

MINI-SYMPOSIUM: CYSTIC FIBROSIS

# Cystic fibrosis psychological issues

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team approach

**Summary** To most parents the diagnosis of cystic fibrosis (CF) in their child represents a severe blow because they are confronted with an unwanted and unexpected disease that completely changes their whole life. Common reactions such as shock, denial, sadness and anger have to be mastered before a gradual adaptation to reality will be possible. The provision of support by a multidisciplinary team, including a psychologist, that offers its services from diagnosis and puts an emphasis on preventive care may help to achieve, maintain and improve physical and mental health, and social functioning in both patients and parents. CF not only affects the individual but the whole family, and the presence of biopsychosocial stressors may add to the burden caused by the disease. CF does not necessarily cause long-term serious family dysfunction, but it changes family structures and often taxes the family system beyond its strength. Even if there is only partial adherence to the demanding and complex treatment regimen, health professionals need to acknowledge the tremendous underlying effort on the part of the families. As a consequence of their continuous endeavours, many individuals with CF do lead remarkably normal lives with the prospect of gene therapy and lung transplantation, maintaining hope in case of severe deterioration.

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## INTRODUCTION

An incurable disease such as cystic fibrosis (CF) certainly has an impact on patients, their families, and their caregivers. There is a body of literature indicating that children with long-term illnesses, including CF, are at greater risk of having significant emotional or behavioural problems. However, despite these problems, individuals with CF do lead remarkably normal lives until the terminal stages of the illness. The term 'normal' refers to those aspects of life where children with CF appear to be very much like other children. As an example, CF does not appear to be a major obstacle to school attendance although disruption of classwork by absence, coughing, and need for medication may cause transient discomfort. Overall, progressive improvement has been described for the whole CF population in terms of quality of life and life expectancy. Although this is promising information, we

must not underestimate or downplay the emotional burden that is associated with the disease.

## COMMUNICATING THE DIAGNOSIS

The majority of children are diagnosed at age 6 months. They grow up with the management of CF being part of their lives and gradually, dependent on age and maturity, become aware of the disease and its implications. In contrast, most parents are adults who are thought to be able to perceive the meaning of CF from the very beginning. To them the diagnosis of CF represents a severe blow, confronting them with traumatic information. In short, parents are exposed to the following facts: you are healthy; you carry faulty genes and that is why CF was transmitted to your child; at present, this disease is incurable, it will last all life and the future health development of your child is hard to predict; provided that your child is in a stable condition, you will spend an average of 1 to 2 hours a day following a multifaceted treatment regimen

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including inhalation, physiotherapy, high-energy diet and medication; compared to the general population, the life expectancy of your child is markedly diminished.

## REACTIONS TO THE DIAGNOSIS

Many parents of a child with CF progress through a predictable sequence of emotional states: shock about the unexpected and mostly unknown disease that completely changes their whole life, denial that the disease really exists, and sadness, anger and disappointment about the child who does not meet parents' initial hopes and expectations. Heavy emotional loads often produce effects on mental processes and thinking. Parents may suffer from dulling of their ability to think clearly, to concentrate or to pose questions. Jedlicka-Köhler *et al.* described initial shocklike reactions in 54% of parents and reported that the occurrence of shock led to a significant decrease in the understanding and recall of the information given.<sup>1</sup> The essence of denial is the refusal to acknowledge certain aspects of reality because it is painful, distressing or threatening. Bad news is almost always difficult to take in, hence to escape the information or to doubt it is a very common first reaction to it. Parents who are in the denying stage may want to seek a second opinion and should not be discouraged from doing so. Only prolonged denial that is maintained despite sufficient evidence that the diagnosis is correct may be harmful to the patient if he or she is prevented from adequate therapy. There is no list that can exhaustively describe parents' feeling states. Intense feelings of fear, disappointment and sadness are the most frequent emotional reactions. Anger against many targets is another common emotion, and sometimes the caregiver is the primary target. Feelings of guilt, despair and depression add to the emotional turmoil. It is a challenge for both the families and the health-care professionals to cope with the described phenomena. The knowledge that parents' reactions are predictable and not pathological and will gradually lessen over time should aid the health professionals in their attempts to provide support and establish a satisfactory relationship. Although it is impossible to determine the length of particular stages of parental reactions, the stage of adaptation will be reached in most cases even if asynchronous courses of the mother and father complicate this process. From then on more energy and strength will be available for the development of coping strategies.

