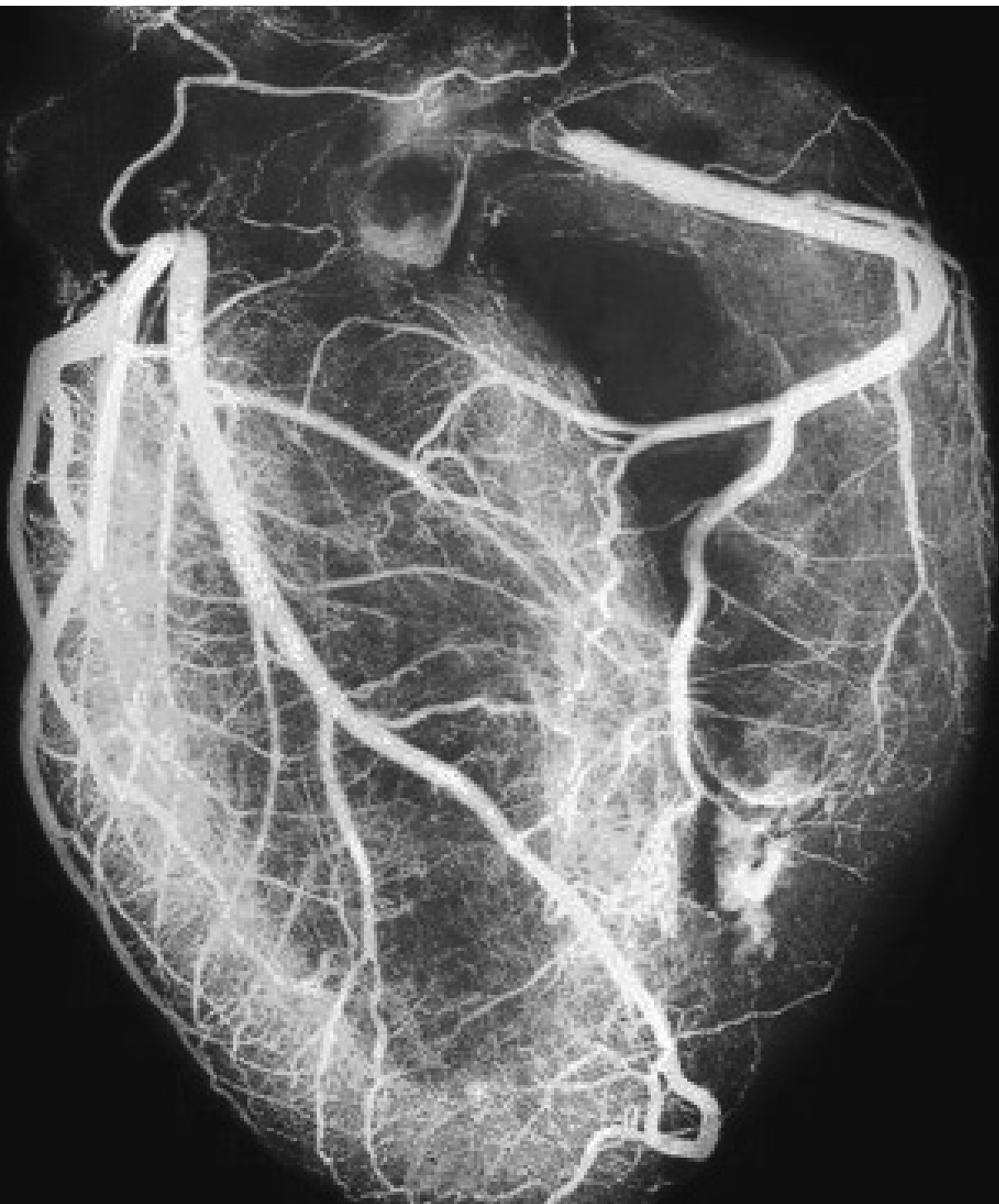


National Audit of Sudden Arrhythmic Death Syndrome 2012



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The Healthcare Quality Improvement Partnership (HQIP) is led by a consortium of the Academy of Medical Royal Colleges, the Royal College of Nursing and National Voices. Its aim is to promote quality improvement, and in particular to increase the impact of clinical audit in England and Wales. HQIP's work includes hosting the Department of Health contract to manage and develop the National Clinical Audit and Patient Outcomes Programme (NCAPOP). The programme comprises more than 30 clinical audits that cover care provided to people with a wide range of medical, surgical and mental health conditions.



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.



