

Understanding the ‘unknown’: a case study approach to explore patients’, carers’ & health care professionals experiences of cancer of unknown primary

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Introduction

Cancer of Unknown Primary (CUP), also known as metastatic malignant disease of unknown primary origin, is a neglected disease in terms of specific, dedicated services and availability of research, despite accounting for 4% of UK cancer cases and 8% of female and 6% of male deaths in 2009. CUP has a 5 year survival rate of only 8%. There is very little research to help health professionals understand the experiences of patients and families affected by CUP. Nor is there research into the challenges health professionals encounter whilst diagnosing and treating this condition. Better information is needed about the experiences of living with, and caring for patients with, CUP so that future care may be more evidence-based and patient-centred. This study was designed in response to this need.

Aims and objectives

To explore patients, families¹ and healthcare professionals' key issues and experiences of CUP; and to work with CUP patients, their families and health care professionals to decide priorities for improving the care and support offered to those living with, or affected by, CUP; in order to inform development of evidence-based, patient-centred care.

Methods

Phase 1– qualitative case studies

It was planned to create 25 case studies of people with CUP, comprising interviews with the CUP patient, as well as a family member and health professional nominated by the patient. Patients were recruited by clinicians in two large acute NHS trusts and a NHS primary care trusts in the Central South Coast Cancer Network and two specialist palliative care services in the same locality. The CUP Foundation website was also used to access patients with experience of other services. Inclusion criteria were: a suspected or confirmed diagnosis of CUP or metastasis where no primary had been identified; over 18 years of age; able to give informed consent; English language speakers; physically well enough to be interviewed; likely to survive at least one month as judged by the health professional involved in care. At recruitment, patients nominated for interview a family member, if they had one, and up to two key health professionals whom they regarded as important to their care.

In-depth interviews with participants, whilst informed by an interview guide, were participant led. Interviews were audio recorded, with consent, and transcribed verbatim. An audit of each patient's medical notes was undertaken to provide context to the interviews. In-depth interpretative qualitative data analysis methods were used. The Research Team met frequently to discuss emergent findings, and aid data synthesis and interpretation. A Study Advisory Group also reviewed evolving themes and interpretation.

Phase 2 – focus groups using an adapted nominal group technique (NGT)

Phase 1 participants were asked to also participate in Phase 2. Health professionals were asked to nominate colleagues with experience of CUP. Focus groups were held within two hospitals within the Central South East Cancer Network. Seven health professionals, one patient and one carer were unable to attend a group, and completed the same exercise by post or email. Five groups took place: 2 health professional groups (N=5, N=6), 1 patient group (N=4), 1 carer group (N=2), and 1 patient/carers group (1 patient, 2 carers).

¹ In this study 'family' is taken to mean those of significance to the person with CUP. It includes friends, as well as those in kinship relationships.

Preliminary analysis of Phase 1 interviews identified a range of potentially important support needs. These were phrased as statements and, following revision and refinement by the Research Team and Study Advisory Group, eleven statements remained. At the focus groups, participants were told that the group's overall purpose was to address the question of 'what is the most important support need experienced by people living with CUP?'. The eleven statements were then presented to the group and discussed in turn. New issues suggested by participants were then discussed by the group. Following the discussion, participants were asked to rate each of the statements on a scale of 0 (not important) to 10 (very important). Once statements had been rated, participants were then asked to choose the 5 issues they thought were most important and to rank them from 1 to 5, with 1 being the most important.

Findings

Phase 1 – qualitative case study

Seventeen case studies were constructed: three consisted of patients, health professional interviews (2 health professional interviews in 2 cases) but no carer interviews; 10 had patient, carer and health professional interviews (2 health professional interviews in 4 cases); and 4 had patient and carer interviews only. Ten patients were women. Their mean age was 61 years (range 41–78). Four lived alone. Nine had retired, three were in paid employment, and five were not currently working because of ill health. Nine had first attended hospital for CUP in 2011, 6 in 2010, and one each in 2009 and 2005.

Ten of the 14 nominated carers were women. Carers had a mean age of 55 years (range 34–81 years). Five oncologists, 2 surgeons, 2 clinical nurse specialists (CNSs), 2 GPs and 2 allied health professionals were interviewed. One health professional was interviewed about four patients, three about two patients, and the remaining nine about one case only.

