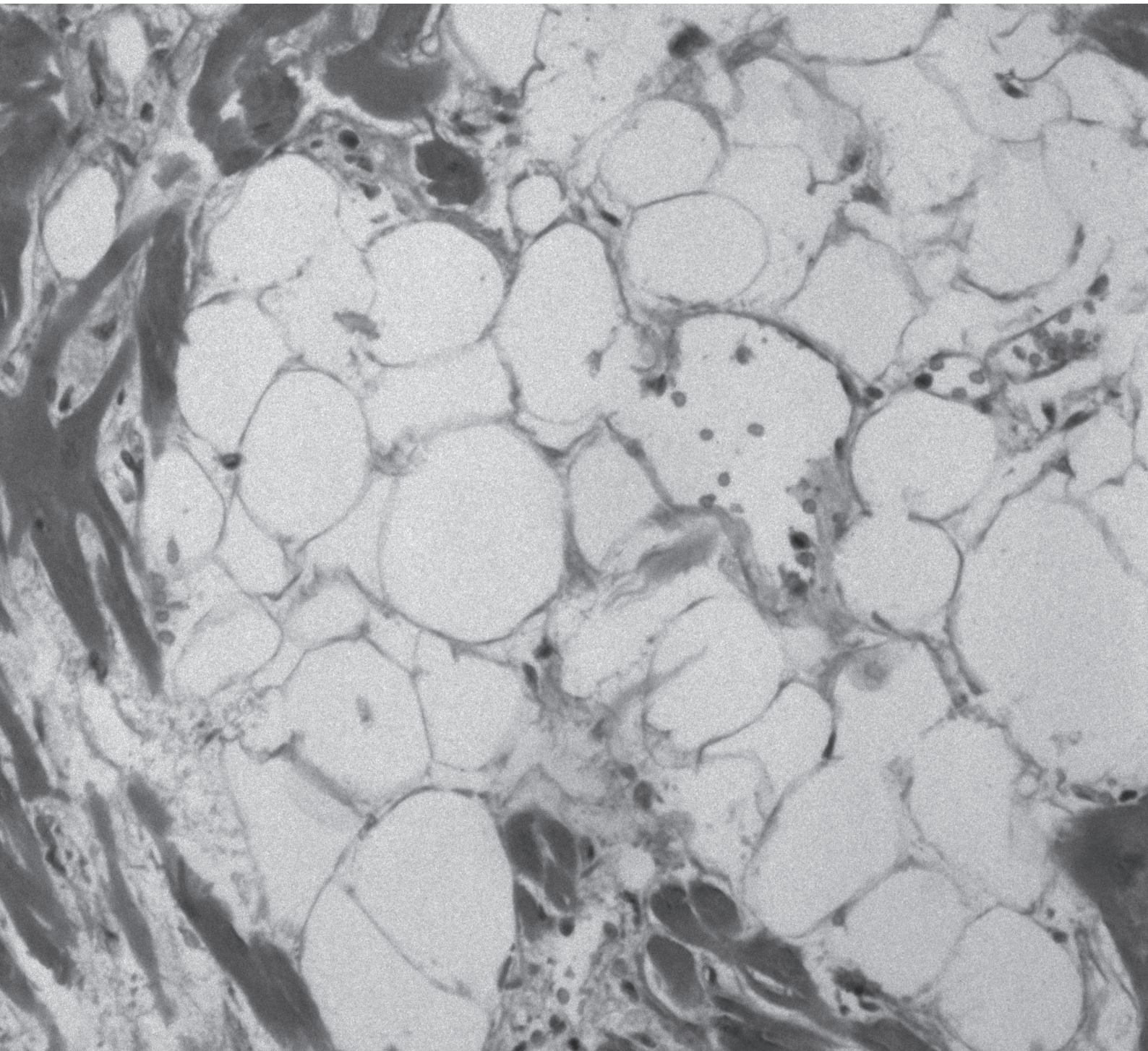
National Audit of Sudden Arrhythmic Death Syndrome 2011



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Prepared in partnership with:

The Healthcare Quality Improvement Partnership (HQIP) promotes quality in healthcare. HQIP holds commissioning and funding responsibility for the National Lung Cancer Audit and other national clinical audits as part of the National Clinical Audit & Patient Outcomes Programme (NCAPOP).

Health and Social Care Information Centre (HSCIC) is England's central, authoritative source of essential data and statistical information for frontline decision makers in health and social care.
The HSCIC managed the publication of the 2011 annual report.

The U.K. Cardiac Pathology Network (UK CPN) is a network of cardiac pathologists throughout England and Wales established to provide local coroners with an expert cardiac pathology service, and for the promotion of best pathological practice in sudden cardiac death cases.

National Audit of Sudden Arrhythmic Death Syndrome 2011

Second annual report:
Key findings from the
National Audit of Sudden
Arrhythmic Death Syndrome

Contents

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Introduction

More than 100,000 people die from cardiovascular disease every year in the UK. In the majority the cause is coronary artery disease or stroke, however in a minority of cases death is caused by less common cardiac diseases, some of which have a genetic basis. Until quite recently these disorders received relatively little priority in health service planning, however the growing realisation that a substantial proportion of cardiovascular disease is caused by genetic mechanisms, and an appreciation of the important role of family screening following a sudden cardiac death, has led to several national initiatives designed to improve cardiovascular genetic services. One in particular, chapter eight of the National Service Framework (NSF) for coronary heart disease, published in March 2005, focused on the issue of sudden cardiac death in young people. An aim of the document was to develop NHS systems that identify family members at risk and provide personally tailored, sensitive and expert support, diagnosis, treatment, information and advice to close relatives. A core component of this initiative was the creation of a national pathology registry that would capture essential epidemiological data and provide a method to audit current coronial and autopsy practice. In response to this report, a voluntary network of interested pathologists and allied professionals, the UK Cardiac Pathology Network (UKCPN), was established.