At the early stage of being informed about the diagnosis, parents lack knowledge about the disease and are predominantly impressed by the threatening facts. They have difficulty imagining that the future may hold positive events. Even if they want to be optimistic, they often lack facts that justify optimism. Therefore, in addition to breaking the bad news, it is important to give positive messages. Parents should learn that intellectual functioning is distributed normally and that children with CF are

typically able to perform at the same academic level as their peers.<sup>2</sup> Adolescents with CF are able to maintain a good self concept and be socially competent.<sup>3</sup> A high proportion of adults with CF live full and productive lives.<sup>4</sup> By citing such results, caregivers may contribute to helping parents develop hope and to broaden their view, inasmuch as life contains many facets, CF being just one.

## THE BIOPSYCHOSOCIAL BACKGROUND OF THE FAMILIES

Children live within an environmental context with the family being the most important component of that environment during the first years of life. The family affects the child's adjustment, and similarly the child's disease affects the family's functioning. Only recently has a relationship between family functioning and a child's physiological functioning been demonstrated. When families emphasized the autonomy and individuality of each family member, the FEV<sub>1</sub> trend of the child with CF declined. When parents employed a broad repertoire of different coping behaviours that equally met individual needs, family needs and medical needs, the FEV<sub>1</sub> trend improved.<sup>5</sup> In a study by Lewis and Khaw<sup>6</sup> children with CF had higher frequencies of behaviour problems than healthy controls. When controlled for the effects of family functioning, this difference was no longer significant, i.e. family functioning significantly contributed to behaviour problems whereas the disease per se did not.

Are families with a child with CF different from other families? It may be difficult and sometimes impossible to distinguish between disease-related problems and problems of alternative origin. In order to gain more insight into potential stressors, it is helpful to take epidemiological information, negative life events and family-oriented theory into account. Such considerations may serve to increase the awareness of both the explanatory power of non-disease-related difficulties and the impact of CF not only on the affected individual but on each family member.

Individuals with CF and their families are heterogeneous. They come from different social backgrounds, economic strata, and educational levels. They only share in common that CF impacts on their lives and that the existence of the disease is an irreversible fact. A common presumption is that family life is happy and harmonious. Another common presumption is that children, and in particular chronically ill children, have the support of loving, caring, physically and psychologically vital parents. Both presumptions may prove right in some families, but may reflect desires rather than reality in others.

Epidemiological data reveal the prevalence of a disease in a large population. Such data are also relevant to a random CF sample, as they give health professionals an idea of diseases other than CF that may affect the families. A few examples may serve to demonstrate this. The lifetime

prevalence of psychiatric disorders, i.e. the proportion of the sample which ever experienced a disorder, was 49% among persons aged 15–54 years.<sup>7</sup> The most common disorders were major depression (17%) and alcohol dependence (14%). The Vienna CF centre reported a lifetime prevalence of psychiatric disorders of 18% among 162 parents. Alcohol dependence was the most frequent disorder in fathers (7%); in mothers recurrent depressive disorders predominated (9%). In all cases of alcohol dependence the child was born into these circumstances. Physical health was impaired in 12% of the parents, including only incurable conditions that required continuous medical monitoring and treatment.<sup>8</sup> Given overlaps, these data clearly show that in approximately 20% of families there is a need to care for an individual with CF and, additionally, for a mentally or physically ill parent. As parents are the primary caregivers of the individuals with CF, it seems reasonable to assume that the resources in such families may easily be dried up. There is evidence that parental psychiatric disorders significantly contributed to poor adherence to outpatient visits<sup>8</sup> thus impairing regular health monitoring and preventive care of the child with CF.

