RESEARCH PAPER 04/24
10 MARCH 2004

The *Cardiac Risk in the Young (Screening) Bill*

Bill 19 of 2003/4

Dari Taylor MP, who drew fifth place in the ballot for Private Members' Bills, has introduced a Bill to require cardiac screening for people under 35 years old. The Bill is due to have its Second Reading on Friday, 12 March 2004. This Paper describes the policy background to the Bill and the medical conditions that have given rise to it.

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ISSN 1368-8456

Summary of main points

Most of the sudden deaths from natural causes that occur in young people under 35 years old are due to sudden cardiac death (SCD)[1]. The majority of these cases are due either to a disorder of the heart muscle (cardiomyopathy), or to an abnormal heart rhythm (cardiac arrhythmia). In a proportion of sudden deaths, no cause can be identified and these cases are classified as death from natural causes. It is now accepted that many of these deaths are attributable to an underlying cardiac disorder, predominantly problems with abnormal heart rhythms.[2]

The aim of Dari Taylor's Bill is to ensure that young people who are close relatives of someone whose death may have been caused by a "genetic" cardiac disease are screened for the condition and that those who have symptoms associated with some of the other causes of sudden cardiac death are advised to see a relevant specialist. In order to achieve this aim, the Bill would place requirements on pathologists who carry out post mortems, GPs and relevant specialists.

The Government, on the advice of the UK National Screening Committee, has so far rejected calls for a national programme but does advise that relatives of people who have died of sudden cardiac death, or who are diagnosed with one of the underlying conditions, should be screened because there is a genetic component in many of these cases.[3]

The Bill is supported by CRY, the All-party Parliamentary Group on Cardiac Risk in the Young and the All-party Parliamentary Group on Heart Disease.

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1 "Sudden cardiac death in the young", *Medical Journal of Australia*, 18 February 2002

2 "Sudden death in children and adolescents", *Heart*, 1 October 2002

3 HC Deb 16 December 2003 c882W

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I Introduction

When Marc-Vivien Foe, the former Manchester City and West Ham player, died during a match in 2003, it became headline news. Newspapers reported that, like several other young sportsmen, his death was caused by a rare heart condition.[4] In his case it was cardiomyopathy, possibly the most common heart condition that can kill young people without apparent warning.

The recent publicity surrounding the well-known sportsmen who have died young has drawn attention to what the charity, *Cardiac Risk in the Young*, considers to be a neglected problem. It argues that in addition to cadiomyopathy, there are several other heart conditions that can cause sudden death in young people. Although many of the young victims had no obvious symptoms, the medical evidence suggests that there may be some danger signs and that some of the conditions have a genetic component. The charity and its supporters are campaigning for an active screening policy focused on those at risk. Dari Taylor's Bill, which is supported by the All-Party Parliamentary Group on Heart Disease, reflects their concern.

The medical conditions that can cause sudden cardiac death and the risk factors that may be associated with them are described in Part 1 of this Paper. Part 11 covers the Bill and related policy aspects.

II Sudden death in young people – medical aspects

A. Sudden Death Syndrome in Young People

1. Definition

Most of the sudden deaths that occur in young people, under 35 years, are due to sudden cardiac death[5]. This is defined as an unexpected natural death due to a cardiac cause occurring in a short time period, generally within 1 hour, from the onset of symptoms.[6] Both the time and mode of death are unexpected. The majority of these cases are due either to a disorder of the heart muscle (cardiomyopathy), or to an abnormal heart rhythm (cardiac arrhythmia).

In a proportion of sudden deaths, no cause can be identified and these cases are classified as death from natural causes. It is now accepted that many of these deaths are attributable to an underlying cardiac disorder, predominantly problems with abnormal heart rhythms.[7]

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4 See, for example, a report of some other deaths that refer back to this one, "Heart defect that strikes the young and fit", by Jeremy Laurance, *The Independent*, 3 March 2004. Cry's website also contains a list of young sportsmen who have died from such heart conditions: www.c-r-y.org.uk

5 "Sudden cardiac death in the young", *Medical Journal of Australia*, 18 February 2002

6 "Sudden cardiac death in the young", *Medical Journal of Australia*, 18 February 2002

7 "Sudden death in children and adolescents", *Heart*, 1 October 2002

If specialist post mortem examination were performed in such cases it is thought that cardiac disorders would be more often identified as the cause of death.