Together with the Health and Social Care Information Centre (HSCIC), the UKCPN created the first UK audit for sudden cardiac deaths in November 2008. In its first report, the audit presented important data on the characteristics of sudden death cases from across England and Wales. In this second report, we update these data and provide additional information on paediatric deaths and the distribution of deaths by coroner's district.

Data Summary

Data entry since last report

17 centres are registered with the HSCIC. Since the last report, 7 centres entered one or more cases onto the database (table 1). A substantial proportion (38 per cent) was paediatric cases from a single national referral centre. The rate of data entry from some centres was substantially lower than in the previous reporting period. Numbers shown only represent the data submitted and may not reflect all cases seen in centres.

Table 1
Submission of data since first annual report

Hospital	n
Gloucestershire Royal Hospital	7
Great Ormond Street Hospital for Children	13
Northern General Hospital	1
Royal Brompton Hospital	9
Papworth Hospital	2
Southampton General Hospital	1
Whiston Hospital	1
Total	**34**

General statistics

The analysis in this report is derived from data submitted from July 2008 up to and including January 2012. A total of 317 cases are currently recorded in the Sudden Arrhythmic Death Syndrome (SADS) audit. The number of cases reported by each registered centre is shown in table 2. As in the previous report, the majority of cases are from four centres. The data presented in this report focus predominantly on the basic characteristics of the index cases, including the timing and circumstances of death and family history when recorded. The data are also dichotomised into adult (more than or equal to 16 years of age) and paediatric cases (less than 16 years). In the adult cohort, additional data on body mass index and history of drug or alcohol abuse are shown.

Table 2
Data submission by registered centres

Hospital	n
Gloucester Royal Hospital	19
Great Ormond Street Hospital	36
Northern General Hospital	1
Royal Brompton Hospital	175
Papworth Hospital	2
Southampton General Hospital	45
Harefield Hospital	33
St Thomas's Hospital	3
Whiston Hospital	2
Arrowe Park Hospital	1
Addenbrooke's Hospital	0
Leeds General Infirmary	0
North Devon District Hospital	0
Queen Alexandra Hospital	0
Queen Elizabeth Hospital, Edgbaston	0
John Radcliffe Hospital	0
University College Hospital	0
Total	**317**

General demographics

The majority (62 per cent) of cases were male (figure 1). The peak age at death was between 30 and 39 years of age (figure 2).

Figure 1
Sex distribution for all entries

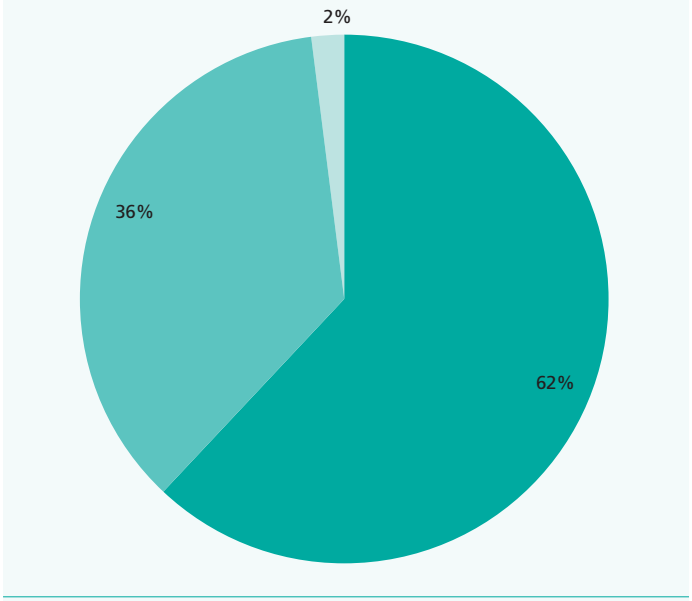
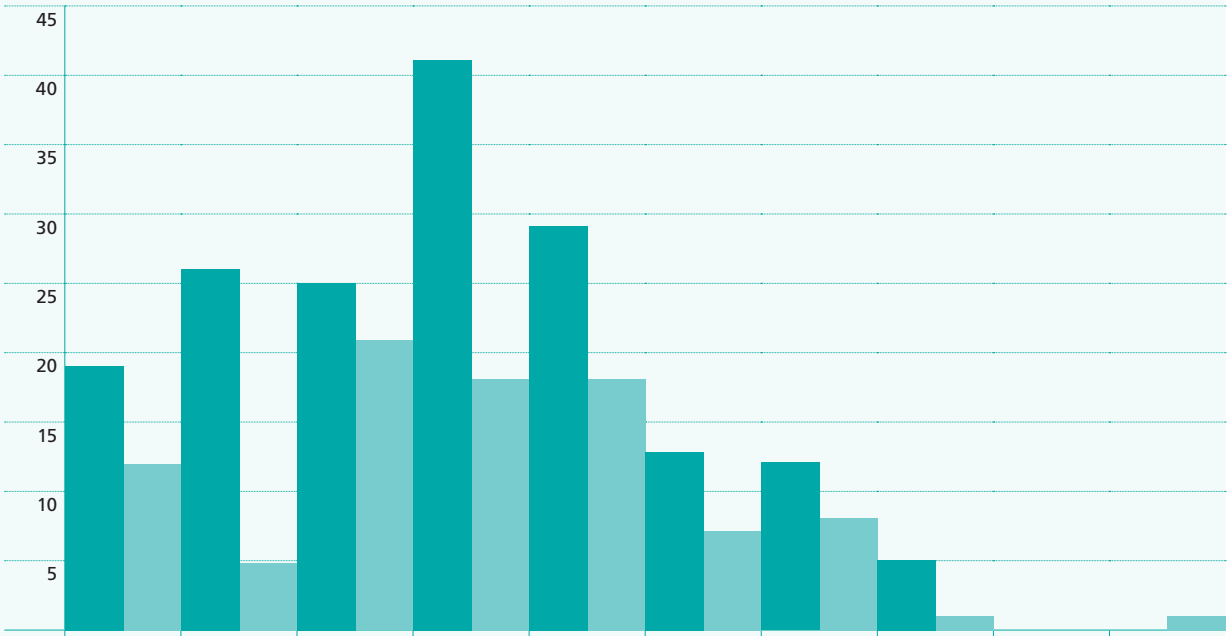


