Legal

Avoidable pitfalls when writing medical reports for court proceedings in cases of suspected child abuse

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Avoiding pitfalls

All paediatricians, paediatric radiologists, paediatric pathologists, forensic pathologists, and many other specialists have to deal with cases of suspected child abuse, and in terms of the generation of complaints from families this is a high risk activity. Many complaints are devoid of merit, but in some cases a complaint is justified because of a faulty approach. This review draws attention to the avoidable pitfalls associated with report writing when child abuse is under consideration (see box 1).

The modern guidance on the duties of experts in cases of suspected child abuse stems in part from a care proceedings case heard in 1990. A 3 month old baby was admitted to hospital with serious injuries comprising subdural haemorrhages of more than one age, damage to the brain, multiple rib fractures, and multiple limb fractures. Despite the evidence pointing to child abuse, a number of experts from various disciplines offered a variety of unjustified or highly improbable alternative medical explanations (including abnormally fragile bones) for the injuries. The judge was highly critical of some of the expert reports and evidence given in court, and in his published judgment he gave clear advice concerning the duties of an expert.[1] This and subsequent advice from various authorities on the work of an expert[2–12] is summarised in box 2. In addition, a High Court judge of the Family Division has written a useful handbook providing simple and practical guidance on all aspects of the duties of experts in care proceedings.[13]

DIFFERENT LEGAL PROCESSES

Care proceedings in England, Wales, and Northern Ireland involve a two stage process. The court must be satisfied, firstly, that a child is suffering or is likely to suffer significant harm attributable to care given to the child or its absence. These are known as the threshold criteria. If they are met, the court goes on to consider whether a care order is warranted on welfare grounds. Sometimes these two aspects are dealt with at separate court hearings. The first hearing, sometimes referred to as a "finding of fact hearing", is devoted to whether or not a child has been harmed, and if so by whom. If the finding of the court is that the child has been harmed, then following a period of further investigation, including a risk assessment, and maybe a psychological assessment (of parents and/or children) and/or a psychiatric assessment, a second hearing (often rather inelegantly called "disposal") decides whether or not a final order should be made, and if so what long term plans should be made to safeguard and promote the child's welfare. The input of paediatric reports is mainly into the first part of the process. Parties to the process are the local authority, the child (whose interests are represented by a children's guardian), the parents, and possibly other relatives. The paramount concern is the welfare of the child.[13 14]

In criminal proceedings, the focus is on the criminal prosecution of one or more individuals who are suspected of harming or neglecting a child.

In some cases, civil and criminal proceedings run more or less side by side, and this carries the potential, discussed later, of directly interfering with the work of a paediatrician whose task is to prepare a report.

PROTOCOL FOR CASE MANAGEMENT IN CARE PROCEEDINGS

To avoid undue delay in making decisions about the future of a child, on 1 November 2003 a protocol for judicial case management was introduced.[14 15] The aim of this protocol is to complete a case within a maximum of 40 weeks, and one effect of this is to put pressure on doctors to provide reports by a specific date. Appendix C to the protocol provides a short but helpful code of guidance for experts.

MOST READERS OF REPORTS ARE NOT DOCTORS

Because most readers of reports in cases of suspected abuse are not medically or scientifically trained, it is essential to avoid or to explain any jargon. It is often helpful to provide a glossary that lists and briefly explains any technical terms.

When writing reports be aware that non-medical readers of the report will not necessarily apply appropriate caution to medical opinions, for example the dating of bruises or fractures. It is worth bearing in mind that some take medical opinion as gospel, rather than appreciating the limits of medical science.

THE OVERRIDING DUTY OF THE DOCTOR IS TO THE COURT

Regardless of whether one is dealing with civil or criminal proceedings, the overriding duty of the doctor preparing a report is to the court, and not to any of the parties such as the prosecution, the defence, the local authority, the parents, or the child. This contrasts with the primary responsibility in clinical care, where the primary duty is to the child. The doctor who cannot come to terms with this fact may with the best of intentions sow the seeds for serious criticism as events unfold. The scenario is simple enough: a child is admitted with what is thought to be abuse; the paediatrician, convinced that the child needs to be protected from the presumed perpetrator, unwittingly or deliberately exaggerates items in the report, such as the likelihood of abuse, the degree of pain suffered, or the amount of force inflicted. The aim of the doctor is to do everything possible to ensure that the child is protected. This is how the caring and concerned doctor can take the first steps on a path that ultimately may lead to criticism. In the face of a child with worrying injuries, it is essential to resist a natural tendency to overstate or exaggerate aspects of a report.

THE MEDICAL CHRONOLOGY; A USEFUL TOOL

It can sometimes be helpful to one's own thinking, and is useful to non-medical people involved, to prepare a medical chronology that lists exactly what happened and when, taking care to include information from typewritten sources (letters, discharge summaries), handwritten medical notes, handwritten nursing records (which often contain information not included elsewhere), and the results of investigations.

Box 1: Common medical issues that may arise in child protection cases

- Precise delineation of injuries such as bruises and fractures
- An attempt to identify the age or likely age range of an injury
- An attempt to indicate the likely type and degree of force resulting in an injury
- Consider the type of mechanism which could have caused the injury
- The differentiation between natural disease and pathology resulting from abuse
- The differentiation between accidental and non-accidental injury
- The identification of neglect
- The delineation of the likely symptoms exhibited by the child in a period prior to presentation to the health or welfare services

Box 2: Duties of an expert witness

- Experts should act independently of the parties and the exigencies of the court process
- Experts must express only opinions which they genuinely hold and which are not biased in favour of one particular party
- Experts should not mislead by omissions. They should consider all the material facts in reaching their conclusions, and they must not omit to consider the material facts which could detract from their concluded opinion
- Experts should state the facts or assumptions on which their opinion is based
- If experts look for and report on factors which tend to support a particular proposition or case, their report should still:
  - provide a straightforward, not a misleading opinion
  - be objective and not omit factors which do not support their opinion
  - be properly researched
- If the expert's opinion is not properly researched because insufficient data are available, that must be recorded and the opinion offered as only a provisional one
- Experts must express only opinions which they genuinely hold and which are not biased in favour of one particular party
- A misleading opinion from an expert may well inhibit a proper assessment of a particular case by the non-medical professional advisers, and may also lead parties, and in particular parents, to false views and hopes
- A misleading opinion from an expert is likely to increase costs by requiring competing evidence to be called at the hearing on issues which should in fact be non-contentious
- An expert should not assume the role of an advocate. Experts have a duty to be objective, and not to mislead. The welfare of the child may be at stake, and an absence of objectivity may result in a child being wrongly placed and thereby unnecessarily put at risk
- An expert who is involved in new research should be conscious of the natural tendency to promote his or her own findings, and should make every attempt to avoid becoming subjectively biased
- Experts should make it clear when a particular question or issue falls outside their area of expertise
- Experts should have sufficient practical experience in the area on which they are commenting
- If the medical evidence points overwhelmingly to non-accidental injury, an expert who advises that the injury has an alternative and innocent causation has a heavy duty to ensure that he has considered carefully all the available material and is, moreover, expressing an opinion which takes that material fully into account and which can be objectively justified

INTENT AND MOTIVATION

The binary terms "accidental" and "non-accidental" and "unintentional" and "intentional" are often employed. However, they mean different things, and both pairs have their drawbacks. There are two key points to be made. The first is that intent to harm a child is uncommon. In most cases of child abuse, the adult loses control of his or her reactions and injures a child. The deliberate and calculated act of setting out, in cold blood, to harm or injure a child is believed to be rare. It is therefore incorrect and needlessly inflammatory to label all cases of child abuse as "intentional", a term that is often equated by lay people with the word "deliberate". The second point is that no-one can look at an injury and know with certainty the intent of the person who caused the injury, although there are a few particularly extreme cases where it is only natural to wonder what was going through the mind of the person who caused the injuries. The key point is that the job of the paediatrician (or radiologist or pathologist) is to distinguish between accident or naturally occurring disease on the one hand, and abuse on the other. The paediatrician cannot know the mindset of the person believed to have caused the injury, and speculation on this aspect should be avoided. Once it has been established by the court that the case is one of abuse, then experts in social work, psychology, and psychiatry can become involved and examine motivation.

Paediatricians should not try to be mind readers. The implication is that the terms "intentional" and "unintentional" should be avoided in paediatric reports on the topic of suspected child abuse.

INTERVIEWING THE PARENTS OR CARERS

When a child is referred to a paediatrician because of a clinical problem, the paediatrician has three main tools: the history, physical examination of the child, and a study of the available data including investigation results and previous records. Most paediatricians would not dream of giving a clinical opinion without taking a history, and yet it is not uncommon for paediatricians to undertake a "paperwork exercise" of preparing a report without ever meeting the child or the family. While there is a desire to avoid needless repetition of physical examination of small children, particularly long after an injury has occurred, it is hard to justify avoiding speaking to the parents or carers.

It is likely that the paediatrician who has been asked to conduct a paper

exercise will have available witness statements from the parents and/or carers and others, and there may also be a transcript of police interviews. There should also be copies of the medical records. Alas, none of these is a substitute for a properly taken history of ones own. However detailed are interviews by police or lawyers, neither have the medical knowledge, training, or expertise to take a medical history. Furthermore, the medical records all too often contain incomplete histories obtained by the admitting junior doctors. The fact is that all clinicians perform their clinical duties with the aid of various routines, and departure from these routines by the use of short-cuts may be associated with a greater risk of error. Interviewing parents or carers is time consuming, and may be inconvenient and cause delay, but preparing reports in cases of possible child abuse is not an activity where a doctor should contemplate second best.

A paediatrician who does not attempt to interview the parents risks being criticised for by-passing the usual routines and failing to consider all aspects of the case. Recent press publicity has indicated that parents may be exceptionally aggrieved by paediatricians willing to make a confident diagnosis of abuse without ever meeting the parents and giving them the opportunity to explain their side of the story.

