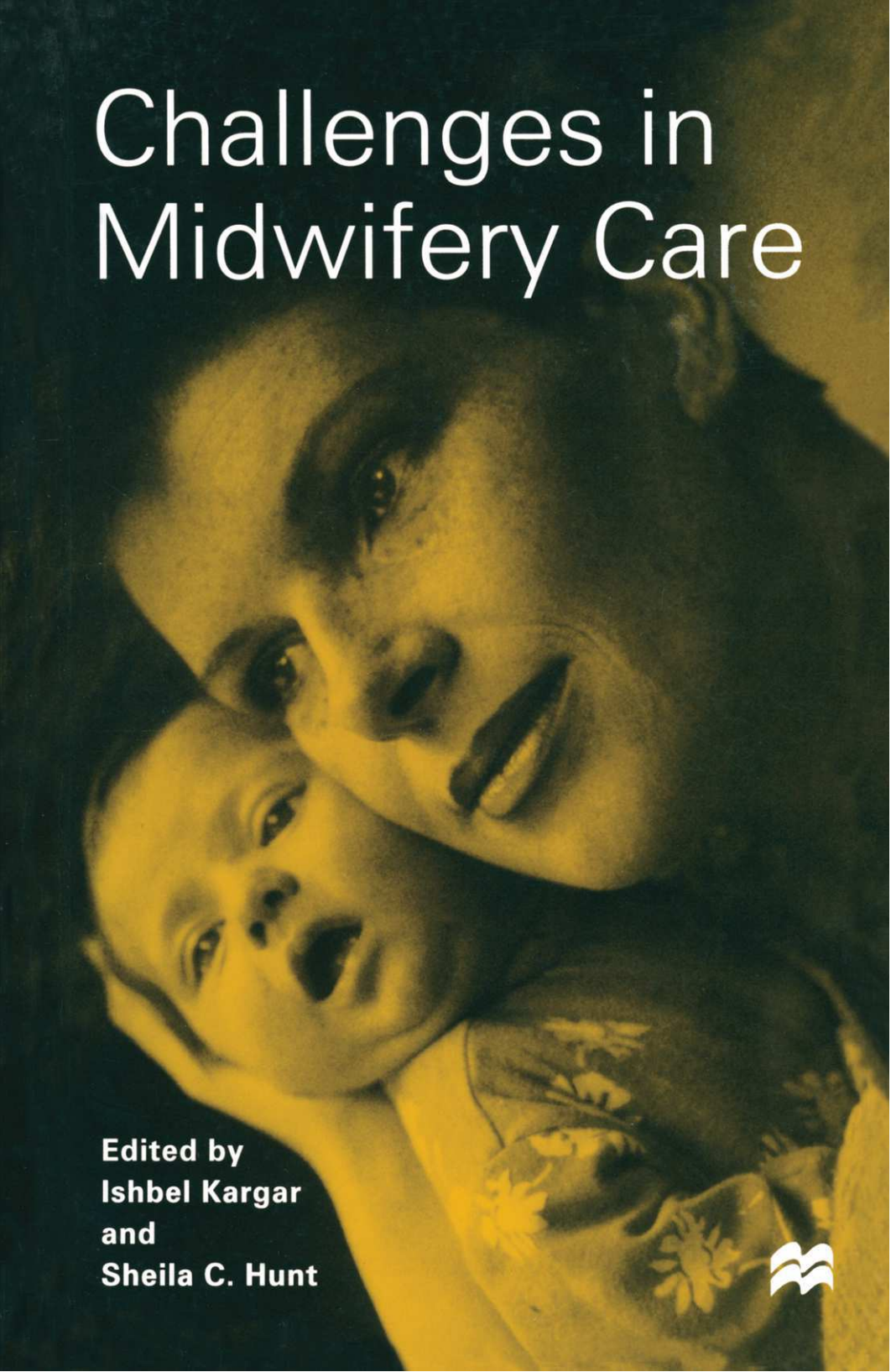
Challenges in Midwifery Care



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FOREWORD

This book emerged from the experiences of midwives as they met together at regional meetings of the Association of Radical Midwives. It is clear that in that supportive environment they thought, felt, listened, reflected and learned more about the practice of midwifery. The ideas were sometimes new, sometimes different, and often about unusual experiences.

In this text are ideas, rich sources of information, references and those 'tricks of the trade' that are often difficult or impossible to locate in standard textbooks.

For midwives who are passionate about their practice and intent on improving the childbirth experience for women, this book is a gem. It will be invaluable for midwives seeking advice and sometimes inspiration on giving care to special people in special circumstances. I commend this book to midwives everywhere.

Caroline Flint
London, 1996

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The editors would like to thank the sterling efforts made by the contributors in bringing the chapters in this book to publication. In addition, we would like to thank Kerry Lawrence for believing in the ideas, and Carrie Walker for her patience, guidance and perseverance.

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This book is dedicated to those special women and their families from whom we have learned so much and who need from midwives that extra special care in childbirth.