Figure 2
Age at death (years)



	0-9	10-19	20-29	30-39	40-49	50-59	60-69	70-79	80-89	90-99
Male	19	26	25	41	29	13	12	5	0	0
Female	12	5	21	18	18	7	8	1	0	1

Min 0.00 Max 96.71 Average 33.30 Median 33.03

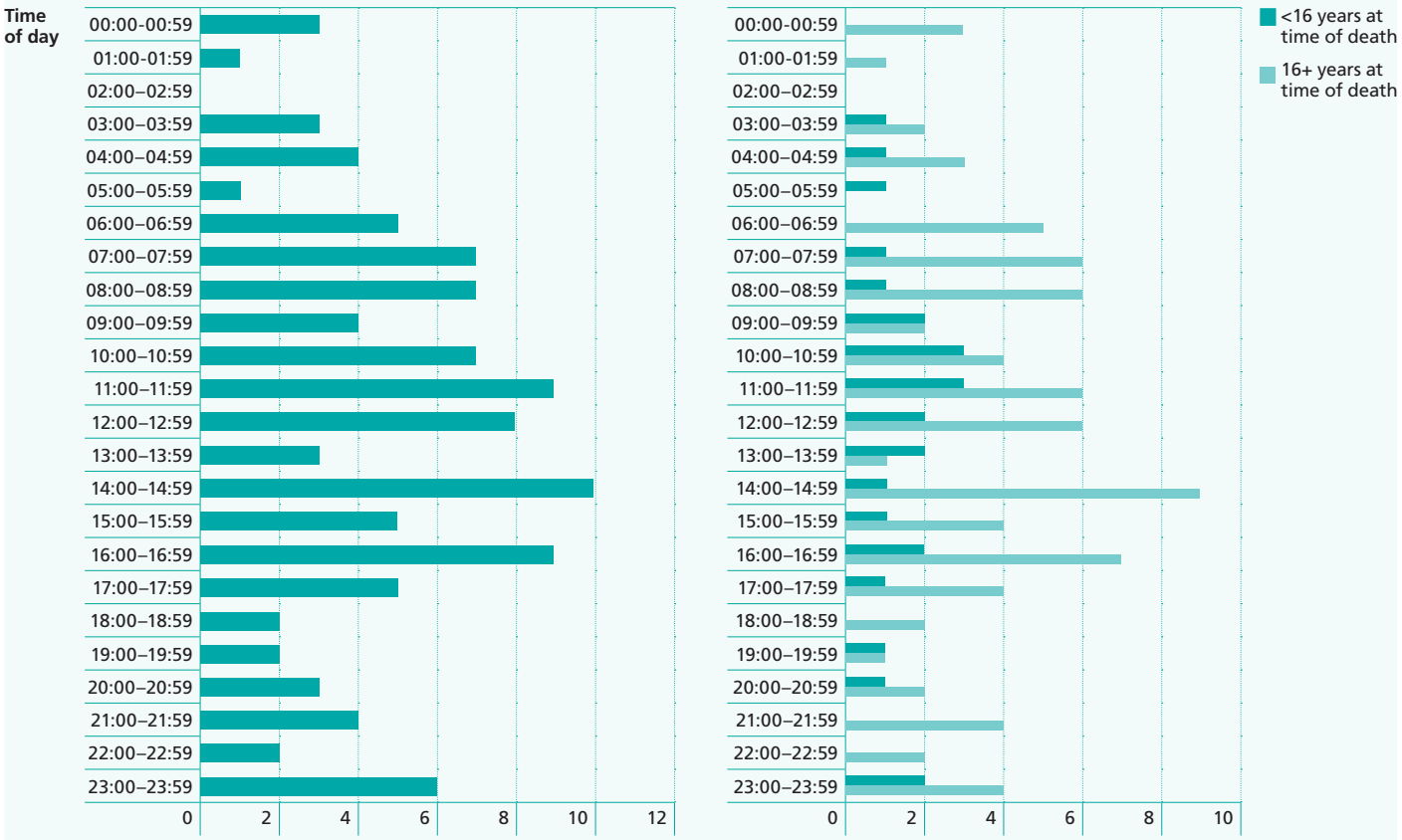
Time and circumstances of death

For the first time we present time and circumstances at time of death.

