

Introduction

For the main part of our response we have followed the format of the Consultation Paper, giving input under headings of each workstream as appropriate. Additional sections give recommendations on areas that have not been covered by the Consultation Paper, and that SADS UK feel are crucial.

The specific views requested in the Introduction to the Consultation Paper (no.5) have been addressed in our submission.

The recommendations of SADS UK are based on personal experience and those of our members, together with information and knowledge gained through the Trust. Personal stories highlight recommendations made, together with medical studies and papers, which further underpin our proposals. For the purpose of our response only a few personal stories have been included, it should be stressed that these are not isolated cases, as is shown in the SADS UK bi-monthly newsletter.

Our document includes an Overview Section, which broadly outlines an ideal procedural plan, in primary and secondary care, for the ongoing detection and monitoring of the conditions under consideration. Contained in this section is our recommendation for a basic screening plan, which we believe should be the absolute minimum & could be readily applied.

In recent years links have been made between one of the conditions that may cause a sudden arrhythmic death, the Long QT Syndrome (LQTS) and SIDS (see Appendix 'I'). In our response we have considered this and made references where appropriate although we understand that there is currently no conclusive evidence.

SADS UK believe that as it becomes available, refined technology and genetic testing for the purpose of accurate and efficient diagnosis of cardiac conditions should be assessed for the purpose of population screening to help save lives.

Definitions

At risk group

A person can be in an at risk group if they experience signs and symptoms consistent with a cardiac condition, a family member suffers with heart rhythm abnormalities, has been a victim of sudden death, or there has been an incidence of SIDS.

Family Member

To include all relatives.

Monitoring

Investigation of heart employing diagnostic equipment as necessary but as a minimum the following; ECG, ECHO, 24-hour monitor and stress test.

Screening

Population - or selected groups of the population – monitoring.

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We have used various terminologies to describe the conditions that will be covered by the new chapter. These include as follows:

- Underlying heart condition
- Abnormal heart rhythms
- Cardiac arrhythmia
- Sudden cardiac death
- Sudden death
- Sudden arrhythmic death
- Heart condition/abnormality/defect

Sudden Cardiac Death & Screening

Raising awareness of signs and symptoms of conditions which may lead to sudden cardiac death

There are many opportunities to raise awareness to the **general public**, and these have been successfully used for other health campaigns.

For the purposes of any leaflet or media campaign it is important to establish for inclusion the signs and symptoms of conditions that may lead to sudden death eg fainting, palpitations, chest pains, etc. Mention should also be made of the drugs that can exacerbate conditions that cause arrhythmia and even the fact that arrhythmia can be induced by certain drugs. (see Drugs Issues pg 21)

- **Leaflets & Literature**

Dept of Health sponsored leaflets should be placed in GPs surgeries, hospital depts and other medical establishments, training environments, schools and youth clubs, swimming pools¹ sports and leisure facilities, corporate medical rooms, etc

Distribution of leaflets by GPs would be preferable at various stages of development in children, eg toddler check-ups. This is currently standard practice for the distribution of leaflets warning of other typical childhood diseases eg Meningitis.

Distribution of leaflets in secondary schools, youth, swimming and sports clubs could be considered. At this age young people are warned in various ways of the dangers of illegal substances and smoking, as well as the importance of safe sex.

Distribution of leaflets in corporate medical rooms and cafeterias where leaflets are currently displayed advising people on smoking issues and healthy eating.

- **Media & Advertising**

There are currently nationwide campaigns on radio and TV, in newspapers and on billboards, advising the general population on healthy eating, exercise and the dangers of smoking. These are all issues which, if not addressed, prove costly to the health service because of extensive health care requirements.

While these campaigns are undoubtedly expensive, an initial media campaign to kick-start the promotion of awareness of arrhythmias would be prudent.

Among **health professionals** the question of how to raise awareness, and in some cases introduce them to conditions and subsequent implications they know little about, may have to be met in many instances with some form of additional training or education. There are many instances where GPs may have not given enough consideration to signs and symptoms, or where hospital consultants may have not given enough urgency to making a diagnosis (although it is accepted that available resources may currently restrict the speed with which tests can be accomplished), sometimes with disastrous consequences. (see Personal Stories/Colin Hankin).

The introduction of new procedures or practice recommendations will automatically bring awareness to the health professionals, however the following could be considered:

- Use of leaflets, as described above, and those supplied by relevant charitable organisations to support guidelines.
(Warwick University use SADS UK leaflets to inform trainees)
- Compulsory post-graduate education eg seminars on cardiac arrhythmias and their causes, to be achieved within a specified timescale, no longer than six months.
- An information pack distributed to all health professionals alerting them to the publication of the new chapter. A summary of the guidelines would be appropriate.
- Training regarding the possibility of drug related arrhythmia, including how frequently medications should be assessed, together with how to use on-line agencies to assist them in this process. With the instigation of so many new drugs, agencies such as the Drug Safety Research Unit Southampton and the Torsades de Pointes website, University of Arizona would be helpful resources as they collate important information about medications. These on-line facilities should be accessed on a frequent basis and specialist advice should be considered where appropriate (see Drug Issues pg 21)

Obviously, review of education of this subject at training level would be expected.