Although civil and criminal proceedings arising out of the same injuries are separate processes, there are various legitimate ways that information can flow between the two. Thus it is that in some care proceedings, lawyers acting for parents may advise them against being interviewed, for fear of disclosures that could assist an ongoing police investigation or criminal prosecution. The practice of giving such advice has been ''strongly deprecated'' by a High Court judge of the Family Division, notwithstanding the fact that the parent sought to be interviewed was suspected of killing two of her children.[16]

Having mentioned the importance of interviewing carers, in the light of recommendation 65 of the Victoria Climbié Inquiry report,[17] it is worth also remembering (in selected cases) the importance of considering interviewing the child.

STUDYING THE MEDICAL RECORDS

It sometimes happens that one is invited to prepare a report on the basis of very limited information such as a telephone call, a few photographs, or a couple of witness statements. Requests to prepare reports in child protection cases without access to vital materials such as the child's medical records should always be resisted; it is absolutely essential that the full medical records are sought.

EXAMINING THE PATIENT

The need for a child to be re-examined specifically for the purpose of preparing a report will depend on the circumstances. It is important to give early notice of a need to examine a child and the reasons for the examination, as children who are the subject of care proceedings cannot be examined without the consent of the court.

CLINICAL PHOTOGRAPHS

It is common for solicitors to fail to provide photographs, sometimes because they do not know of their existence. A study of the nursing section of the hospital medical records may uncover the fact that a hospital or police photographer visited, and one may have to ask for these photographs to be obtained. One should insist on good quality glossy prints of original photographs, enlarged if necessary, and not make do with laser prints or colour photocopies.

It is worth remembering that photographs, particularly those of very extensive injuries, have the potential to be highly prejudicial, which is why in many abuse cases photographs of the dead and injured child may well be withheld from the jury. Paediatricians are only human, and require considerable self-discipline to stand back and try to make an objective analysis of photographic evidence.

Photographs or video recordings of the home, the scene, and any domestic items should be viewed if they are available. They may be helpful in determining, for example, whether or not a particular fall was from a low level.

It is important to bear in mind that for a variety of reasons, photographs of suspected injuries can be misleading. Photographs may fail to show lesions that were undoubtedly present, either because the quality of the image is poor or because the lesion was particularly difficult to photograph. In some situations photographs can suggest the presence of lesions or injuries that do not exist, and in a recent case no less than five experts were seriously misled into incorrectly diagnosing abuse because two-dimensional colour photographs failed to accurately reflect a three-dimensional normal variant appearance.[18]

ESTIMATING THE AGE OF BRUISES

It is common to be asked to provide an opinion on the age of bruises. Textbooks have provided schedules of colour change of bruises over time. However, these schedules are unreliable,[19] and should not be used. A few rather limited studies of the colour changes of bruises, some using colour photographs and some using direct observation, all indicate that attempts to age bruises based on their colour is fraught with difficulties.[20–23] The time course of the appearances of bruises may vary with the location, depth, extent, and nature of a bruise. The only established fact is that the presence of a yellow colour within a bruise indicates that it is at least 18 hours old.[20 21]

It is perfectly reasonable for an experienced doctor who has examined the child to state that a purple bruise ''looked fresh'', meaning it was unlikely to have been inflicted more than two days previously,[19] but considerable caution is required when estimating the upper and lower age limits for bruising.

Histopathology of bruises is worth performing in some fatal cases. Histology can rule out skin lesions such as blue naevi and can confirm bruising in those with dark skin. Histology of a bruise may enable the pathologist to say it is very fresh (no cellular reaction), recent (infiltration with neutrophils), or more than approximately two days old (presence of haemosiderin).

BLOOD COAGULATION TESTS

A frequent error is to believe that a normal ''clotting screen'' (such as a full blood count, prothrombin time, and partial thromboplastin time) excludes the presence of a coagulation disorder. It does not. It only excludes the commonest conditions that may cause spontaneous bleeding or serious bleeding following trauma.[24] A ''clotting screen'' is no more than a screening test, and if there are pointers to a coagulation disorder (such as a history of a bruising or bleeding tendency in the patient or the family), referral to a haematologist and further investigation will be required. There are a number of coagulation disorders, individually but not collectively very rare, that may pass undetected by a ''coagulation screen''. Just one example is the Hermansky-Pudlak syndrome, normally a mild disorder of platelet function, but which in one 7 week old infant was associated with both subdural and retinal haemorrhage.[25]

THE INTERPRETATION OF RADIOGRAPHS

It is essential that radiographs are reported by radiologists who are familiar with the broad brush of children's radiology, particularly in relation to the different patterns of injury in accidents. It is also essential that the radiologist has knowledge and experience of paediatric

disease, its effects on the radiographic appearances, and the numerous radiological appearances which are normal variants. In practice this means that the films need to be seen by a paediatric radiologist. Copy films vary in quality, and numerous errors have been made as a result of relying on copy films rather than ensuring that the original films are supplied.

Caution is required when interpreting routine clinical reports on radiographs. The original request form for the investigation will usually be very restricted (for example ''? fits ? cause'') and often omit essential clinical information. The report provided by the radiologist at the time may not have been written with forensic investigations or legal proceedings in mind. The author of the report may need to be approached to clarify matters or provide further information.

FORCES NEEDED TO CAUSE INJURY

A 3 month old infant is found to have unexplained healing rib fractures and metaphyseal limb fractures, and one is asked to comment on the forces required to cause the injuries. One is well advised to exercise caution and avoid dogmatic statements about how flexible are the bones of babies and how enormous the forces must have been. The truth is that the requisite scientific studies, in which human infants are deliberately injured in different ways and with different degrees of violence in order to establish precisely the nature and extent of the forces required to produce injuries, have not been performed and never will be. The only exception to this is the unusual studies by Weber in which dead babies were dropped on to various surfaces from a height of 82 cm in order to study the genesis of skull fractures.[26–28] Biomechanical studies using theoretical constructs or various models cannot provide the required human data. The nature of child abuse is such that, unlike accidental injuries, the reliable accounts of independent witnesses are rarely available. Confessions of perpetrators may tend to understate the degree of force. The only really solid ground is that in an infant with healthy bones, normal handling and normal activities do not produce fractures, and domestic accidents (such as short falls) rarely produce significant injury. It is self evident that significant force must be needed to break a bone, but in the absence of any reliable hard scientific data it is wise to avoid overstating the amount of force that is likely to be involved.

PAIN RESULTING FROM FRACTURES

Fractures cause two kinds of pain. One is acute pain resulting from the forces applied to the bone and the pain resulting from the bone breaking. The other is ongoing pain occurring in the days and weeks after a fracture has occurred. The immense variability means that overconfident assertions are worth avoiding. While the occurrence of the fracture itself is certain to cause significant immediate pain, the way that this pain is communicated to carers or parents can vary between different children and at different ages.

In some cases the ongoing pain is obvious for all to see, and the limb of a child is manifestly not being used for some days after a fracture has occurred. However, ongoing pain after a fracture is highly variable, and, for example, in infancy, rib fractures and metaphyseal limb fractures often produce no detectable ongoing pain at all, presumably because the bone is stable and the periosteum often little disturbed. These injuries commonly pass undetected by clinicians examining the child and the nursing staff looking after the child. This is sometimes because the fractures are in a state of healing, and sometimes because something else is dominating the clinical picture. The point is that caution is required before concluding that a reasonable carer should have known that something was seriously amiss in a child with rib or metaphyseal limb fractures.

BIAS

It requires constant vigilance to avoid bias creeping into a report. On reading the medical records of a child with a suspected combination of abuse and neglect, one finds that the patient has failed to attend appointments in the hospital or the community, and accordingly (and quite innocently) one adds to the list of problems ''three failed outpatient appointments''. What is at fault is that probably quite inadvertently one has made a point of including negative information without putting it into context, and without providing the available positive information. In fact, although our hypothetical patient missed three appointments, he or she kept 15 others. Furthermore, when the child was unwell the mother always made a point of seeking medical help. Finally, the data for the family have omitted the normative data. Many other babies in the neighbourhood may be missing just as many appointments; this is not an excuse for missing the appointments, but it is important contextual information.

Selective extraction of negative information is one of the most common faults in medical reports. Vigilance and a strong sense of fair play are needed to avoid this trap. There are few human beings for whom one can find nothing positive to say.

A doctor who is involved in new research should be conscious of the natural tendency to promote his or her own findings, and should make every attempt to avoid becoming subjectively biased. It is essential that an expert considers and mentions in a report all relevant material, including that which tends to throw some doubt on the expert's conclusion.[11]

PARENTING ISSUES

Instructions to experts rarely invite conclusions about the quality of parenting, but often include a catch-all instruction inviting comment on any issues not covered in the instructions. The doctor preparing a report notices in the papers that one or more previous children, siblings of the child under consideration, had to be placed in foster care a number of times. In addition, the mother has failed to take the child to a number of appointments, and there are suggestions that the children have not been fed regularly, are filthy dirty, and are understimulated. After careful study, the doctor becomes anxious about this mother's abilities to look after children. The actual evidence of abuse is far from clear, but the doctor is seriously worried about the welfare of the child. The pitfall for the caring doctor, fearful that in the absence of evidence of abusive injuries the child is going to be left in the care of this apparently inadequate mother, is to try to compensate for the lack of hard evidence of abuse by over-stressing the general concerns. This is the beginning of a slippery path that leads to the final but wholly inappropriate conclusion, along the lines of ''this mother is totally unfit ever to have the care of children again''. However experienced, doctors should never make recommendations of this sort. This particularly applies to experts parachuted into the case who have never met the family or visited the home! Decisions about the child's future are for the court to determine, after appropriate input from those with the relevant skills. Paediatricians must remember they are not social workers, clinical psychologists, or child psychiatrists.

It is perfectly valid, indeed quite proper, for a doctor to highlight areas of concern. But however grim the circumstances, and however valid the criticisms of the child's care, a parent is likely to react with great bitterness when sweeping recommendations are made by doctors, particularly if the doctor has not met the family, or visited the home, or been involved in the care of the children in recent years. Furthermore, courts may

accord an exaggerated respect to a doctor's views about non-medical matters, making it all the more important to avoid excessive comments about poor parenting.

