The care of childhood leukaemia in general practice

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Leukaemia occurring in a child under 15 is a rare illness. In Great Britain about 400 new cases are reported each year, or about one in 35,000 children. There are slightly more boys than girls. With luck a general practitioner may never see such a child throughout his years in practice. Nevertheless, childhood malignancies, of which leukaemia is the commonest form, are now the second commonest cause of death (after accidents) for the age group 1–14 years in this country. This paper looks at the current practice and place of the primary care team in the care of children with leukaemia.

The treatment of childhood leukaemia has demonstrated the success of multiple cytotoxic chemotherapy to induce and retain remission of cancer (Medical Research Council Working Party, 1971; Spiers, 1972a). In addition, a series of elegant studies in America has shown that a course of prophylactic radiotherapy, given shortly after bone marrow remission has been obtained, can often prevent the later onset of leukaemic meningitis. The combined treatment of induction with prednisolone and vincristine, a course of prophylactic cranial irradiation, and subsequent ' cyclic' chemotherapy, can produce sustained remission in a high proportion of children with acute lymphatic leukaemia. The possibility of ' cure ' is suggested, although as yet the follow-up is too short for confidence (Spiers, 1972b). For acute myeloid and monocytic leukaemias the outlook is less bright, but considerable advances in remission have been obtained.

Method

A study of the medical care of a population-based group of children with leukaemia was therefore undertaken to describe the current pattern of care and the particular problems of the families. With the permission of the Chief Medical Statistician, a group of 75 children in whom acute leukaemia had been diagnosed during the nine months April-December 1971 was identified from the cancer registers of the four London Metropolitan regions. The consultant in charge was approached and permission sought to write to the general practitioner of the child. Permission was then asked of the general practitioner to interview the mother directly. In all, 64 mothers were visited at home between December 1973 and March 1974. These children, 85 per cent of the total group, are subsequently termed the sample and form a follow-up of two to three years from the child's original diagnosis (figure 1).

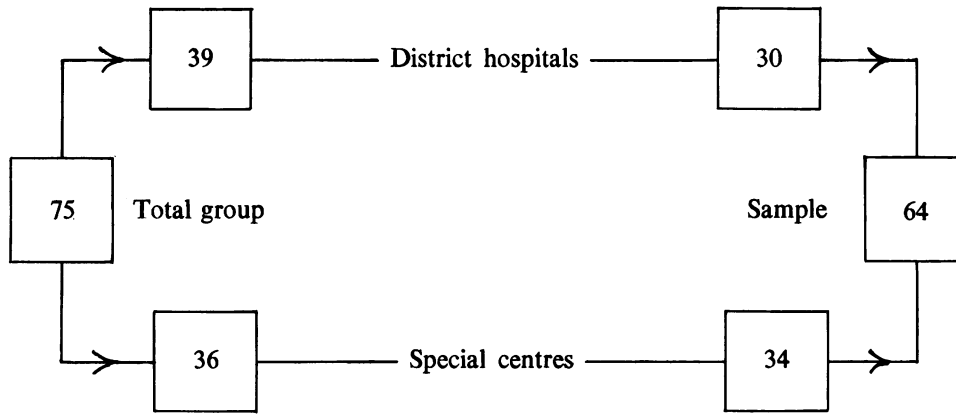

Figure 1.
Classification of the group.

Primary treatment was given at local hospitals (sometimes this was a teaching hospital with a district commitment) to 39 children, forming 30 mothers of the survey sample; and 36 children were treated at ' special centres ' (four hospitals in the London area with units claiming special skill in treating this illness) forming the other 34 mothers in the survey sample. As well, 55 of the general practitioners looking after children in the total group were interviewed at their surgeries.

Findings

Presentation to the general practitioner

The children varied in the length of time with symptoms before consulting a general practitioner. It was at times difficult to identify a specific ' onset ', since intercurrent illness events such as a tooth abscess, were sometimes felt to have been related, even causally. Some form of upper respiratory tract symptom was often mentioned, but these are very common in this age group, and mothers would often add, for instance, " she'd suffered from colds ever since she was a baby." It seemed reasonable to take as the start of the illness the first event recalled by the mother as being unusual for the child, and which led sequentially to the diagnosis of the leukaemia. Using this retrospective assessment, mothers felt that symptoms were present for an average of 2·7 weeks before attending the doctor, though the range was 1–50 weeks.

Presenting symptoms are shown in table 1, where they are grouped into:

(a) those with a specifically haematological connotation,

(b) those which were non-specific,

(c) those which might more readily be associated with other diagnoses.