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 2010 and 2011 annual reports.

National Audit of Sudden Arrhythmic Death Syndrome 2012

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

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Introduction

This is the third annual report from the National Audit of Sudden Arrhythmic Death Syndrome (SADS). The SADS audit database was designed and launched by the UK Cardiac Pathology Network (UKCPN), in collaboration with the Health and Social Care Information Centre (HSCIC), under the direction of Dr Perry Elliott and Dr Patrick Gallagher. Its realisation is a testament to their hard work and dedication and the importance that we attach to sudden cardiac deaths in young adults. I would like to convey my thanks to them both for their work and continuing support and mentorship. For the database to be a useful tool to collate data from sudden deaths, to understand the prevalence of these conditions and to foster opportunities for research and audit, it is important that data are collected from as many cases as possible.

Sudden cardiac deaths will form part of the work of all pathologists with an autopsy practice. Limitations on time and resources for autopsy practice place increasing pressure on pathologists. The UK Cardiac Pathology Network aims to support all colleagues by providing support in the investigation of sudden cardiac deaths. It is not within the capacity of the network to examine every heart and therefore, most importantly, the network leads on education through meetings to update colleagues and advise on best practice. The network also provides a second opinion on individual cases either through the submission of blocks and slides or whole hearts for specialist examination.

The retention of tissue at autopsy remains a difficult issue, with overlapping needs of the coroners and future needs of the family for possible genetic testing. This matter was the subject of a meeting held at the Royal Society of Medicine and led by Dr Mary Sheppard from the Brompton Hospital, London. The meeting included representation of coroners, the Human Tissue Authority (HTA), cardiologists, geneticists and pathologists and has resulted in guidance being published in the Royal College of Pathologists Bulletin in October 2012.¹

Dr Martin Goddard
Chair, UK Cardiac Pathology Network

Data summary – all cases

Data entry since last report

Table 1 shows the numbers of cases entered into the database since the production of the last report.² For each centre, the number shown only represents the data submitted and may not reflect all cases seen.

Table 1
Submission of data 1 February 2012 - 31 January 2013

Hospital	n
Gloucestershire Royal Hospital	1
Great Ormond Street Hospital for Children	10
Southampton General Hospital	36
Total	47

Although the overall number of cases submitted is higher this year than last, when 34 new cases were added to the database, the number of centres submitting data is low. A number of factors may have contributed to this situation and consideration is given to these in the summary at the end of this report.