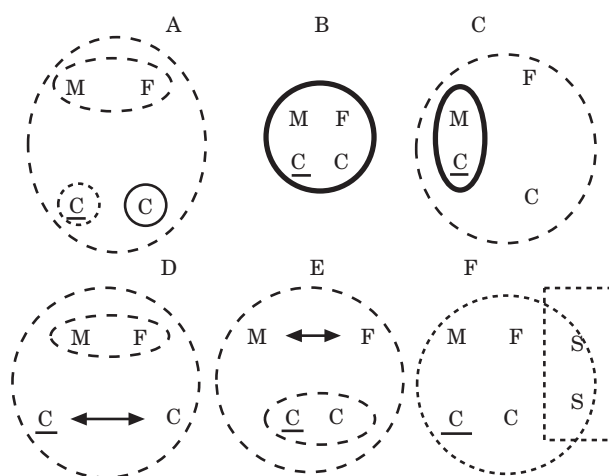
Most people have to cope with negative life events such as divorce, periods of unemployment, financial problems, etc. Divorce rates have reached historically high levels over the last years. At present, in many European countries as well as in the USA one out of three marriages ends in divorce. At the Vienna CF centre, 28% of 179 individuals with CF grew up in less-than-ideal family circumstances, i.e. in incomplete families. They lived in single parent households (5%), experienced — mainly in childhood — divorce (15%), witnessed ongoing marital discord (5%), or had been placed in foster care (3%).<sup>8</sup> These results show that the family environment itself can be a considerable source of stress, meaning that individuals with CF have to cope with a variety of problems in addition to CF. Therefore it is important to know and recognize the impact of the family background because this knowledge may alleviate identification and interpretation of problems presented within the complexity of CF. Health-care professionals who have been following families for many years would probably agree that in some or many cases disease-related problems are outweighed by psychosocial problems. As with psychiatric disorders, the presence of the stressor 'incomplete family' also significantly contributed to poor adherence at clinic visits.<sup>8</sup> Hence, already the basis of regular medical treatment may be weakened by less-than-ideal family circumstances.

## THE IMPACT OF CF ON FAMILY STRUCTURES

Lessons learned from structural family therapy may serve health professionals to better understand common

interaction patterns related to a chronic disease. According to Minuchin,<sup>9</sup> family structure is the organized pattern in which family members interact. A family carries out its functions by differentiating into subsystems of members who join together to perform certain functions. The enduring family subsystems includes the spouse subsystem (for marital functions and adult mutual support), the parental subsystem (for parental functions and nurture of children), and the sibling subsystem (for peer socialization). Individuals, subsystems, and whole families are demarcated by clear (---), diffuse (.....) or rigid (—) boundaries (Fig. 1A). Boundaries are invisible barriers that regulate the amount of contact with others and manage proximity and hierarchy. CF does not necessarily cause long-term serious family dysfunction, but it often taxes the family system beyond its strength. Some examples may illustrate how family functioning is challenged by the need of adjusting to CF.

The newly diagnosed family who feels overwhelmed and fears communication about CF may be surrounded by a rigid boundary (Fig. 1B). Such a boundary can serve to protect the family members, but in the long run withdrawal and isolation would hinder interaction with the environment. CF usually requires time-consuming and demanding treatment. A mother and her child with CF may form such a tightly bonded subsystem that it excludes the father's involvement in parenting and disease management (Fig. 1C). As a consequence, disengagement on the part of the father may result. Recurrent episodes of weakness or hospitalization may result in disruption of the sibling's subsystem, with both children temporarily losing their playmate (Fig. 1D). Sometimes the healthy sibling needs to be sent away from home or left in the hands of unfamiliar persons. Marital satisfaction (Fig. 1E) is jeopardized if caring leaves too little opportunity to