An overview of how the detection and management of all cardiac conditions could be incorporated into Primary Care and Secondary Care is attached, and this includes ways of raising awareness.

Support for those who have lost relatives or friends

Due to the current lack of awareness and understanding of sudden cardiac death, the bereaved need sources of information about these conditions as well as compassionate support. Some people find it psychologically impossible to come to terms with a death if they have no reason for that death. This lack of information can be detrimental to their mental and physical health.

For compassionate support however the Dept of Health should make available to relatives lists of charitable organisations, support groups and counsellors. Immediate access to NHS bereavement counsellors (see * below) should be a priority as currently relatives of sudden death victims are usually given no precedence and must join what can be a three to six month waiting list. For parents suddenly losing an otherwise energetic and healthy child, this resource should be more readily available.

Ideally, the health professionals involved at the time of the death and those subsequent to it eg Coroner's Officer, would have improved training, including bereavement training, to deal with these cases and improved education of the conditions being dealt with. Currently, there is very little face-to-face contact with health professionals after a sudden death. What relatives really need is support from the people whom it is perceived should know what has happened to their loved one. Avoiding discussing the death with relatives is not an option.

The following are recommendations for a support and inform procedure:

(* Throughout this procedure the term Coroner's Officer is used. It may be that another better placed and qualified individual can be employed eg local NHS counsellor)

- Immediately, or as soon as possible before post-mortem, Coroner's Officer should visit relatives and advise them of the process to take place and why. It is important to be sensitive and assure the relatives that establishing a cause of death could help them in the long term to come to terms with the death. This would obviously require particular training. Leaflets could be offered that have information on sudden death and support groups, even if already done so by for example the hospital. Leaflets should also be offered at this time supporting the verbal information provided on post-mortem procedure. The bereaved are rarely able to ask pertinent questions and must be given every opportunity to review information received.
- During this consultation, or as soon as possible before post-mortem but in person, the CO should offer to take information that may help to establish a reason for the sudden death eg recent food intake, any prescribed medications or over-the-counter drugs taken, recent activity. A questionnaire could be used to ensure consistency of information gained throughout the country, but obviously this would not be delivered as such to the family. It cannot be emphasised enough how sensitively this should be handled, but the data could provide important indicators at post-mortem and could go on to be used in a pathology database for research purposes, providing important data relevant to public health safety issues. (see Pathology Issues pg 20)
- Subsequent to post-mortem relatives should be visited again by the CO to have the results explained. Opportunity should be given to allow the relatives to ask questions, both during the consultation and after.

- During PM there may be reason to make additional investigations that require the permission of the relatives eg organ retention. As before, a face-to-face consultation should take place in order to explain the procedures and reasons for them. Again, sensitive handling of the request and information given is a necessity.
- Once a cause is known or after specialist cardiac pathology where no cause of death can be determined, the family GP should be informed and first-degree relatives contacted to commence a procedure to establish any genetic condition.

These steps may sound insensitive but the feedback received from the majority of SADS UK members is that the question of why their loved one died can become all-consuming until a cause of death is forthcoming (see Appendix 'A' & Personal Stories/Tom Gosling). Any opportunity to assist with discovering cause of death would have brought some comfort and very importantly would have established whether other family members were also at risk of sudden death. Some families have suffered several bereavements before a genetic condition was suggested. They have since been diagnosed with a condition and are receiving treatment (see Personal Stories/Michelle Griffiths/Nadine Bennett)

Schools & Colleges should have robust procedures in place to ensure the welfare of students after a bereavement. Currently support and understanding in schools appears to be patchy or non-existent, with some schools placing precedence on the child's education rather than their welfare. This needs to be addressed.

Good Practice - Cheltenham Coroner's Office

In the case of unascertained deaths, Cheltenham Coroner's Office contacts families within 48 hours and will make an appointment to either visit them or for them to meet at the Coroner's Office (see Cheltenham Coroners Office pg 25)

Considering the feasibility of guidelines on how these deaths are certified

It is believed that many sudden cardiac deaths are not accurately certified as such, and that this is due to incomplete/insufficient post-mortem investigations or pathology.

The current system of post-mortem and coroner's review is set up to look for evidence of death by unnatural causes, foul play. (see Pathology Issues pg 20).

The actual cause of death under the present Coroner's System is not given enough importance; this can lead to misdiagnosis and therefore inaccurate statistics. Funding therefore not being allocated to the most appropriate public health area.

An effective death certification and investigation procedure is crucial. This will validate the amount of research that needs to be placed in any specific area, to protect the general public, to help inform about medications and help prevent medical error.

This Coroner's System is currently under review and with the advent of a medically qualified Medical Examiner to be assigned to Coroner's Offices in the future to scrutinise death certificates and verify cause of death, hopefully the above issues will be addressed.