Crude analysis of time of death shows that in the cohort as a whole, the majority of deaths occurred during waking hours (figure 3). The time of death data were also dichotomised according to age however missing data mean that this analysis could only be completed in 110 patients.

The majority of deaths occurred in bed or at rest (table 3).

Figure 3
Number of deaths by time of day



The left panel shows time of death for entire cohort; the right panel shows time of death dichotomised by age.

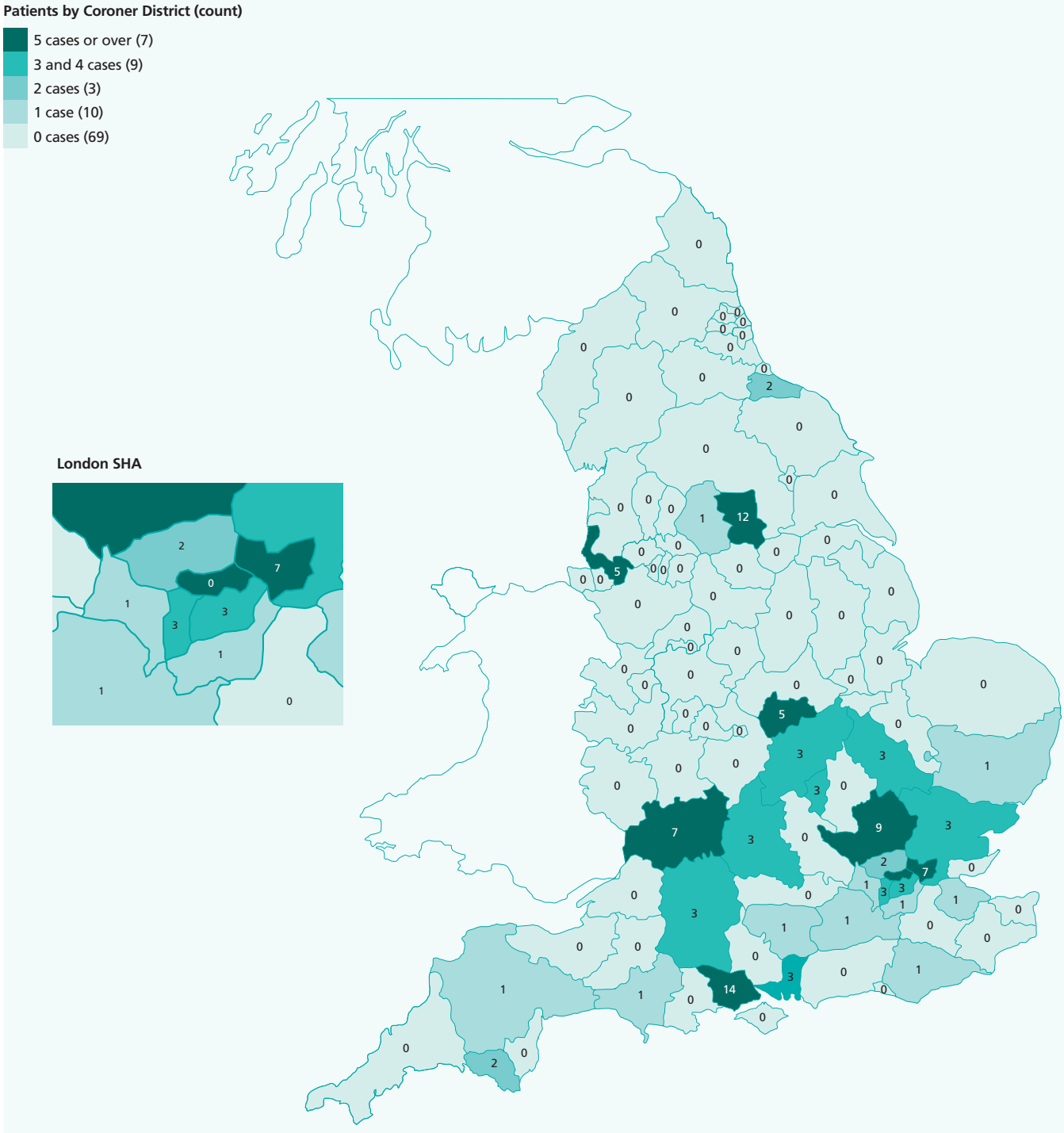
Table 3
Circumstances of death

Hospital	n	%
Died in bed	59	18.6
Died in sleep	22	6.9
Died at rest	104	32.8
Died during exertion	13	4.1
Died during mild exertion	10	3.2
Died during moderate exertion	15	4.7
Died immediately after exertion	7	2.2
Died during severe exertion	9	2.8
Died circumstances other	56	17.7
Died during emotion	6	1.9
Missing	16	2.0
Total	**317**	**100.0**

Distribution of deaths by coroner's district

Data on referring coroner's district were available in 105 cases. The data for the 102 cases where the coroner's district was in England are presented in figure 4. The remaining 3 cases were from the Jersey coroner's district. In none of the cases was the referring coroner's district in Wales.

Figure 4
Distribution of deaths by coroner's district (England)



Toxicology

Toxicological screening (table 4) was recorded in the majority of cases, however no results were noted.

