CYSTIC FIBROSIS MANUAL – PSYCHOSOCIAL MANAGEMENT - CHW

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

The chapters of the CHW Cystic Fibrosis Manual have been divided into individual documents. The chapters are:

1. Making the Diagnosis	2. Management of the newly diagnosed patient	3. Respiratory Management
4. Gastrointestinal Management	5. Nutritional Management	6. Psychosocial Management
7. Adolescent Management and Transitioning to Adult Care	8. Outpatient Management	9. Endocrine Management
10. Palliative Care	11. Cross Infection	12. Miscellaneous Associated Conditions
13. Therapies for Aberrant CFTR Function	14. CF Pharmacopoeia	

Psychosocial Management

- Psychosocial wellbeing is an increasingly recognised area of importance within CF care
- Age specific factors of note should be appreciated when evaluating CF children

Approved by:	SCHN Policy, Procedure and Guideline Committee	
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Team Leader:	Clinical Nurse Consultant Cystic Fibrosis	**Area/Dept:** Respiratory

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CHANGE SUMMARY

- Incorporation of recent assessment of mental health issues amongst our clinic
- Updated roles within the team across the CF social worker and psychologist are outlined

READ ACKNOWLEDGEMENT

- Cystic Fibrosis Multidisciplinary team members are to read this guideline.

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Introduction

- CF has a significant impact on the emotional and physical development of the child, young person and their family.
- The TIDES study is a recent multicentre study across 8 countries that investigated rates of depression and anxiety in >1000 adolescents and >2000 caregivers[1, 2]. It found that:
  - 25% of adolescents expressed depression and 20% expressed anxiety
  - 35% of caregivers expressed depression
  - Rates of 2-3 times the general population
  - Detrimental effects were seen on adherence and clinic visits
- A similar, but smaller, study was conducted in the under 12s at CHW. Our results showed that:
  - 8% school-aged children (8-12 year olds) experienced depression and 21% expressed anxiety
  - 0% of younger children (under 7 years) experienced depression and 2% expressed anxiety
  - 8% of caregivers experienced depression and 28% of caregivers expressed anxiety

1 General Considerations

- **Emotions Experienced**
  - Throughout the course of the child's life, it is usual to expect that the child and adolescent with CF, their parents and perhaps other family members will experience a wide range of emotions.
  - These can include feelings of sadness, grief, anger, confusion, anxiety, fear, guilt and resentment.
  - Different family members may feel different emotions at different times, and each family member's reactions might change over time.
- **Religious and Cultural Beliefs**
  - A family's religious and cultural beliefs with regards to illness need to be considered.[3]
  - This may influence aspects such as their attitude to medical intervention, feelings about hospitalisation, and feelings about end of life care and death.
- **Relationship Issues**
  - CF may place enormous strain on relationships. It is vital that areas of potential conflict be identified immediately and possible solutions sought.

- **Family Issues**
  - CF impacts on family life. All family members need to adjust to the treatment demands of having a child with CF, and some may achieve this adjustment more readily than others.
  - Higher levels of distress and an avoidant coping style are associated with poor psychological outcome.(4)
  - Support may be necessary for family members other than patient and parents.
- **Support Systems**
  - A family's available support networks influence their long-term adjustment to CF.
  - Low levels of family support are associated with poor psychological adjustment.(4)
- **Developmental Milestones**
  - Major developmental stages and transitions often pose a crisis for children with CF and their families and may precipitate the re-emergence of emotions such as grief and loss.
  - Ongoing assessment and psychological support are vital during these stages.
- **Childhood and adolescence**
  - The diagnosis and impact of living with CF has a significant impact throughout the lifespan.
- **Peer Relationships and Activities**
  - Consideration should be given to a child or young person's life outside the family, particularly with regard to school environment and peer relationships.
  - Many have problems with interpersonal relationships which can result in isolation and social maladjustment.(5)
- **Interaction with Hospital Staff**
  - Some families may feel they are constantly being "watched" and judged by health professionals.
  - Parents in particular need to be affirmed in their "expert" role with the child, and encouraged to actively participate in decision making with regard to treatment.
- **Adjustment Issues**
  - Sometimes children and adolescents with CF require individual counselling if they experience major periods of non-adherence, profound distress and difficulties adjusting to their illness.
  - Referral for assessment and support around adherence, adjustment, transition and family issues can be helpful.
  - In some instances families, children and adolescents may experience major psychiatric disorders.