However, there is also concern regarding the confusion of classification and recording of deaths. The current categories appear ambiguous and we believe diffuse the statistics for sudden arrhythmic and unexpected death, therefore hiding the extent of these types of deaths.

There needs to be a change in the classification system to clarify and classify sudden deaths under their cardiac origin within an umbrella category in order that we gain more accurate statistics with regard to how many people are dying suddenly and unexpectedly from these types of conditions.

When baby cot deaths were officially labelled sudden infant deaths, SIDS, there was a 70% fall in the number of deaths within a period of 10 years (Foundation of SIDS, 2002 – BHF Study March 03). Once these deaths were under one heading the statistics proved shocking and measures were taken to advise on how to avoid this type of death.

BHF Study March 03 (see Appendix 'B')

The Lancet Nov 03 (see Appendix 'C')

See improbable cause of death – (see Personal Stories/Ashley Jolly/Colin Hart).

Setting standards or practice recommendations around how suspected cases are treated in primary care

See 'Comments on GMS Contract 2003' (pg 23)

See 'Overview Section' (pg 15) - An overview of how the detection and management of cardiac conditions could be incorporated into primary and secondary care, recommendations for screening, monitoring and raising awareness at various stages of development are given.

It is believed that as a minimum, as a screening exercise, the following should be procedure for children prior to starting secondary school, and again just before completing secondary school.

Minimum Screening requirements (as currently practical)

At the ages of 11yrs and 15yrs all children to be invited to their doctor's surgery for the following investigations:

- To be examined by stethoscope by the GP to detect any heart murmur.
- A questionnaire to be completed (with parental assistance) to establish if the child is in an at risk group and/or has suffered signs or symptoms consistent with a cardiac condition. Enquiries should also be made into any

medications the patient may be taking (for any reason). Should the patient be in an at risk group the medication should be checked urgently with specialised agencies for possible contra-indications and specialist advice sought as appropriate.

If there is any suspicion that the child may be suffering from a cardiac condition an ecg test should be undertaken. This is to be sent on-line or by fax to a cardiologist who must return a recommendation within 48 hours.

They must be referred immediately for further tests to be carried out. The minimum tests should be:-

Resting ECG
24-hour monitor
ECG Stress test
Echocardiogram

As paediatric consultants do not specialise, these tests must be referred to a cardiologist, with results made available within 2 months of the primary care consultation.

In addition to the above tests, the patient is to be referred to a genetic counsellor to compile a detailed family tree and assess risk. Genetic testing is especially important if the family history revealed by the questionnaire already suggests a possible hereditary link.

It is recommended that the increased use of the above described questionnaire, together with ECG testing and 24 hour heart monitoring in primary care would be beneficial, supported by a cardiologist on-line as appropriate. A procedure for any person, any age, presenting with, or having had signs or symptoms would be:

- An ECG would be taken during the first visit to the GP, and this would be checked by a cardiologist (via fax or email). A recommendation should be returned by the cardiologist within 48-hours. An abnormal ECG would mean commencement of investigations in secondary care employing all diagnostic procedures, as before, with emphasis on achieving results within 2 months.
- A questionnaire would be completed in primary care to discover any family history of signs and symptoms, or incidence of sudden death. If any history is revealed a referral to a genetic counsellor would be offered, in addition to any secondary testing that may be needed.
- In the absence of an abnormal ECG or evidence in the family history, secondary testing is still recommended if symptoms are shown, as many conditions require the use of other diagnostic equipment to be detected.
- Enquiries should also be made into any medications the patient may be taking (for any reason). Should the patient be in an at risk group the

medication should be checked urgently with specialised agencies for possible contra-indications and specialist advice sought as appropriate.

It is believed that the GPs surgery could be a lifesaving first stop for many patients presenting or having had signs and symptoms, and their contribution should not be overlooked.

The education and training of health professionals working within primary care about conditions that may cause sudden cardiac death is crucial. Practice nurses who will require additional training could carry out the procedures above although a cardiac nurse assigned to each surgery would be preferred. GP's should also have the complete support of resources and information from secondary care.

The GP's surgery is the first place people go when feeling unwell and patients place enormous trust in them. If the patient presents with symptoms consistent with a cardiac condition and the GP overlooks or dismisses them, referral to a specialist is not made and treatment is not established, which may have fatal consequences.

Promoting models of good practice for local use

If by this it is meant providing evidence of current good practice, which may be replicated, our organisation cannot assist with examples.

Design of an evidence-based protocol which clarifies when it is recommended that patients and/or their relatives should ideally be invited for screening

See 'Overview Section' – An overview of how the detection and management of cardiac conditions could be incorporated into primary and secondary care

There are studies that suggest that cardiac conditions are genetic (see Appendix 'D'); it is thus important to offer screening to first-degree relatives of a patient who has a cardiac condition or to family members where there has been a cardiac death. There is a 50/50 chance that another family member will have the same condition. (see Personal Stories/Barrie Irons/Michelle Griffiths/Nadine Bennett)

¹ Swimming is a typical trigger for the congenital cardiac condition Long QT Syndrome 1 causing a dangerous cardiac arrhythmia, often death (See Appendix 'E')

Acute Care

Design of a care pathway showing the ideal treatment of those who require emergency care for these conditions

It is expected personnel in this area will be trained in the treatment of the cause and effects of arrhythmia and, as with GPs and consultants, have full knowledge of the implications of these.