INTRODUCTION

One of the guiding principles of the Association of Radical Midwives when planning its quarterly meetings around the UK is that speakers and skill-sharing workshop leaders can usually be recruited from local midwives. Those doubting this, feeling that 'experts' from elsewhere would have been better, have often been pleasantly surprised that a really informative, instructive, interesting and stimulating day has been enjoyed by everyone, listening and learning from local colleagues with hidden talents.

This book is a demonstration of this principle and proof of the valuable knowledge and experience possessed by 'ordinary' midwives who have accepted the challenge presented by a particular aspect of their practice. They have all seen that some women in their care have special needs, over and above the usual maternity care, that must be met. These colleagues of ours made it their business to learn more about the particular difficulties of such women and have, in the process, built up an impressive body of practical expertise and theoretical knowledge, which they now share with us.

Many midwives will read the contents page and comment that they are already dealing with such 'special' circumstances quite adequately. We have no doubt that this is so, and would hope that such is the case for the majority of practitioners. Nevertheless, midwives will surely agree that they can never stop learning, and if the special situations dealt with in these chapters highlight aspects of care that have not previously been addressed, the book will have served its purpose in challenging midwives to enhance the care they provide.

Some of the problems dealt with are less common, and midwives may be forgiven for reading these chapters with only professional curiosity, rather than as examples of situations they may have to deal with themselves. However, midwifery is continually widening its knowledge base, and just as we learned about Bandl's ring, probably never actually to see one, we also need to learn how to help a woman whose needs may be out of the ordinary, but who may be the next client in our practice.

The book is aimed at midwives in clinical practice caring for women with special needs, for whom the usual approach to maternity care has to be modified.

Until comparatively recently, good midwifery texts were few in number, and although one or two of these have almost attained the status of a 'bible' of midwifery knowledge, they contained only passing reference to a few special needs in pregnancy and childbirth, such as some physical handicaps, and specific medical or social conditions presenting an element of risk to the pregnancy or to the woman. None of these has so far addressed in any depth the requirements of particular groups of women, or offered substantial assistance to midwives caring for these women by way of information about support groups, extensive reference texts and help on a more practical level.

This situation is now changing, as midwives develop the skills and discover the joys and rewards of research, often inspired by problems encountered in their own clinical practice. Such studies are welcomed by their colleagues, not least because the situations dealt with often reflect their own practice and are therefore highly relevant.

There is a growing recognition that there is no single 'best' system of maternity care for all women, and that not only individual women, but also certain *groups* of women, have special needs. The contributors to this book realise that while addressing the problems of particular 'groups', there must always be an awareness that the women concerned are first and foremost individuals, with their own needs and desires, albeit having many of those needs in common with other women with similar difficulties.

The authors examine these needs with particular reference to midwifery care, and in exploring the problems try to offer a deeper understanding than has hitherto been available to those seeking it. It is acknowledged that almost all of these 'special needs' groups have already been identified within society, and some suggestions for improving their maternity care have been made, either in passing while discussing other problems, perhaps in a textbook, in research study, in occasional professional journals or in the information leaflets produced by campaigning and self-help groups. This book gathers a significant amount of this scattered but valuable information, plus additional material, into one text which we hope will become a book of choice for midwives wishing to learn more about special needs.

Most of the chapters have been written by practising midwives in their spare time, drawing on their own experiences and clinical practice, using case studies where possible to illustrate their work. One author is not a midwife but has made a valuable contribution to the book, which will be welcomed by midwives encountering the problems she deals with. Some of the writers are putting pen to paper for the first time, having been encouraged to make their work more widely known, often after speaking at a local meeting.

The final chapter considers a relatively new challenge, faced by all midwives – the challenge of change in the organisation and philosophy of

midwifery as a result of 'Changing Childbirth' and other documents. This chapter analyses the origins of change and explores key management theories. It considers why individuals and organisations resist change and looks at strategies for 'coping with it'. It seeks to support and understand midwives as they support and understand women during a period of unprecedented 'upheaval'.

Throughout the book, the authors' deep understanding of the feelings, frustrations, emotions and needs of childbearing women is evident, encouraging fellow midwives to accept the challenge, to practise through listening to the women in their care with patience and empathy, and really to be 'with woman'. Only thus will midwifery move forward to the woman-centred, sympathetic service that is becoming recognised as the only right way to practise.

We are very proud to have been associated with these people and shall be eternally grateful for the opportunity of helping them to bring their special experiences, studies and findings to the attention of an even wider audience.

Ishbel Kargar
Sheila Hunt
January 1997

CHAPTER ONE

Haemoglobinopathies and pregnancy

Rachel Celia

Haemoglobinopathies (HbOs), including sickle cell disease and thalassaemia, are inherited abnormalities of haemoglobin manufacture. They can occur in people of all races and origins, although they are more common in people who have non-northern European ancestry. All women at risk of having inherited an HbO should be offered antenatal testing, and appropriate laboratory and counselling services must be provided to enable them to make informed choices.