USING A PROFILE OF A CARER TO MAKE A DIAGNOSIS

A 4 month old infant is found to have multiple rib fractures; the radiological appearances (different stages of healing) suggest at least three separate episodes of injury, accompanied by two metaphyseal fractures of the long bones and a human bite mark. The carers are unable to offer any explanation as to how the injuries might have occurred. The fact that the mother's current partner is not the father of the child, was himself in care in his own childhood, and has a record of violence, is of no value in proving that the diagnosis is one of abuse, a diagnosis which must be based solely on the features of the injuries and the lack of any explanation.

It is well recognised that stepfathers can sometimes harm a child, but the fact that a child with an injury has been in the care of a stepfather is not an indicator that injuries are the result of abuse. As with cigarette smoking and lung cancer, being a stepfather, having been in care in one's childhood, and a previous criminal record for violence are risk factors for abuse. Risk factors cannot be used to make a diagnosis, but they can help explain why a condition has occurred, and they may inform a risk assessment of future harm.

These simple facts are sometimes overlooked in cases of Munchausen syndrome by proxy (the current fashion is to call this ''fabricated and induced illness''). Instead of focusing on the primary issue, such as ''has this child been suffocated, causing attacks of apnoea?'', or ''has the history of haematuria been fabricated?'', the mistake is to attempt to make the diagnosis by constructing a personality profile of an alleged perpetrator and then show that the adult carer in question falls into the described pattern. This is a serious error.

BE PREPARED TO DISSECT AND ANALYSE INDIVIDUAL COMPONENTS

When making a diagnosis, the treating paediatrician may look at the overall picture, but those investigating a child with multiple injuries (such as the police, or social services) will almost certainly consider each injury separately, as will the courts. To anticipate this, it is advisable in a report to discuss the nature and possible causes of each injury, one by one. The evidence for abuse may be weak or non-existent for individual lesions, but the overall pattern may nevertheless point to non-accidental injury, maybe because of the extent or distribution of the injuries.

Another reason for considering each injury separately is that in some criminal cases one will find that for legal reasons information about certain injuries may be inadmissable during a court hearing. In this situation the ability to consider an injury independently of other injuries may be essential.

YOUR AREA OF EXPERTISE

It is a mistake to comment on an area that is outside one's expertise. An expert should have sufficient practical experience in the area.[11] This means, for example, that a histopathologist whose expertise is in conducting postmortem examinations and dealing with the examination of tissue samples is not in a position to comment on the symptoms, diagnosis, treatment, or prognosis of living children. Lawyers may have a poor understanding of the different areas of medical expertise, and a question not in one's domain should be firmly redirected. This can be difficult if the relevant expert (such as a paediatric haematologist) has not been instructed. The temptation to try to be helpful and ''have a stab'' at the question must be resisted.

QUOTING REFERENCES TO THE MEDICAL LITERATURE

It may be relevant to refer to the medical literature. For example, when asked to provide an estimate of the age of some bruising, it may be helpful to refer to the published literature on the subject, if only to show how little objective data is available. What is not acceptable is to selectively provide a few references that bolster the point of view that one wishes to advance, while ignoring all material that points in other directions. The volume of medical literature is such that one can usually find some articles to support almost any point of view, however outrageous. The court needs a balanced assessment of the possibilities.

It is important to bear in mind that a high proportion of the medical literature on child abuse is deeply flawed. Much of the published material is contained in case series, and these always tend to over-represent particularly severe cases. It is unsafe to generalise from this kind of uncontrolled and often highly selected data.

NEW MEDICAL THEORIES

The place for new medical theories is medical journals and scientific conferences. Everyone agrees that court is the very worst possible place in which to float new theories. Doctors who use a court case of suspected abuse as a testbed for a new unproven theory do the court a disservice and are inviting complaints from colleagues.

READILY ADMIT UNCERTAINTY

A number of publications on the subject of child abuse divide a case series (say of toddlers with fractured femur) into two categories, those who were abused and those who were not. Such publications are to be distrusted, as they fail to acknowledge the existence of a large category of ''uncertain'' cases in whom one never really knows whether or not the injury was the result of accident or abuse. The reality is that there are many cases in which doctors can do little more than voice suspicion or anxiety. When writing a report, one may feel (or actually be placed) under considerable pressure to express an undue degree of boldness and confidence, for fear that to do otherwise may weaken the case for the local authority or the prosecution. Regardless of pressure, never be afraid to say that one is simply not sure. ''Unexplained bruising'' or ''unexplained bruising ? accidental ?non-accidental'' are acceptable terms that plainly indicate a genuine diagnostic difficulty.

EVERYONE WILL READ YOUR REPORT

In clinical practice one may be accustomed (rightly or wrongly) to saying or writing things that are not shared with the parents. Bear in mind that whatever you write in a report for court will be disclosed to the parents, and in a criminal case to the person who is accused. In the case of care proceedings, this applies not only to one's report but to anything else that one writes or says to any of the parties. It is good practice to assume that one will be asked to justify anything that one has said or written. In a report one should make a point of providing the reasons for having reached particular conclusions.

POINT OUT ANY LIMITATIONS OF YOUR REPORT

Through no fault of one's own, there may be all sorts of limitations to a report. Certain papers may have been unavailable. Other reports may be awaited. One may have been unable to interview the parents. The photographs or radiographs supplied may have only been copies rather than originals. Be frank about any limitations.

DISAGREEMENT; MAINTAINING A PROFESSIONAL APPROACH

When dealing with a case of suspected abuse, one may be confronted by the report of a colleague with which one profoundly disagrees. Anger or bewilderment

are common reactions. Here is some advice:

- Legitimate disagreement can occur, a legacy of the lack of objective controlled studies of what happens when someone injures a child.
- However strongly one may feel about a case (and maybe *particularly* if one feels strongly about a case), do not reject the alternative viewpoint out of hand, but give it the most careful consideration, and maintain a balanced and professional approach. It does no harm to ask oneself whether it is just possible that there is at least some merit in the opinion that has been advanced.
- Never ever consider a child protection case in terms of winning or losing. That is for advocates, not doctors. The challenge for the doctor writing a report is not to help win a case, but to do a careful, thorough, and honest piece of work to the very best of one's ability.
- If, on reflection, one realises that one has arrived at an incorrect conclusion, it is a strength and not a weakness to readily acknowledge this. It is a common experience at experts' meetings (a topic that is discussed elsewhere[29]) that opinions change when one has a chance to better understand the reasoning of a colleague, or when one learns of new facts of which one was unaware. Experts who change their opinions for good reason on receipt of fresh information are respected by the court rather than criticised. However, if one changes one's opinion, one should always explain the reasons for the change.[13]
- All doctors make mistakes. The most serious error is to refuse to admit one has made a mistake, even when it is pointed out.
- Remember (and take comfort from the fact) that the ultimate responsibility for making decisions rests with the court.

CONCLUSIONS

The task of distinguishing between natural disease or accident on the one hand, and abuse on the other, is often difficult. However skilfully the situation is handled by the doctors involved, there will always be a risk that parents or carers will react badly on learning that a diagnosis of abuse is under consideration. However, reports prepared for court hearings will inevitably be exposed to the very closest scrutiny, and it is worth being extra careful and cautious when preparing such reports.

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REFERENCES

1 **Cazalet J** Re J (Child abuse: expert evidence). FCR 1991:193–227.
2 **Cazalet J** Re R (A Minor) (Experts' evidence). FLR 1991;**1**:291–3.
3 **Wall J** Re AB (Child abuse: expert witnesses). FLR 1995;**1**:181–200.
4 **Cazalet J** Re C (Expert evidence: disclosure: practice). FLR 1995;**1**:204–11.
5 **Bracewell J** Re S (Care proceedings: split hearing). FLR 1996;**2**:773–6.
6 **Stuart-Smith LJ**, Evans LJ, Thorpe LJ. Vernon v Bosley (Expert evidence). FLR, 1998;**1**:297–303.
7 **Wall J** Re CB and JB (Care proceedings: guidelines). FLR 1998;**2**:211–36.
8 **Singer J** Re X (Non-accidental injury: expert evidence). FLR 2001;**2**:90–117.
9 **Wall N**. Judicial attitudes to expert evidence in children's cases. *Arch Dis Child* 1997;**76**:485–9.
10 **Butler-Sloss E**, Hall A. Expert witnesses, courts and the law. *J R Soc Med* 2002;**95**:431–4.
11 **Williams C**. Expert evidence in cases of child abuse. *Arch Dis Child* 1993;**68**:712–14.
12 **Williams C**. A controversial expert witness. *Fam Law* 2000;**30**:175–80.
13 **Wall N**. *A handbook for expert witnesses in Children Act cases*. Bristol: Family Law, 2000.
14 **The Law Society**. *Good practice in child care cases. A guide for solicitors acting in Public Law Children Act Proceedings including cases involving adoption*. London: The Law Society, 2004.
15 **Anon**. Protocol for judicial case management in public law children act cases. FLR, 2003;**2**:719–812.
16 **Wall J** Re AB (Care proceedings: disclosure of medical evidence to police). FLR 2003;**1**:580–614.
17 **Laming H**. *The Victoria Climbé Inquiry. Report of an Inquiry by Lord Laming*. Norwich: The Stationery Office, 2003.
18 **Holman J** Re Y (Evidence of abuse: use of photographs) [2003] EWHC 3090 (Fam). FLR, 2004;**1**:855–70.
19 **Knight B**. *Forensic pathology*. London: Arnold, 1996.
20 **Langlois NEI**, Gresham GA. The ageing of bruises: a review and study of the colour changes with time. *Forensic Sci Int* 1991;**50**:227–38.
21 **Stephenson T**, Bialas Y. Estimation of the age of bruising. *Arch Dis Child* 1996;**74**:53–5.
22 **Munang LA**, Leonard PA, Mok JYQ. Lack of agreement on colour description between clinicians examining childhood bruising. *J Clin Forens Med* 2002;**9**:171–4.
23 **Bariciak ED**, Plint AC, Gaboury I, *et al*. Dating of bruises in children: an assessment of physician accuracy. *Pediatrics* 2003;**112**:804–7.
24 **Khair K**, Hann IM, Liesner R. The investigation of easy bruising. In: David TJ, eds. *Recent advances in paediatrics*. London: Royal Society of Medicine Press, 2003:81–97.
25 **Russell-Eggitt IM**, Thompson DA, Khair K, *et al*. Hermansky-Pudlak syndrome presenting with subdural haematoma and retinal haemorrhages in infancy. *J R Soc Med* 2001;**93**:591–2.
26 **Weber W**. Experimentelle untersuchungen zu Schadelbruchverletzungen des sauglings. *Z Rechtsmed* 1984;**92**:87–94.
27 **Weber W**. Zur Biomechanischen Fragilität des Sälingsschädels. *Z Rechtsmed* 1985;**94**:93–101.
28 **Weber W**. Prädilektionsstellen infantiler Kalottenfrakturen nach stumpfer Gewalt. *Z Rechtsmed* 1987;**98**:81–93.
29 **David TJ**, Hershman DA, McFarlane AE. Pretrial liaison between doctors in alleged child abuse. *Arch Dis Child* 1998;**79**:205–6.