The Public Access Defibrillators initiative has saved many lives and providing life saving equipment in places that are populated is a potential lifesaver to victims of cardiac arrest. Data collated by the Resuscitation Council UK shows that the lives of 20% of victims have been saved due to rapid response.

Bringing Community First Response teams into the community is also of great value, especially to areas which may be more difficult for ambulances to access, eg., in more rural areas.

Defibrillators should be placed in leisure centres, golf clubs, etc., and regular training in the use of these and action plan in the event of an emergency should be undertaken.

Outlining what information patients require when in hospital to help them successfully manage their condition after discharge

It is recommended that patients aren't discharged from care once a cardiac condition has been diagnosed. Databases should be kept and patients should be alerted when new medical advice and/or potentially life saving treatments become available about their condition - they should be recalled immediately. In the current situation discharged patients should be recalled for re-assessment. (see Personal Stories/Louise Hurrell)

For some causes of arrhythmia there are definitely ways the patient can minimise the incidence of arrhythmia, and it is hoped the consultant will discuss these in depth with the patient. It is also hoped the patient will have future opportunities to consult, ie., regular monitoring and advice, in order to detect and adapt to changes in the condition.

Treatment options should be discussed ie., medication, ablation or ICDs, as if the patient were having a typical consultation after referral from their GP. Procedures that have been carried out must be fully explained, together with any complications that may occur, in order that the patient may be aware of these and know what action they should take if complication occurs.

Inform the patient of the Expert Patient Programme which helps them to manage their condition. (see Appendix 'H')

Considering the feasibility of setting standards or practice recommendations around re-admission for those conditions which could be better managed.

If patients are re-admitted on several occasions this is unacceptable. These patients are obviously at risk and their consultant should be negotiating more effective treatment for the patient and discussing it with them.

Development of a set of audit indicators and suitable performance indicators that might be used locally around emergency care for those with arrhythmias and conditions which may lead to sudden cardiac death

We are not aware of how performance is monitored in acute care compared with General Practice (see comments on GMS Contract pg 23) but believe this area of healthcare requires regular assessment and monitoring.

Cardiac Interventions

Design of a care pathway for the diagnosis, management and treatment of arrhythmias and conditions which may lead to sudden cardiac death (considering cardiomyopathies, atrial fibrillation and electrical conduction disorders)

We believe some of the considerations here are covered previously. Please see 'Overview Section'

In Summary

- Speedy, thorough and conclusive diagnosis
- Offer of relevant treatment where necessary, instructions for self-management, details of Expert Patient Programme and programme of on-going monitoring
- Diagnosed patients should not be discharged from further care at any time
- Screening and support of relatives

With regard to management and treatment of arrhythmias we believe specialists in this field of cardiology should lay down guidelines as appropriate, to be assessed on a regular basis and updated including NICE recommendations and/or new medical evidence.

Identification of Models which deliver the appropriate interventions reliably

A questionnaire carried out by SADS UK regarding people living with cardiac conditions showed the Implantable Cardioverter Defibrillator (ICD) to be the treatment of choice by most people – (see Appendix 'F'). The efficacy of the ICD has been trialled by the National Institute of Clinical Excellence and has been shown to be more efficient in saving lives than purely anti-arrhythmic medication. See Excerpt from 'Making the Economic Case for Medical Technology', The Medical Technology Group (see Appendix 'G').

Development of audit measures and suitable performance indicators around the care and treatment of people with these conditions, which might be used locally

As with the indicators used within primary care, there should be certain procedures that are observed throughout the country to assist consultants to decide when further or different treatment should be investigated.

Local audit channels should be put in place and The Healthcare Commission should monitor overall.

Patient Involvement and Support

Design of care pathway for management and control of specific conditions

We believe some of the considerations here are covered in 'Overview'

Patients should be made aware of the type of lifestyle that would alleviate their condition and what medications etc., may be dangerous to them. In addition GP's computer systems should 'flag up' any treatments that may be dangerous to the patient's condition.

Development of guidance on monitoring patients with chronic conditions

Guidance laid down by professionals must be assessed on a regular basis and health professionals must collaborate and share new knowledge and evidence regarding the monitoring of patients with chronic conditions.

It is important that this information, together with new advice or warnings regarding medications and interventions used are rapidly disseminated.

Patients must be reviewed regularly.

Identifying effective ways of providing emotional support to patients and their relatives where appropriate

- Counselling to be available quickly, when needed.
- Provide lists of support groups and charities for specific conditions, helping to dispel the feeling of isolation and enabling them to share their thoughts and feelings.
- Provide information where possible, as this is facilitative in providing support to patients and their families.