2 Infants, Children, and Adolescents

- CF may create a wide range of issues and feelings for infants, children, and adolescents, with some of these issues also being common to children without CF.
- Living with a chronic illness interacts with normal development and increases the complexities of the issues that arise.

These issues can change across development and include:

Infancy and early childhood

- The development of the parent-child relationship, otherwise known as attachment, often coincides with the time of CF diagnosis which can be linked to stress, anxiety, and parental distress[6].
- Maladaptive attachment in CF infants can lead to poorer nutritional status and lower BMI[7].
- Parents are often adjusting to the role of parents versus the "medical parent"
- Young children start to become more aware of their CF and daily treatments, e.g., up to 40% have poor compliance with ACT physiotherapy[8].
- Potential to develop behavioural reactions and anxiety reactions around CF treatments and medical procedures.
- Nearly half of young children with CF have moderate to large sleep and/or eating problems[8].

Children

- A feeling of being different from their peer group due to comparisons such as physical differences, school absences, taking medication.
- Experiencing negative peer reactions such as teasing but also including overprotection
- Increasing awareness of treatment burden
- Being jealous of their healthy siblings.
- Anxiety about medical procedures e.g. needles etc.
- Difficulties with compliance that first emerged in early childhood continue in this developmental period, generally with even greater oppositionality.
- Potential to develop maladaptive coping behaviours e.g., trying to hide their illness which leads to poorer adjustment in adolescence and adulthood.

Adolescents

- Difficulties balancing the demands of their illness with school, family and peer responsibilities .e.g., managing peer group pressure around risky behaviours such as drinking, drugs, vaping.
- Being self-conscious or embarrassed about their illness e.g. having to take enzymes, coughing, body image etc.[9]
- Concerns about falling behind academically due to frequent hospital visits or periods of hospitalisation.
- Potential to develop psychological issues, especially anxiety and depression.
- Feeling of having no control over their lives.
- Feeling that decisions are made without their input e.g. by doctors, nurses, social workers, parents.
- Concerns about their future and their own mortality.
- Loss/death of friend/sibling or relative from CF.
- Concerns about the burden their disease places on their parents.
- Concerns about medical intervention and the impact this will have on their appearance (insertion of port and gastrostomy).
- Fear of complications e.g. CF related diabetes or liver disease.
- Issues relating to fertility, sex, drugs and alcohol.
- Fears, anxieties and dreams about what the future holds, including the potential negative impact of CF on future employment opportunities [10] and fertility.[11]
- Non-adherence is a commonly encountered problem in CF, present in over 50% in one adult CF study.[12]
- May be accidental or intentional, with simple forgetfulness reported as a common cause.[13]
- Postulated reasons include inadequate knowledge, psychosocial resistance and educated non-adherence. [14]
- For further information see Cystic Fibrosis Manual - Adolescent Management Issues and Transition to Adult Care Guideline.

3 Parents

- Parents with children who have CF may struggle at different times with different aspects of the disease and how it impacts on family life.
- Mothers may become overly involved and protective, leading to enmeshment, whilst fathers may be involved less leading to withdrawal.[15]
- Depression has been reported in 30-35% of mothers of children with CF.[1, 16] Decline in a child's health can lead to increased family stress.[15]
- Some of the issues encountered by parents may include:
  - A belief that the disease is somehow their fault, given the genetic component of CF.
  - Grief issues related to the loss of a "healthy child".
  - Continual vigilance around preventing their child from developing CF related bacteria e.g. *Ps. aeruginosa*, *B. cepacia*.
  - A belief that CF can be controlled e.g. by doing more physiotherapy, not allowing children to mix with peers.
  - Fear of the future and long-term implications of the disease.
  - Achieving a balance between being supportive and structured around their child's treatment however not being over protective.
  - Financial constraints compounded by the illness
  - The process of "letting go " i.e. handing over responsibility for management of treatment to their adolescent.
  - Communication difficulties between parents.
  - Concerns regarding genetic counselling, further pregnancies and antenatal diagnosis
  - Balancing the needs of the child with CF and the needs of other family members.[17]
  - Disruption caused by clinic visits and periods of hospitalisation.

4 Siblings

- Chronic illness impacts on all family members.
- It is not unusual for siblings to experience struggles related to living with a person with CF, and it may be necessary to provide family support and intervention to assist in the resolution of such issues.
- These may include:
  - Feeling jealous about extra time and attention the sibling with CF requires.
  - Frustration with the restrictions CF places on family routines and lifestyle.
  - Feeling concerned about their sibling particularly at periods of deterioration Feeling embarrassed about their sibling having Cystic Fibrosis.
  - Feeling confused about what is happening in the family, when other family members are distressed.
  - Confusion about health and illness, in particular, unintentionally seeing illness as the only way to gain attention in the family.

