway on a diverse array of topics. Patient enrollment has increased from <5/year in 2006-2007 to 56 in 2009. The review panel was impressed by such a marked improvement in a relatively short time span.

In addition to a developing therapeutics research program, the Centro Regionale Veneto per la Fibrosi Cistica has a long history of extensive high quality CF clinical research with multiple areas of active research including; clinical epidemiology, molecular biology, genetics and CF diagnostics. The clinic boasts an impressive array of publications in these areas.

The clinic is notable for its long and extensive history in neonatal screening programs for CF. Indeed, a screening program has been in effect in variable forms since the 1970s. The newborn screening program has an excellent infrastructure with advanced genetics support. It is well staffed and very well regarded internationally. It was very interesting to note that in its time, the birth incidence of CF has fallen markedly in the region of Trento.

2.15 Future Developments

At present the there are renovations underway to expand and improve the delivery of outpatient pediatric care. These renovations have been occurring over some time and are near completion. This will serve to further segregate adult and pediatric care within the clinic.

The review team noted that the composition of the clinic – while primarily composed of individuals from within the catchment area of Verona still has a large cohort of patients that travel to Verona for their CF care (~30%). The review panel was impressed by the attempts of the clinic staff to repatriate long term attendees to clinics within their originating region in the past. The review panel noted significant time and effort had been spent on teaching and mentoring local services to ensure continued high quality care. The lack of reliable CF care in Millano continues to limit further repatriation attempts in this area. We would encourage the clinic to continue with their previous efforts of developing, educating and mentoring a Millano local CF service.

In terms of future challenges the Verona CF team raised three great concerns. 1. How to best provide the best individual care possible to a growing patient population which has already surpassed 800 in number? The Centro Regionale Veneto Per La Fibrosi Cistica is to the best of our knowledge the largest CF clinic in the world. As such the clinic is subject to unique challenges. 2. How to reconcile the increasingly divergent needs of an aging CF population and the pediatric patients which constitute its historical patient base. 3. How best for a team of pediatric trained CF physicians to continue to provide optimal

care to an ageing CF cohort who are increasingly encountering problems unique to an adult population.

3 The Core Team

3.1 CF Physicians

There are eight physicians employed full time in the Centro Regionale Veneto per la Fibrosi Cistica. The center director is supported by external grants and his role is focused on administration and research. Of the eight physicians all have come to CF via a pediatric training route reflecting the evolution of care within the Centro Regionale Veneto per la Fibrosi Cistica. The aging of the CF patient cohort has enabled them to interact more and more with adult patients. Unlike other CF clinics, no member of the team has received training in adult medicine. Prior areas of specialization among CF physicians include nephrology, gastroenterology and medical genetics. One physician has received prior training in lung mechanics and within his responsibility of CF care he also manages the pulmonary function laboratory and all bronchoscopic studies in CF patients.

CF consultant physicians provided dedicated "in house" coverage 7 days a week, 24 hours a day. This is quite an unusual model compared to other countries where designates (ie senior trainees, fellows, staff physicians) would be in house for direct care with consultants available from home for support as required. All physicians provide overnight coverage to all CF patients (from newborn to post transplantation). While all physicians have a broad core knowledge of CF, many attending physicians have areas of special expertise within the CF team. These dedicated special interest areas include; CF-related diabetes, post transplantation management, gastroenterology, medical genetics and diagnostics/new born screening.

Team members are heavily focused on CF care. Only one member of the medical team also routinely treats individuals outside of the spectrum of CF (gastroenterology). While this allows for the opportunity for the development of excellent core knowledge, several detractors need also be mentioned. Focus on CF does not allow for the cross fertilization of ideas, nor the incorporation of new ideas or practices from other disciplines within CF model of care delivery. With this pattern of practice the peer review team would imagine it would be very easy to develop a unique practice pattern that did not conform to other practice patterns.