Human rights

Discrimination against children

E Webb

Developing a conceptual framework

In the last few decades in Britain successive acts of parliament have attempted to tackle discrimination and promote equality of opportunities for women, members of ethnic minority communities, and disabled people. Currently there is discussion about eradicating "ageism", including within health services, although this term is used only to describe discrimination against older people.[1] One group—children—despite experiencing profound discrimination within society, are omitted from the general equality debate. Indeed many would think it ridiculous to include them. In fact, as will be illustrated in this paper, children experience significant discrimination, from both individuals and institutions. This discrimination affects both their health and the quality and delivery of child health services.

BACKGROUND

There was a growing and global commitment to the promotion of children's rights in the last half of the twentieth century, culminating in the UN

Convention on the Rights of the Child in 1989 and the establishment of Children's Rights Commissioners in several, mainly Western, countries. Among industrialised countries, only the USA did not ratify the Convention. Although the UK government did so (with some reservations) it has, relative to many of its European neighbours, been slow to respond to these developments, not adequately addressing the undoubted disadvantages children experience as a result of what is, in effect, a society inherently discriminatory against them.

At local and regional levels there has been more encouraging activity, with many local authorities, NHS Trusts, public health authorities, and schools making use of the Convention to inform strategies and services. Even so children, and children's services, remain marginalised. Even when there is a genuine commitment to children's rights, poor understanding of how discrimination against children—that is, "*childism*", is manifest, is compromising efforts to develop policies and services that are truly centred around the Convention.

This paper will explore what forms this discrimination can take, the mechanisms via which it affects children, and its impact on health and health care. A conceptual framework will be developed (fig 1), using current understanding of racial discrimination (for which the debate is much more advanced) to better explain the concepts developed.

The definition of "a child" is as in the UN Convention—that is, those aged less than 18 years.

THE NATURE OF DISCRIMINATION

Discrimination can be direct or indirect. Indirect discrimination is the inequitable treatment of one group disadvantaging another, as opposed to direct discrimination in which the focus of discriminatory attitudes, actions, and policies is the group itself. Discrimination can act at the level of the individual, but can also be institutional. Institutional discrimination occurs when the structures or operating policies of organisations result in certain sections of the community being disadvantaged. This concept is most familiar as institutional racism,[2] but can apply to any group disadvantaged by stigma and discrimination, including children.

Direct discrimination

This can be manifest in the following ways.

Overt discrimination

The late nineteenth and early twentieth centuries saw the beginnings of the exclusion of children from adult spaces, not for their safety or wellbeing but for the convenience of adults. This separateness of the child's world is now seen as natural. The resulting discrimination is so much a norm that it is both ubiquitous and unrecognised, with hotels in the UK routinely refusing access to children (and dogs).

Marginalisation

> "Through their constructed otherness, children's status in British society is as non-persons relegated to a social, economic and political marginalisation".[3]

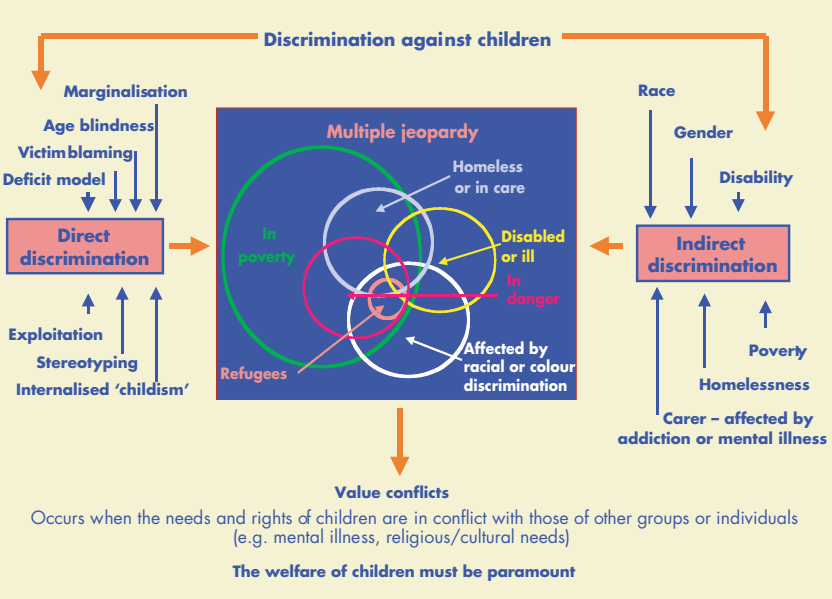

Figure 1 Discrimination against children.

Marginalisation is when a group experiencing discrimination is not seen as part of the core business or service. In the context of health care it is not only a modern phenomenon:

> "… when sick children are admitted promiscuously with adults, the former never have so much attention paid them as the latter".[4]

In 1994 both the confidential local audits performed by the Audit Commission (S Farnsworth and B Fitzsimon, personal communication, 1994) and other research[5] revealed that the needs of children were not prioritised in the commissioning process. Little has changed since.[6 7] When government first outlined plans for National Service Frameworks, they did not include the health of children. The implications of this marginalisation in policy, at local and national levels, for the health and welfare of children are fully explored by Aynsley-Green and colleagues.[8]

Children are similarly under-represented in funding for research and development, resulting in an inadequate evidence base for much paediatric practice,[9 10] particularly evident in the development of new therapeutic drugs. Over two thirds (67%) of 624 children admitted to wards in five European hospitals received drugs prescribed in an unlicensed or off label manner.[11] Although the problem is complicated by the ethical problems encountered around consent for child participation in trials,[12 13] it is largely profit driven. Drugs are not tested in children and thus not licensed for paediatric use. Even licensed drugs are prescribed off label "resulting in children becoming therapeutic orphans sometimes with tragic consequences".[12]

Age blindness

This is equivalent to colour blindness in racial parlance—treating everyone in the same manner, so ignoring or denying different needs. Such an approach can exclude children: for example, marina developments with inadequate barriers between toddlers and deep water.

Although huge strides have been made within the health sector to respond to the accommodation needs of children—providing facilities for play, and ensuring parents can accompany their children—there are still examples of poor practice, with shared waiting rooms in primary and secondary care in which carers have to spend considerable time with very young children in surroundings that are unsuitable and stressful.

The deficit model of childhood

Children are seen as immature—that is, incapable or unfinished; simply on the road to adulthood rather than people in their own right.

Children as incapable

We continue, for the most part, to exclude children from decision making or, at best, fail to take their input seriously. Although there is a commitment within the NHS to children's participation in decision making[14] and no shortage of guidance,[15–19] participation in health service development remains exceptional. For example, of 509 Trusts and health authorities in the UK, only 27 consulted children on services for chronically ill or disabled children, and only 11 of these went beyond consultation to meaningful participation in policy.[20] Young people with serious illness have reported feeling marginalised in decision making.[21] Although there are problems in achieving full and meaningful participation, not least the competing rights of children and parents,[21 22] children can be effective partners in the management of their own treatment,[23] and there is plenty of convincing evidence from the Child-to-Child programme showing how children can contribute effectively to health alliances and transform their lives and health.[24]

Children as ''immature''

Delinquency conceals two distinct categories, each with a unique natural history and aetiology. One group engages in antisocial behaviour at every life stage—''life course persistent'', whereas another is antisocial only during adolescence—''adolescence limited''. Adolescent onset delinquents do not share the pathological backgrounds found for those with life course persistent antisocial behaviours. Moffit and Caspi[25] suggest that this phenomenon is a consequence of what he terms ''a contemporary maturity gap'' in which, in modern post-industrial societies, essentially mature individuals are infantilised by extended education and delayed work opportunities, resulting in antisocial behaviours ''that are normative and adjustive''. This is an important issue. Around 25% of British men under 25 will have accrued criminal records to accompany them through their adult life, of which over half will have been adolescent onset delinquents.

Victim blaming

This term describes the phenomenon in which a vulnerable group are blamed when they experience disadvantage or harm.

Pedestrian injuries, a leading cause of childhood mortality, provide a good example. Children are blamed, with prevention strategies continuing to stress child behaviours, rather than addressing necessary and more effective changes in the structure of transport systems.