Key features of the CUP patient journey

Patients did not follow a "typical" journey, characterised by a predictable set of events shared in common with other CUP patients. They were a diverse group with a variety of presenting symptoms, treatments received, and prognoses. However, key features of their journeys included the initial presentation with symptoms to GPs and referral to hospital; tests and investigations to try and reach a diagnosis; diagnosis itself; treatment (including chemotherapy for 13, radiotherapy for 9 and surgery for 7); and, for eight at the time of interview, the post-treatment period.

Patients presenting to GPs with clear signs were usually referred urgently to specialist services or for investigations, whilst, when symptoms were less clear several GP visits were usually needed before referral. Patients' experiences of tests and investigations also varied significantly, from smooth progress through a series of tests towards a diagnosis and treatment plan, to disrupted progress characterised by delay, poor coordination and lack of accountability. The number of tests did not appear to be problematic, as patients did not raise objections to the number of investigations they had undergone or to their futility if they were all negative; tests seemed to have a symbolic importance as well as clinical value in that extensive testing demonstrated the clinical team's commitment to discovering what was wrong and finding the best treatment. However, nearly half of patients experienced lengthy delays during this phase of their patient journey which was problematic and led to anxiety and frustration for them and their carers. When it came to diagnosis, again patients' experiences varied significantly, with some prepared early for a diagnosis of cancer (if not CUP) whilst others had a long period of investigations during which communication was limited or did not clearly communicate the possibility of cancer,

making the eventual diagnosis more shocking and difficult to absorb. Variations were also found in the experiences of treatment, with some experiencing frustration about the general lack of knowledge on CUP treatment, concern about the necessity

or effectiveness of treatment, confusion about why treatments were being given and doubt as to whether treatment had been worth it. Two 'exemplar cases' in the Report demonstrate these variations in individual patient experience: Ian experienced a smooth journey, with an oncologist and CNS giving him the CUP diagnosis, taking early responsibility for his care and negotiating with other MDTs on his behalf; in contrast Karl had over three months of initial tests, with no-one in overall control, leading to him being completely unprepared for a cancer diagnosis and its blunt delivery. He was shunted from one MDT to another, and neither he nor his partner felt they had anyone they trusted or could talk to – he was a member of 'a lost tribe'.

Themes

Uncertainty

Uncertainty was a core part of the experiences of patients, friends, family members and health professionals.

The majority of patients and carers had not heard of CUP before being diagnosed. The diagnosis of CUP itself was unexpected, unfamiliar and a source of great uncertainty. Uncertainty about treatment led to concerns about the effectiveness of treatment, confusion about why treatments were being given, doubt as to whether treatment had been worth it and frustration about the general lack of knowledge of CUP treatment. Perceived uncertainty among health professionals was sometimes a source of distress for patients. There was also considerable uncertainty about recurrence because of the lack of evidence on the disease's likely incidence and location.

Most health professionals viewed a CUP diagnosis in negative terms. They associated it with insufficient or vague knowledge and information, unpredictability and complexity – all aspects of uncertainty. The lack of evidence and accurate information on which to base treatment decisions and prognoses, and from which to advise and inform patients, was a source of concern for them. They felt patients who received a CUP diagnosis were at a disadvantage compared to those with a known primary, the diagnosis being harder for the patient to understand, and prognoses more ambiguous. While uncertainty is prevalent in all areas of medicine, it is amplified in CUP. Health professionals were concerned that their own uncertainty could be communicated to patients with potentially negative effects. The tension between treating a CUP patient based on the limited information available, or waiting for the results of further tests, but delaying treatment, was discussed by a number of health professionals. They described having to balance hope that further testing would lead to a diagnosis, with concerns about the deteriorating health of a patient waiting for treatment to begin.

Strategies used to cope with uncertainty depended on factors associated with the illness such as stage of illness and prognosis, as well as individual characteristics, such as level of emotional distress.

In most cases patients and their carers talked about seeking information about their illness. Information was sought from health professionals, books, leaflets, newspapers, the internet – in particular the CUP Foundation website, friends and family members and other people with cancer. For some patients and carers, finding information about CUP led to an increased understanding of the condition, helped interpret events, and structured expectations of their illness. This helped reduce uncertainty. Others, however, did not wish to seek information, found the information they were given confusing and were worried by the information they found. Having faith in health professionals was an important strategy for reducing uncertainty among study participants. In trusting health professionals, patients gave

them responsibility for aspects of their illness such as decisions about treatment, understanding of the illness and expectations about outcomes. A small number of patients described using minimising, avoiding and denying strategies for coping with their illness.