2. Incidence

The actual number of sudden cardiac deaths in those under 35 years of age is relatively low but concern is still raised as many of these deaths are thought to be preventable.[8]

Accurate statistics for the incidence of sudden cardiac death are not available. This is, in part, due to the difficulty in correctly identifying the cause of death at post mortem. CRY (Cardiac Risk in the Young) claim the figure could be as high as eight deaths a week but other studies believe the figure is considerably lower.[9]

The incidence of sudden cardiac death during competitive sport was estimated from a population survey of high school and college athletes in the United States. This suggested that 1 in 200,000 competitors were affected.[10]

3. Risk factors

The following factors are recognised as increasing the risk of sudden cardiac death.[11]

- Family history of sudden premature death
- Family history of a known cardiac disorder such as a cardiomyopathy, or an abnormal cardiac rhythm.
- Certain recognised genetic mutations which are known to be linked with cardiomyopathies.
- Previous cardiac arrest
- Recurrent syncope (fainting)
- Palpitations or any other evidence of an abnormal cardiac rhythm
- Other physical symptoms such as breathlessness or chest pain

In addition, there is a growing body of evidence to indicate that physical exercise can be a precipitating factor for sudden death in those individuals who have an underlying cardiac abnormality. Up to 80% of sudden cardiac deaths in athletes occur during or immediately after vigorous exercise.[12]

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[8] "Sudden cardiac death in the young", BMJ. 22 March 1997

[9] "Sudden death in children and adolescents", *Heart*, 1 April 2000

[10] "Sudden death in the young competitive athlete", *JAMA*, 1996;276:199-204

[11] "Sudden cardiac death in the young", *Medical Journal of Australia*, 18 February 2002

[12] "Risk of competitive sport in young athletes with heart disease", *Heart*, 1 July 2003

4. Screening

The majority of sudden deaths occur in individuals who have underlying genetic or inherited cardiac disorders.[13] Over 200 gene defects on at least 10 different genes encoding for the proteins which make up the cardiac muscle have already been identified.[14]

All the cardiac conditions causing sudden death are rare, meaning that screening of the general population would be impractical. A more useful approach would be to target screening at those who are at highest risk. As there is this strong familial link for many of the conditions, the most effective method of screening would be to identify individuals with a family history of sudden death or inherited cardiac conditions. The presence of any other risk factors, as listed previously, may also provide a strong indication for screening[15].

B. Cardiac diseases causing sudden death

1. Cardiac function

For the heart to work effectively, both the cardiac muscle and the electrical conduction system of the heart must be functioning normally. The pumping action of the heart is brought about by the contraction of the cardiac muscle wall, the myocardium. The myocardial contraction is initiated by electrical impulses originating in the pacemaker area of the heart, the sinoatrial node. These impulses are transmitted by specialised nerve fibres in the conduction pathway and then spread through the myocardium. Disorders of the myocardium or disturbances of the conduction system are responsible for the majority of cardiac disorders linked to sudden deaths in the young.

Post mortem studies of sudden death victims reveal an abnormality in the structure of the heart in about 90% of cases.[16] In most of the remaining cases it is thought that death is due to an electrical disorder of the conduction pathway.

2. Cardiomyopathies

Cardiomyopathy is the term given to any disease of the myocardium, which results in a decrease in the force of contraction of the heart. They are classified into three main groups.

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[13] "Risk of competitive sport in young athletes with heart disease", *Heart*, July 2003;
[14] "Sudden cardiac death in the young", *Medical Journal of Australia*, 18 February 2002
[15] "Sudden cardiac death in the young", *BMJ*, 22 March 1997
[16] "Sudden cardiac death in the young", *Medical Journal of Australia*, 18 February 2002

3. Hypertrophic cardiomyopathy

This is the single most common disorder causing sudden cardiac death in people under 35 years.[17] The disease is characterised by thickening of the myocardium, usually of the left side of the heart. The heart muscle fibres also become abnormal.

