

The role of the general practitioner in cystic fibrosis

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The current emphasis in the management of patients with cystic fibrosis (CF) is the provision of centralized specialist care with the back-up of a well defined multidisciplinary team. Is the role of the general practitioner (GP) or the rest of the primary care health team anything more than to issue repeat prescriptions and file correspondence from the hospital? Does it make sense for the GP to suddenly become responsible for the terminal care of a chronically ill patient and family who up till then have been totally looked after by a specialist centre? Is it right for a GP to issue repeat prescriptions for a patient who he/she has not seen for many years? The potential role of the GP and the primary health care team is the topic of this review.

BACKGROUND

The centralization of care of patients with CF has encouraged a preoccupation with illness, and may be leading to a neglect of the patient and the family¹⁻⁴. In 1990, the Royal College of Physicians report on CF in adults commented on the role of the GP⁵:

'With this complex and ever-changing disorder the general practitioner has a central role in providing family support, encouraging compliance with all aspects of treatment, sharing care with the hospital clinic and organising terminal care at home where this is appropriate.'

The Clinical Standards Advisory Group (CSAG) surveyed 177 GPs who had had a patient with CF who had been admitted to hospital⁶. Only 83 (46%) returned questionnaires. They were generally satisfied with the level of expertise of hospital care, but less satisfied with the degree of communication or education provided for the primary care team, and a sub-group of GPs felt less involved in the care of the patient than they would have liked.

GENERAL PRACTITIONERS CAN AND SHOULD HAVE A POSITIVE ROLE

Our starting point is that GPs can and should have a positive role in the care of the individual and the family. There is a need to overcome the negative attitude towards GPs having responsibility for a small number of patients with a

specialized condition. There are skills in primary care that could be recognized as providing a valuable support to these patients and their families, to fill an apparent gap in care that appears to exist at present.

We suggest a move away from the mind-set of needing 'complete central' care and ownership' of these patients, especially as there are implications to their health care in the impending epidemiological changes in CF. Whilst hospital paediatric services usually arrange 24 hour-a-day instant access to services for chronically ill children, with patients often having the home telephone numbers of one or more members of the hospital team, this is not an arrangement that is to be found in adult medicine. Children with CF and their families, who have become accustomed to gentle cossetting by the paediatric services, may find the transition to a slightly less welcoming arrangement for adults something of a shock, especially if they have been in the practise of bypassing the GP. The changing epidemiology of CF⁷⁻⁹ means that by the turn of the century, almost 50% of the CF population will be in the adult age group.

As there is no clearly defined role for GPs in the management of CF, their current relationship with patients and their family is often fortuitous, and may simply depend on the individual interest of a GP. With increasing emphasis on the importance of care by specialized centres^{10,11}, the GP has become increasingly sidelined as the specialization and complexity of treatment has grown¹².

Under existing arrangements, there is little opportunity in the standards of care set at local level to define a role for and incorporate GPs. The concept that GPs are not experts in CF distorts and is restrictive. GPs should be seen as the experts in primary care that they are. The principle of a GP's involvement should be that he/she will be providing care for this chronic condition in a way that has become well established for other chronic conditions in primary care. A level of expertise in dealing with CF is something that can develop over a period of time. There is an established link between longitudinal care and accumulated knowledge¹³. Despite this being a rare condition, it is justifiable to involve GPs as there is a link between long-term care and a doctor's sense of responsibility to their patient.

BARRIERS TO INVOLVING GENERAL PRACTITIONERS

There appear to be a number of barriers to involving GPs in the care of patients with cystic fibrosis and these are shown

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Box 1 Barriers to involving general practitioners in the care of patients with cystic fibrosis

General practitioners are seen by specialist centres as non-specialists who lack essential specialist skills and knowledge.

General practitioners are likely to lack detailed knowledge about cystic fibrosis and its management.

General practitioners feel reluctance/fear of having a role in the management of such a 'specialized condition'.

Motivation in care is heavily focused on trying to improve chances of survival.

There is reluctance to change the present system of specialized care, in the belief it is working well.

The mistaken assumption exists that specialized care equates to 'total' care.

Little knowledge or agreement has been reached on the expectations of a general practitioner by either the patient, the family, or the specialist service.

No agreement on areas of responsibility that would be appropriately fulfilled by general practitioners has been reached.