General statistics

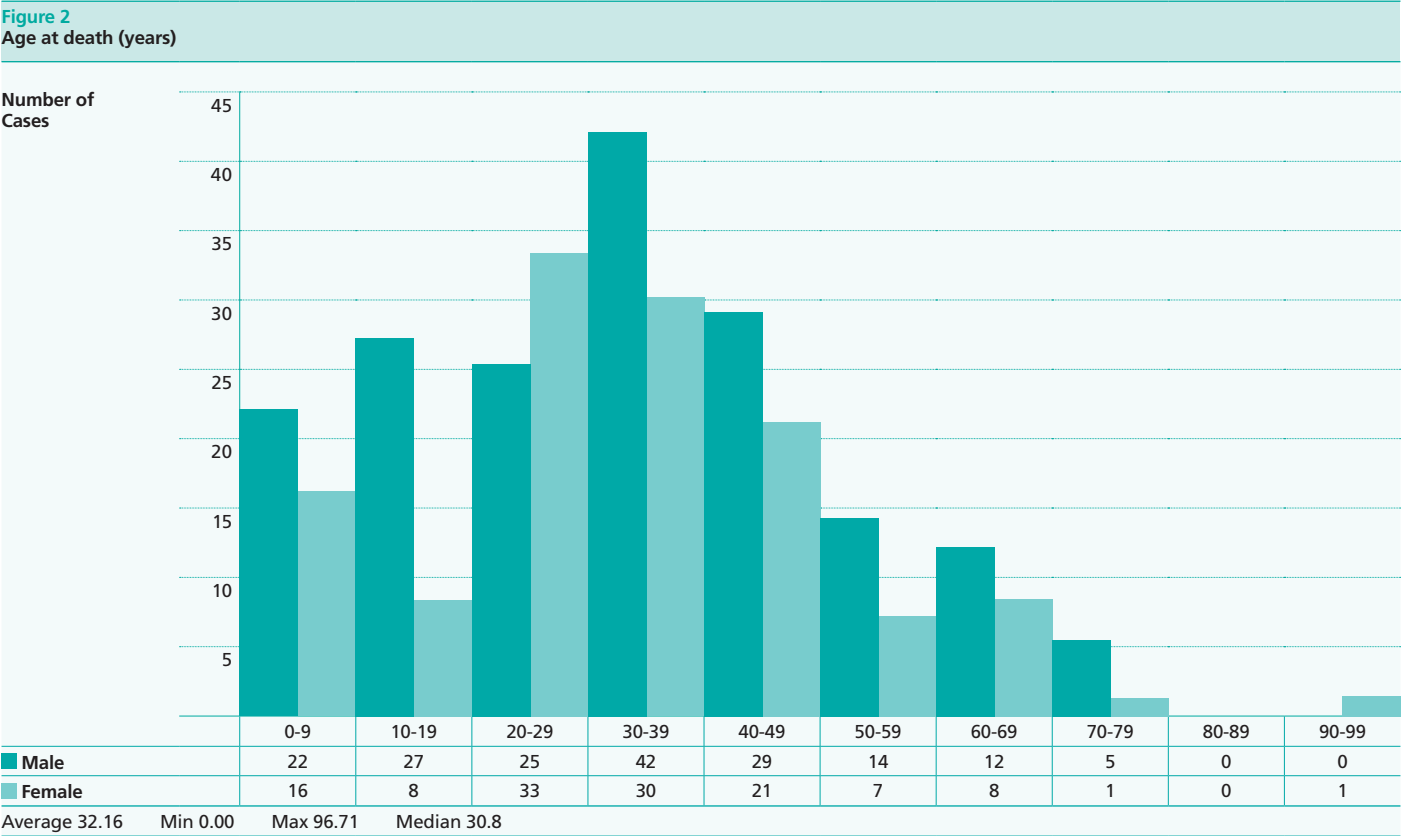
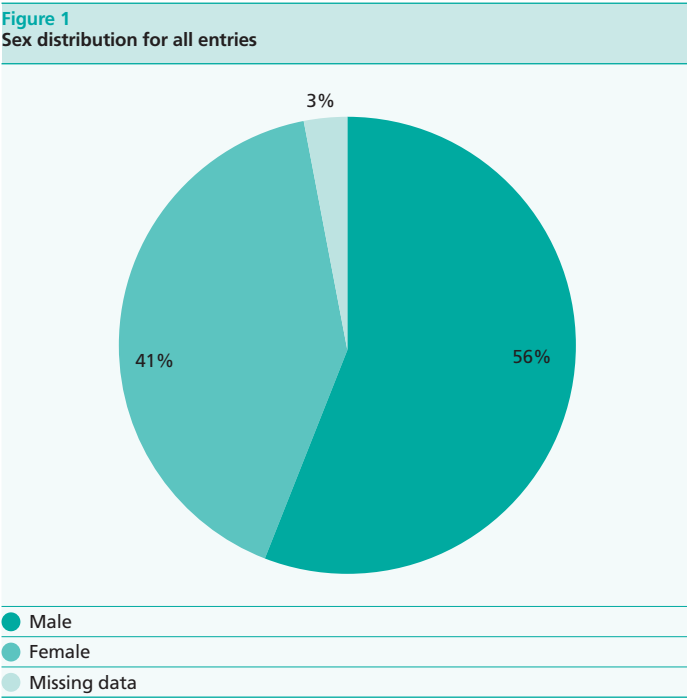
The analysis presented in this report is based on the 364 cases currently recorded in the database, which represent all submissions from July 2008 up to and including January 2013. The number of cases reported by each registered centre is shown in table 2. The number of registered centres (17) remains the same as last year.

Table 2
Data submission by registered centres

Hospital	n
Gloucester Royal Hospital	21
Great Ormond Street Hospital	46
Northern General Hospital	1
Royal Brompton Hospital	175
Papworth Hospital	2
Southampton General Hospital	45
Harefield Hospital	33
St Thomas's Hospital	39
Whiston Hospital	1
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	364

General demographics

The demographic data (Figures 1 and 2) shows the majority of cases being entered into the database are below the age of 50 with a male predominance. This is the expected distribution for non-coronary deaths and it is reassuring that the appropriate case mix is being added to the database.