a. Incidence and causes

The incidence of hypertrophic cardiomyopathy, (HCM), is about 1in 500.[18] There is an average annual mortality rate of 1-2%.[19] It is caused by a defect in one of the genes coding for the contractile proteins of the myocardium. There is a familial link in up to 90% of cases, [20] with an autosommal dominant pattern of transmission, which means that a parent carrying the gene for hypertrophic cardiomyopathy has a 1 in 2 chance of passing the disorder onto their children. However, the severity of the disease will vary even between people carrying the same gene defect. Children who carry the gene defect may not develop the disease until adolescent growth is completed.

b. Symptoms

Hypertrophic cardiomyopathy is frequently asymptomatic. Children and adolescents are most often diagnosed with the condition during the screening of affected families. Approximately 50% of adults present with symptoms of breathlessness, syncope (fainting), palpitations and chest pain. The remainder are diagnosed after detection of an unexpected abnormality during a routine medical examination or following a screening test. In all groups, sudden death may be the first presenting symptom of the disease.

c. Diagnosis

- *Electrocardiography:* A 12 lead electrocardiogram, ECG, which measures the pattern of electrical activity in the heart muscle, will show changes due to the increased thickness of the heart wall. The ECG may also show cardiac arrhythmias.
- *Chest x ray*: This will show enlargement of the heart shadow.
- *Echocardiogram*: This is a non invasive test and uses ultrasound waves to produce an image of the heart. It provides information on the structure and function of the heart. In hypertophic cardiomyopathy it enables the thickness of the myocardium to be measured, to provide some assessment of the severity of the disease.

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[17] "Sudden cardiac death in the young", *Medical Journal of Australia*, 18 February 2002

[18] "Sudden cardiac death in the young", *BMJ*, 22 March 1997

[19] Hypertrophic cardiomyopathy: management, risk stratification, and prevention of sudden death", Heart, February 2002

[20] "The cardiomyopathies", *Oxford Textbook of Medicine, 4th Ed*

- *Genetic diagnosis*: Recent advances have identified several genes involved in hypertophic cardiomyopathy. It is possible to identify some of these genetic defects by DNA testing.

Problems in the diagnosis of hypertophic cardiomyopathy can occasionally occur in young athletes. This is because many of the anatomical and physiological changes that result from strenuous physical training, such as an increase in the muscle bulk of the heart, are similar to the early changes of cardiomyopathy. Further screening tests can be used to differentiate such cases.

d. Treatment

Treatment is aimed at improving symptoms and preventing complications of the disease as the underlying condition can not be changed. Treatment options include:

- *Drugs:* These can be used to improve symptoms of breathlessness and control cardiac arrhythmias
- *Implantable cardiverter defibrillators:* These are similar to pacemakers and will deliver an electrical impulse to the heart if significant arrhythmias occur, to restore the heart to a normal rhythm.
- *Surgery:* In a small number of cases this is used to remove a section of cardiac muscle to enable the heart to pump the blood out of the heart more effectively. Very rarely a heart transplant is a suitable method of treatment.
- *Lifestyle changes:* More than 60% of sudden deaths in HCM occur during or immediately after mild or moderate exercise.[21] Individuals with HCM should be advised not to undertake vigorous activity.

Other cardiomyopathies are considerably less common.

4. Arrhythmogenic right ventricular dysplasia

This disorder has only been recognised since the 1970s but is now known to have an incidence between 1:1000 and 1:5000. It is characterised by replacement of the normal muscle cells in the wall of the right side of the heart with fibrous or fatty tissue.

There is familial link in at least 30% of cases and specific genetic defects have been found.

The usual presenting symptoms are palpitations and syncope due to cardiac arrhythmias.[22] Sudden death related to exercise may be the presenting symptom in the young.

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[21] "Hypertrophic cardiomyopathy: management, risk stratification, and prevention of sudden death", *Heart*, February 2002

5. Dilated Cardiomyopathy

The disease is characterised by the dilatation of the main pumping chamber of the heart associated with impaired contraction of the heart wall. This results in a low cardiac output that will eventually produce signs of cardiac failure.

The incidence quoted in the United States is 3-10 per 100,000 of the population[23] and it is likely the incidence is similar in this country. At least 20% of cases have a familial link and specific genetic defects have been identified. The genetic expression is variable so people carrying the same gene defect will be affected with various degrees of severity. A number of other factors are also known to be associated with dilated cardiomyopathy. These include certain viral infections, auto immune diseases and pregnancy.