**Figure 1** The family subsystems, demarcated by clear (---), diffuse (.....) or rigid (—) boundaries. M=mother, F=father, C=child, C=child with CF, S=support person.

constantly renew the partnership. The spouse subsystem needs a clear boundary to separate it from parents and children, but sometimes parents lose sight of their need to sustain a loving couple relationship. If the tasks of caring for the child with CF exceed the available resources (Fig. 1F) the involvement of members of the extended family (particularly grandparents) in executive functions may be necessary. This may result in diffuse boundaries between parents and grandparents and lead to conflict if their greater power and influence cannot be handled appropriately.

## A MAJOR PROBLEM: ADHERENCE TO TREATMENT

To increase awareness of the difficulties regarding adherence, it makes sense if caregivers ask themselves the following question: why do we expect families to cope with a disease that came unexpectedly and completely changes one's life? A further question could address the complexity and intensity of treatment: why do we expect individuals with CF to adhere to a time-consuming and often boring regimen that interferes with daily activities, reminds the patient of the disease and is often associated with the problem of disclosing CF to others? It is likely that the caregivers' answers point to the fact that appropriate treatment seems to be the only alternative to stop progression of CF. Presumably, many patients would approve. Yet neither knowledge about the disease nor its severity increased adherence in adult patients with CF.<sup>10,11</sup>

Given the priority and necessity of medical treatment, the crucial point is that following treatment recommendations is predominantly a psychologically and socially determined act. Whether or not a patient adheres to prescribed treatments depends on many variables, including attitudes, motives, subjective interpretation of information, locus of control, and anticipated benefits. In addition, the clinician's estimation may not be equivalent to the patient's estimation of disease severity, resulting in an often unrecognized discrepancy. A recent study showed that adults with CF considered the disease to be less severe than their physicians. While 83% of patients rated their health as above or well above average compared with others with CF, only 35% of physicians considered the patients to have mild disease.<sup>12</sup>

Patients are seldom completely non-adherent if all important aspects of daily CF management are assessed. Adherence with components of treatment varies according to the perceived unpleasantness and degree of infringement on daily activities.<sup>11</sup> Patients with CF adhere better, for instance, to taking pancreatic enzymes than to physiotherapy and taking of vitamins.<sup>10</sup> The fact that many patients only partially adhere to medical recommendations and that so many variables influence adherence need not create pessimism. If the caregivers are willing to understand that in the light of never-ending demands

even partial compliance is a result of tremendous effort, they will enhance their effort to individualize treatment and to adjust it to realistic goals.

Lask suggested five main principles for communicating and improving adherence: empathy, enthusiasm, exploration, education, and expression of emotion. In contrast to coercion, the emphasis is on a warm, non-judgemental approach by a caregiver who communicates confidence, takes time to explore the patient's reason for poor adherence, provides repeated information, and allows the patient to express all sorts of feelings.<sup>13</sup> For the promotion of adherence in asthma, Taggart<sup>14</sup> suggested five strategies summarized by the 'five Rs for teaching': reach agreement on goals, rehearsal, repetition, reinforcement and review. In a step-by-step process the first aim is that the doctor and patient together establish realistic goals and design a realistic treatment plan. The patient then needs to learn and rehearse management skills which should be supported by repeated information and demonstrations. Reinforcement by means of praise and thanks, reward systems, reassurance-calls to the patient, support groups, etc. is critical to shape and maintain the desired behaviour. By constantly reviewing the success of the management plan with the patient, barriers to adherence can be identified and alterations discussed.