Both the stand alone nature of Centro Regionale Veneto per la Fibrosi Cistica and its incorporation with non-University affiliated hospital result in limited opportunities for medical trainees to involve themselves with CF care. A few medical students have specifically approached individual team members to be involved in research projects. Furthermore, the trainees that are interested in CF are usually associated with pediatric training programs. **Increasing the CF teams profile within other disciplines is required in order to generate a familiarity and knowledge regarding the intricacies of CF management for other medical trainees.** Furthermore, the recruitment of future generations of CF physicians is going to continue to be difficult owing to the limited exposure opportunities during their training.

3.2 Nursing

There are eighteen registered nurses and 5 nursing assistants employed full time at the Centro Regionale Veneto per la Fibrosi Cistica. There is one Capo Sal (equivalent to a charge nurse in the UK) who oversees the nursing staff. The nurses all receive on the job training and education around cystic fibrosis. In limited discussions with the nursing staff the review panel came away with the general impression that nursing staff were content with their role in the larger team structure. Nursing staff did feel they had the opportunity to contribute in many ways both on inpatient ward rounds as well as in the multi-disciplinary ward rounds.

The role for nursing staff is focused on the provision of inpatient care. There are three shifts of nurses each with two to four nurses operating. Nurses are assigned to an individual ward and work within a multi-disciplinary team with a physician and a physiotherapist. Nurses are responsible for the administration of all medication and patient / parent education. The majority are competent in accessing totally implantable access devices and insertion of temporary vascular access devices

There are two nurses who assist in the outpatient management of the clinics operation. They are involved in call screening and triage, organizing appointments and patient follow up. A dedicated period exists each day in which patients can phone in to speak to a nurse for advice or to arrange for an urgent assessment.

The review panel was concerned that with the current complement of only eighteen nurses that the system is currently working at or exceeding capacity. Discussions with nursing staff suggested that even one maternity leave or sick day can result in tremendous disruption to the work schedule. Furthermore, with very heavy patient loads there was little opportunity for the nurses to expand the current scope of their practice. There was evidence of significant volume of staff turnover over the last several years. This had created some difficulties as junior staff required education around the unique needs of CF patients. The review panel notes that high staff turnover rates are often related to a high stress work environment.

Continuing medical education opportunities for nursing staff were extremely limited. The nurses reported that on average only three nurses were able to attend national CF conferences each year. There was no opportunity for the nurses to attend international meetings.

There is no current analogous position to a CF nurse specialist within the Centro Regionale Veneto per la Fibrosi Cistica. This position has been seen as extremely advantageous in many CF centers across the UK. CF nurses specialists play a crucial role in many respects including; triaging patients, coordinating on going home care, providing support and education to ward nurses regarding CF. These nurses act to support and supplement physician services, even leading clinics focusing on nursing issues. Some of the CF specialist nurses across the UK can operate as independent prescribers – the review panel was informed that no similar system currently operates in Italy.

3.3 Specialist CF Physiotherapists

There are currently four specialist physiotherapists and one physiotherapist coordinator employed at the CF center. Their sole role and responsibility is in the provision of care to patients with CF, both on an inpatient and outpatient basis.

It was explained to the review panel that physiotherapy is offered twice a day for inpatients and the physiotherapists are also responsible for ensuring physiotherapy support is adequately provided at outpatient appointments. Referral for outpatient physiotherapy can be made both by physicians assessing patients and by physiotherapists based on perceived needs from prior interactions. CF physiotherapists provide assisted physiotherapy 6 days/week to the inpatients. Physical therapy includes manual chest physiotherapy, assisted chest physiotherapy, and exercise therapy.

The review panel did note that inpatient physiotherapy remained primarily supportive – and noted that assisted physiotherapy was preferred by most centers to be superior for individuals with pulmonary exacerbation. The current limited staffing complement may limit this practice. Patients did comment that they noted a reduction in the availability of physiotherapy services. Furthermore, physiotherapy services are limited on the weekend to a single session on Saturday mornings – with no coverage for Sunday. **Efforts to make this service increasingly available should be undertaken.**

Physiotherapists are responsible for assessing all nebulized medications, including; teaching and technique observation, tolerance studies with first doses, and nebulizer maintenance and trouble shooting. Physiotherapists are also involved in the administration and adjustment of non-invasive ventilation for use both as an aid for airway clearance and as a tool in end stage respiratory disease as a bridge to transplantation. Patients did report that over time there seemed to be less assisted physiotherapy available, presumably owing to increasing work load and patient numbers. Staff are committed to the on-going provision of care to CF patients.