Identifying effective means of involving patients in their own care and in influencing the way that services are provided.

- Patients carrying with them lists of medications they are taking.
- Wearing Medic alert indicators where possible
- Becoming involved with support groups and charities for their condition
- Becoming an Expert Patient via Expert Patient Programme – (see Appendix 'H')

- Information and choice empowering patients to speak with healthcare providers regarding their needs will ultimately influence how services are provided. We are aware that in primary care, patients have an opportunity to complete a written assessment questionnaire regarding their doctor's surgery and GP's practice. Face to face negotiations with health professionals would be of greater value to both patient and health professional and would be instrumental in providing patients with the services they require.

Overview Section

An overview of how the detection and management of cardiac conditions could be incorporated into primary and secondary care

Minimum Screening requirements (as currently practical)

At the ages of 11yrs and 15yrs all children to be invited to their doctor's surgery for the following investigations:

- To be examined by stethoscope by the GP to detect any heart murmur.
- A questionnaire to be completed (with parental assistance) to establish if the child is in an at risk group and/or has suffered signs or symptoms consistent with a cardiac condition. Enquiries should also be made into any medications the patient may be taking (for any reason). Should the patient be in an at risk group the medication should be checked urgently with specialised agencies for possible contra-indications and specialist advice sought as appropriate.

If there is any suspicion that the child may be suffering from a cardiac condition an ecg test should be undertaken. This is to be sent on-line or by fax to a cardiologist who must return a recommendation within 48 hours.

They must be referred immediately for further tests to be carried out. The minimum tests should be:-

Resting ECG
24-hour monitor
ECG Stress test
Echocardiogram

As paediatric consultants do not specialise, these tests must be referred to a cardiologist, with results made available within 2 months of the primary care consultation.

In addition to the above tests, the patient is to be referred to a genetic counsellor to compile a detailed family tree and assess risk. Genetic testing is especially important if the family history revealed by the questionnaire already suggests a possible hereditary link.

Overview

An ideal procedural plan, in primary and secondary care, for the ongoing detection and monitoring of the conditions under consideration

During Pregnancy

During routine primary and secondary check-ups a family history can be established, as is currently, for genetic conditions to include arrhythmia causing conditions, including signs and symptoms.

At Birth

Promotion of awareness begins with leaflet to parents explaining signs and symptoms of conditions which may lead to arrhythmia, in the same way information is distributed about meningitis and other typical diseases. Usually from the GP or health visitor.

Information on family history to be gathered if not done so by this stage.

At this point, monitoring of baby in secondary care if in an at risk group would be preferable.

Toddler Stage/2 Year development check

Awareness promotion to parents continues as with the promotion of awareness of many ailments at this stage; leaflets, advice from health visitor and GP.

Monitoring at secondary care if in at risk group, and regular secondary care monitoring for diagnosed patients.

3 – 10 Years

Regular monitoring in secondary care for diagnosed children, regularity to be determined but with no more than two years elapsing between each check. Once diagnosed, patient is not to be discharged from care.

Regular monitoring in secondary care for at risk groups, again regularity to be determined but with no more than two years between each check.

Awareness to continue at every opportunity in GPs' surgeries, pre-schools, nurseries and primary schools, swimming, sports and youth groups, in a manner similar to the way other life-threatening diseases are communicated to parents.

Puberty – 10/11 Years

Screening at age 11, implemented by means of a questionnaire, which will highlight any abnormalities or family history.

Monitoring in secondary care if in at risk group, and regular secondary care monitoring for diagnosed patients.

This is a particularly stressful time for the body, with the onset of hormonal and physical changes. There are also emotional stresses to consider as most children start secondary school.

Awareness to continue through to secondary school medical staff, Guides/Scout groups, swimming¹, sports and dance clubs.

Years 12 to 18

Screening at age 15, implemented by means of a questionnaire, which will highlight any abnormalities or family history.

Minimum screening recommended, prior to leaving secondary education, as described above. Regular monitoring of at risk groups and diagnosed patients to continue, with a recommendation that these remain at yearly intervals until the age of 18.

Awareness campaign to parents and children via school, sports and youth clubs (young people are constantly reminded of the dangers of illegal substances, smoking and unsafe sex at this time and cardiac conditions should be no exception)

This is still a dangerous time for young people, if a condition has not yet come to light, due to continued and dramatic hormonal and physical changes. Emotional stresses are also increased because of, for instance, exams and typical teenage considerations.

Adults

Diagnosed patients

These should continue to be monitored in secondary care, but at a frequency negotiated between patient and consultant. A 'care pathway' should already be in place providing guidelines enabling the consultant to advise the patient on future monitoring and treatment as the need arises. At no time should the patient be discharged from care.

At risk groups

Secondary care monitoring should be continued at a frequency negotiated between patient and consultant. The fact that regular monitoring has taken place during earlier years, and there is a record of family history taken during previous consultations, is a good basis for the patient to make an informed decision about the continuation of care.