> ''The strength and pervasiveness of the ideology of victim blaming in child pedestrian injuries is explained by the special position that the road transport system holds in relation to dominant economic interests. Victim blaming ideology is a strategy that serves to maintain these interests at the expense and suffering of children''.[26]

Another example is the ''Lolita'' syndrome, in which children are blamed for their own sexual abuse. In 1993 a man found guilty of the rape of a girl, aged 9, was given two years' probation. The presiding judge said: ''I have been provided with information which leads me to believe that she was not entirely an angel herself''.[27] Although Lord Taylor stated on appeal that this comment should not have been made, it is a view met elsewhere. On Alice Liddell, the girl with whom both John Ruskin and Lewis Carroll were infatuated, Prose writes: ''what seems clear is that Alice was by no means a frail flower attracting these predatory bees; she pursued and actively encouraged their attentions''.[28]

Stereotyping

Children can be viewed as poor witnesses, more likely to lie than adults. This has had serious consequences for vulnerable children in care:

> ''The negative response (to complaints) especially in relation to reports of physical abuse, justified the pervading cynicism of most residents in care about the likely outcome of any complaints that they might make''.[29]

There is also a pervasive stereotype of children, particularly poor children, as inherently naughty, with distress frequently mistaken for ''badness''. Research evidence shows clear links between life course persistent delinquency and abuse, poor parenting, poverty, and socially disorganised communities.[30 31] Many of these young people also have neurodevelopmental problems, of which the most common is attention deficit hyperactivity disorder (ADHD).[29 30] Growing up with abuse and violence can also lead to anxiety and attachment disorder, both of which may lead to children fulfilling criteria for ADHD. Some children with primary hyperkinetic disorders will also be abused, or subject to poor parenting. There is thus a complex inter-relationship between abuse/poor parenting, poverty, delinquency and neurodevelopmental difficulties[32] which ought to, but does not, inform both preventive and responsive strategies to this problem. Instead governments of all persuasions in the UK have tended to focus largely on a punitive approach.

Internalised discrimination

Discrimination can be internalised. A member of a group experiencing discrimination adopts and shares the views of a hostile society, thus seeing him/herself as inferior. A powerful example of internalised racism is provided by Nelson Mandela in his autobiography.[33] He describes an incident during a period of exile in which he panics on noticing that the pilot of an aeroplane in which he is travelling is black—even Mandela had internalised the view that a black person could not be capable of such a task.

Children also take on society's view of themselves—as someone adults can pass in a queue unchallenged, as people having nothing to say worth hearing, as lawful victims of physical assault.

Exploitation

As with any powerless group, children are vulnerable to exploitation by the powerful—that is, adults. This may be private and secret, for example, the sexual exploitation of children within families. It may be commercially driven, for example, child labour (including sexual exploitation), advertising aimed at, or using, children; or politically driven—consider the exploitation of athletic prodigies in former Eastern Europe, given anabolic steroids in adolescence with serious consequences for their health.

Child labour is traditionally seen as a problem of low and middle income countries, but Field argues that we see emerging another equally exploitive form of labour—a tests and outcomes dominated education system, an ''insatiable schooling industry'' with education as ''endless labour''.[34] Although Field is writing about Japan, her work makes for uncomfortable reading.

Indirect discrimination

As children are dependent and powerless they are particularly vulnerable to indirect discrimination, in which their carers are disadvantaged as result of gender discrimination, racial discrimination, or the disadvantage many marginalised groups experience because they are poor, ill, disabled, or stigmatised for other reasons. Table 1 provides

examples of how indirect discrimination affects children.

Children may of course experience, for example, racism and sexism directly—a possible explanation for the high exclusion rate of African-Caribbean boys in UK schools,[40] and certainly the cause of the excess mortality of girls in India.[41] However, the focus of response strategies would still be to combat racism and gender discrimination, not childism per se.

For children, indirect discrimination always compounds direct discrimination, with some children experiencing multiple jeopardy. For example, a child may be disabled, belong to an ethnic minority community, be living in poverty, and have a parent with mental health problems. Such a child will be victim to layers of discrimination, all of which will affect his or her circumstances. The effects of such multiple disadvantage may not be simply additive, but act in synergy to paralyse services and leave children in danger.[42]

Racial discrimination as an example of indirect discrimination

Poverty

As a result of societal racism many black or ethnic minority (BEM) communities in the UK are at risk of poverty with some, notably the Pakistani and Bangladeshi communities, in ''serious poverty''.[43] Many adults in BEM communities are either unemployed or in low paid work, with their children more likely to attend poorly resourced inner city schools, be in the public care, and/or excluded from school. All these factors are linked with adverse health outcomes.

Access to information

Many parents in BEM populations, particularly mothers, do not have a working knowledge of English, and may not read. Without adequate provision of interpreters these parents are unable to access information crucial to their ability to make informed choices, to liaise with health, welfare, and education, and to advocate for their children when they are in need. Their situation is somewhat analogous to that of an illiterate mother in the developing world, a factor long known to be linked to high infant mortality.[44] It would seem reasonable to hypothesise that the language status of unsupported migrant parents is likely to impact on the health of their children.

Access to health services

BEM communities do not have equality of access to services.[45] There are examples of institutional racism. For example, services are dependent on postal addresses, which disadvantages asylum seekers and travellers. Services are often planned using whole population data. For BEM populations there is a mean age shift to the left. Nineteen per cent of white British people are aged under 16, but 38% of British Bangladeshis are under 16 (see table 2). Using whole population data to plan services ensures that areas with high BEM populations are undermanned and under-resourced for children's services, despite the increased needs of these communities as a result of poverty.

Services are also discriminatory in that they are culturally inappropriate, inaccessible, and with BEM clients stereotyped in ways that interfere with their care.[44]

Table 1 Indirect discrimination

Primary focus of discrimination	Mechanisms via which children are disadvantaged
Girls/women	Low pay; single mother households trapped in poverty Poor maternity provision: working mothers returning to work when babies are very young Reduced educational opportunities/expectations (in some communities)
Parents	No or little paternity leave[35] No parental leave for child illness (in the UK) Little acknowledgment of dual role of working parents in occupational law[36] Inadequate or poorly organised childcare services for very young children Inadequate provision for prams in public transport—difficulty in accessing appointments
Victims of domestic violence/homeless families	Stigmatised; poor access to services; low uptake of surveillance and immunisation[37 38]
Disabled	Access difficulties; marginalised in policy
Mentally ill carers	Stigmatised; unsupported—children acting as carers[39]

Table 2 UK % of ethnic groups aged under 16[46]

White	19
Mixed*	55
Indian	22
Pakistani	35
Bangladeshi	38
Other Asian	22
Black Caribbean	25
Black African	33
Other Black	35
Chinese	18
Other	20

*A UK census category designating someone with antecedents from two or more ethnic or racial groups.

VALUE CONFLICTS

These arise when the rights of one group are in conflict with those of another. The rights of children may conflict with those of other groups or individuals. This may be as a population; for example, the right of children to play in a safe place is often in conflict with the needs of car drivers. On an individual level there may often arise situations in which parental rights, religious rights, or cultural needs may appear to be in conflict with the wishes of children, or even their best interests. For example, a teenager may be in conflict with her parents with regard to whether or not to terminate her pregnancy. Such value conflicts are not easy to resolve, and may require the help of an independent advocate.[47] In the context of child abuse, value conflicts can be extremely difficult to resolve, with the safety of children at odds with, for example, the right of a severely mentally ill parent to care for her children, or the right of religious or ethnic minority groups to discipline children in ways they regard as appropriate to their culture.[36] Even within the Convention itself there may be conflicts; for example, the right of a child to be safe may conflict with the right to family life. It is imperative in such situations to focus firmly on Article 3 of the UN Convention of the Rights of the Child—what is in the best interest of the child?

CONCLUSIONS

In this paper a conceptual framework has been developed to describe the nature of discrimination against children. By applying this framework to the literature, and current political developments within the UK and beyond, examples of direct and indirect discrimination have been related to the health of children.

Children's advocates, and that includes paediatricians, need not only to identify *when* children are being disadvantaged but *why and how* they are disadvantaged. Without an understanding of how discrimination affects children it is not

possible to identify the appropriate focus for action in any particular case. Is a child being disadvantaged because she is a refugee, because she is disabled, or because all children's services are not prioritised in a Primary Care Trust? The conceptual framework developed here provides a tool to improve the recognition of discrimination against children, and help identify the precise mechanisms by which any child, or group of children, is disadvantaged. This will enable advocates to more effectively combat discrimination against children and successfully implement the UN Convention.