Continuity: timeliness, co-ordination and accountability

Delays were experienced by patients in referrals, investigations and decisions about treatment. Most delays occurred either in the period between the development of initial symptoms and referral to specialist hospital services, or during the investigation phase prior to diagnosis. Protracted investigations were associated with patients being transferred between different teams or clinicians. Some patients who experienced delays in investigations worried that their health would deteriorate as the start of treatment was held up, and some did experience worsening of symptoms. Others felt abandoned, “left in the middle” and unsure of what would happen. In most cases, by the treatment phase of the patient journey patients were in a more stable routine with a definite treatment plan, and in regular contact with a smaller number of health professionals which allowed therapeutic relationships to develop.

Some patients experienced excellent coordination of care, with rapid referral to an appropriate specialty, a structured process of investigation and diagnosis, and treatment provided by health professionals working as a team with good lines of communication. Four out of the five patients whose cancer was discovered in the head and neck area described good coordination of care, which took place within one MDT. Other patients, however, experienced poorly coordinated care, characterised by delays in referral to a specialty, a muddled process of investigation and diagnosis, disagreements between health professionals about treatment and poor communication. Patients and friends and family members were often aware of poor co-ordination of care, leading to sadness and frustration.

‘MDT tennis’ or ‘MDT bounce’ was the term used by health professionals to describe the phenomenon of CUP patients being passed from one MDT to another with both reluctant to take responsibility for the case: rather than starting treatment when the primary was still uncertain, health professionals chose to refer a patient to another MDT for further testing. For patients who experienced ‘MDT tennis’ during the investigation phase, poor communication was a salient feature. When patients were aware of disputes about ‘ownership’ this led to feelings of abandonment and powerlessness, with patients left wondering who was taking responsibility for their case. Health professionals in this study recognised this issue. Some voiced concerns about CUP patients not being assigned a CNS. Patients assigned CNSs or other key workers, such as a radiographer, were positive about the support they received. Most patients developed and sustained a relationship with an oncologist during the treatment phase and the majority were very positive about the oncologists in charge of their treatment.

Information

Patients and carers often found explanations of CUP given by clinicians very confusing, especially the idea that clinicians were uncertain of the primary. They were sometimes unsure whether they had received all the information available from clinicians or what questions they should ask. Some were happy with the level of information provided by clinicians, but others would have preferred much more. Carers often had more questions than patients and they expressed greater frustration at the point of diagnosis when questions could not be adequately answered. Changes in MDT team and meeting different doctors who sometimes provided contradictory information often led to increased uncertainty and anxiety, especially amongst those who wished to be more informed. Some patients, particularly those with little prior knowledge of cancer and those less inclined to search for information, found it difficult explaining their condition to others.

Most patients indicated their preference for limiting their source of information to what their doctor told them rather than seeking information for themselves. In some instances the need to preserve hope could drive patients to seek further information, to take control of their disease and actively participate in decisions about their treatment. Others, however, avoided information in their effort to maintain a sense of hope: the greater degree of uncertainty that CUP involves may mean that in some cases any information is 'unsafe', and those wanting to remain hopeful avoid it. Patients also took a 'charitable' approach to other patients which restricted the time they were willing to spend asking questions: they were aware that asking for more information involves time that is in short supply, and by taking more time with a clinician this restricted the time they could spend with other patients. The 'charity', or sympathy, of patients also appeared to extend to the clinicians themselves, whose time they did not wish to waste by asking questions.

Several described how they found support in hearing about other cancer patients' experiences, often over the internet, and expressed a desire for some form of peer support groups for CUP patients, where they could learn from the experiences of other patients with a similar diagnosis. Several found the CUP Foundation website valuable, both as a source of information and for peer support.

Consensus exercise on priorities for improving care and support

Including six additional themes generated by the focus groups, the statements included in the nominal group ranking and rating exercises were as follows: need for a key person to give consistent information and support; need for a key person to co-ordinate the efforts of different health professionals; need for CUP and what it might mean to be explained; need to know that health professionals have not given up or abandoned patients and carers; need to know that everything appropriate has been done to find the primary cancer; need for patients and carers to know their needs are recognised and addressed; need for information to understand more tests may not be the best thing; need for patients to understand reasoning behind investigations and tests; need for patients and carers to talk about causes of cancer; need for early involvement of specialist palliative care services; need for comprehensive written information about CUP; need for patients and carers to have a support group; need for good lines of communication with health professionals when treatment has finished; need for a lay person to accompany patients on the treatment journey; need for health professionals to be honest about their own uncertainty; need for carers to have access to information independent of the patient; need for a treatment and follow up plan at the beginning of treatment.