Continuing medical education was available to the physiotherapists. Members of the physiotherapy team intended to attend the European CF meeting and were going to present local data.

3.4 Specialist CF Dietitian

The area of greatest concern that was observed by the review panel was dietitics. There is only one full time CF dietician employed at the CF center. She has been in this post for just over one year after replacing a colleague who left abruptly. Unfortunately she has had limited opportunity for CF specific education and furthermore no significant opportunity for mentoring from experienced CF dieticians exists to date. This post is not funded through the regional hospital framework but rather supported by grant moneys from a local CF charity. As such, it is a non-permanent post with no job security.

We did have the opportunity to meet with the sole CF dietician. She was very committed to her role in the CF team. However, she admitted that the sheer volume of work and the pressure put on the dietetic service can be extremely stressful. She is very busy on a day-to-day basis and is often unable to cope with the demand on the service. Furthermore, the

great diversity of patients and problems that a dietitics service providing care to 800 individuals from neonate to post transplantation must be overwhelming.

The dietitics service urgently needs assistance. It is markedly understaffed and under resourced. The practice patterns for advanced nutritional deficiency in the Verona clinic are markedly different than those established patterns of national CF organizations from United Kingdom, United States and Canada. The peer review committee asked for detailed data with respect to BMI on all adult patients. Of the data provided for 464 adult patients, median BMI is 20.8. This value is lower than that observed in other epidemiologic cohorts from the US (21.4)[2], UK (21.6)[4] and Canada (22.1)[1]. BMI was shown to improve with increasing age as with other datasets. In particular, 10.3% of the adult clinic population had a BMI in the less than 18 kg/m^2 category and 36% < 20 kg/m^2. These values are markedly lower then values reported from other CF clinics.

The dietetic service of the CF clinic strives to improve the nutritional status of patients. There is quite correctly an emphasis placed on "real food" being the appropriate way and means of weight gain. However, there the review team gathered there was a strong disincentive for the use of nutritional supplements even in those individuals failing conventional management. In the UK 32.4% of patients will receive supplemental feeds after failing to optimize nutritional therapy through traditional means. Furthermore, in the UK 8.4% of patients receive supplemental nutrition through either a gastrostomy tube (6.6%), or via nightly nasogastric tubes (1.8%). **The review panel were very surprised to hear that in ten years no patients had received enteral nutrition via a PEG.** Apparently, for advanced nutritional failure particularly in those individuals awaiting transplantation we were told that home TPN would be common place. The review panel were concerned that this practice markedly increases risk of infectious complications and rapidly consumes limited financial resources. In contrast only 0.03% of adult patients in the UK would have received TPN owing to the perceived extreme nature and cost of this intervention.

3.5 Social Workers

The review panel did not have the opportunity to meet with the social worker of the Centro Regionale Veneto per la Fibrosi Cistica. As such, our understanding of their role within the team is limited. This individual is primarily responsible for assisting patients in the process of applying for and receiving state sponsored income support. We were unable to assess their involvement in a number of areas including; family and peer support, employment advice and assistance, home visits and patient advocacy. Compared to our own experience this service would seem to be understaffed.