Symptoms are due to cardiac failure with breathlessness as the most common presenting complaint. Arrhythmias are common and can cause sudden death. Survival is improved by early treatment and drugs used to control arrhythmias will reduce the incidence of sudden death.

6. Myocarditis

Myocarditis is an inflammation of the heart muscle. The condition can be acute or chronic but is almost always caused by a viral infection.

Mild cases are often asymptomatic and spontaneous recovery will occur. In a small proportion of cases, the muscle cells remain inflamed and become infiltrated with areas of fibrosis. This area of fibrosed tissue prevents normal electrical through the heart muscle, so producing arrhythmias, which can lead to sudden death. The disease can progress to cardiac failure and a dilated cardiomyopathy in about 10% of cases.

The condition is more common in the young. A retrospective review of United States Air Force crew found an estimated sudden death rate of 4 per 100,000 in the population aged 17-28 years.[24] A recent French communication claims that the death rate due to myocarditis could be as high as 5% of all sudden out of hospital cardiac deaths.[25]

Treatment involves total rest as even moderate exercise can trigger arrhythmias and sudden death.

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[22] Cardiac arrhythmias cause an irregular heartbeat. The individual will experience palpitations. Certain irregular heart beats prevent the blood being pumped normally from the heart. This reduction in blood flow to the brain can cause loss of consciousness.

[23] "Sudden cardiac death in the young", *BMJ*, 22 March 1997

[24] "Diseases of heart muscle", *Oxford Textbook of Medicine*, 4th edition

[25] "Myocarditis as a cause of sudden death", *Circulation*, 103-e12 2001

7. Conduction disorders

In conduction disorders, the heart appears structurally normal but there is an abnormality in the electrical conduction system of the heart. This means that following a sudden death these conditions will often not be recognised at post mortem.

Some of the conduction disorders implicated in sudden cardiac death are;

- Wolff-Parkinson-White syndrome
- Congenital long Q-T syndrome
- Brugada syndrome
- Idiopathic ventricular fibrillation syndrome

Each of the conduction disorders produces characteristic ECG changes.

The prevalence of these conditions in the population is impossible to determine. There is a known genetic link in many cases and screening of affected families will help identify cases. With the increasing recognition of the genetic abnormalities, it has been suggested that blood samples saved following sudden death might allow retrospective identification of the cause of death.[26] This would also be of benefit in recognising and screening affected families.

The most common symptoms are of palpitations and syncope. However, individuals are often asymptomatic and sudden death may be the first presentation. A number of cases will also be diagnosed as an incidental finding during a routine ECG testing.

It is important to diagnose these disorders as the majority can be successfully treated either surgically or with drugs thus minimising the risk of sudden death.

8. Others

Several other medical conditions are known to cause sudden cardiac death.

- *Coronary artery disease*
  Although rare in those under 35 years, some people will develop coronary artery disease at an early age, especially if there are genetic factors involve.
- *Marfan syndrome*
  This is an inherited connective tissue disorder, with an incidence of about 2 per 100,000 people. Of these affected individuals 90% will develop problems with the heart or aorta. If the cardiac problems are diagnosed at an early stage they can usually be successfully treated with surgery
- *Coronary artery anomalies*

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[26] "Sudden death in children and adolescents", *Heart,* April 2000

> The coronary arteries supply blood to the heart. An anomaly of the vessel can cause an interruption in the blood supply to the myocardium, resulting in a heart attack or an arrhythmia. The charity CRY, believe that this disorder is the second commonest cause of death in athletes under 25 years of age.