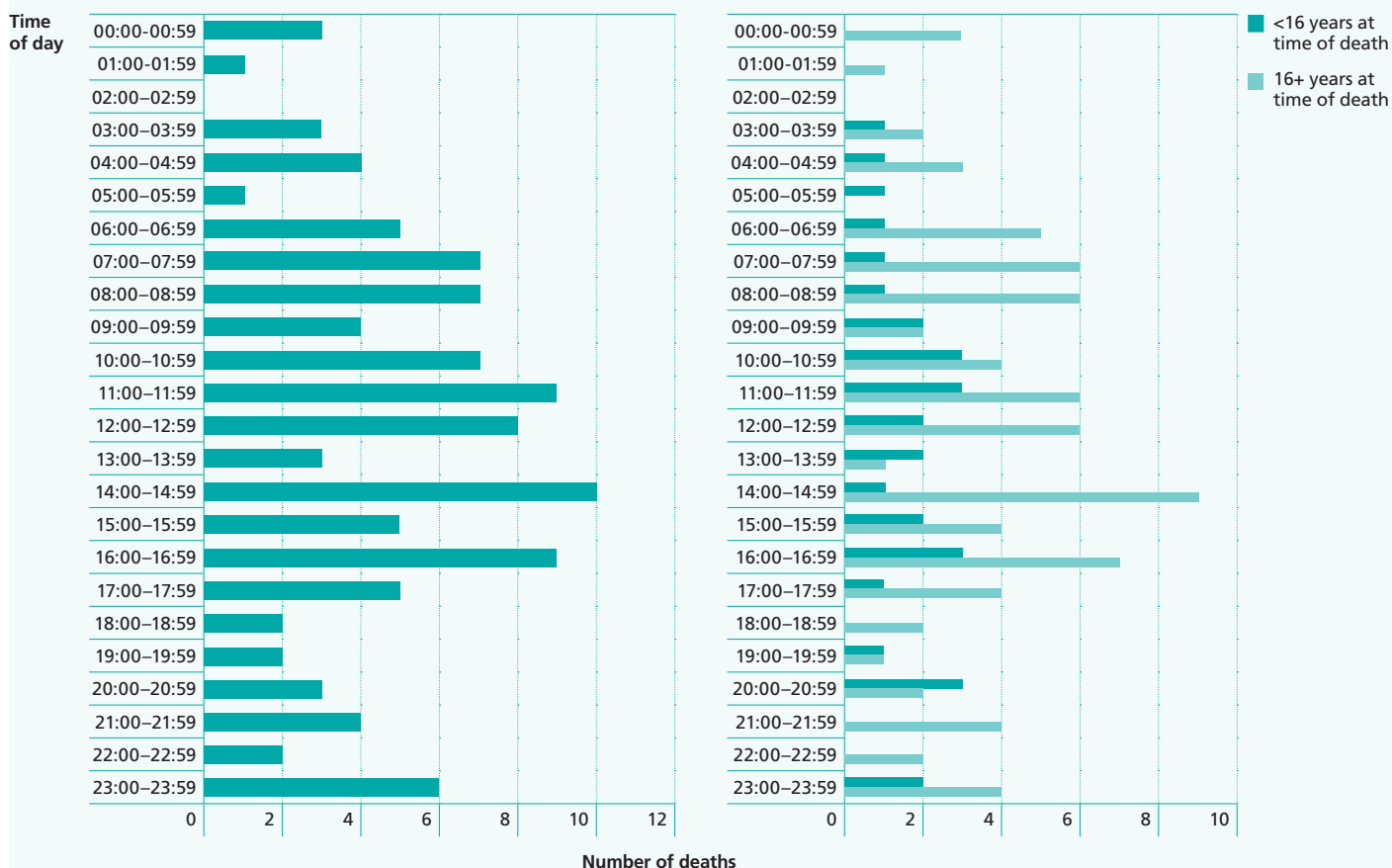


Time and circumstances of death

Figure 3 presents the number of deaths by time of day, firstly as a single group then in two groups according to age at death. Table 3 presents data on the circumstances of death.

Figure 3

Number of deaths by time of day



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

Table 3
Circumstances of death

	n	%
Died in bed	59	16.2
Died in sleep	30	8.2
Died at rest	115	31.2
Died during exertion	15	4.1
Died during mild exertion	10	2.7
Died during moderate exertion	15	4.1
Died immediately after exertion	7	1.9
Died during severe exertion	9	2.5
Died circumstances other	81	22.3
Died during emotional stress	6	1.6
Missing	17	4.7
Total	364	100.0

The SADS group of sudden cardiac deaths is unheralded and often occurs at rest or during sleep. Pathologically, the heart is morphologically normal and these often represent the group of cases most commonly referred for a second opinion, as much to reassure that no other significant pathology has been missed. These may on clinical parameters be slightly over represented in the cohort because of their referral and hence more likely to be put into the database.