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REFERENCES

1 **Macdonald S**. Age discrimination difficult to identify, says King's Fund guide. *BMJ* 2003;**326**:355.
2 **Macpherson W**. The Stephen Lawrence Inquiry. Volume 1:46.25 1999. www.archive.official-documents.co.uk/document/cm42/4262/sli-46.htm (accessed 27 March 2003).
3 **James A**. *Childhood identities: self and social relationships in the experience of the child*. Edinburgh: Edinburgh University Press, 1993.
4 **Armstrong G**. *An account of the diseases most incident to children, from birth to the age of puberty, with a successful method of treating them*, 3rd edn. London: T Cadwell, 1783 (cited in Dunn PM. George Armstrong MD (1719–1789) and his dispensary for the infant poor. *Arch Dis Child Fetal Neonatal Ed* 2002;**87**:F228–31).
5 **Webb E**, Naish J, MacFarlane A. Planning and commissioning of health services for children and young people. *J Public Health Med* 1996;**18**:217–20.
6 **Viner RM**, Keane M. *Youth matters: evidence-based best practice for the care of young people in hospital*. London: Caring for Children in the Health Services, 1998.
7 **NCB**. *Improving children's health. A survey of 1999–2000 health improvement programmes*. London: NSPCC, Children's Society, National Children's Bureau, 2000.
8 **Aynsley-Green A**, Barker M, Burr S, *et al*. Who is speaking for children and adolescents and for their health at the policy level? *BMJ* 2000;**321**:229–32.
9 **Smyth RL**. Research with children. *BMJ* 2001;**322**:1377–8.
10 **AHCPR**. Policy on the inclusion of children in health services research. *NIH Guide* 1997;**26**(15).Agency for Health Care Policy and Research. http://www.ahcpr.gov/fund/nih5997.htm (accessed February 2003).
11 **Conroy S**, Choonara I, Impicciatore P, *et al*. Survey of unlicensed and off label drug use in paediatric wards in European countries. *BMJ* 2000;**320**:79–82.
12 **Stephenson T**. New medicines for children; who is protecting the rights of the child? *Curr Paediatr* 2002;**12**:331–5.
13 **Sutcliffe A**. Testing new pharmaceutical products in children. *BMJ* 2003;**326**:64–5.
14 **DoH**. Principle for participation of children, young people and families in the children's task force, 2002. http://www.doh.gov.uk/childrenstaskforce/participation.htm (accessed 20 October 2003).
15 **BMA**. *Consents rights and choices in healthcare for children and young people*. London: BMA, 2000.
16 **Brook G**. *Help me make choices too! Developing and using a framework to help children, with their families, to contribute to decisions about treatment*. Cascade; Issue 26. Action for Sick Children, 1997.
17 **Brook G**. Children's competence to consent; a framework for practice. *Paediatr Nurs* 2000;**12**:31–5.
18 **DFES**. *Core principles for involvement of children and young people—Children and Young People's Unit*, DFES, 2001.
19 **NSPCC**. Two-way street training video and handbook about communicating with disabled children and young people. London: NSPCC, 2001.
20 **Sloper P**, Lightfoot J. Involving disabled and chronically ill children and young people in health service development. *Child Care Health Dev* 2003;**29**:15–20.
21 **Young B**, Dixon-Woods M, Windridge KC, *et al*. Managing communication with young people who have a potentially life threatening chronic illness: qualitative study of patients and parents. *BMJ* 2003;**326**:305–10.
22 **Walker NE**, Doyon T. Fairness and reasonableness of the child's decision: a proposed standard for children's participation in medical decision making. *Behav Sci Law* 2001;**19**:611–36.
23 **Emilio J**, Sanz EJ. Concordance and children's use of medicines. *BMJ* 2003;**327**:858–60.
24 **Pridmore P**, Stephens D. *Children as partners for health: a critical review of the child-to-child approach*. London: Zed Books, 2000.
25 **Moffit TE**, Caspi A. Childhood predictors differentiate life-course persistent and adolescence-limited antisocial pathways among males and females. *Dev Psychopathol* 2001;**13**:355–75.
26 **Roberts I**, Coggan C. Blaming children for child pedestrian injuries. *Soc Sci Med* 1994;**38**:749–53.
27 Attorney-General's Reference No. 13 of 1993. *Karl Justin Gambrill* 1994;**15**:Cr. App. R. (S.) 292 CA (Crim Div).
28 **Prose F**. The lives of the muses: nine women and the artists they inspired. Aurum, cited in Hughes K. Pas de deux. *Guardian Review* 8 November 2003:15.
29 **Waterhouse**. *Lost in care—Report of the Tribunal of Inquiry into the Abuse of Children in Care in the Former County Council Areas of Gwynedd and Clwyd since 1974. Part VI: 29.30*. London. The Stationary Office, 2000 (also at http://www.doh.gov.uk/lostincare/20131.htm; accessed 27 March 2003).
30 **Moffit TE**, Harrington HL. Delinquency across development: the natural history of antisocial behaviour in the Dunedin multidisciplinary health and development study. In: Stanton W, Silva PA, eds. *The Dunedin study: from birth to adulthood*. Oxford: Oxford University Press, 1994.
31 **Farrington DP**. The development of offending and anti-social behaviour from childhood: key findings from the Cambridge study in delinquent development. *J Child Psychol Psychiatry* 1995;**36**:929–64.
32 **Webb E**. The role of the paediatrician in the management of delinquency. *Curr Pediatr* 2003;**13**:279–83.
33 **Mandela N**. *Long walk to freedom*. Abacus, 1995.
34 **Field N**. Education as endless labour. In: Montgomery H, Burr R, Woodhead M, eds. *Changing childhoods: local and global*. Haddington, UK: Open University/Wiley, 2003:35–7.
35 **MacDonald R**. Childcare for working parents (website of the week). *BMJ* 2003;**326**:170.
36 **Leach P**. *Children first*. London: Random House, 1995.
37 **Webb E**, Shankleman J, Evans MR, Brooks R. The health of children in refuges for women victims of domestic violence: cross sectional descriptive survey. *BMJ* 2001;**323**:210–13.
38 **Tischler V**, Vostanis P, Bellerby T, *et al*. Evaluation of a mental health outreach service for homeless families. *Arch Dis Child* 2002;**86**:158–63.
39 **Wilson J**. *The illustrated mum*. London: Corgi, 2000.
40 **ONS**. Permanent school exclusions by ethnic group: Social Focus in Brief: Ethnicity, 2001. http://www.statistics.gov.uk/STATBASE/ssdataset.asp?vlnk=6220&More=Y (accessed 7 November 2003).
41 **Khanna R**, Kumar A, Vaghela JF, *et al*. Community based retrospective study of sex in infant mortality in India. *BMJ* 2003;**327**:126–30.
42 **Webb E**, Maddocks A, Bongilli J. Effectively protecting Black and minority ethnic children from harm: overcoming barriers to the child protection process. *Child Abuse Review* 2002;**11**:394–410.
43 **Modood T**, Berthoud R. *Ethnic minorities in Britain: diversity and disadvantage*. London: Policy Studies Institute, 1997.
44 **Cleland JG**, Van Ginneken JK. Maternal education and child survival in developing countries: the search for pathways of influence. *Soc Sci Med* 1998;**27**:1357–68.
45 **Webb E**. Health care for ethnic minorities. *Curr Pediatr* 2000;**10**:184–90.
46 **ONS**. Census data. Ethnicity: age distribution—ethnic groups have younger age structure. 2001. http://www.statistics.gov.uk/cci/nugget.asp?id=272 (accessed 20 October 2003).
47 **Webb E**. Health services: who are the best advocates for children? *Arch Dis Child* 2002;**87**:175–7.

New guidelines

Hereditary spherocytosis; new guidelines

P H B Bolton-Maggs

Hereditary spherocytosis

Hereditary spherocytosis (HS) is the commonest cause of haemolysis in northern Europe. Most children have mild disease with little interference with lifestyle. Presentation with parvovirus B19 infection causing transient severe anaemia is not uncommon. The laboratory diagnosis of HS is usually straightforward and additional tests are rarely required. A new test, EMA binding, will probably replace the time consuming and non-specific osmotic fragility test. Splenectomy leads to improved red cell survival and is indicated for severe and probably moderate disease; gallstones may occur in the first decade, and if symptomatic are an indication for both cholecystectomy and splenectomy. Splenectomy is associated with a life long increased risk of sepsis, which has not been completely eradicated by preoperative vaccinations and post-splenectomy antibiotic prophylaxis.

Haemolysis is an uncommon cause of anaemia in children. The commonest cause of anaemia is iron deficiency, globally a major problem; worldwide, haemolysis is most commonly associated with a red cell enzyme deficiency, glucose-6-dehydrogenase deficiency (G6PD). Haemolysis produced by G6PD deficiency is usually episodic and self-limiting; in contrast the haemolysis associated with hereditary spherocytosis, a red cell membrane disorder, is chronic, and prone to exacerbations with intercurrent infections. Hereditary spherocytosis (HS) is the commonest cause of inherited haemolysis in northern Europe and the USA; the incidence is in the order of 1 in 5000 births, but rises to 1 in 2000 if milder forms are considered.[1] It has been reported in most ethnic groups, and can come to light at any age, mild cases often being diagnosed in adulthood. Although HS is relatively common, there has been little published advice concerning management; new guidelines have been produced[2] and this article summarises some of the relevant issues.

PATHOLOGY

The defects in hereditary spherocytosis lie in the red cell membrane (fig 1).[2] The proteins essential for the integrity of the membrane structure lie immediately under the lipid bilayer; horizontal alpha and beta spectrin molecules form heterodimers with linkage to vertical elements—ankyrin, proteins 4.1 and 4.2, and band 3 (which is a transmembrane protein). Different genes code each of these proteins, thus hereditary spherocytosis is a heterogeneous disorder, which can result from a defect in any one of these proteins. The resultant destabilisation of the membrane leads to both abnormal morphology and a reduced red cell life span (from the normal 120 to a few days). The shorter the red cell life span, the worse the clinical effects. The defect, and therefore the clinical severity, tends to be fairly constant within a given family, but between families varies from mild asymptomatic haemolysis to severe continuous anaemia with jaundice. Inheritance is usually dominant (75%).

CLINICAL FEATURES

The classical clinical features of haemolysis (whatever the cause) are anaemia, jaundice, and splenomegaly. However, anaemia is often mild or absent when the haemolysis is well compensated (about a third of cases), as the bone marrow is able to increase red cell output some 6 to 8-fold; the only finding in some affected individuals is a raised reticulocyte count reflecting the increased marrow turnover. Jaundice is variable, often absent, and likely to increase when the marrow is stressed by intercurrent infection. Splenomegaly is usual, being generally mild, and a finding of massive splenomegaly should prompt a search for an alternative diagnosis. The enlarged spleen is not more prone to rupture than in a normal person, and the size is neither an indication for splenectomy nor for any restriction of activity.

Hereditary spherocytosis can present at any age from the neonatal period to the ninth decade, depending on severity. In the neonatal period jaundice is common, occasionally requiring exchange transfusion. It is important that affected families are aware of this. The severity of the jaundice (which may develop over several days) does not necessarily predict a severe subsequent course, and is perhaps not dependent on the severity of the membrane defect alone.

Children with hereditary spherocytosis may be usefully classified by clinical features as ''severe'', ''moderate'', or ''mild'' using the criteria shown in table 1.[3] This assessment should be made when the child is in a stable baseline state, as with intercurrent illness the severity may be overestimated. Children with severe hereditary spherocytosis (rare, about 5%) are constantly anaemic, and may be transfusion dependent, especially in the first few years of life. Treatment with erythropoietin may be beneficial and reduce transfusion requirements in the first year of life.[4 5] After this time, regular transfusion is rarely required; most children can tolerate a low haemoglobin level and this alone should not be a transfusion trigger. Severe HS is the exception rather than the rule; most children with HS have a normal or only slightly reduced haemoglobin and lead a

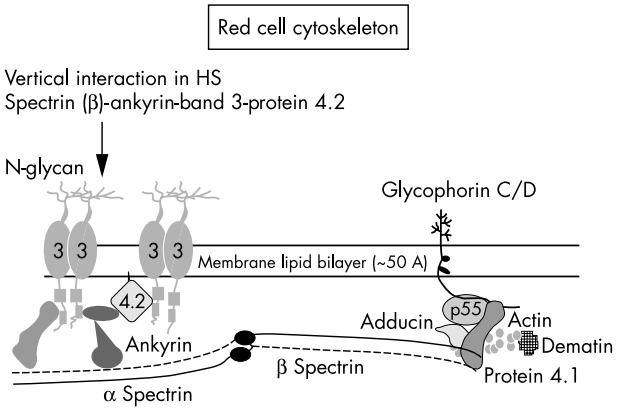

Figure 1 Schematic presentation of the structural organisation of red cell cytoskeleton. β Spectrin is the key component in that it pairs with α spectrin to form a heterodimer, and it has binding sites for ankyrin and protein 4.1. The common protein defects are associated with spectrin (α and/or β), ankyrin, band 3 protein, and protein 4.1.

normal life. Haemolysis is associated with increased red cell turnover and an increased pigment load for the liver – this may result in gallstones developing as early as the first or second decade of life, and the risk is increased with co-inheritance of Gilbert syndrome.[6] Generally, the higher the reticulocyte count, the higher the risk of developing gallstones. This is therefore one of the parameters which may guide a decision for splenectomy.