The commonest non-specific symptoms at initial presentation were tiredness and body pains. These latter were often in the legs, brought on by walking, but also intermittent abdominal pains, and headaches. Bruising and pallor were often mentioned as initial complaints. Bruises, particularly on the legs, tended to be discounted by the mother as due to the child's rough play, but several mothers felt the general practitioner had noted, and sometimes mentioned, pallor without taking further diagnostic action.

By the stage of referral to hospital, specific haematological symptoms had become more frequent and were present in 51 out of the 64 children.

TABLE 1

CUMULATIVE LIST OF SYMPTOMS NOTED BY MOTHERS IN CHILDREN WITH LEUKAEMIA BEFORE DIAGNOSIS (N = 64)

Symptom	*Number of children having symptom*	
	At presentation to general practitioner	*At referral to hospital*
a. Pallor	18	19
Bruising	14	19
Bleeding	5	7
Petechiae	3	6
b. Tiredness	36	31
Body pains	21	18
Low appetite	3	5
Weight loss	2	2
Fainting	2	2
c. Lumps	2	3
Walking difficulty	2	5
Vomiting	4	4
Diarrhoea	1	0
High temperatures	0	1
Respiratory difficulty	0	1
Swollen eye	2	2
' Hard stomach '	1	1

Forty per cent of children were referred to hospital within a week of presentation. For the sample as a whole, however, there were on average 3·3 consultations over 5·3 weeks before referral. About one third of mothers expressed some resentment about delay in diagnosis or referral, and nine of the 64 subsequently changed their doctor because of this.

Of the total sample, 25 out of 75 of the general practitioners had done a blood test before hospital referral, sometimes at the first presentation of the child. Several of the mothers mentioned that the doctor had visited the next day to arrange admission. Nine of the 64 mothers were first informed of the diagnosis by their general practitioner. But for some the blood count alone proved unrewarding, as is shown in this extract from a general practitioner's referral letter to a special centre:

". . . She has been anaemic for about 3 months. Her first blood count, taken in February showed a W.B.C. of 4,300, with a lymphocytosis and some myelocytes and metamyelocytes, and a Hb of 50 per cent. On iron she improved to 70 per cent in a month and her WBC reverted virtually to normal, without primitive cells. However, despite further iron therapy she has deteriorated again in the past month and now, for the first time, has an enlarged liver and spleen and lymph glands in both groins. She looks pale again and her Hb has probably again gone down. She has a few purpuric spots on the legs."

Delay in diagnosis also occurred occasionally at the hospital. One paediatrician, for a child presenting with pallor, tiredness, and pins and needles, told the mother he had "slight anaemia" and prescribed iron without doing a blood test. It was three weeks later, after four further visits by the general practitioner, that the child was admitted to hospital. Another child had had a palpable spleen recorded a year before diagnosis during admission for vomiting and tiredness, but the peripheral blood count was normal and he was not followed up.

In all, 15 out of the 64 mothers felt they had had to press their doctor for hospital referral or a blood count. One mother said, " The general practitioner thought I was fussing. We had to push him to get the child to hospital. He came into the hospital after the diagnosis was made and said he'd never seen it presenting as lumps."

The parents of only one child went as far as complaining to the executive council, the particular grievance being that the doctor had refused to see their child one day at the surgery as an emergency because she had no appointment. The child had a swollen inflamed eye. " I persisted and persisted with the receptionist and the doctor, and he eventually saw me. He said it was an abscess and gave her some medicine; but there was no improvement, so we took her to the accident centre, and they admitted her to the eye hospital. The doctors there upset me by asking lots of questions about the bruises." The eye swelling was a lymphomatous manifestation of the leukaemia. Such resentment may be exacerbated by anxiety and fear when the underlying diagnosis is revealed.

Subsequent care

Sixty children, four fifths of the total sample, were referred to the district hospital by their general practitioner, either to outpatients or casualty depending on urgency and custom. Ten children were referred directly to a special centre; for four of these it was acting as the local hospital, for one referral was made without a specific diagnosis, and for five the diagnosis was already known or suspected after a blood count.

Four patients went to a casualty department directly, without referral, but all had been to their general practitioner several times first. No patient presented directly to a special centre. One general practitioner, working in an immigrant area of London, sent the child to a local authority children's clinic for symptoms of respiratory difficulty. A chest x-ray indicated the cause to be a large intrathoracic lymphomatous mass.

Once a diagnosis of leukaemia has been established at the hospital, the decision for further care rests with the consultant in charge. In the total group, 25 of the 60 children referred to district hospitals were sent on for treatment by special centres soon after the diagnosis was made. The other 35 were treated by the consultants themselves. However, including children referred direct to special centres, and other routes, just over 50 per cent of the children were primarily treated at a special centre (table 2).