Overall, there was a considerable degree of consensus during the rating and ranking exercises. Although the order was different, the three themes rated highest amongst all participants were the same three ranked highest overall: the need for CUP and what it might mean to be explained; the need for a key person to co-ordinate the efforts of different health professionals; the need for a key person to give consistent information and support.

Discussion

We hope that our research will enable health professionals to better recognise, understand and address the needs of people with CUP – a group aptly referred to by one of the health professionals as the ‘lost tribe’. The challenges patients and carers face centre around the incomprehensibility – in an age of technology assisted diagnostic techniques – of the fact that a primary cancer cannot be found and identified. This is amplified by ambiguities in determining what might constitute optimal treatment, and having no clearly identified professional responsible for management. Our research serves to emphasise the consequences for patients when they are left floundering when no one health professional takes responsibility for co-ordinating their care, and where there is a failure to evaluate and treat them in a systematic and timely fashion.

Our findings support the NICE CUP Guideline in pointing strongly to the need for a health professional to assume responsibility for the management of this group. A health professional needs to step forward to take responsibility for co-ordinating care and ensuring patients get the support and information necessary to enable them to better face difficult dilemmas such as deciding on a further set of investigations or whether to have chemotherapy or not.

There is however the question of when might be the optimum point on the patient pathway to involve a CUP team. The difficulty of identifying patients with CUP in order to make an effective referral to a CUP team should not be underestimated. We would suggest assignment of a named oncologist and CNS should occur at the point at which the first set of investigations fails to locate a primary in the presence of metastatic disease. The findings suggest early involvement of a professional with an interest in CUP often led to patients feeling they had someone ‘fighting their corner’ working to ensure appropriate and timely investigations were undertaken in order to make an informed decision about which (if any) treatment options to pursue. Health professionals need to consider how best, and when, to feed CUP patients into appropriate site-specific teams where management by a site-specific team would be in a patient’s best interests.

Attention to communication, continuity and information will help achieve a positive experience for patients and carers. Our research endorses the need for patients to be assigned a specialist nurse who will undertake the functions recommended in the NICE CUP Guideline. How healthcare providers will respond to the need to establish another cancer-related specialist nurse service in a cost-constrained environment is not at all clear. The need for a key worker to co-ordinate the effects of different health professionals, and to provide consistent information were two of the three statements agreed upon in the consensus exercise. Given the limited prognosis of many CUP patients and the increasing emphasis in oncology of the role of palliative care alongside acute care, the role of palliative care specialists in CUP teams and, where appropriate, as key workers in CUP, warrants further investigation.

Uncertainty is a significant aspect of CUP. It involves an illness state where many of the uncertainties that accompany cancer are amplified and accompanied by a unique set of clinical ambiguities. No information exists concerning the effects of uncertainty about diagnosis and treatment on distress levels and psychosocial adjustment in this population. Future research should be directed at comparing patients’ levels and sources of uncertainty and its effect in terms of emotional distress and adjustment in patients with CUP as compared to those in whom the site of the cancer is known. The impact of health professionals’ own uncertainty on patients also needs to be explored.

CUP is clearly a challenging diagnosis for health professionals. Breaking bad news is a complex communication task and can affect a patient’s comprehension, satisfaction with care and level of hopefulness. This task is made all the more difficult in CUP because of the discomfort some clinicians felt around the uncertain diagnosis and consequently the lack of a clear treatment plan or prognostic information. How health professionals and patients deal with, and can be supported

in managing, clinical uncertainty and ambiguity merits further research. This should include exploration of the best ways of communicating information in the face of uncertainty and ambiguity.

Conclusion

The experience of patients with CUP and their families is characterised by ambiguity and clinical uncertainty. Some patients receive excellent care. Others experience 'MDT tennis' as they are bounced between site-specific MDTs while tests are undertaken, cases reviewed and opinions sought about treatment possibilities without anyone taking responsibility for them. Patients and families need to be able to access a health professional who is knowledgeable about CUP and who has been nominated as responsible for co-ordinating the care of all professionals involved in their care, providing consistent information and ensuring they receive necessary support and information. The most efficient, effective and appropriate model of providing a key worker for people with (or suspected to have) CUP needs further investigation, as do other aspects of the CUP patient pathway, including the appropriate point of referral to a MDT with experience in CUP and optimum use of specialists in palliative care.