Sickle cell disease has an effect not only on the bodily structures of the individual, but also on every aspect of that person's life. She comes to her pregnancy not only with particular health needs, but also with complicated psychological needs. Childbearing has additional risks for both the mother and the baby, and the mother should be cared for by experienced obstetricians and haematologists.

The midwife's role is crucial, however, in providing a responsive and appropriate environment. The challenge to her as a professional is to facilitate a situation within which the woman's complex and changing needs can be determined, respected and met. In order to achieve this, and to empower her client, the midwife will need to use all her skills of communication and observation:

> These are special patients needing special care... for optimum results what is needed are eyes trained to see, ears trained to hear, hands trained to feel, a mind trained to monitor, interpret and act, and a kind heart. (Klufio *et al.* 1977, quoted by Konotey-Ahulu 1991)

A successful response to the midwifery challenge will provide the best possible opportunity for a healthy mother and baby to form the essential relationship base they need in order to support them through the many difficult times that they are likely to have to deal with in their life together.

A basic knowledge of the physiology of blood formation will be useful in understanding this topic. For more information, see the Further Reading list at the end of this chapter.

WHAT ARE HAEMOGLOBINOPATHIES?

Sickle haemoglobin (HbS) is one of several hundred inherited abnormalities of haemoglobin (Hb) manufacture collectively called 'haemoglobinopathies', which are mostly just different forms of the common adult Hb (HbA). HbOs are determined by recessive genes, so do not usually manifest themselves as symptomatic illnesses unless they are inherited from both parents. They are not sex linked, although it has been suggested that sickle cell diseases may affect males and females slightly differently (Leavel & Ford 1983).

People who have inherited the HbS gene from only one parent are said to have sickle cell trait (HbAS) or to be carriers. Such people are symptom free because their HbA protects them. It is similarly possible for a person to have a trait of any of the other HbOs.

There are two kinds of HbO. One affects the structure of the Hb molecule, and this group includes HbS and HbC which is another common HbO. The other kind affects the amounts made of the component parts of the Hb molecule; this group includes the thalassaemias.

HbOs can be inherited in any combination. HbS, for example, can be inherited in combination with HbS, HbC or beta thalassaemia. This results in the conditions sickle cell anaemia (HbSS), HbS/HbC disorder and sickle/beta thalassaemia (HbS/B thal) respectively. The expression of these disorders can vary from person to person, and from one period of life to another for the same individual. HbSS and one form of HbS/B thal generally, but not always, cause the most clinically serious problems.

The many different HbOs are generally believed to be associated with people from different parts of the world. It is important to remember when providing health care that most HbOs are much more widely distributed than is commonly believed, and that those at risk can certainly not be identified by visual appearance. HbS, for example, is just as common in parts of Greece as it is in parts of Nigeria.

SICKLE CELL DISEASE

In HbS, the genetic mutation causes an amino acid substitution in the beta-globin chains of the Hb. This changes the solubility of the Hb when it is deoxygenated, resulting in the formation of polymer chains, which deform the cells from within. This in turn causes the characteristic sickle-shaped cells after which the condition is named, which can become lodged in the capillaries, causing vaso-occlusion.

As deoxygenation of the Hb is a major factor in sickle cell disease, anything which increases oxygen requirements is a risk. So too are dehydration, changes in the pH of the blood, infection, fever and extremes of, or sudden changes in, temperature.

The vaso-occlusive episodes may occur at any time and at any site in the body, where they cause excruciating pain and progressive deterioration of body structures. The pain may be of such severity for people to believe that the pain will kill them. These painful crises are therefore not only experienced as torture, but also as life-threatening or even near-death experiences. This has a profound effect on the person. The acute episodes, or 'crises', are identified by the nature of the individual episode. For example, if pain is the presenting problem, it is called a 'painful crisis'; if the lungs or chest are involved, it is called a 'chest crisis'. These crises can be life-threatening and can cause intense fear and anxiety. Body organs (some of them vital) will be progressively damaged, resulting in further health problems.

The deformed red cells have a greatly reduced lifespan (10–14 days), resulting in a profound lifelong haemolytic anaemia. People with HbSS usually have an Hb level of 7–9 g/dl. However, the lifelong nature of this state results in an adaptive process that allows the more efficient utilisation of the available Hb. Maternal anaemia can never be taken lightly, however, and requires diligent monitoring during pregnancy.

In addition to these effects, many other medical emergencies may arise, and sufferers are particularly susceptible to overwhelming infection. Sudden and/or early death may occur at any time.

Reports regarding mortality rates vary. In infancy, they are as high as 30 per cent in some studies (Horn *et al.* 1986). Throughout childhood, the mortality rate falls, only to rise again steeply in the mid-thirties.

When well, people with sickle cell disorders are said to be in 'steady state'. Apart from penicillin prophylaxis for children under 16 and folic acid (to facilitate red cell production), treatment is unlikely to be necessary. Treatment during a crisis is symptomatic (analgesia, antibiotics, rehydration and so on, as appropriate).