5 Grandparents

- Grandparents and extended family are also be affected by the diagnosis of CF.
- They may experience issues such as:
  - Feeling responsible and guilty that they passed on the disease.
  - Feeling isolated due to lack of current knowledge about CF.
  - Feeling fearful about minding their grandchild due to the treatment they require.
  - Feeling a strong sense of identification with their own child and the grief they may be experiencing.
  - Becoming over-involved and overly concerned about their grandchild.
  - Unintentionally creating conflict especially around issues of nutrition and physiotherapy, due to their lack of understanding about CF.
- In addition to the above, the presence of pre-existing personal, family and situational problems can be exacerbated by an illness like CF.
- The early identification of these issues and the prompt intervention by a social worker is therefore paramount.
- Grandparents can be very supportive and greatly add to the quality of the family system.
- To enable this to happen, involve them in CF education, attend clinic visits, learn physiotherapy techniques and become familiar with and reinforce the treatment routine

6 Referral for assessment

- A number of teams provide input to screen and manage psychosocial issues as they arise.
- Within the CF team, there is a social worker and clinical psychologist who work closely with patients and their families when psychosocial issues arise. Broadly speaking, the CF social worker focuses on:
  - Adjustment to illness across development e.g., grief, anger, fear
  - Parenting support and strategies across development
  - Practical support such as Centrelink
- The CF clinical psychologist provides support on:
  - Parent-child difficulties in early childhood
  - Psychological issues in children and adolescents e.g., anxiety, depression, medical trauma
  - Family therapy
  - Yearly mental health screening at annual review
- At times, the child/family will be referred to the Adolescent Medicine Unit or to the Department of Psychological Medicine, when appropriate, as outlined below:

Adolescent Medicine Unit

- Issue of compliance and adherence
- Family distress and coping
- Minor mental health concerns, not deemed to require cognitive behavioural therapy or psychiatric medication

Department of Psychological Medicine

Kids with severe disease likely to require lung transplant.

Kids with complex social and /or mental health needs

Kids outside Metro Sydney with complex psychosocial needs.

- Prior to being seen by psychological medicine, the following must be provided or ensured for all referred adolescents:
  - A clear mental health question articulated by the referring physician.
  - Referral is made in consultation with the CF social worker or psychologist.
  - That no other psychosocial team is currently involved with the child/family, to avoid undue replication (e.g. AMU, ICAHMS).
  - If another team are involved, there must have been adequate discussion, and endorsement of any planned referral.

- There is an expectation that this team will liaise with CHW psychological medicine as well, during their period of involvement.
- In specific circumstances CHW psychological medicine will review and hand back to the local services when appropriate.
- Reasons for referral have been discussed with the family and the family are willing to engage.

7 Medication options[18]

- Whilst several medication options exist, the choice of medication, if indicated, should be made after discussion with Psychological medicine/AMU.
- Options include Selective serotonin reuptake inhibitors (SSRIs) such as Fluoxetine, which has the best available evidence base and is most widely used in adolescents.
- It is important to consider the use of medication in conjunction with other therapies or strategies which are additionally useful (e.g. cognitive behavioural therapy, family based therapy)
- Close monitoring for adverse events and response to therapy is essential
- Consideration of potential interactions with existing medication should also be considered.
- Further details about medication options, side effect profiles and drug interactions are given in below in Table 1

Table 1 Anti-depressants Used in Adolescents *(**Note**: These medications must be initiated and doses changed by an adolescent medicine specialist or child psychiatrist.)*

Drug	Initial Dosage	Preparations	Notes
Citalopram	10mg daily	Tablet 10mg Tablet 20mg Tablet 40mg	Considered second line Usually given in the morning May cause QT prolongation
Escitalopram	5mg daily	Tablet 10mg Tablet 20mg Oral solution 20mg/mL	Considered third line Usually given in the morning May cause QT prolongation
Fluoxetine	5mg daily	Dispersible Tablet 20mg Capsule 20mg	Considered first line Usually given in the morning Not recommended in liver disease
Fluvoxamine	25mg daily	Tablet 50mg Tablet 100mg	Give with food to reduce GI upset Usually given in the morning but sedation is a common side effect; if this occurs, give at night.
Sertraline	25mg daily	Tablet 50mg Tablet 100mg	Usually given in the morning. Dose reduction may be required in liver disease.

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