Nonetheless, as this group of deaths may be inheritable in more than 50 per cent of cases and related to channelopathies, it further demonstrates that the database is being used to collect data on the appropriate cases.

Accessibility – distribution of deaths by coroner's district

Table 4 presents the distribution of deaths by referring coroner's district for the 145 cases where this information was recorded.

Table 4
Distribution of deaths by coroner's district

Coroner's district	n	Coroner's district	n
Berkshire	2	Mid Kent and Medway	2
Bournemouth, Poole and Eastern Dorset	2	Milton Keynes	3
Cheltenham	5	North East Hampshire	1
East Sussex	1	Northamptonshire	3
Essex No. 1	5	Oxfordshire	3
Exeter and Greater Devon	1	Pembrokeshire	1
Gloucester	3	Plymouth and South West Devon	2
Greater Suffolk	1	Portsmouth and South East Hampshire	3
Hertfordshire	12	South and West Cambridgeshire	4
Jersey	3	South Yorkshire (Western)	1
Leicester City and South Leicestershire	5	Southampton and the New Forest	14
London Eastern	10	St. Helen's and Knowsley	5
London Inner North	1	Surrey	1
London Inner South	15	Teesside	2
London Inner West	8	West Yorkshire (Eastern)	12
London Northern	4	West Yorkshire (Western)	1
London Southern	4	Wiltshire and Swindon	3
London Western	2	Total	145

There is a wide distribution of coroner's districts from which referrals have been received. Given the relatively small number of centres that have entered cases, this may reflect under-reporting in certain areas. However, if the distribution reflects the referral patterns for second opinions, then some areas may not be utilising the availability of the cardiac network. This may be down to local pathologists' practice or due to the practice of the local coroner and the process of obtaining permission to send organs or slides and blocks away. The data field is incomplete in many cases but this issue warrants further investigation.

Toxicology

In all sudden cardiac deaths, particularly where the heart is morphologically normal, best practice is for toxicology screening to be undertaken. The use of recreational drugs in young people can be associated with sudden death and the relevant history may not always be obtained.

Toxicology screening was undertaken in just over 50 per cent (table 5) of cases with a nearly 20 per cent positive rate in the 57 cases where the outcome of screening was recorded (table 6). Without looking at these cases on an individual basis it is not possible to say whether the toxicological findings were contributory to death.

Table 5
Toxicology

	n	%
Yes	192	52.7
No	147	40.4
Unknown	9	2.5
Missing	16	4.4
Total	364	100.0

Table 6
Toxicology results

	n	%
Negative	46	80.7
Positive	11	19.3
Total	57	100.0

Retention of tissue

Slides and blocks were retained in approximately 66 per cent of cases (table 7), whilst the whole organ was retained in just 18 per cent (table 8).

Table 7
Tissue blocks/slides retained

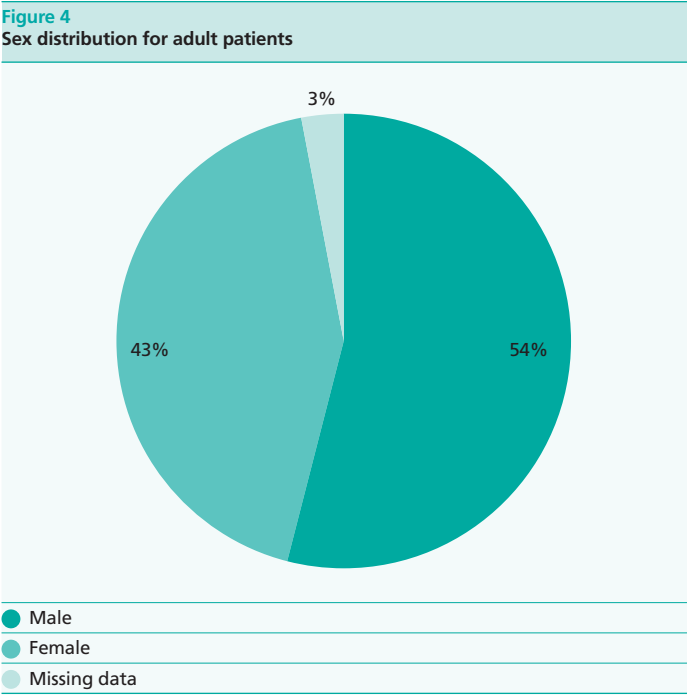
	n	%
Yes	241	66.2
No	104	28.6
Unknown	3	0.8
Missing	16	4.4
Total	364	100.0