3.6 Psychologist

The review panel did not have the opportunity to meet with the psychologist of the Centro Regionale Veneto per la Fibrosi Cistica. As such, our understanding of their role within the team is limited. This individual has been with the team for close to three decades. We were unable to assess their involvement in a number of areas including; individual and family counseling, diagnosis and referral to psychiatric services for

pharmacologic management of mental illness. **We understand from prior patient surveys it was psychosocial support that patients noted the greatest deficiency in center resources.**

3.7 Pharmacist

No clinical pharmacist is employed by the Centro Regionale Veneto per la Fibrosi Cistica. All prescribing in hospital is completed by physicians. Medications are made up in the general pharmacy and administered by nurses. For the provision of medications to outpatients, center physicians write prescriptions that are then faxed to local hospital pharmacies to fill. Local hospital pharmacies are responsible for independently negotiating prices with each drug supplier.

The panel felt that taking into consideration the number of patients cared for at the Centro Regionale Veneto per la Fibrosi Cistica, it is important to secure a dedicated CF clinical pharmacy service. We would estimate that up to three pharmacists would be required to appropriately staff this service. These individuals should be fully incorporated into the CF multi-disciplinary team and be able to participate in all elements of care, including outpatient consultations and annual review. **The panel confirmed that in their experience a CF pharmacist usually pays for themselves in terms of the savings that can be made by close monitoring of medications**, including adherence to treatments (reducing wastage), consideration of alternative treatments (sometimes resulting in more cost-effective provision), sourcing of bulk purchases and negotiating incentives, etc. Certainly a centralized CF pharmacy could enable significant cost savings.

3.8 Database maintenance

The review panel was particularly impressed by the database developed and maintained by the Centro Regionale Veneto per la Fibrosi Cistica. While originally started in 1993, several revisions and new iterations of the database have come in this time. At present all patient interactions are captured. Some data is manually inserted and others (such as laboratory values) are automatically updated. The database is comprehensive and includes historical data such as genetics, family history, means of diagnosis, sweat chloride values, etc. The database is clinically useful and generates combined reports at the end of clinics, coordinates appointments and ensures patients and care providers get copies of each note. Data is maintained on an independent server and maintained by two qualified information technology technicians.

4 Patient Interviews

The review panel had the opportunity to meet with only two patients and did not conduct an independent survey prior to the review. Both patients felt quite confident they received top quality care at the Centro Regionale Veneto per la Fibrosi Cistica. Patients felt particular strong bonds with the physicians whom they saw on a regular basis – and admitted less confidence in those team members whom they had less prior exposure. Patients admitted that nursing practice had improved in the last several years, though there was still concern about some nurses were unable to access ports. The patient who is also followed by one of the satellite clinics admitted greater confidence in Centro Regionale Veneto per la Fibrosi Cistica staff. Both patients also commented on the difficult nature of access to the center, and the very limited availability of parking. For those individuals on supplemental oxygen therapy, handicap parking passes were available. All other individuals had to find their own arrangements.

An anonymous survey of patients was conducted by Centro Regionale Veneto per la Fibrosi Cistica staff in 2006. We did not have access to this data, however, reported concerns were predominately regarding lack of psychosocial support and parking.

5. Quality Assurance Officers

The review panel did have the opportunity to meet with the Clinical Director and Quality Manager. They reiterated that the CF service operates autonomously within the hospital structure. They explained that hospitals are certified according to regional and national standards. Quality is monitored internally within the hospital structures. Both managers were confident that the CF unit was delivering a high quality service with a high level of patient satisfaction. They did report that in six years they had not received any complaints regarding Centro Regionale Veneto per la Fibrosi Cistica.

6. Funding

Funding for the CF service comes directly from federal government. It was estimated that the service costs approximately 3,000,000 euro per annum to run, with the recurrent annual funded budget currently 1,000,000 euro. The resulting deficit of 2,000,000 euro is currently met at end of year. **The service however is grossly underfunded on a recurrent basis.** Although not clearly stated by the CF team, the peer review committee would question the implication of this underfunding, for the service. There does not appear to be a clear mechanism for forward business planning, in particular, making the case for additional staffing requirements. It has been previously noted that the dietitian post is funded by a patient association on a non-recurrent basis.

5. References

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3. Standards of Care: Standards for the clinical care of children and adults with cystic fibrosis in the UK. May 2001 Cystic Fibrosis Trust.

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