For others

Increased awareness among the general public and health professionals will enable people to look for warning signs.

Awareness

It must be the responsibility of GPs and hospital cardiac departments to display leaflets and posters at all times, and of course to have improved their own knowledge and training.

The Dept of Health to continue campaign of national press releases and advertising.

At All times

A procedure for any person, any age, presenting with or having had signs or symptoms would be:

- An ECG would be taken during the first visit to the GP, and this would be checked by a cardiologist (via fax or email). A recommendation should be returned by the cardiologist within 48-hours. An abnormal ECG would mean commencement of investigations in secondary care employing all diagnostic procedures, as before, with emphasis on achieving results within 2 months.
- A questionnaire would be completed during the primary care appointment to discover any family history of signs and symptoms, or incidence of sudden death. If any history is revealed a referral to a genetic counsellor would be considered, in addition to any secondary testing that may be needed.
- Anyone in an at risk group must have any medication being taken checked urgently with specialised agencies for possible contra-indications and cardiac specialist advice sought as appropriate.
- In the absence of an abnormal ECG or evidence in the family history, secondary testing is still recommended for people where symptoms have been shown that may be consistent with a cardiac condition, as many conditions require the use of other diagnostic equipment to be detected.
- Diagnosed patients, no matter what level of condition they are believed to have, should not be discharged from secondary care at any time. We believe as an immediate priority patients who have been discharged should be recalled in the light of any new medical evidence or advice that could affect them.

- Monitoring of relatives of at risk and diagnosed patients. Refer to section on an evidence-based protocol for screening of relatives.

¹ Swimming is a typical trigger for the congenital cardiac condition Long QT Syndrome 1 causing a dangerous cardiac arrhythmia, often death (See Appendix 'E')

Pathology Issues

Pathology offers the opportunity to reach a conclusive cause of death, however, at present sudden cardiac death is not systematically well investigated in the UK due to lack of resources.

At the present time pathology is under funded and underutilised, containing procedural discrepancies across the country. It appears the most basic elements to help with post-mortem investigations such as height and weight of the deceased appears not to be recorded in all PM procedures/reports. It is important that where there is difficulty in ascertaining a cause of death these deaths are referred to specialised cardiac pathologists. Incorrect verdicts have left some families at risk from life-threatening hereditary conditions and some have led to further deaths and dangerous incidents within families. (see Personal Stories/Barrie Irons/Michelle Griffiths/Nadine Bennett).

There are cases where no cause of death has been ascertained, again exacerbating the distress of families and leaving family members at risk (See BHF Study March 03) (see Personal Stories/Jenny Legg).

Presently there appear to be causes of death certified which appear highly improbable, ie., asthma, epilepsy, where no previous incident or family trait is apparent. (see Personal Stories/Ashley Jolly/ Ria Hickerton).

It is understood that to improve pathology procedures certain contentious actions would need to be in place, eg., taking of blood samples, tissue samples and maybe organs. These issues may be addressed by the recommendations in the section on 'Support of relatives' where a coroner's officer could sensitively ask permission for these actions to be taken. As stated previously, it is a form of support for grieving families to know why their loved one has died.

We believe if resources were made available for accurate pathology and guidelines were in place to ensure a cause of death was found if at all possible, these improvements would assist with the section 'Considering the feasibility of guidelines on how these deaths are certified'

Even if the recommendations of SADS UK regarding changing and clarifying the death classifications were not considered, improved pathology will still assist in accurately allocating deaths to the classifications as they presently exist. The result of these improvements will be to provide greater statistical evidence of specific cardiac conditions and the possible extent of arrhythmic deaths, also the incidence of incorrect classification or certification and the subsequent distress and possible risk to families who have a hereditary condition is alleviated.

Refer back to main document Sudden Cardiac Death and Screening
'Considering the feasibility of guidelines on how these deaths are certified'

Drug Issues

Making Health Professionals Aware

The issue of drug related arrhythmia is currently being investigated in a BHF study launched in April 2003 (Appendix 'J'). The concern is that over-the-counter and prescribed medications for common complaints could aggravate existing heart conditions that can cause arrhythmia, or even instigate arrhythmia where no condition has been present. For instance, some prescribed drugs can prolong the LQT interval in some patients sometimes causing dangerous and fatal cardiac arrhythmia (see Personal Stories/Ashley Jolly). We believe that the risks to patients must be understood and assessed adequately by GP's. (see Appendix 'L')

For at risk groups and diagnosed patients there should be more efficient systems in place of alerting prescribing GPs and consultants to the drugs currently thought to pose a risk to these groups. At the moment, for new information on adverse reactions in any drug these professionals rely on updates from the Committee on Safety of Medicines and, for urgent safety issues, a cascade system of notification is in place. Although the British National Formulary publication provides a source of reference for the main groups of drugs, it is believed this is not referred to often enough. It is recommended that, included with a possible information pack for health professionals to raise awareness, a list of drugs that may cause problems for at risks groups and diagnosed patients be drafted. This information could then be placed with every relevant patient's notes, even if computerised. Updates could then be made to those notes as necessary.