Table 4
Toxicology

	n	%
Yes	183	57.7
No	109	34.4
Unknown	9	2.8
Missing	16	5.0
Total	**317**	**100.0**

Retention of tissue

Tables 5, 6 and 7 show the data on retention of samples for heart, tissue blocks and spleen respectively.

Table 5
Heart retained

	n	%
Yes	67*	21.1
No	235	74.1
Missing	15	4.7
Total	**317**	**100.0**

Table 6
Tissue blocks retained

	n	%
Yes	199**	62.8
No	103	32.5
Unknown	2	0.6
Missing	13	4.1
Total	**317**	**100.0**

Table 7
Spleen retained

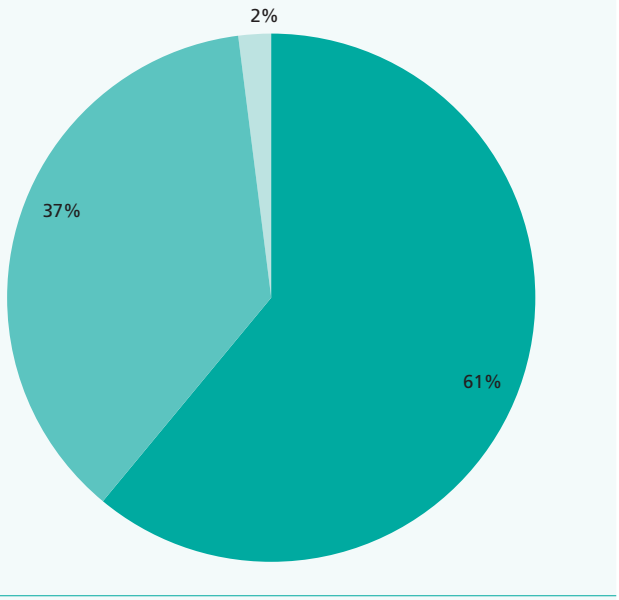
	n	%
Yes	16	5.1
No	80	25.2
Missing	221	69.7
Total	**317**	**100.0**

* Including three paediatric hearts
** Including 35 paediatric cases

Adult Patients

Two hundred and sixty-nine cases (84.9 per cent) were aged 16 years or more at the time of their death. The characteristics of the adult deaths are shown in figures 5 and 6 and tables 8-12. The final diagnosis (ICD code) is shown in table 13.

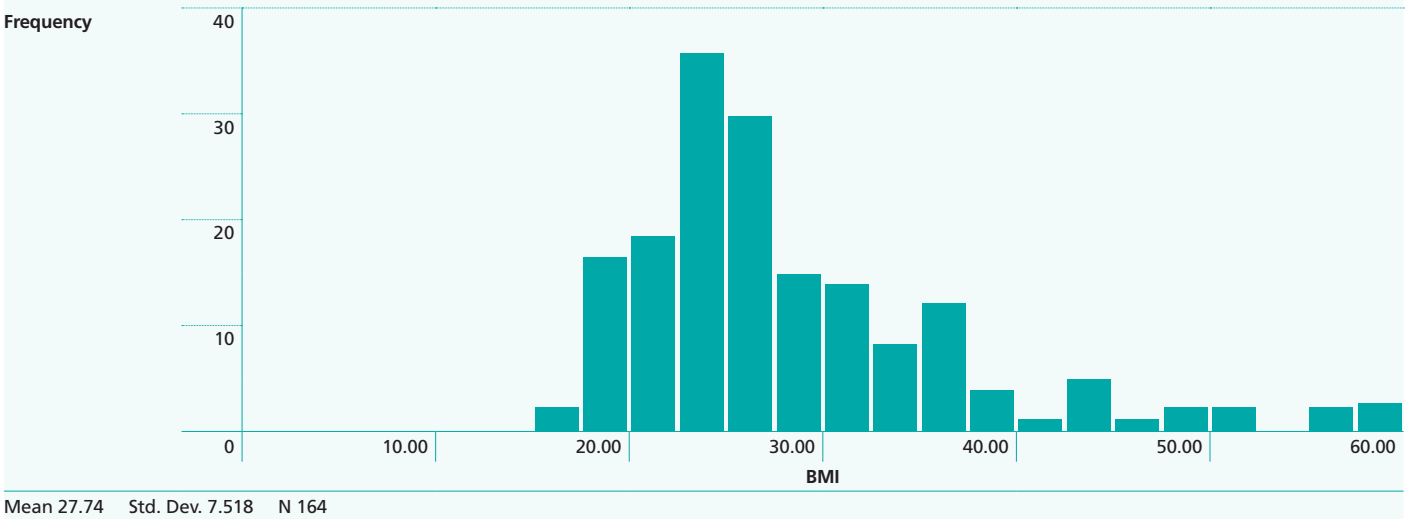
Figure 5
Sex distribution for adult patients



Body Mass Index

Body mass index (BMI) could be calculated from height and weight in 164 adult cases. The distribution of values is shown in figure 6.