No encouragement exists for the active involvement of general practitioners in articles, books, or standards of care.

Some patients have previous bad experience with general practice (e.g. difficulties obtaining adequate quantities of medicines with repeat prescriptions).

in Box 1. Hitherto, general practitioners have not been an established part of the multidisciplinary team. Other than perhaps in a bereavement, a clear opportunity for involvement of GPs has not existed.

When there is a delay or failure to diagnose CF, there may be difficulties for the GP in becoming involved at what is a crucial time for the family. It is not unusual for the family's grief and anger to be directed at the GP, in whom they have lost trust and confidence. This predicament needs positive handling from the specialist team. In most situations, the parents' negative views about their GP are inappropriate, and the specialists' role is vital in rescuing the relationship between the GP and the family.

Both experts and policy-makers have made reference to the use of GPs in CF care, though there has been no clear development of a recognized programme to involve GPs. Having responsibility for a patient with CF would not be common in general practice. An average-sized practice would not expect to have more than 1 or 2 patients with CF at any one time¹⁴.

A postal questionnaire sent to 1052 adults with CF in the UK in 1990 achieved an 82% response rate¹⁵. A number of areas of dissatisfaction were noted, including dissatisfaction with GPs, attributable to difficulties in obtaining repeat prescriptions, prescriptions for sufficient amounts of treatment, the correct treatment as advised by the specialists, and access for urgent appointments. Patients

were also found to feel uncomfortable when issues concerning the cost of their care were discussed by the GP¹⁶.

In many regions shared care has become established between secondary and tertiary centres, although the emphasis remains on a centralized multidisciplinary scheme of care. We contend that, within the concept of shared care, the GP should be involved as a partner in this multidisciplinary team.

THE POTENTIAL ROLE FOR GENERAL PRACTITIONERS

The most appropriate way to involve GPs is for their potential to be realized by themselves and their patients and families, and this should be encouraged by the specialist units. Their involvement would also enhance the value of the multidisciplinary team, which is a concept that appeals to these patients.

Currently, patients with CF and their families may not have a clear idea of how to utilize their GP and primary care services. The hospital units could begin to develop involvement of GPs by ensuring regular review by a GP, and by advising patients on when to make contact with the GP and when it is most appropriate to make direct contact with the hospital. Patients may be advised that the provision of continuity of care from their GP will contribute to their overall management.

The following are potential roles for a GP in the care of a patient with CF and their family.

Role in continuous assessment

Medical attention from a GP, in between visits to the specialist CF centre, would enhance true shared care, and would allow interim monitoring of progress of either general or specific matters. This contact may be patient-led once trust and confidence have been achieved in the GP, or as directed by the specialist. Adults with CF are poor at assessing their own health status, and tend to underestimate disease severity¹⁷. An opportunity to observe patients in primary care may assist; the GP is well placed to say, 'You are not as well as you think you are', and reinforce concerns of the CF centre. The GP can also help the patient decide if an earlier outpatient appointment is needed. The interim medical contact has the potential to contribute to improving compliance, both in terms of medication and reinforcing the benefits of the system of care provided by the whole multidisciplinary team. Regular contact would also contribute to the development of the GP's experience in the management of CF, and to a successful relationship with the patient and family.

Interpretative role

The opportunity would exist for informed GPs to interpret and explain to patients and their families the treatment that is being received and advised from the specialist unit. Involvement of GPs in care would also help the patient and family in making decisions that are relevant to their overall management, e.g. helping the family to decide to opt for home intravenous antibiotic therapy or enteral nutrition.

Additional support complementing the multidisciplinary team

Involvement of a GP can benefit the patients and families as they feel more care and attention is being given to them. Parents have reported the inadequate provision of supportive services, which needs to be addressed if comprehensive care is to be provided². Involvement of the GP and the primary health care team can help to overcome the centralization of technical expertise that so often entails the fragmentation of responsibilities and comparative neglect of supportive aspects of care¹⁸⁻²².

Providing continuity of care

The concept of cradle to grave care still exists in general practice. It is the degree of continuity of care that family physicians provide that differentiates them clearly from other physicians²³. By providing personal continuity, care of patients with CF may be enhanced, as GPs can recognize the individuality and uniqueness of their patients and families. It is recognized that this helps their coping mechanisms, and the approach can be geared to their need.