III Screening policy

A. Government policy

The Government's general approach to screening programmes was summarised in a recent Written Answer:

> **Miss Melanie Johnson**: The national breast and cervical screening programmes operate in each strategic health authority area. A number of other national screening programmes are currently being developed. These include screening for Down's syndrome where presently 75 per cent., of maternity units have a screening programme in place. 210,000 babies have been screened under the newborn hearing screening programme since it started. Newborn screening for sickle cell disease is in place in London and West Midlands. Raising public and professional awareness is a key aspect in the development of all screening programmes. Funding to implement screening programmes is mostly included in baseline financial allocations to primary care trusts. In keeping with the decision to enable local PCTs to decide their priorities in the light of their own needs, PCTs decide the allocation of resources locally.[27]

Although some screening programmes have developed locally and patchily over time, national screening programmes for medical conditions of people who have no symptoms are now generally undertaken following advice from the UK National Screening Committee. This advice is not binding on Ministers but in practice it is influential. The website of the UK National Screening Committee says:

> The UK National Screening Committee (NSC) is chaired by the Chief Medical Officer for Northern Ireland and advises Ministers, the devolved national Assemblies and the Scottish Parliament on all aspects of screening policy. It has two sub-groups dealing with antenatal and child health screening issues. In forming its proposals, the NSC draws on the latest research evidence and the skills of specially convened multi-disciplinary expert groups, which always include patient and service user representatives.
>
> The NSC assesses proposed new screening programmes against a set of internationally recognised criteria covering the condition, the test, the treatment options and effectiveness and acceptability of the screening programme. Assessing programmes in this way is intended to ensure that they do more good than harm at a reasonable cost. In 1996, the NHS was instructed not to introduce

[27] HC Deb 6 January 2004 c312W

> any new screening programmes until the NSC had reviewed their effectiveness. This was communicated in EL96(110).

The UK Screening Committee, which is concerned with national programmes, uses the following definition of screening:

> …a health service in which members of a defined population, who do not necessarily perceive they are at risk of a disease or its complications, are asked a question or offered a test, to identify those individuals who are more likely to be helped that harmed by further tests or treatment.[28]

In practice tests on people who do not have symptoms are not necessarily undertaken as part of a national screening programme as defined or recommended by the UK National Screening Committee and the word 'screening' may be used in a number of ways. For example, an individual GP may judge that an individual needs a test (or screening) even though an individual does not have symptoms. Testing symptomless people for communicable diseases is also undertaken in certain circumstances for the protection of the public. The National Institute for Clinical Excellence, which issues guidance about a variety of clinical subjects, has in practice included advice about screening as part of its broader guidelines on routine care for the healthy pregnant woman.

The major existing national screening programmes, such as those for breast and cervical cancer, have not been introduced directly by legislation. More generally, patients do not have a "right" to particular treatments. However, in practice there are various methods by which individuals and governments may seek to enforce a policy, not all of which would be relevant to all circumstances or in all countries of the UK. For example, the Government encourages or prioritises policies in various ways depending on the type of policy, such as through the performance assessment framework that monitors Primary Care Trusts, the National Service Frameworks for particular diseases, such as the one for coronary heart disease, and the new GP contract.

None of these policies currently includes screening for sudden cardiac death in young people. In fact the UK National Screening Committee has considered this form of screening and rejected it but is re-examining the evidence (see below). The criteria that it uses for recommending a programme,[29] current pilot studies and its policy positions on a wide range of possible programmes and details of its work are available on its website on and the National Electronic Library for Screening.[30]

The Government's attitude to screening for sudden cardiac death in particular was given in response to several questions about heart disease at the end of 2003. In answer to a

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28 http://www.nsc.nhs.uk/glossary/glossary_ind.htm
29 http://www.nsc.nhs.uk/uk_nsc/uk_nsc_ind.htm
30 http://www.nsc.nhs.uk/ and http://www.nelh.nhs.uk/screening/vbls.html

question about advice given to health professionals about signs and symptoms consistent with underlying cardiac conditions that could cause unexpected death, the Minister said:

> **Miss Melanie Johnson**: Relatives of people who have died of sudden cardiac death, or who are diagnosed with one of the underlying conditions, should be screened because there is a genetic component in many of these cases.[31]

However the Government did not believe that this warranted a national programme. Its reply to a question on the same day about a British Heart Foundation's study into sudden cardiac death[32] is set out below:

> **Miss Melanie Johnson:** The British Heart Foundation (BHF) study suggested that sudden cardiac death should be classed as a separate condition.
>
> We do not believe that sudden cardiac death should be classified as a syndrome in its own right. This is because it has many possible causes – neurological, metabolic or cardiac problems, or an underlying infection.
>
> The UK National Screening Committee has advised there is insufficient evidence to warrant a national screening programme for sudden cardiac death. This recommendation is based on a report produce din 1999 by Dr Stuart Logan of the Institute for Child Health. Dr. Logan has now updated his report in the light of recent research and, after consideration of the report by the Child Health sub-committee, the position remains the same.
>
> The evidence base is due to be re viewed in March 2004.[33]