Figure 6
Body Mass Index (kg/m²) in adults



Ethnicity

Table 8
Ethnicity

	n	%
White British	103	38.3
Mixed White/black Caribbean	2	0.7
Unknown	4	1.5
Indian	1	0.4
White other	2	0.7
Other ethnic group	1	0.4
Black African	1	0.4
Black Caribbean	3	1.1
Missing	152	56.5
Total	**269**	**100.0**

Co-existing illnesses

Table 9
Co-existing illnesses*

	n
Unknown	83
Neuromuscular	3
Other	60
Postpartum	2
Pregnancy	3
Liver disease other	3
Congenital heart disease	9
Asthma	25
Major depressive illness	1
Schizophrenia	1
Epilepsy	5
None	38
Missing	38

* More than one entry per patient in some cases

Family History

Table 10
Family History

	n
Unknown	158
Other	10
Diabetes	1
Premature SCD	15*
Cardiomyopathy	2
None	38
Missing	43
Arrhythmia	1
Death > 60 years	1
Total	**269**

* Two with additional history of cardiomyopathy, two with history of epilepsy

History of illegal drug and alcohol use

These data were confined to adult cases. In the case of illegal drug use, the drug was not recorded in any case. The amount of alcohol in those patients in whom alcohol use was noted was recorded in only 11 patients.

Table 11
Adults drug use

	n	%
Yes	20	7.4
No	227	84.4
Unknown	14	5.2
Missing	8	3.0
Total	**269**	**100.0**

Table 12
Adults alcohol use

	n	%
Yes-unspecified	38	14.1
Yes- Heavy	7	2.6
Yes-Light	1	0.4
Yes-degree unknown	5	1.9
No	134	49.8
Unknown	53	19.7
Missing	31	11.5
Total	**269**	**100.0**

Cause of death

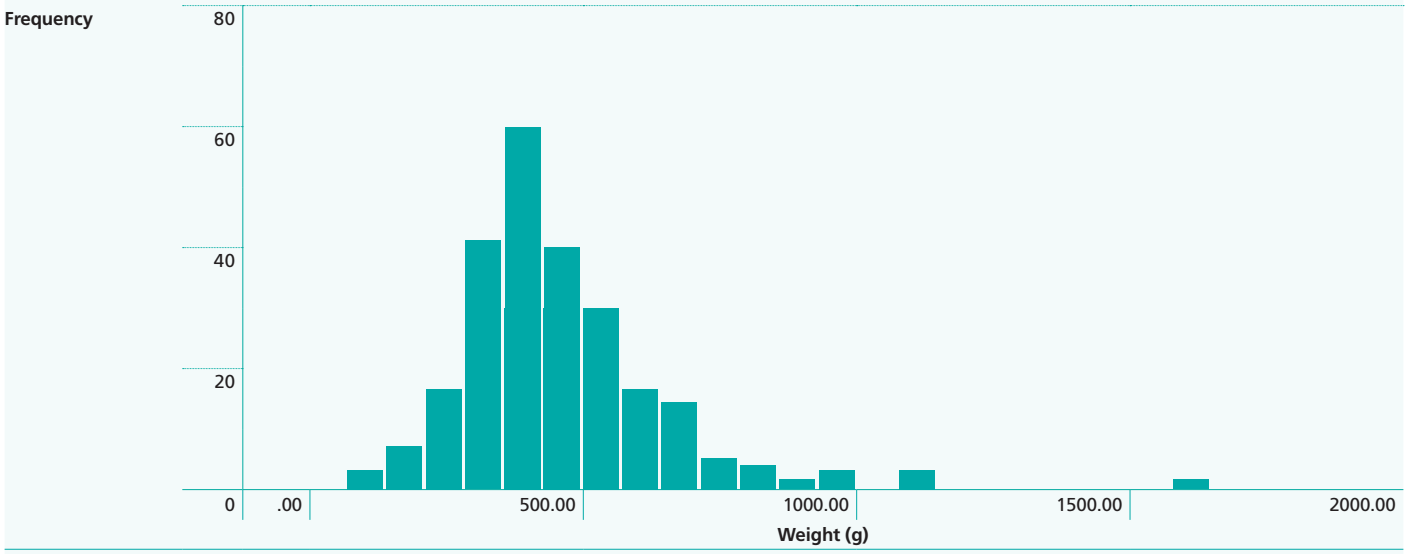
Table 13
Final diagnosis - ICD10 codes

	n
I011. Acute rheumatic endocarditis	1
I050. Mitral stenosis;I35. Nonrheumatic aortic valve disorders	1
I078. Other tricuspid valve diseases	1
I083. Combined disorders of mitral- aortic and tricuspid valves	1
I11. Hypertensive heart disease	1
I110. Hypertensive heart disease with (congestive) heart failure	1
I219. Acute myocardial infarction- unspecified	2
I249. Acute ischaemic heart disease- unspecified	1
I269. Pulmonary embolism without mention of acute cor pulmonale;I270. Primary pulmonary hypertension;I517. Cardiomegaly;I518. Other ill-defined heart diseases	1
I27. Other pulmonary heart diseases	2
I279. Pulmonary heart disease- unspecified	1
I312. Haemopericardium- not elsewhere classified	1
I328. Pericarditis in other diseases classified elsewhere;I426. Alcoholic cardiomyopathy	1
I340. Mitral (valve) insufficiency	1
I341. Mitral (valve) prolapse	1
I358. Other aortic valve disorders;I390. Mitral valve disorders in diseases classified elsewhere	1
I371. Pulmonary valve insufficiency	1
I40. Acute myocarditis	2
I400. Infective myocarditis	1
I401. Isolated myocarditis	2
I418. Myocarditis in other diseases classified elsewhere	1
I42. Cardiomyopathy	6
I420. Dilated cardiomyopathy	13
I421. Obstructive hypertrophic cardiomyopathy	9
I422. Other hypertrophic cardiomyopathy	3
I424. Endocardial fibroelastosis	1
I426. Alcoholic cardiomyopathy	4
I428. Other cardiomyopathies	11
I429. Cardiomyopathy- unspecified	10
I432. Cardiomyopathy in nutritional diseases	2
I438. Cardiomyopathy in other diseases classified elsewhere	1
I447. Left bundle-branch block- unspecified;I45. Other conduction disorders	1
I456. Pre-excitation syndrome	1
I458. Other specified conduction disorders	1
I46. Cardiac arrest	1
I461. Sudden cardiac death- so described	77
I469. Cardiac arrest- unspecified	1
I50. Heart failure	2
I509. Heart failure- unspecified	1
I514. Myocarditis- unspecified	3
I516. Cardiovascular disease- unspecified	5
I517. Cardiomegaly	13
I519. Heart disease- unspecified	1
I71. Aortic aneurysm and dissection	1
I710. Dissection of aorta [any part]	6
I711. Thoracic aortic aneurysm- ruptured	1
I740. Embolism and thrombosis of abdominal aorta	1
I99. Other and unspecified disorders of circulatory system	1
Missing	67
Total	**269**