Prescribing

The GP should understand and take responsibility for the medication advised following the patient's contact with the specialist carers. The surgery should have the facility to respond to both their repeat prescribing requests and an awareness of the need for appropriate amounts of medication to be supplied. Part of the learning curve for GPs is to appreciate, for example, how much pancreatic enzyme replacement patients need, which would facilitate appropriate prescribing. Many expensive drugs are used in the care of CF. Through liaison with the specialist centre and an awareness of prescribing protocols agreed with health commissions, GPs can ensure patients get the best treatment.

Anticipatory role

GPs can use their skills to anticipate the needs of the patient and the family and aim to meet them (Box 2). This can often be done opportunistically, a skill used every day by GPs. Seeing the patient and family as a whole puts the GP in

Box 2 Some of the areas in which medical care may be anticipated

- Contraceptive care
- Immunization
- Travel needs:
 - precautions
 - drug needs
 - salt supplements
 - risk of travel by air
- General health education:
 - smoking
 - drinking
- Genetic implications to the patient and family

a good position to play a part in anticipating the care that is needed. This has the potential to contribute to preventing, for example, the social, psychological and academic disturbances that can be associated with chronic physical disorders. Knowing the family well can allow an estimate of the extent and kind of supportive services required. There is a risk that if care is preoccupied only with illness, it will lead to a neglect of the whole patient. Anticipatory care is important when considering the difficulties these patients may have. We should not forget that some of these patients can be considered as being 'well but sick' at one and the same time.

Accessibility to care

It is essential that a surgery provides a facility for the acute medical and prescribing needs of patients. Consideration also needs to be given to increasing the allocation of time for routine appointments in order to consider the wider needs of these patients. The GP should be reassured that their role in this care should not be onerous and that the time spent providing care for this condition is likely to be of greater value than time spent in some other activity. An appreciation of the patient's needs should also be understood by all surgery staff including receptionists, which would allow the appropriate access for patients and families to the GP and would enhance the relationship with the primary care team.

Responsibility to the family

GP support in providing understanding shares the burden of illness and can reduce the family's vulnerability to stress. This is especially important as there is no recognized or organized provision of care just for the families by the specialist units. Enhancing the health of the family can only contribute to the health care of the patient with CF.

Home intravenous antibiotics

The GP has a key role in assessing the suitability of the patient and family for home intravenous antibiotic

treatment, and in some cases the GP can provide practical help and support, for example in the management of annulae.

The annual assessment

Annual assessment of patients with CF has become a routine in many of the larger CF centres. The GP could have a role in this process, for example providing the hospital team with feedback about problems at home or within the family, as well as providing data (based on repeat prescriptions) about compliance with treatment.

Terminal care

All too often the GP becomes responsible for terminal care having had no involvement with the patient for many preceding years. This whole role is bound to be facilitated if the GP has been actively involved in care of the patient throughout his or her life.

Non-cystic fibrosis needs

Patients are going to have non-CF needs. Feeling comfortable to attend for these at the surgery can only enhance and develop the relationship between patient and GP.

Antenatal/neonatal screening for CF

Harris *et al.* have shown that GPs can successfully integrate genetic counselling and CF screening into the provision of antenatal care²⁴⁻²⁶. This has support amongst GPs, particularly in the context of family planning services. Involvement in neonatal screening for CF, in the diagnosis of CF, and early involvement in management is likely to be an advantage for the future care of the family by the primary care team.

REQUIREMENTS FOR SHARED CARE IN CYSTIC FIBROSIS

We suggest the following principles:

- It is essential that care links the specialist hospital-based multidisciplinary team with primary care, and with patients and their families.
- Allocation of responsibility in management, covering all areas, must be agreed.
- Patients and families should be given clear advice as to when they should contact the hospital or the primary care team.
- Patients' and families' views/concerns/needs must be taken into account.

CONCLUSIONS

Involving GPs in an effective structure of care for patients with CF would be a major change. The current practice of largely excluding GPs from involvement in CF management is a major barrier to the provision of a comprehensive service to patients and families. Hospital services need to appreciate that they cannot provide for all the health care needs of patients. GPs and the primary care team need to be incorporated into, and networked with, the specialist services. The respective roles need to be clearly identified and agreed between the health care professionals themselves, and between the professionals and the families. Progress requires two key ingredients. Hospitals must cease to sideline GPs, and GPs need to take an active interest in CF.

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