## TEAM APPROACH

The care for patients with CF should be viewed within the context of a biopsychosocial concept of health. Appreciating the biological, social, and psychological factors and their interdependence may increase the efficacy of treatment and improve quality of life. The weighing of the above factors over the course of the disease will then determine the type and priority of interventions. The nuclear team which delivers the main part of care usually includes paediatricians/adult physicians, nurses, psychologists/psychiatrists, physiotherapists, nutritionists, social workers, and lung function personnel. All health professionals involved should provide their services from the very beginning, that is at the time of diagnosis, and all should put emphasis on preventive care. Although the focus of attention is quite different between the professionals involved, the aims of the caregivers are identical: to help achieve, maintain and improve physical health, mental health and social functioning. Ward rounds and team conferences are essential for appropriate communication and effective multidisciplinary care. In addition to the nuclear team, a number of other disciplines, e.g. transplant surgeons, geneticists, andrologists, function as experts on request. Their services are intermittent and case-dependent.

A few considerations may serve to outline basic principles of the psychologist's approach. It seems reasonable to provide preventive care and anticipatory guidance rather than perform crisis intervention only. If comprehensive care starts from diagnosis, psychologists

have the opportunity for early intervention and can support the families before significant emotional or behavioural problems develop. Regrettably, not all health professionals recognize the fathers' influence on health care and disease management. Lack of paternal support and mothers carrying the major responsibility for the child's health needs are two common patterns that mirror the blind spot of the caregivers rather than reluctance in the fathers. Therefore, to motivate both parents, mothers and fathers, to share the responsibility for the child's health needs may well be a psychological task. Psychologists can help to disclose all the capabilities, strengths and resources the families have as a means to increase their confidence in their potential and efficacy to fight CF. Given the vulnerability to any possible stressful life event, a thorough and continuously updated documentation of the family background may be the basis for identifying the origin of problems presented to the CF team.

Health professionals simultaneously treat patients at all stages of the disease, including severely ill and dying patients. Similar to the families with CF, health professionals may also find themselves under pressure at times. Lewiston *et al.*<sup>15</sup> investigated 'burnout' in CF caregivers. They reported that caregivers who spent more than 50% of their professional time with CF had a significantly increased level of emotional exhaustion compared to those with 20% or less. Emotional exhaustion was defined as a depletion of the emotional energy available to maintain the caring and commitment needed to the job. When the first group (>50%) was divided by burnout scores, those with greater scores than the mean for the entire group had a significantly higher percentage of time spent with sick hospitalized patients.

## CONTROVERSIALLY DISCUSSED ISSUES

Apart from daily concerns, there are a range of controversial issues that challenge both families and health professionals. Examples are: living donor lobar lung transplantation, pre-natal diagnosis, reproduction, and gene therapy. They represent scientific advances, which may be perceived as a ray of hope, as a benefit, but also as a threat. Apparently, the offer of additional choices not only enriches but aggravates decision making.

Lung transplantation (LTX) has become a viable option for end-stage lung disease. Although it is sometimes misperceived as a cure instead of another treatment modality and will only be available to selected patients, it gives hope to many. Thinking of LTX caused anxiety in most parents (89%) of children with CF, but at the same time was viewed as progress in medicine (98%) that warranted the expensive research (93%).<sup>16</sup> Due to medical and financial factors many patients do not survive to transplant. In addition, the supply of replacement organs cannot meet the demand and it is likely that cadaver

organs will remain in short supply for the foreseeable future in most countries. The most recent development was successfully aimed at reducing this problem. Lung lobes from living donors have become an additional source of lungs, with two donors each giving one lobe to the recipient. Donor groups are parents (32%), other relatives (50%) and non-related donors (18%).<sup>17</sup> Reasons for concern addressed the initially anticipated undue pressure on family members and the potential harm to support persons. In order to avoid coercion and to guarantee objective information, adult donor candidates undergo a medical examination and a psychological evaluation separately and independently of the CF team. There have been no major complications to the donors.<sup>18</sup> Yet the dilemma to donate or not, the psychological burden imposed on potential donors, and the uncertainty as to what the right decision might be for both the donor and the recipient must not be neglected. To be involved in the question of prolonging someone's life is an extreme challenge per se, and is inevitably associated with a wide range of emotions independent of the resulting decision.