People with HS (both children and adults) may remain undiagnosed for years or decades if haemolysis is mild. In both childhood and adulthood, parvovirus infection is an important initial presentation. Parvovirus B19 leads to red cell aplasia for a few days. In people with a normal red cell life span this is barely noticeable, but in people with HS or any other form of haemolytic anaemia, parvovirus infection leads to a sudden profound fall in haemoglobin level to as low as 20 or 30 g/l.[7] Such individuals often feel unwell, and may have an associated mild leucopenia and thrombocytopenia leading to fear of a more sinister diagnosis. The diagnosis may be suspected from the blood film (spherocytes) and the clinical finding of splenomegaly. The reticulocyte count is characteristically low at the onset, but will increase rapidly in the recovery period. Once recovered from the infection (within a week or two), parvovirus B19 infection does not recur; it is then important to establish the usual baseline haemoglobin and reticulocyte count as often people who present in this way have otherwise mild disease. An increase in haemolysis (and therefore a drop in haemoglobin) may be produced by stress or other infections, but such severe anaemia due to aplasia is unlikely to recur; no other infection produces such a profound fall in the haemoglobin. Helpful pointers to the diagnosis of HS include a family history of others with similar ''aplastic'' crises, jaundice, or splenectomy. It is surprising that people who have had a splenectomy in the past may not know that the reason was an abnormality in the red blood cells. People with known HS who have not had parvovirus infection, and their family doctors, should be warned of possible future infection, as the degree of anaemia and symptoms can be alarming. Transfusion support may be required (usually a single transfusion episode). It is helpful to confirm the diagnosis by showing an increasing IgM parvovirus antibody titre or demonstration of parvovirus DNA in the blood. At present there is no way of preventing infection, but a novel recombinant parvovirus vaccine has successfully shown seroconversion in adults in a phase 1 trial.[8] Parvovirus infection is readily spread so that several members of a family may be infected at the same time.[9]

People with mild HS may remain undiagnosed for decades and only be discovered when spherocytes are seen when a blood count is done for another indication, or the person is found incidentally to have an enlarged spleen. Some individuals will be diagnosed in pregnancy as a result of routine blood counts.

LABORATORY DIAGNOSIS

The key features are spherocytes on the blood film and a raised reticulocyte count with or without anaemia. The bilirubin level (unconjugated) is often raised. The red cells may show a reduced red cell volume (MCV) and increased red cell haemoglobin concentration (MCHC). The differential diagnosis is from autoimmune haemolytic anaemia (AIHA); a direct antiglobulin test (for the detection of antibodies on the red cells) will be negative in HS and usually positive in AIHA. The clinical context is important. People with HS are usually clinically well and often have a positive family history; AIHA is rare in children and most commonly associated with an acute viral infection. With the classical clinical picture and these simple laboratory tests, there is usually no need for further investigation. The osmotic fragility test (OF) is time consuming, labour intensive, and adds nothing to the diagnosis if there are obvious spherocytes on the film. The OF does not distinguish between the causes of spherocytosis and so will be positive in AIHA. It can also be falsely negative in the presence of iron deficiency and obstructive jaundice, and is difficult to interpret in neonates who have a different normal range. (Normal neonates may have spherocytes and it may not be possible to confirm a diagnosis of HS for some months.) Confirmatory tests for HS are rarely required. A particularly promising test, now being introduced in many laboratories, is the eosin-5-maleimide (EMA) binding test performed by flow cytometry.[11 12] EMA binds to band 3, a skeletal protein, and the test has a high sensitivity (92.7%) and specificity (99.1%) for HS. The test can be performed rapidly (within two hours) on a small sample of blood.

Although it may be possible to identify the genetic basis of the HS in any given family (that is, to see which protein is defective), this is usually unnecessary for clinical management. Genetic analysis is a research investigation, and is only helpful in very unusual cases. The majority of individuals with HS are easily diagnosed with routine methods, providing the laboratory staff are given the appropriate clinical information. Doubt may occasionally arise in the context of an atypical appearance of the blood film; there are some important and rare disorders of red cells that can resemble HS. Atypical cases must therefore be carefully reviewed with the haematologist. In these cases, further investigation may be necessary.[2]

CLINICAL MANAGEMENT

Once the diagnosis is made, it is important to reassure parents and children that HS is not generally a serious disorder, and no restrictions are required on activity or lifestyle. If a child has been diagnosed as a result of parvovirus infection, both parents and medical staff may have a false impression of the severity, so it is important to review the blood count some months later to obtain a more accurate reflection of the normal status. Family studies will be appropriate. There may be adults in the family who have had splenectomy in the past for HS. This raises other issues; in the past splenectomy was performed more readily than it is now (see below) and adults may not be aware of their long term infection risks. Splenectomy in other family members does not necessarily predict for the newly diagnosed child because criteria for surgery are now stricter.

Children with severe HS may have significant anaemia (for example, Hb 60 g/l) but are usually remarkably well and active. Transfusion is rarely required and should not be based on the haemoglobin level alone. In general, once the diagnosis and baseline severity of HS in a child are established, it is not necessary to perform repeated blood tests unless there is an additional clinical indication (such as intercurrent infection and pallor, or an increase in

Table 1 Classification of hereditary spherocytosis (modified from Eber *et al*[3])

Classification	Mild	Moderate	Severe
Haemoglobin (g/l)	110–150	80–120	60–80
Reticulocyte count (%)	3–6	>6	>10
Bilirubin (μg/l)	17–34	>34	>51
Splenectomy	Usually not required	Indicated during school age, usually before puberty	Necessary—delay until 6 years if possible

jaundice). A routine annual review is usually sufficient together with an open door policy for potential complications such as parvovirus infection, or abdominal pain, which may trigger investigation for gallstones.

It is traditional for children with chronic haemolysis to receive oral folate supplements, but most children in developed countries consume well above the minimum daily requirement. There is no strong evidence to support universal folate supplementation in HS, and it is likely that it is only required in children with severe and moderate HS, but for all patients during pregnancy. There is no standardised regimen; a reasonable daily dose is 2.5 mg up to the age of 5 years and 5 mg daily thereafter.

SPLENECTOMY

The most important question in the management of HS is to decide whether splenectomy is likely to be of benefit, and when it should be carried out. Once it was recognised that splenectomy abrogated the clinical symptoms of HS, it was universally applied; however, this was followed by the recognition of the risk of severe overwhelming post-splenectomy sepsis, often fatal,[13 14] usually caused by pneumococcal species. The risk is age related, being highest in the youngest children and within the first few years after surgery. Current UK guidelines recommend pre-splenectomy vaccination against pneumococcus, haemophilus, and meningococcus, together with long term (lifelong) postoperative penicillin prophylaxis.[15 16] These measures do not completely eliminate the risk.[17 18] There remain many unanswered questions: How long does pneumococcal immunity last? Which is the best vaccine? How long should penicillin prophylaxis be continued? The increased susceptibility to serious pneumococcal sepsis is lifelong,[19] but there are no trials showing benefit of long term antibiotics, and other guidelines are more cautious, recommending penicillin prophylaxis for at least two years in adults, and at least five years in children,[20] rather than lifelong. This is partly an acknowledgement of the increase in penicillin resistant pneumococci.[21] The decision for splenectomy needs to be carefully made, weighing up advantages against the small but real risks, which must be carefully discussed with the child and family. Chronic anaemia is debilitating; although the young child tolerates significant chronic anaemia well in terms of activity, rarely it can lead to increased cardiac output with cardiomegaly, and leg ulcers. Children with chronic anaemia may show a fall-off in growth rate and weight gain as they approach puberty. Chronic severe haemolysis is associated with a significant risk of gallstones, which may become symptomatic as early as the first or second decade of life. Since the spleen is the site of destruction of the abnormal red cells, splenectomy produces a significant increase in the red cell life span to normal or near normal in most cases. The red cell morphology does not improve, but red cell destruction is reduced, leading to improvement in the haemoglobin level, and reduction of the reticulocyte count. The increased risk of gallstone development is also corrected.[22] There are therefore significant advantages in proceeding to splenectomy in children with severe HS, and probably most with moderate HS. Splenectomy should be avoided if possible in the young child, and if possible, postponed until the child is at least 6 years of age. Some centres advocate partial splenectomy for the most severe transfusion dependent children; this ameliorates the haemolysis, but experience with this technique is limited to a few centres, and many of these children subsequently require repeat surgery to complete splenectomy.[23 24]

It is often convenient to perform splenectomy before the child moves into secondary education; the decision for elective surgery can be made over a period of time and it may be helpful for children and parents to talk to other families who have experience. Parents find it particularly difficult to decide for elective surgery when a child seems very well, particularly in the face of some risks. However, the child or young adult often reports a considerable improvement in wellbeing after surgery. Traditionally, surgery has been open, laparotomy, but there is increasing experience in many centres with laparoscopic surgery, which has the advantage of more rapid recovery, a shorter hospital stay, and a better cosmetic result. The decision must rest with the surgeon, and is dependent on experience and availability of suitable equipment.