Blood transfusions will be used to save life as an emergency measure, as a long-term attempt to arrest the deterioration of a vital organ and occasionally (but only as a temporary measure) to improve the quality of life, for example to reduce mother–baby separation resulting from frequent admissions to hospital, or to enhance wellbeing during a course of study or a much-needed holiday abroad. This treatment is embarked upon reluctantly, not only owing to the many commonly known risks of blood transfusion, but also because these people, as a result of repeated transfusions, may run into problems with iron overload and are particularly at risk of becoming untransfusable due to the accumulation of antibodies. This obviously has implications for pregnancy, as well as for long-term management and prognosis.

SERVICE PROVISION

For professionals to respond adequately to women with HbOs who are pregnant or seeking to become so, some essential services must be available. These include:

- laboratory services for diagnosis in adults and the fetus;
- genetic counselling services;
- expert medical care throughout life for the treatment of people with the disease states;
- psychotherapeutic counselling services to support individuals and their families, at any stage of life, with the many difficult emotions and adjustments with which they may have to deal.

Service providers need to be aware of the needs of clients and ensure that services meet these needs. Appropriate literature should be provided in the relevant languages. Counsellors must be acceptable to the clients. All staff must be rigorously educated about the dangers and implications of racism, for both themselves and their clients (Adams 1994).

SCREENING IN THE MATERNITY UNIT

Different maternity units have different policies depending on the population considered to be at risk and therefore needing to be screened. It is important to remember that all screening programmes have a percentage of failures (usually around 10–20 per cent) and that selective screening programmes are the most fallible. It is essential, therefore, for midwives to maintain an awareness of the relevance of screening for their clients throughout the entire reproductive process. They should not assume that, because screening tests were meant to have been carried out earlier, they have necessarily been done.

In an ideal world, everyone would know and understand their HbO status long before they embarked on a pregnancy, but this situation is still a long way off. At the booking visit, every woman who is likely to be at risk should be screened for HbO. Such an approach is however unlikely to be effective without extensive education of the relevant staff and 'at-risk' communities and careful administrative structures (Adjaye *et al.* 1989).

Some hospitals (such as St Thomas' in London) have developed units within the computerised booking package that request information about the country of origin of the woman, her parents and grandparents. Although this does not guarantee that every woman who is at risk of HbO will be identified, it has greatly increased this likelihood. Screening should also be offered to all partners who attend the booking appointment. This is only a beginning, however, as this information is quite worthless unless the appropriate response is made.

Normal results

Each woman who has been screened should be informed of her result and be given sufficient explanation to ensure that she can understand its significance and be reassured.

Unclear results

Difficulties in obtaining a definitive diagnosis for alpha and beta thalassaemia (which are often almost indistinguishable from iron deficiency anaemia) are not uncommon. It is, however, vital that a diagnosis is made in order to achieve an appropriate response. Genetic counselling must be provided for those with thalassaemias, and iron therapy for those with iron deficiency. Haematologists will advise on the additional blood tests needed to obtain clearer results. In the meantime, a sensitive balance must be maintained between dealing honestly with the woman and not making her unnecessarily anxious.

Positive results – women with traits

It is important that there are clear policies specifying the actions to be taken when a blood test result indicates a diagnosis of any trait. The aim of this response is to ensure that each woman obtains all the information, support and access to facilities that she needs and wishes. To ensure that this happens the following must be available.

Partner testing

The offer of screening for the woman's partner has the most likely benefit of demonstrating that he does not have an HbO and that the baby is therefore not at risk of inheriting a major HbO. The partner may, however, be unavailable or unwilling to be tested. If this is the case, it is important that encouragement to take the test is not at a cost of greater anxiety on the part of the woman, especially as women are, unfortunately, often not offered counselling unless both partners are known to have HbOs. If the partner's result is unknown, many women will need even more support than other women until the baby has been tested. Some partners may be encouraged to attend for testing if a simple letter explaining the request is provided. This should be given to the woman herself to avoid breaches of confidentiality, unless she specifically requests that it be sent directly to her partner.

Counselling

All women with an HbO should be offered an appointment for genetic counselling with a person trained in the specialty. If no trained HbO counsellor is available, referral should be arranged to a haematologist experienced in the management of HbOs, or to the nearest sickle cell and thalassaemia information centre, as a matter of urgency. In the meantime, information, support and realistic reassurance should be given.

Genetic counselling should include information about the nature and behaviour of the relevant HbO, the possible results of its interaction with other HbOs, a clear description of the choices open to the woman and referral for any requested procedures (such as chorionic villus sampling) or to other avenues of support. This must be done by trained counsellors who are able to provide this information free from personal bias. The object of this exercise is not that the woman does what the health professional thinks is best, but that she is enabled to make the best possible decision for herself, her baby and her family. Whatever the outcome, it is imperative that she experiences as much control over that choice as is possible.

SCREENING OF THE NEWBORN

There are two main reasons for screening for HbOs in the newborn. The first is to identify those babies with major HbOs and thus to be able to reduce the risks to them by providing the necessary medical care (see below). The second reason is to identify children with HbOs so that their parents can be informed and the family can be offered genetic counselling. It is essential to be able to ensure that adequate follow-up is available and provided, although this is not always the case (Milne 1990).