Pre-natal diagnosis by DNA analysis is associated with the question of whether a life with CF is worthwhile. Due to the nature of this question, views and attitudes in society are many and determined by a variety of reasons. Adults with CF supported pre-natal screening in antenatal clinics (88%), pre-natal testing in couples who already have a child with CF (89%) and the option of terminating an affected pregnancy (68%).<sup>19</sup> For parents with a child with CF, the greatest concern centres around the difficulty in having to make a decision about giving birth to an affected child. Before its availability, the majority of parents with a child with CF favoured the development of fetal diagnosis and intended to have recourse to it in further pregnancies. However, when actually presented with a decision about pre-natal testing, this procedure was only arranged in 21% and performed in 17% of cases. The anticipated fear of an unsolvable conflict in case of an affected fetus, along with the fear of possibly not being prepared to terminate the pregnancy, was the most frequent reason (39%) among parents who rejected pre-natal diagnosis. A feeling of disrespect for the living child with CF (33%), and, to a lesser extent, moral constraints (22%) and religious beliefs (17%) together with a variety of personal reasons mirror the tremendous distress caused by the element of choice.<sup>20</sup>

The issue of reproduction is associated with concern about the potential impairment of health, the additional demands of raising and nurturing a growing child, the limited lifespan, and the genetic risk to the offspring. Because of the potential for deteriorating health, along with increasing demands on the child-rearing family, it is reasonable to counsel the whole family, including prospective grandparents or other significant persons, in order to increase awareness for the need for support. Attitudes between spouses, significant family members and doctors

may show considerable differences and complicate decision-making. However, counselling on reproductive issues should be open, informative, and non-judgemental. Caregivers need to examine their own attitudes and beliefs regarding childbearing and must refrain from allowing their own attitudes to interfere with optimal counselling. The final decision as to whether to attempt to bear and raise children remains with the individual with CF. Their needs and desires need to be respected even if they do not correspond to medical advice.

Gene transfer holds the promise of overcoming the condition, but the uncertainty about its applicability has led to disappointment in the CF population. However, the prospect of this potential treatment keeps many adults and parents optimistic and helps them to feel less frightened when thinking about the future. It is also helpful to communicate to patients that the discovery of the gene responsible for CF has greatly increased knowledge about the molecular pathophysiology of CF, and that this in turn has led to the discovery of other treatment strategies. In a pilot study of 16 adults with CF of a phase I safety trial of a single application of gene transfer, most patients had a realistic understanding that no personal clinical benefit could be expected from participation in the safety trial. For most, altruism was the motivating factor for taking part in this novel procedure, with family and friends, who were often more optimistic about gene transfer than the patients, being the driving force behind the decision.<sup>21</sup>

## SUMMARY

Individuals with CF and their families have no choice other than to live with the disease. However hard they try, they cannot get rid of it. Many patients and families incessantly battle with the disease. They face an uncertain future and have to adapt to many changes in disease management brought about by scientific progress. Health professionals can support and enhance the families' efforts by paying equal attention to physical, psychological and social aspects of their lives and by offering them their total respect for their ongoing endeavours.

### PRACTICE POINTS

- CF affects not only the patient but the whole family.
- Families with CF differ from other families because, in addition to the negative life events most families face at times, they have to manage disease-related problems.
- Knowing the biopsychosocial background of the family may alleviate identification and interpretation of problems presented within the complexity of CF, some of which are wrongly attributed to CF.

### PRACTICE POINTS

- Patients need to undergo time-consuming and often boring treatments that interfere with daily activities and act as a constant reminder of the disease.
- Many treatments resemble hard work.
- Adherence to one component of the treatment programme does not predict adherence to another treatment component.
- Even partial adherence is a result of tremendous effort on the part of both the patient and the family.

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