A similar answer was given a year earlier in January 2002:

> **Yvette Cooper**: the United Kingdom National Screening Committee (NSC) advises Ministers, the devolved National Assemblies and the Scottish Parliament on all aspects of screening policy. The NSC does not currently recommend screening for cardiomyopathy but is keeping its position under review.
>
> Current policy is that the relatives of a family with a known high risk of contracting this disease should receive regular cardiovascular examinations.
>
> The Department is working closely with the medical profession and voluntary organisations with a view to producing clear clinical guidance, which will play a very important part in raising awareness and improving diagnosis and testing of people at risk.

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31 HC Deb 16 December 2003 c882W

32 British Heart Foundation Press Notice 23 March 2003, *Urgent Call For Greater Research Into So-Called Adult "Cot Death" Leading charity asks if mystery adult deaths could be just the 'tip of the iceberg*" http://www.bhf.org.uk/news/index.asp?secondlevel=241&thirdlevel=765&artID=3514

33 HC Deb 16 December 2003 c883W

The UK National Screening Committee's findings on this subject was summarised in its Second Report:

> *5. Hypertrophic Cardiomyopathy*
>
> *5.1 Background*
>
> …Screening could consist of a family and personal history and physical examination by a doctor with a low threshold for proceeding to further investigation either in the general population or among the relatives of people who have developed or died from HCM. The latter activity is not screening as defined by the NSC.
>
> *5.2 Progress*
>
> The NSC considered a preliminary review of the evidence (2) for screening for hypertrophic cardiomyopathy prepared for the Institute of Child Health at its meeting in June 1999. The Committee accepted the conclusions of the report. This stated that the absolute requirements for a possible screening programme were a clear case definition linked to prognosis, and good evidence that intervention can improve the prognosis of the condition if instituted before cases would have been expected to present without screening. Hypertrophic cardiomyopathy fulfils neither of these criteria. The case definition misses many of those at risk and identifies many others whose life span is likely to be no different from the general population. There is currently little evidence that treatment changes the course of the disease if instituted before the onset of symptoms. Screening of adolescents and young people who intend to engage in competitive sport can similarly not be justified at present. Most people identified by the screening process would be individuals who are destined to live a normal lifespan, most of whom would remain asymptomatic at least until much later in life.
>
> *5.3 Current advice*
>
> There is no justification to introduce screening for this condition.[34]

B. The Bill and its supporters

Soon after Dari Taylor presented her Bill to Parliament at the beginning of 2004, she wrote to colleagues saying that she was confident that they would be successful in

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[34] Department of Health et al. UK National Screening Committee Second Report: http://www.dh.gov.uk/assetRoot/04/01/45/60/04014560.pdf

enacting a Bill or, if not, in exerting sufficient pressure on the Department of Health to prevent unnecessary and untimely deaths. She wrote:

> …Some of the case studies are truly shocking: five deaths in one extended family; repeated misdiagnoses as epilepsy or asthma; people found dead in their beds after doctors dismissed their condition as no more than a "nuisance".
>
> If successful, my Bill will enhance the screening process and establish an automatic right to screening for all relatives of people who have died of sudden cardiac death, and all those diagnosed with the symptoms. By raising the profile of sudden cardiac death, the Bill will at least encourage doctors, patients and screening technicians to really take these cases seriously….
>
> ….The ideal outcome is of course to enact the Bill – but public awareness and political pressure could be almost as important. To the extent that the Private Member's Bill raises awareness amongst GPs, cardiologists and families it will have achieved something; if it influences the Department's view of screening in 2004, it will have been a success.

In her letter, Dari Taylor argues that

- Department of Health policy lacks any sense of urgency and does not provide any indication that the condition that could be fatal.
- Its advice is not as widely known as it needed to be, by either doctors or patients.
- The screening process itself is problematic. The technicians and medics who perform the screen have frequently not been trained to look for the conditions that lead to sudden cardiac death.