The risk of sudden and preventable death in infants with sickle cell disease is significant. Early diagnosis and institution of prophylactic measures can prevent this. It is imperative, therefore, that effective policies and procedures are in place to ensure the safety of these children (Eboh & van den Akker 1995). The authors' study at St Thomas' hospital demonstrated clearly the unreliability of selective screening. This unreliability appeared to increase as the selectivity increased. For example, screening all black women produced a 7 per cent failure rate but screening all babies of black women with traits produced a failure rate of 20–30 per cent. Selective screening also places great responsibility on the midwife. In spite of frequent claims to the contrary, usefully reliable results can be obtained from cord blood by a committed and experienced laboratory. Some units are now using the Guthrie spot for HbO testing. This has the benefit of using a tried and tested collection system, although even the Guthrie testing is likely to show a failure rate. All tests should be repeated (usually at 3 months), but the early test means that the baby's status is identified, the parents can be

informed and therefore the baby's chances of receiving the appropriate follow-up are increased. The early diagnosis means also that there is a sufficiently long time to educate and counsel parents before there is any danger of the baby becoming ill. It is a tragedy to lose this opportunity.

Babies with traits

The baby's HbO status remains the same for life. It is sensible therefore to have a system for communicating this information to the health visitor and family doctor. This can be done via routine discharge papers, employing special standard format letters. All neonatal HbO results could be sent to the Child Health Records Department, which could be briefed to relay them to the appropriate health visitor. If such an arrangement is instituted, it is important that parents are informed of it.

Although few health authorities have the resources to provide counselling to every family, it is imperative that suitable literature is available, with contact addresses of organisations from which additional information can be obtained.

It is important that the baby's HbO status is known so that every attempt can be made to ensure as healthy a life as possible for the growing child. Medical problems encountered by children with sickle cell disease are discussed below. The families of babies with traits will need to be aware of the implications for any other children they may have, and indeed for the children that their child may have in the fullness of time.

Babies with a major HbO

Ideally, each parent of an affected child will not only be aware that a major HbO is a possibility before the baby is born, but will have already met the HbO counsellor. However, the real world is not always quite like this.

Parents should be informed as soon as possible after the diagnosis is made, by a person confident about the information who is able to provide appropriate counselling. Shock, denial, depression and anger are all normal responses. An intensive follow-up counselling programme must be available and must include the repetition of information already given, as well as a place for coming to terms with the difficult feelings involved. This must be sensitively provided in response to the needs of the family. Each member of the team must behave towards the parents with understanding. Parents of children with special needs frequently report the distress caused by the insensitive behaviour of others. They rarely wish for the subject to be ignored, or to be blindly 'reassured', or to be advised 'not to think about it'. It is nearly always possible to acknowledge that one is aware of the news, offering an opportunity to talk, and it is kind to do so. One does not have to be very knowledgeable, as just listening quietly and

with acceptance is valuable. People have different needs for companionship and solitude at different times, and withdrawal does not always mean that a person wishes to be left alone.

In spite of the enormity of the news for the parents, the baby is not at any risk from its blood disorder at this stage. At birth, and for at least 4 months, babies are protected by their fetal Hb. It is not until they are about 4–6 months post-term, when they switch to making their adult version of Hb (which in their case will be HbSS or some other combination) that they are at risk. As far as their HbO is concerned, they can be treated exactly the same as any other baby. Guilt, grief, anger and other feelings may make this difficult for the parents to appreciate, but it can be helpful to have this pointed out.

Children with sickle cell disease are likely to be at particular risk from overwhelming septicaemia, most often caused by pneumococcal infection to which their antibody response is greatly impaired, and from splenic sequestration. Both of these events can cause death in a matter of hours. However, they are entirely preventable through the simple measures of penicillin prophylaxis throughout childhood and by teaching mothers the signs to look out for. Mothers can be taught to palpate the spleen, the consequent early warning resulting in a dramatic reduction in mortality from this complication. These two measures illustrate the vital importance and benefit of the early diagnosis of these conditions.

PREGNANCY AND SICKLE CELL DISEASE

Early papers on this subject suggested that patients not only had reduced fertility, but were also more likely to spontaneously abort, have small babies and show raised perinatal and maternal mortality rates (Serjeant 1985). Although more optimistic figures are now available, it is clear that pregnancy is a hazard for these women and their babies (Konotey-Ahulu 1991; Howard & Tuck 1995). It is also very unpredictable and the quality of maternity care may be a crucial factor (Adams 1994). Pregnancy may also have a deleterious effect on the woman's symptoms, and life-threatening events may be more common (Serjeant 1985; El-Shafei *et al.* 1992; Howard & Tuck 1995).