Heart weight

In this analysis we present the heart weight as recorded in 248 adult cases (figure 7 and table 14).

Figure 7
Heart weight*



Mean 457.62 Std. Dev. 181.351 N 248

* An outlying value (1637g) was noted, however it was not possible for it to be verified.

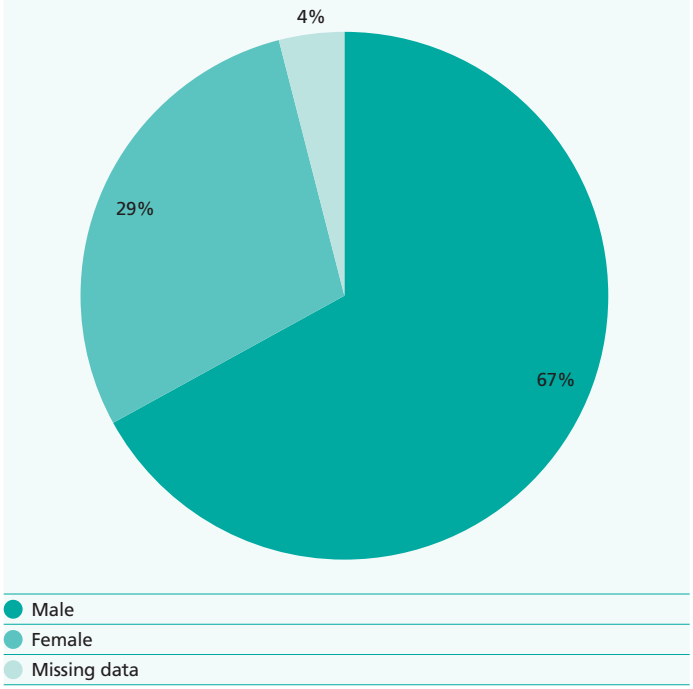
Table 14
Heart weight

	n	%
<400g	102	41.1
400 - 449g	41	16.5
450 - 499g	27	10.9
>500g	78	31.5
Total	**248**	**100.0**

Children and Adolescents

Forty-eight cases were less than 16 years of age at the time of death. The sex and ethnicity are shown in figure 8 and table15. Eleven children (22.9 per cent) had congenital heart disease as a reported coexisting illness (table 16) and three had a family history of sudden death (table 17).

Figure 8
Sex distribution for paediatric cohort



Ethnicity

Table 15
Ethnicity in children

	n	%
White (British)	24	50.0
Black African	3	6.2
Pakistani	3	6.2
Unknown	2	4.2
Other	1	2.1
Missing data	15	31.2
Total	**48**	**100.0**

Co-existing illnesses

Table 16
Co-existing illnesses in children

	n
Congenital Heart Disease	11
Epilepsy	1
Asthma	3
None	15
Other	6
Unknown	13

Family History

Table 17
Family History of Sudden Death in children

	n	%
Yes	3*	6.3
None	5	10.4
Unknown	39	81.3
Missing	1	2.0
Total	**48**	**100.0**

* Two at age of 35 years; one aged 35-65 years

Cause of death

Table 18
Final Diagnosis - ICD10 codes

	n
I24. Other acute ischemic heart diseases	1
I249. Acute ischaemic heart disease- unspecified	1
I270. Primary pulmonary hypertension	1
I342. Non rheumatic mitral (valve) stenosis;I350. Aortic (valve) stenosis	1
I348. Other non-rheumatic mitral valve disorders;I37. Pulmonary valve disorders	1
I350. Aortic (valve) stenosis	1
I37. Pulmonary valve disorders	2
I370. Pulmonary valve stenosis	1
I372. Pulmonary valve stenosis with insufficiency	1
I40. Acute myocarditis	3
I42. Cardiomyopathy	2
I420. Dilated cardiomyopathy	2
I421. Obstructive hypertrophic cardiomyopathy	2
I422. Other hypertrophic cardiomyopathy	2
I429. Cardiomyopathy - unspecified	1
I431. Cardiomyopathy in metabolic diseases	1
I46. Cardiac arrest	1
I461. Sudden cardiac death- so described	10
I490. Ventricular fibrillation and flutter	1
I514. Myocarditis- unspecified	2
I516. Cardiovascular disease- unspecified	1
I517. Cardiomegaly	1
Not recorded	8
Total	**48**

Notes:
One additional case coded as "I058. Other mitral valve diseases; I080. Disorders of both mitral and aortic valves;I350 Aortic valve stenosis; I390 Mitral valve disorders in diseases classified elsewhere."