There currently seems to be no database that filters out expected adverse reactions of a drug to specified groups, and the arrangement of this information would be recommended.

It should be made compulsory that in the case of an adverse drug reaction a yellow card is immediately sent to the Medicines Control Agency and the Committee for Safety of Medicines are notified.

It should be made compulsory that after a sudden and unexpected death an adverse drug reaction form is sent as detailed above.

Patients often perceive their doctor to be omnipotent and they are put in a position of trust and respect, sometimes disproportionately. It is important that they discuss medication prescribed with their patient and warn of any dangerous side effects that mean the patient should cease taking the medication and return to the surgery. It is also important that the doctor sees their patient regularly when they give them repeat prescriptions. It is very unlikely that the patient will read the medication insert each time they purchase the medication and side effects may occur at any time. The doctor must understand their responsibility and remain vigilant.

Worldwide Parity

It is hoped that information about drugs is shared with similar organisations to the CSM/MHRA in other countries are an issue here and in the US. The safety concerns of Triludan (Seldane) in this country, for instance, occurred after the US had withdrawn this drug due to safer options being available (see Appendix 'K'). We are concerned at which point these safety warnings lead to withdrawal from worldwide circulation.

Comments on GMS Contract 2003

On initial investigation of the new GMS Contract, considering we have limited knowledge of its implications, it seems that there is currently little consideration for the detection and management of arrhythmia, although it is accepted that the new contract includes outlines for incentives available rather than good practices necessary.

It is expected that a section giving indicators for the detection and management of arrhythmia will be inserted, hopefully based upon the procedures recommended in the Overview attached to this response. However, one major concern about the reward system within the Quality and Outcomes Framework is that the attraction of gaining points for what would appear to the layman as procedures that should be carried out as a matter of course may deflect attention from the conditions we are concerned with in this paper.

It would appear from the tables of points available in each clinical area that there is a bias towards the routine recording of blood pressure, and maintaining it at specified levels, in patients with Hypertension, CHD and Diabetes. It is hoped that these checks, while crucial to controlling the overall health of the patient, would be routinely done without the need for incentives. The cynical view would be that practices will concentrate on these procedures to the detriment of others of lower points value. The document states that each practice can choose which areas of the framework to concentrate on.

This approach also reinforces the impression that the Dept of Health's main concern is to reduce the costs of caring for those mature people who may have made unhealthy lifestyle decisions, which probably accounts for a large proportion of available funds. While this may be a good economic tactic, the incentives currently available do not encourage practices to monitor for and manage the lesser known diseases that do not have lifestyle links, especially in the young. There appears to be no incentives for checking the cardiac health of anyone except under the category of CHD, which generally occurs in the older population, and does not include arrhythmia. There are other cardiac conditions that can induce potentially fatal arrhythmias and these can occur in any age group, including children.

There appears to be no points available for patients with known heart defects to have regular ECGs or heart monitoring of any kind.

Unfortunately, the document states that the indicators are evidence-based, which highlights again how it is believed incidences of fatal arrhythmia are not accurately recorded and how these statistics will not encourage improvements.

The document also confirms the importance of Echocardiography in the diagnosis of Left Ventricular Dysfunction, and goes on to acknowledge that this resource is limited in parts of the country. It will be seen from the Overview that we recommend extensive use of this piece of equipment. This is undoubtedly an obstacle with regard to the Department of Health's priorities regarding CHD at the moment and will become even more urgent with the implementation of the New

Chapter regarding arrhythmias and sudden cardiac death. SADS UK believes improving diagnostic equipment availability must be viewed as an immediate priority.

There seems to be no mention made of the GPs relationship with the local hospital and its consultants, and how referrals impact on the practice. This may be a procedure referred to in other primary care guidelines and we believe this is of great importance to the role of the professional in primary care. It is felt that currently there is room for improved communication between primary and secondary care, especially with regard to the flow of information about a patient's condition. The GP relies heavily on the letters received from a patient's consultant and we wonder if these adequately explain a condition and how a GP might deal with signs of deterioration or progression. It is understood that signs and symptoms of a fatal arrhythmia may be overlooked by the GP (see Personal Stories/Louise Hurrell), especially when suffered by a young, energetic person. We believe the GP would benefit from as much secondary care information as possible including face-to-face discussions between the professionals.

Good Practice

Cheltenham Coroner's Office

The following procedure is followed for inquest cases:-

Cheltenham Coroner's Office work very closely with the pathologists at their local hospitals. The Pathologists inform the Coroner's office of what they need to retain and the Coroner's Office passes on the information to the next of kin. It is their policy that when they meet with the family they take a 'registration statement' (sometimes called an antecedant statement by other Coroner's offices). This statement contains all the information needed for the Coroner's Office to register the death when the Inquest is concluded. It also contains all the information needed to start their investigation ie. a little of the family history, childhood illnesses of note, interests, work/school and recent health or allergies etc. They also ask for a detailed description of the last day or so of the deceased's life to try to ascertain if there was anything unusual. The statement can be anything up to 5/6 A4 pages and will probably take 1 - 2 hours. At the end of the statement they write in details of the organ and/or tissue retention and ask the informant to sign each page, thereby confirming the details but also confirming that they have been informed of the organ/tissue retention. They also include the wishes of the next of kin re disposal of the organs when they are no longer required by the pathologist. This information is passed on to the pathologist and the statement will be included in the Notes of Evidence at the Inquest. The informant is offered a copy of the statement.