The specialty of consultants at district hospitals making decisions about treatment for

TABLE 2
THE PATTERN OF REFERRAL TO HOSPITALS FOR CHILDREN WITH LEUKAEMIA

	Primary treatment by special centre	*Primary treatment by district hospital*	*Total*
General practitioner →District hospital	25	35	60
General practitioner →Special centre	10	0	10
Patients ——————→District hospital	1	3	4
Patients ——————→Special centre	0	0	0
Other route	0	1	1
	36	39	75

the child also varied (table 3). Although commonly this was the paediatrician, for seven children a haematologist was directly in charge, and for three a radiotherapist. When in hospital, however, the children were all in children's wards. For six children the paediatrician and haematologist acknowledged joint treatment decisions, combining their perspectives of the bone marrow state, practical therapeutic regimens, and the family situation.

TABLE 3
CONSULTANTS MAKING DECISIONS ABOUT TREATMENT FOR CHILDREN WITH LEUKAEMIA

District general hospital	
Paediatrician	23
Haematologist	7
Joint care	6
Radiotherapist	3
Special centre	36
	75

Interviews with general practitioners

Most general practitioners regarded the overall care for this illness as hospital-based. Several of the doctors who declined to see me gave the reason that the hospital had " taken over the child completely ". Nevertheless, of the general practitioners interviewed, one third had seen the child over ten times in the intervening period since diagnosis, and another third between four and ten times. And of the mothers interviewed, fifty per cent stated that if their child had a sudden illness such as a temperature, they would either contact their own doctor, or the doctor and the hospital together.

The general practitioners were asked about communication from the hospitals. The large majority of doctors were quite satisfied with this, and felt the information provided in letters, or directly to the parents, was sufficient. Twenty-five (45 per cent) of the doctors had given one or more injections to the child, in the surgery or home. Significantly more of these were children under the special centres, reflecting problems of travelling for daily injections. Several doctors were pleased to have been contacted by telephone by the hospitals, and mentioned that one special centre wrote progress letters directly to them, with a copy sent to the district hospital paediatrician.

Apart from the ill child itself, 50 per cent of general practitioners considered that they gave medical attention more often than usual to other members of the family, most commonly, supportive care to the mother. One third of the mothers interviewed also recalled receiving prescriptions, usually benzdiazepine tranquillisers.

Only one fifth of the doctors had made arrangements for a health visitor or district nurse to visit, but 40 per cent of the mothers interviewed said they had been visited. This difference

is perhaps partly related to varying degrees of attachment of these workers to the practices, and also to health visitors attending for other children in the family.

Although most of the children attended school at some period during remission, no general practitioner had himself contacted the school medical officer. A few local authority education departments wrote directly to the hospital giving treatment, and for three children a home tutor was arranged during periods of stress—particularly when a boy had lost his hair after radiotherapy.

For just over half of the doctors interviewed the leukaemic child had died, but in only one third of these had any member of the primary care team been involved in the bereavement. A few mothers said that their doctors had come round to see them within a few days, and two practices routinely sent a health visitor. Two other doctors made special arrangements for the mother to see them weekly, and one of these mothers subsequently needed hospital admission for depression.

Discussion

Presentation

The presenting symptoms were clearly recalled by mothers after two years, perhaps because they had been frequently rehearsed to medical people and kin, and because most parents would have tried to recall every event around this time in looking for some precipitating event. For instance, the mother who mentioned the tooth abscess as a preliminary episode in the child's illness remarked " I still wonder whether this poison was the cause of the illness."

It can be seen in table 1 that in the interval between onset of symptoms and referral to hospital, there is only a moderate trend towards an increase in specific symptoms, which emphasises the problem of making this diagnosis in the setting of general practice. Indeed, from a clinical point of view, analysis of the presentation of the illness illustrates a fundamental difficulty of this branch of the profession—being alert to the possibility of an extremely rare but serious illness, yet without being over cautious for the many patients presenting with similar symptoms who do not have this illness. Management of these patients represents a greater test of clinical ability than dealing with more classical, clear-cut illness presentations.

One approach to the problem is to have guidelines for action in particular situations. For example, anaemia in a child is relatively uncommon. But estimating the haemoglobin alone does not give an aetiological diagnosis, so that a full blood count and blood film would also be advisable. If the anaemia remains unexplained it is useful to know that an iliac crest bone marrow biopsy, in experienced hands, is no more difficult in a child than a lumbar puncture, and may be as life-saving. An interesting point here, however, is that, for several of the blood tests that general practitioners arranged, the mother stated that the doctor " did the blood test because he thought it was glandular fever ", for which a blood film is often requested. Nevertheless, despite these problems in presentation, it has been found in both the present and previous studies that the length of pre-hospital symptoms does correlate significantly with the length of survival (Pierce *et al.*, 1969). In this study, this is true both for the period before presentation to the doctor, and also the time under general-practice care.