There are also significant risks to the fetus. Miscarriage (in around 19 per cent of pregnancies), premature labour, low birthweight (in approximately 31 per cent of babies) and raised perinatal mortality have all been reported (El-Shafei *et al.* 1992; Brown *et al.* 1994; Howard & Tuck 1995). Brown *et al.* (1994) found the most common complications in fetuses and newborns with sickle cell disease to be jaundice (25 per cent), fetal distress (13 per cent), anaemia (10 per cent) and respiratory distress (6 per cent). It is of paramount importance, therefore, that those caring for women whose pregnancies are complicated by sickle cell disease are well informed,

meticulous in their observations and sensitive to their clients' individual concerns and needs.

These pregnancies must be supervised by an experienced team comprising midwives, haematologists, obstetricians and an HbO counsellor. The woman's haematological and obstetric progress must be carefully monitored throughout the pregnancy, particular attention being paid to any changes in her general health and the development of her baby. In addition to other obstetric care, Doppler velocimetry (ultrasound) may be helpful in early detection of babies at risk of low birthweight (Billett *et al.* 1993).

Medical factors can be particularly closely intertwined with social and psychological factors in this disease. Stress affects the circulatory system in ways that make sickling more likely to occur. This is commonly misunderstood, and patients are likely to be accused of coming into hospital 'just because they've got problems at home'. They may well have problems at home, but now they are in hospital as well. Help must be provided to resolve those problems if the woman's health is to improve. It is therefore imperative that a holistic approach is used and referral to and liaison with other agencies fully implemented. To fail to do this is to fail the woman and jeopardise her health and her baby's future. All aspects must be closely monitored in order to ensure the best possible outcome for the woman and her baby.

THE ROLE OF THE MIDWIFE

The midwife has a crucial role in the care of women with HbOs. In caring for those with a chronic illness, it is important to remember that they have a significant history with their illness within the health care system, and that they often know more about their illness and what they need than do many of their carers. Past experiences may have left them ambivalent towards health professionals, and repeated periods of hospitalisation may make them reluctant to attend clinics unless it cannot be avoided. In addition, having sickle cell disease may have had an important impact on a person's self-image, beliefs, self-efficacy, relationships and support structures (Leavel & Ford 1983; Morgan & Jackson 1986; Gil *et al.* 1989; Midence & Shand 1992). It is vital that a good, trusting relationship with the woman is initiated as early as possible, and that every effort is made to ensure the appropriateness of her care.

It must be remembered that the woman with sickle cell disease has limited energy that must not be wasted by inefficient systems; hospital appointments, for example, must be arranged to coincide whenever possible. She needs to understand why her lifetime pattern of care might now have to be changed. She must be enabled to feel confident about pathways of admission, and how to obtain advice about any concern she may have regarding herself or the baby.

Many people with sickle cell disease tend to postpone hospital admission until the last possible moment. A woman who is not pregnant and who is familiar with her own body will not usually be placing herself in serious danger by this approach. During pregnancy, however, the situation is different. Both the severity and the speed of onset of a crisis during pregnancy may catch the woman unawares, endangering her own life and/or that of her baby. She therefore needs to understand the requirement for earlier presentation, and efforts must be made to encourage her to do so.

Preconception care

Genetic counselling should ideally be provided long before reproduction is embarked upon, so that the woman is aware of her own and her partner's blood test result and the possibilities for their children. Where a pregnancy has been planned, it is much more likely to commence with the woman in possession of all necessary information of the possible courses and outcomes. Unfortunately, this is not always the case (Howard *et al.* 1993).

If there is a risk of the baby having a serious HbO, the mother should be counselled by a trained HbO counsellor to elicit her feelings and wishes regarding prenatal diagnosis and termination. There is no 'right' or 'wrong' decision. What is important is the opportunity for informed choice on the part of the parents. An additional consideration for a woman with sickle cell disorder who is considering prenatal diagnosis is the stress of looking after a sick child when her own health is not good, and the effect on her and the baby of repeated separations due to the illness of one or the other.

The booking visit

A careful history must be taken, as always, with special attention being paid to the family history of sickle cell disease and other illnesses. It is not at all uncommon for women to have lost other children with sickle cell disease, or to have sisters, cousins or friends who have died of it, sometimes during pregnancy or the puerperium. This is bound to have a profound effect on the woman's confidence, increasing her anxiety, and should be sensitively borne in mind throughout her care.

It can be most useful at this point also to obtain a history regarding the usual course of the woman's illness. This can provide a useful comparison later when trying to gauge the effect of the pregnancy on the course of her condition. A deteriorating picture will indicate the need for more intensive observations, or even intervention. This could be medical but may be simply a matter of support to make life less stressful. All concerns should be immediately and aggressively responded to, as there is rarely a place for 'wait and see'.

Partners should be tested at this stage, if this has not already been arranged, but it must be remembered that if the partner is unavailable, anxiety may be caused by the suggestion of testing. The most common benefit of knowing the partner's Hb status is that if he does not have an HbO, there is reassurance that the baby will not be seriously affected.

Sources of support

The booking visit is not too soon to discuss the support upon which the woman will be able to call after the baby is born. This needs to be re-examined throughout the pregnancy as her circumstances may change. At the booking visit, it is important to identify any possible shortfalls and begin at once to address these by working with her and other agencies. Caring for a new baby is an exhausting job for anyone, but it poses an additional strain on the precarious health of women with a sickle cell disorder. If catastrophes and potentially disastrous early separations of mother and baby are to be avoided, extra support may well be needed, and early planning for this provision is essential.