The letter also criticises conventional cardiovascular screening and describes what she would like to see instead:

> Conventional cardiovascular screening includes blood lipids, blood pressure, auscultation of heart sounds, and occasionally exercise ECG. Generally it does not examine specific diseases that cause sudden cardiac death (SCD) in the young. The causes of SCD in the young are predominantly congenital and familial, therefore specific disease recognition and family screening are imperative in the identification of underlying disease.
>
> A more detailed screening than present would be required, including a comprehensive family history and expert interpretation of ECG and echocardiography. The echo would require imaging of areas that may be outside a routine interrogation including right ventricular structure and function, origins of the coronary arteries and aortic dimensions. Further investigation may be indicated if ECG and/or echo are positive.

Dari Taylor's Bill has the support of Ian Botham, who is Honorary President of the CRY Centre for Sports Cardiology and launched the All Party Parliamentary Group for Cardiac

Risk in the Young, which is also supporting the Bill.[35] Although the precise details of the Bill are not supported by all relevant organisations, there appears to be widespread support for the Bill's general purpose. The Bill is supported by CRY, which has long been campaigning for the introduction of a national programme to screen for cardiac abnormalities in adolescents.[36] The all-party Parliamentary Group on heart disease has also given support and, although not actively involved in this Bill, the British Heart Foundation has shown concern about the issue of "so-called adult 'cot death'" and has published research that suggests that the number of known deaths may represent the tip of an iceberg.[37]

The Bill itself contains four clauses and a schedule:

Clause 1 would place requirements on pathologists carrying out post mortems following the death of a young person. Where the post mortem has been unable to identify the cause of death, or has identified the cause as a cardiac disease that has a genetic basis, the pathologists is required to append a form and statement to the post mortem recommending that first degree relatives of the deceased person be screened to see if they have a cardiac disease. The Clause also requires the GP of the deceased young person, once he has received the post mortem, to attempt to contact first degree relatives of the deceased and invite or recommend that they be screened to see if they have cardiac disease.

Schedule 1 sets out the minimum requirements for the form and statement. It includes a statement that it would be desirable for the parents, children and full siblings of the deceased person to be screened for the condition from which the deceased may have died.

Clause 2 would place requirements on GPs to whom a young person presents him/herself. Where the young person has certain symptoms or has a first degree family member who, at under age 40 years, has or had a cardiac disease with a genetic basis, the GP is required to advise him of the desirability of consulting a relevant specialist. The relevant symptoms are listed a breathlessness disproportionate to activity, palpitations, chest tightness on exertion, and frequent faintness or giddiness.

Clause 3 would place requirements on the relevant specialist. Where a relevant specialist finds that a young person has a cardiac disease that has a genetic basis, the specialist is required to a) advise the young person that it would be desirable for his first degree relatives to consult their GPs, and b) to issue to the young person a letter for his first

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35 Dari Taylor Press Release, *Stars rally behind bill on young cardiac death*, 6 January 2004

36 Further information about CRY's views and the history of its campaigning is on its website: www.c-r-y.org.uk

37 British Heart Foundation Press Notice 23 March 2003, *Urgent Call For Greater Research Into So-Called Adult "Cot Death" Leading charity asks if mystery adult deaths could be just the 'tip of the iceberg*" http://www.bhf.org.uk/news/index.asp?secondlevel=241&thirdlevel=765&artID=3514

degree relatives to give to their GPs if they so wish. The letter must state that a first-degree relative of the GP's patient has a cardiac disease that has a genetic basis and it would be desirable for the patient to be referred to a relevant specialist.

Clause 4 deals with definitions.

Cardiac disease that has a genetic basis is defined as: a cardiac disease that close relatives are likely to suffer from, including but not limited to: hypertrophic cardiomayopathy, coronary artery anomalies, arrhythogenic right ventricular cardiiomyopathy, Long QT/Brugada's syndrome, Marfan's, mitral valve prolapse, Wolfe-Parkinson-White syndrome, dilated cardiommyopathy and premature coronary artery disease.

Relevant specialist is defined as: a person with clinical or academic knowledge of conditions predisposing to arrhythmia and sudden cardiac death in the young.

Young person is defined as: a person under 35 years of age.