Support

The need for primary care support to the mother and family varies with individual personalities, family groups, and financial circumstances. Nevertheless, in general, mothers were appreciative of support given by the general practitioner, particularly when travelling some distance to a special centre for treatment.

For several mothers also the health visitor's support and practical help was valuable; when perhaps the marital situation was tense, or the child was ill at home and sheets or bowls were needed.

The emotional needs of the parents of the children with leukaemia have been considered in several studies in America (Binger *et al.*, 1969; Friedman *et al.*, 1963; Solnit *et al.*, 1959). At a time when the chance of long-term survival was almost nil, many parents recounted that learning the diagnosis initially was the most shocking time, more distressing than the treatment or subsequent death. Parents were noted to go through a sequence of three states, sometimes termed " anticipatory grief " because of the similarity to bereavement after a sudden illness. The phases include (a) *protest*, often with self-blame, and a search for meaning by finding

' causes ' in their child's upbringing or environment; (b) *despair*, particularly sadness and social withdrawal, often with autonomic somatic accompaniments; (c) *detachment*, when during the child's terminal illness on the ward, parents often become involved with other children on the ward and other activities such as fund-raising.

This simplified pattern of events, however, varies necessarily with individuals, their personal resources and the course of their child's illness. Several mothers in this study had manifest anxiety symptoms, often mixed with depression, at stages both before and after their bereavement. Psychological mechanisms for coping with unpleasant material include both ' intellectualisation ', a desire for extended information and discussion, and ' denial ', when for instance fear of future events is limited by concentrating on the short-term, living from day to day (Verwoerdt, 1966).

Mothers with surviving children expressed both these view points, and it is necessary that the doctor be sensitive to the underlying needs of his patient. Limited but truthful statements accompanied by hope and an expression of sympathetic support seem the most valuable prescription a doctor can offer. A useful factual booklet on the illness and its treatment is available from the Leukaemia Society and it seems helpful for all parents to be aware of this Society, even if they do not feel they wish to attend any meetings.

Another problem is the ' naming ' of the illness. In the present study ten out of the 69 mothers visited said they had openly spoken with their child of ' leukaemia', though for three of these the mother felt the child was still too young to understand the meaning. One of the problems voiced by several mothers was the difficulty of emphasising the importance of their childs' condition to a receptionist, locum, or partner when the child was present and alert to every innuendo.

A " problem with the blood " was the commonest euphemism. Whilst this is primarily a policy decision taken by hospital staff, it is worthwhile reflecting that some of the mothers' anxiety recorded at interview was directly related to the fear of the child's knowing the name of an illness he was already suffering.

In addition, the question of whether to tell a child growing into his teens who has apparently been ' cured ' has now arisen. Despite the difficulty of other peoples' reactions to the word leukaemia, resultant on their own connotations, a more liberal use of the word would seem defensible in developing public expectations and reducing isolation and stigma to the mother and child.

Pattern of care

Lastly, what course of action should a general practitioner take for a child newly diagnosed with acute leukaemia? As this study shows, most doctors who have requested a blood test or opinion from the district hospital leave this decision to the paediatrician in charge. Just under half of these children were referred on for care at a special centre.

The survival of the children in the study can be divided into three groups. Survival was longest for those attending the major children's hospital; the other special centres had survivals similar to those attained by district hospitals giving modern treatment regimes; and children given less modern regimes had poor survival (McCarthy, 1974).

This picture represents the results of treatment initiated in 1971, and it is probable that regimen have now become more standardised in the special centres and at some district hospitals. The advantages of local care include minimised interruption of schooling and family life, low travelling costs, and personal contact between the paediatrician and the general practitioner. The optimum management policy for a child would, therefore, seem to be either local care by a paediatrician or haematologist using up-to-date protocols, perhaps in the context of a multi-centre co-operative research trial, or immediate referral by the paediatrician for care at a special centre.

Conclusion

At several points during the illness of a child with leukaemia the primary care services can be beneficially involved. Alertness at presentation, support during a period of stress such as the radiotherapy or a child's relapse, and the period of bereavement if this occurs, are all sensitive and require concerned care.

From this study, it is clear that a number of general practitioners are giving first-class service, with corresponding gratitude from the parents and child. For this standard of care to be spread to all mothers, it is necessary for the primary care team to understand its role in this illness, and for hospitals to give greater encouragement to local community support services.

Addendum

The address of the Leukaemia Society is: 45 Craigmoor Avenue, Bournemouth. Dr MacCarthy's present address is now: Health Services Research Unit, University of Kent at Canterbury.

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