The woman's current social situation also needs to be explored. Poor or insecure housing can have a deleterious effect on her health, as can financial worries, poor diet and insufficient rest. Referral to a social worker should be made and the importance of these issues made clear. Strong letters to official bodies, such as housing departments, made by a health professional can be extremely effective, and midwives should consider writing them; persuading the consultant to countersign them may further increase their impact.

The need for additional blood tests should be discussed with the haematologists so as to avoid unnecessary venepuncture. Venous access is precious and venepuncture unpleasant.

Follow-up visits

The woman should be seen at each visit by a consultant obstetrician or a senior registrar experienced in the care of sickle cell pregnancies (Serjeant 1985; El-Shafei *et al.* 1992). Continuity is especially important with such women, as the signs of deteriorating health may be quite subtle and may be missed or ignored by someone unfamiliar with the condition or the patient.

Being more than usually susceptible to infection, women with sickle cell disease are especially prone to urinary tract infections (Serjeant 1985). These can make them extremely ill and must be diagnosed and treated as soon as possible. A midstream specimen of urine should therefore be obtained at each visit.

There is a low threshold for action if intrauterine growth retardation is suspected (El-Shafei *et al.* 1992; Brown *et al.* 1994; Howard & Tuck 1995), and regular Doppler scans should be carried out.

Blood transfusions

There is a seemingly endless debate regarding the relative value of blood transfusion in pregnancy (Koshy & Burd 1991; Howard 1994). Not only does blood transfusion carry the risk of introducing infection, but repeated transfusions may in addition cause accumulation of antibodies in the recipient, which will eventually render her untransfusable. Largely agreed criteria for recommending transfusion would seem to be deterioration in maternal health, life-threatening events, twins or a catastrophe in a previous pregnancy. Each case must be considered individually, although there are a few centres where blood transfusions are offered routinely to all pregnant women with sickle cell disease.

Crises

The most common crisis to occur is the painful bone crisis. This is rarely dangerous but is an indicator of ill health and, more importantly to the patients, can cause excruciating pain, for which enormous doses of analgesia are required. It is commonplace for doses of pethidine as high as 200 mg 2 hourly to be required for days or even weeks at a time, causing great anxiety to carers. The current literature does not identify drug dependency in the neonates of women with sickle cell disease. This may be due to the reduced risk of physical dependency when the drugs are used appropriately, that is, to control pain. Accusations and/or threats of 'addiction' are among the most common and distressing experiences for people with sickle cell disease and are often made by ill-informed people at times of immense personal distress for the person concerned.

Labour and delivery

Labour is a time of extreme vulnerability for a woman with sickle cell disease. Stress, both physical and psychological, hypoxia, dehydration and changes in body temperature are all likely occurrences in any labour and are the very situations most liable to precipitate a crisis. In addition to this, the woman may have deep and unexpressed fears about the risks to her own and her baby's life.

It is of paramount importance to form a trusting relationship with the woman as soon as possible and to take every opportunity to offer reassurance. Midwives are often reluctant to address the concrete fears of the women they care for, and find it hard to know how to do this, but it would

be cruel to ignore these fears. It is easy to ask, 'Are you scared?' Whatever the answer, this can be followed up with a clear statement that although it is understandable to be frightened, the woman is not in danger, and that you, her midwife, are taking personal responsibility for her safety. Details of what measures are being taken to ensure she is safe should then be given.

Koshy and Burd (1991) have suggested that labour should be managed as for a patient with congestive heart failure. The following are vital considerations.

1. *Adequate analgesia.* This is important in order to reduce stress and maintain trust. It must be remembered that the woman may have a lifelong and tortured relationship with pain, and that she may have some level of tolerance to analgesics, thus requiring higher doses than would normally be expected. Epidural analgesia is effective and encouraged for these women.
2. *Oxygen therapy.* Many centres prescribe this routinely, but it is certain that there should be a low threshold for its initiation.
3. *Hydration.* Dehydration must be avoided at all costs. Again, many centres set up intravenous therapy regimes at the onset of labour. It may be possible to maintain adequate hydration by the oral route, but in view of the copious amounts required, the intravenous route is probably necessary. Renal damage, leading to an inability to concentrate the urine, means that an oral intake in excess of 3 l per day is required in normal circumstances. Drinks should be fresh and acceptable – 3–4 l per day of tepid tap water is not pleasant! It must be remembered that fluid intake and output should be closely monitored.
4. *Fetal monitoring.* Perinatal morbidity and mortality rates are significantly raised in these cases, so fetal monitoring should be continuous and meticulous.

As long as there are no contraindications, a vaginal delivery is preferable. Pelvic contracture, however, is a risk in these women, resulting in cephalopelvic disproportion. Should a caesarean section be necessary, postoperative care should be as for any other woman – early mobilisation, adequate analgesia and sufficient rest.