When a child has had symptoms of gall bladder disease, most surgeons would remove the gall bladder at the time of splenectomy; conversely, if a child requires surgery for gall bladder disease complicating HS, the spleen should be removed at the same time.[2]

It is reasonable to perform ultrasound examination of the gall bladder towards the end of the first decade, as the finding of gall stones, even without symptoms at this age may influence the decision for splenectomy. A recent review describes 44 patients aged between 1 and 22 years who underwent annual ultrasound examination from the age of 4 years or at the time of diagnosis.[25] Overall 18 (41%) developed gallstones, with a higher incidence in those with moderate and severe HS; however, stones developed in 4/14 (29%) with mild HS at a median age of 13 years, and overall 94% of stones developed by this age. Five patients had symptoms suggestive of cholelithiasis prior to detection by ultrasound.

It should be noted that it is usual for the platelet count to rise post-splenectomy, sometimes to levels higher than 1000×10^9/l, but there is no evidence that this on its own is a risk factor for thrombosis in people with HS in the short or long term, and in this context, high platelet counts do not need any treatment. People with some forms of hereditary stomatocytosis have an increased risk of thrombosis after splenectomy;[26] this emphasises the importance of careful evaluation of cases with atypical red cell morphology by a haematologist.

CONCLUSION

HS is the commonest form of haemolytic anaemia seen in northern Europe. Most children have mild disease, can live a normal life, and do not require splenectomy. Parvovirus B19 infection can cause an acute aplastic phase, but does not recur. Splenectomy is reserved for those with severe disease or who develop symptomatic gallstones, when cholecystectomy should be performed at the same time. The lifelong risk of post-splenectomy sepsis must be discussed fully with the family, and adequate prophylaxis undertaken.

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REFERENCES

1 **Eber SW**, Pekrun A, Neufeldt A, *et al*. Prevalence of increased osmotic fragility of erythrocytes in German blood donors: screening using a modified glycerol lysis test. *Ann Hematol* 1992;**64**:88–92.
2 **Bolton-Maggs PHB**, Stevens RF, Dodd NJ, *et al*. Guidelines for the diagnosis and management of hereditary spherocytosis. *Br J Haematology*. In press. Also accessible at www.bcshguidelines.com.
3 **Eber SW**, Armbrust R, Schroter W. Variable clinical severity of hereditary spherocytosis: relation to erythrocytic spectrin concentration, osmotic fragility and autohemolysis. *J Pediatr* 1990;**177**:409–11.
4 **Delhommeau F**, Cynober T, Schischmanoff PO, *et al*. Natural history of hereditary spherocytosis during the first year of life. *Blood* 2000;**95**:393–7.
5 **Tchernia G**, Delhommeau F, Perrotta S, *et al*. Recombinant erythropoietin therapy as an alternative to blood transfusions in infants with hereditary spherocytosis. *Hematol J* 2000;**1**:146–52.

6 **del Giudice EM**, Perrotta S, Nobili B, *et al*. Coinheritance of Gilbert syndrome increases the risk for developing gallstones in patients with hereditary spherocytosis. *Blood* 1999;**94**:2259–62.
7 **Brown KE**. Haematological consequences of parvovirus B19 infection. *Baillieres Best Pract Res Clin Haematol* 2000;**13**:245–59.
8 **Ballou WR**, Reed JL, Noble W, *et al*. Safety and immunogenicity of a recombinant parvovirus B19 vaccine formulated with MF59C.1. *J Infect Dis* 2003;**187**:675–8.
9 **Murphy PT**, O'Donnell JR. B19 parvovirus infection causing aplastic crisis in 3 out of 5 family members with hereditary spherocytosis. *Ir J Med Sci* 1990;**159**(6):182–4.
10 **Korones D**, Pearson HA. Normal erythrocyte osmotic fragility in hereditary spherocytosis. *J Pediatr* 1989;**114**:264–6.
11 **King MJ**, Behrens J, Rogers C, *et al*. Rapid flow cytometric test for the diagnosis of membrane cytoskeleton-associated haemolytic anaemia. *Br J Haematol* 2000;**111**:924–33.
12 **King M-J**, Smythe J, Mushens R. Eosin-5-maleimide binding to band 3 and Rh-related proteins forms the basis of a screening test for hereditary spherocytosis. *Br J Haematol* 2004;**124**:106–13.
13 **Eraklis AJ**, Filler RM. Splenectomy in childhood: a review of 1413 cases. *J Pediatr Surg* 1972;**7**:382–8.
14 **Walker W**. Splenectomy in childhood; a review in England and Wales. *Br J Surg* 1976;**63**:36–43.
15 **BCSH**. Guidelines for the prevention and treatment of infection in patients with an absent or dysfunctional spleen: Working party of the British Committee for Standards in Haematology Clinical Haematology Task force. *BMJ* 1996;**312**:430–4.
16 **Davies KA**, Beynon HL, Walport MJ. Long-term management after splenectomy. Consider prophylaxis in systematic lupus erythematosus. *BMJ* 1994;**308**:133.
17 **Eber SW**, Langendorfer CM, Ditzig M, *et al*. Frequency of very late fatal sepsis after splenectomy for hereditary spherocytosis: impact of insufficient antibody response to pneumococcal infection. *Ann Hematol* 1999;**78**:524–8.
18 **Spickett GP**, Bullimore J, Wallis J, *et al*. Northern Region asplenia register—analysis of first two years. *J Clin Pathol* 1999;**52**:424–9.
19 **Evans DI**. Postsplenectomy sepsis 10 years or more after operation. *J Clin Pathol* 1985;**38**:309–11.
20 **Beytout J**, Tournilhac O, Laurichesse H. [Antibiotic prophylaxis in splenectomized adults]. *Presse Med* 2003;**32**(28 suppl):S17–19.
21 **Gallagher PG**, Lux SE. Disorders of the erythrocyte membrane. In: Nathan DG, Orkin SH, Ginsburg D, Look AT, eds. *Nathan and Oski's hematology of infancy and childhood*, 6th edn. Philadelphia: Saunders, 2003:561–684.
22 **Sandler A**, Winkel G, Kimura K, *et al*. The role of prophylactic cholecystectomy during splenectomy in children with hereditary spherocytosis. *J Pediatr Surg* 1999;**34**:1077–8.
23 **Rice HE**, Oldham KT, Hillery CA, *et al*. Clinical and hematologic benefits of partial splenectomy for congenital hemolytic anemias in children. *Ann Surg* 2003;**237**:281–8.
24 **Bader-Meunier B**, Gauthier F, Archambaud F, *et al*. Long-term evaluation of the beneficial effect of subtotal splenectomy for management of hereditary spherocytosis. *Blood* 2001;**97**:399–403.
25 **Tamary H**, Aviner S, Freud E, *et al*. High incidence of early cholelithiasis detected by ultrasonography in children and young adults with hereditary spherocytosis. *J Pediatr Hematol Oncol* 2003;**25**:952–4.
26 **Delaunay J**, Stewart G, Iolascon A. Hereditary dehydrated and overhydrated stomatocytosis: recent advances. *Curr Opin Hematol* 1999;**6**:110–14.

IMAGES IN PAEDIATRICS

BCG lymphadenitis

A 3 month old, otherwise well baby of Chinese origin, who had received BCG vaccine in the left deltoid region on the second day of life, presented with a one month history of a lump in the left axilla (see fig). Examination revealed a fluctuant mass (5×8 cm) at the left anterior axillary fold. Ultrasonography confirmed enlarged but non-suppurative lymph nodes. A presumptive diagnosis of BCG lymphadenitis was made and treatment was started with isoniazid. A month later the swelling increased in size and became more fluctuant. Surgical incision and drainage was performed, which revealed caseous material characteristic of tuberculous infection.

Isoniazid was continued for a total of three months and the lesion healed uneventfully.

On a prolonged culture, Bacille Calmette-Guerin was grown that was resistant to isoniazid but sensitive to ciprofloxacin, ethambutol, and rifampicin.

Although BCG lymphadenitis is a well recognised condition, it is not commonly seen in UK district general hospitals. Treatment of BCG lymphadenitis remains controversial. In one series,[1] surgery was proposed as the most effective treatment in advanced cases with no reported recurrence or fistula formation. Goraya and Virdi,[2] however, suggested that no treatment was required in non-suppurative lymphadenitis. In the case of suppuration, surgical treatment either by needle aspiration or surgical excision could be undertaken. In our case, treatment with isoniazid was probably non-contributory.

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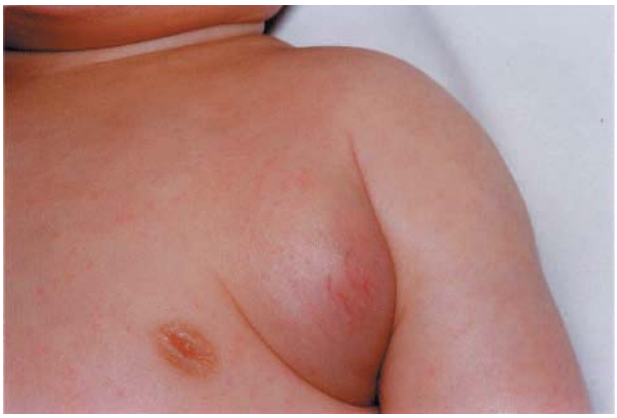

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References

1 **Garcia Merida M**, Miguelez Lago C, *et al*. Regional adenitis following BCG vaccination. *Anales Espanoles de Pediatria* 1979;**12**:597–602.
2 **Goraya JS**, Virdi VS. Bacille Calmette-Guerin lymphadenitis. *Postgrad Med J* 2002;**78**:327–9.

Dr Mohammed Al Moudaris died after a short illness on 15th April 2004. For 13 years he had been a general paediatrician with a special interest in allergy at the Hospital of St. Cross, Rugby and Walsgrave Hospital, Coventry. He was a very popular member of staff and will be sadly missed by patients and colleagues. He leaves a wife, Nudhar Hadid (a consultant radiologist with the same Trust), a daughter, May (an architect), and a son, Al, who graduated as a doctor this summer.