Coronary arterial anomalies (including fistula to the right ventricle, and dysplasia) were reported in four individuals.

Key Points

Progress since the first annual report

The total number of cases entered since the last report was smaller than expected and far below that expected based on current population estimates. Following publication of the first report, all trusts in England and Wales were contacted to inform them of the audit and to encourage participation by their local pathology services. The informal feedback that was received by the HSCIC suggested that there are a number of perceived barriers to data collection:

- **Lack of clear responsibility for the capture of data relating to sudden cardiac deaths.**
  The audit team received a number of communications from lead cardiologists expressing concern that they were unaware of the audit, however had been asked to respond to the request for information by their chief executives. Following establishment of the national audit, it was publicised through the Royal College of Pathologists, the UKCPN website and the HSCIC. Given the focus on pathological data pertinent to sudden cardiac death cases, the original target specialists were general and cardiac pathologist rather than cardiologists. Over the next period, further work will be necessary to understand the role of pathology services in local cardiac networks and to identify local champions for the national audit.

- **Priority**
  A number of trusts and professionals expressed the view that as participation is voluntary it is of lower priority than other national data collections. Also, as the project is funded as a registry, it will not appear in lists (e.g. NCAPOP, Quality Accounts) of national projects identified as compulsory or high priority for providers.

- **Resources**
  Many pathologists expressed a concern about the human resources required to capture and enter data. This is a common barrier to participation in all national audits, however is one that has been overcome with much larger national data collections. Further discussions with individual NHS services may be required to understand this limitation.

- **Data protection**
  A number of centres expressed concern about the legality of recording confidential patient level data in the audit and cited this as a reason for non-participation. This audit has section 251 approval from the National Information Governance Board, which allows collection of patient identifiable data without explicit patient consent.

Data trends

At this early stage of the audit, detailed analyses of the data are inappropriate. Nevertheless, some trends have emerged. These include a male predominance in adults and children, a peak in age at death between 30-39 years of age, and the fact that very few deaths are related to moderate or severe exertion.

Comorbidities were common, although their significance in relation to final cause of death requires further analysis. Congenital heart disease was common in childhood deaths.

The recording of a family history of sudden cardiac death is a clear deficiency in the audit. Previous studies in relatives of sudden death victims have shown that a history of sudden death is relatively common. The fact that the overwhelming majority of entries for this field were unknown or missing probably reflects the fact that pathologists performing the heart examinations frequently have no direct contact with family members and have to rely on information provided to them by other parties.

Use of illegal drugs and alcohol are an important potential contributor to sudden death. A known history of drug use was reported in less than 10 per cent of cases; the picture with respect to alcohol use was less clear. As with family history, it is likely that pathologists face an even greater problem determining the likely extent of prior drug and alcohol intake. Assessment of drug use is assisted by the performance of a toxicology screen (performed in almost 60 per cent of cases) however the results were not specifically recorded in the audit.

Retention of tissue and spleen for subsequent pathological review and DNA testing are invaluable in the evaluation of the risks faced by living relatives of sudden death victims. The human tissue act is frequently cited as an impediment to the retention of tissue, however the audit shows that retention of tissue blocks from the heart is feasible. Retention of spleen on the other hand is more problematic and was recorded in only 5 per cent of cases.

The registry contains a considerable number of fields related to the detailed pathological examination of the heart. These data are not presented in this report, with the exception of heart weight. A striking finding in this study is the relatively high proportion of patients with increased heart mass. This may reflect the underlying pathology of the heart (i.e. the presence of a cardiomyopathy) and comorbidities such as hypertension and obesity. A more detailed analysis of this observation will be conducted in future reports.

Future aims

Collection of data in sudden death victims is a particular challenge as the initial evaluation of sudden death victims take place under the jurisdiction of the Coronial service and not the NHS. However, failure to gather data at this stage can substantially hamper the effort to counsel, diagnose and treat the relatives of sudden death victims who are subsequently referred to NHS clinics for assessment. The

SADS audit represents an attempt to bridge this gap between the legal requirements of coronial autopsy and the needs of surviving family members.

The aims for the next data collection period include:

- **Encourage greater participation of centres in other regions of England and Wales through determination of the barriers to data collection and entry and engagement of professional societies**
- **Enhance geographic analyses to determine variation in coronial and pathology practice**
- **Review database fields to exclude extraneous data fields and to identify areas of interest not covered in present data-set**
- **Secure long-term funding of database and national audit status**

The NHS definition of an audit is: *a quality improvement process that seeks to improve patient care and outcomes through systematic review of care against explicit criteria and the implementation of change*.

(http://www.nice.org.uk/media/796/23BestPracticeClinicalAudit.pdf).

Most national audits including the cancer audits and some of the heart disease audits began with the collection of baseline information on current practice with a view to setting explicit criteria/standards. This is true for SADS, as there are no current standards. The intention of the SADS register is to collect baseline information and thereby facilitate standard setting. The purpose of the SADS audit is to improve patient care and to implement change as a result of the audit.

Health and Social Care Information Centre (HSCIC) is working to make information more relevant and accessible to the public, regulators, health and social care professionals and policy makers, leading to improvements in knowledge and efficiency.

The HSCIC is a special NHS health authority that collects, analyses and distributes data to reduce the burden on frontline staff, releasing more time for direct care.

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