These statements are usually taken at the home of the next of kin but increasingly they have to ask people to travel to Cheltenham to their office. They don't like to take this action but do not always have time to travel 30 odd miles to the end of the county - if people can travel to them it means that they can use the time to see another family.

They inform them that investigation can take many weeks and that they may not hear from the Coroner's Office during that time. At the end of their investigations, whether or not a cause of death is found, they contact the next of kin by phone to discuss their findings and if it is appropriate they invite them to come to the Coroner's Office again to go through the file and inform them of the findings. This allows them to make an informed choice as to whether they wish to attend the Inquest. They inform the next of kin of the possibility that they may not remember all the details of the Inquest procedure and offer them the facility to contact them at any time. They ensure that the next of kin is aware that if witnesses are called then the family are allowed to ask relevant questions in court and they provide the family with some idea of the format of the Hearing, informing them they are entitled to purchase the Notes of Evidence after the Inquest is concluded and this information is repeated in the 'notification of Inquest' letter that they send out to everyone when they have set the Inquest date.

Very occasionally, if families are particularly distressed and vulnerable they arrange to meet the family at the court a week or so before the Inquest so that they can see the empty court and will not be overawed by the venue, although unfortunately they cannot offer this option to all those who have been bereaved due to lack of resources.

Obstacles

- Lack of awareness with the general public and health professionals
- Statutory Primary Care heart monitoring equipment inadequate
- GP made responsible for their own training
- Lack of specific training
- Lack of staffing at primary care level
- GP Budget
- GP Contract (doesn't encompass arrhythmia)
- Human Tissue Bill
- Inconsistent and inadequate investigation at post-mortem
- Lack of cardiac specialist pathologists
- Present outdated Coroner's System
- Coroner's under jurisdiction of the Home Office & Pathologists working in the NHS
- Lack of funding in the area of electrophysiology
- Lack of research
- Inaccurate statistics for sudden cardiac death
- Lack of specific guidelines and integrated pathways
- Healthcare Resource Groups (how will payment for innovative technologies be assessed as payment rates are relevant to data collated from hospitals)
- Insufficient recording of adverse drug reactions from doctors and pathologists
- Insufficient communication between doctors and drug agencies regarding medications

Many of these obstacles are covered in our document, the above expands on our observations.

Conclusion

The Overview included in this document describes certain primary and secondary care procedures that could be put in place quickly and with immediate impact. The basic screening requirements would be an immediate priority, as would the improved vigilance of the general public and health professionals in the primary care sector to understand signs, symptoms and family histories that indicates cardiac investigation should be undertaken. It would make sense that more knowledge and resources are made available in this area and that they have easy access to secondary or tertiary care for further investigation of patients. An immediate improvement in the flow of information between primary and secondary care professionals would benefit GPs when faced with patients demonstrating signs that are not immediately recognised as giving cause for concern.

Correctly reporting and collating data with regard to people who die suddenly and unexpectedly from cardiac conditions is also crucial. Stringent reporting and recording mechanisms need to be put in place quickly in order to ascertain the extent of sudden cardiac death; an umbrella category would also be helpful in this respect. Further resources need to be put into this area of public health and we believe accurate statistics would validate this action being taken. It's a chicken and egg scenario.

Pathology is an area requiring an immediate full review, although it is accepted that this is in need of in-depth assessment, and changes would take time to implement. Consistent, robust procedures for pathologists, throughout the country need to be put in place, including referring difficult cases to specialised cardiac pathologists, and details of medications taken prior to a sudden death to the Medicines Control Agency. Accurate pathology will help give a cause of death at post-mortem instead of reaching a verdict of 'unascertained', thus alleviating to some extent the distress caused to relatives when they don't know why their loved one has died. More resources need to be made available in the whole area of pathology.

It is hoped that accurate pathology, the new Coroners System and the impending negotiations WHO are undertaking with regard to clarifying death classification will be instrumental in achieving accurate statistics.

A long term priority, and an ideal scenario, would be to screen the population for cardiac conditions using accurate means to do so. At the moment accurate means are not available and this is not feasible. Currently family history and signs and symptoms are the best way of assessing whether a person may be at risk of suffering from a cardiac condition.

SADS UK believe that as refined technology and genetic testing for the purpose of accurate and efficient diagnosis of cardiac conditions becomes available it should be assessed for population screening.

A clear set of national standards, sustained levels of investment and reform and assessment should improve the services provided to those living with cardiac arrhythmia and help diminish the risk of sudden cardiac death.