The puerperium

Postnatal care is the same as for other women, with the proviso that the midwife is aware of potential problems and is prepared to take any necessary action earlier rather than later. Many women with sickle cell disease are well during labour and the first few postnatal days, only to become seriously ill between the fourth and seventh days. This process is not fully

understood but may be a delayed manifestation of the stress of the labour, the result of an untreated infection, a response to hormonal changes or the effect of the exhausting work of caring for a new baby.

For this reason, it is necessary to practise the utmost vigilance while caring for the woman and to provide as much help as possible with the baby. A single room may also enable the woman to obtain more rest. A woman with sickle cell disease should never be discharged home before the seventh day, and then only if both she and the baby are completely well. Home visits by an experienced community midwife who is aware of the issues should, at minimum, be made at least once a day until the 14th day.

Throughout this period, attention should be paid to the mother's general health and energy, any signs of infection or slow healing and whether she has any pain. Before discharge, the midwife should ensure that adequate support is available, and if it is not, social services should be informed and encouraged to help.

Contraception

Oral contraceptives are often withheld from women with sickle cell disease in the belief that the medication is dangerous for them (Howard *et al.* 1993). However, there is little or no evidence for this belief, and the real risks of a pregnancy for these women must be remembered. The theoretical risks of oral contraceptives do not constitute sufficient reason to withhold them from women for whom they are the most appropriate method of contraception (Serjeant 1985).

Intrauterine devices are also often discouraged because they are thought to cause infection. There is no evidence of an increased rate of intrauterine infection in women with sickle cell disease (Serjeant 1985), but it might be advisable to bear in mind that any infection might have rather more serious consequences for these women.

Many women are now using condoms, as they provide protection from sexually transmitted infections as well as pregnancy. They can, however, be unreliable, as can other barrier methods. Depot preparations of medroxyprogesterone acetate can be a useful and effective contraceptive, as its absorption is not affected by gastrointestinal disturbances, to which this group of patients is prone. It has also been suggested that it might have a beneficial effect on the course of sickle cell disease and thus might have the dual benefit of providing both effective contraception and an improvement in the woman's health (Serjeant 1985).

For permanent contraception, tubal ligation is the procedure of choice if a permanent male partner is unwilling to undergo vasectomy.

Contraception is an important issue for a woman with sickle cell disease, and any decision that she makes should be the result of correctly informed choice (Howard *et al.* 1993).

THALASSAEMIAS

There are two common forms of thalassaemia: alpha thalassaemia (A thal) and beta thalassaemia (B thal), affecting the production of alpha- and beta-globin chains respectively. Fewer chains are made, so less total Hb can be made. On routine blood tests, A thal traits have a picture almost identical to that of iron deficiency, with which they are commonly confused. It is vital, however, to differentiate between these two conditions if correct responses are to be made. People with A or B thal trait require expert genetic counselling.

Babies who inherit A thal from both parents are unable to make either adult or fetal Hb. This is incompatible with fetal life, and the baby is usually born prematurely as a hydrops fetalis or stillbirth.

If B thal is inherited from both parents (beta thalassaemia major), the baby will be unable to make any adult Hb. This is obviously incompatible with life, and the individual will need blood transfusions for life. In addition to this, iron chelation therapy will be required to prevent the life-threatening complications of the resulting iron overload. Currently, the only way of undertaking this is by subcutaneous transfusion, which takes about 12 hours a day, 6 days a week throughout life. It is therefore vital to obtain a correct diagnosis so that these families can be given the opportunity of receiving all the information and support that they require.

SUMMARY

Sickle cell disease and thalassaemia are inherited abnormalities of Hb manufacture. They affect every aspect of the sufferer's life. This is particularly pertinent during pregnancy when all women, not only those with additional health problems, have additional needs for understanding and support.

All women at risk of having inherited an HbO should be offered preconception and genetic counselling. If they are already pregnant, antenatal testing with appropriate counselling follow-up must be offered.

All pregnant women with sickle cell disease should be regarded as being at 'high risk' during pregnancy, as should their unborn children. The midwifery challenge is to provide responsive, woman-centred care to this special client group. The midwife must develop a trusting and empathic relationship with the woman as early as possible and monitor her complex and changing needs. She should take every opportunity to empower the woman and facilitate informed choice. She should acknowledge that the woman herself is likely to be an expert on her own medical condition and should respect her accordingly. In addition, she must have the courage to learn to address the issues pertinent to her patients, such as racism or the fear of dying or losing the baby, so that real trust can exist, and reassurance and comfort be given.

The midwife must be aware of the obstetric and medical complications to which the woman may be prone. She should bring all her skills of observation to bear and be vigilant for signs of problems. Once the woman is discharged back into the community, midwives and social service staff should be made fully aware of her situation.

Discharge from midwifery care is not an end – it is only the beginning of the mother's and baby's life together. If one or both are prone to serious illness, there will be many difficulties to be encountered. Midwives have the privilege of being in a position to facilitate a good relationship between them in the early days, in order to sustain them through the difficult times – this too is part of the midwifery challenge.

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