Table 8
Heart retained

	n	%
Yes	67	18.4
No	281	77.2
Missing	16	4.4
Total	364	100.0

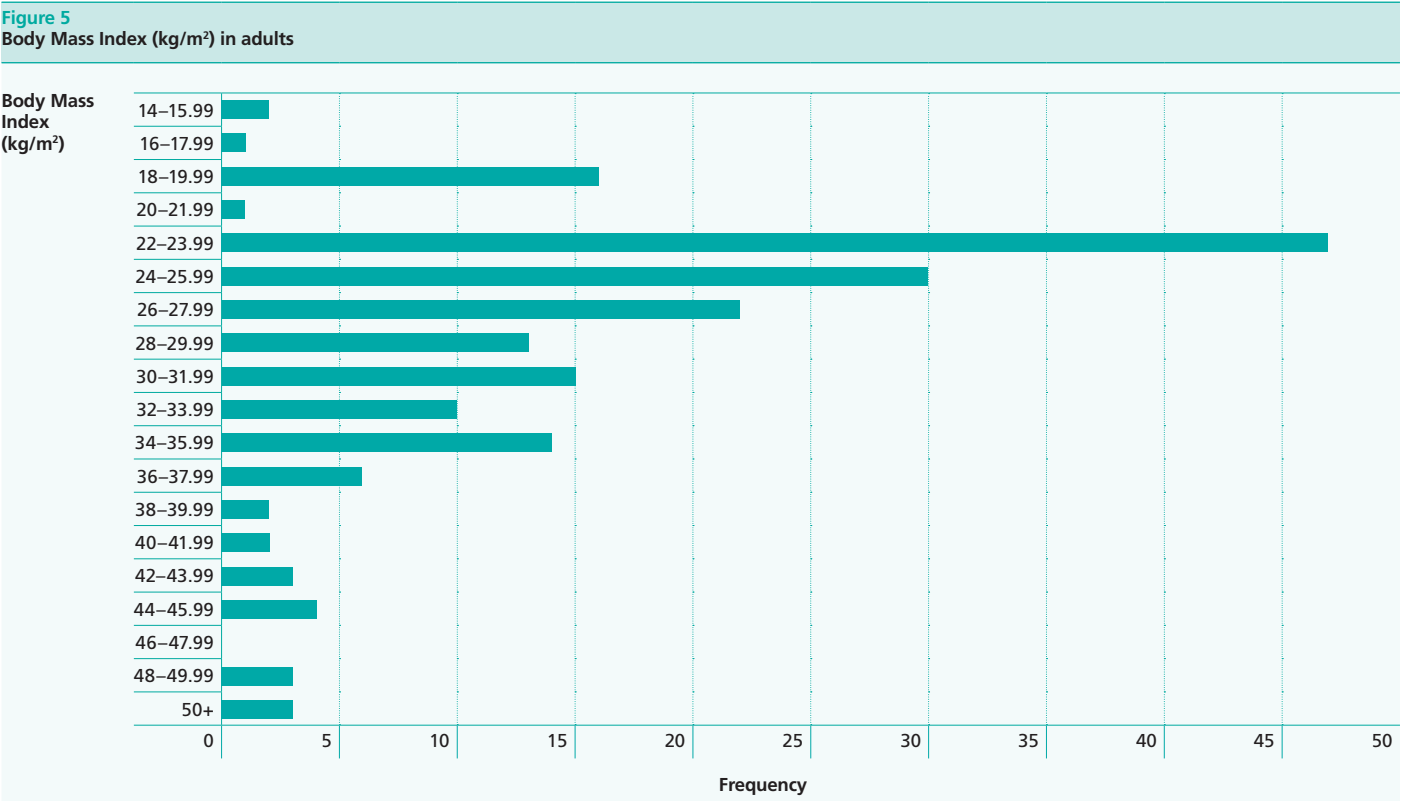
Adults

Three hundred and six cases (84.1 per cent) were aged 16 years or more at the time of their death. The characteristics of the adult deaths are shown in figures 4 and 5 and tables 9,10,11,12,13,14 and 15. The final diagnosis (ICD-10 code) is shown in table 14.



Body Mass Index

Body Mass Index (BMI) could be calculated from height and weight in 194 adult cases (figure 5). The mean value was 28.12



Ethnicity

Table 9
Ethnic group

	n	%
White (British)	122	39.9
Black Caribbean	3	1.0
Black African	1	0.3
Other Ethnic Group	1	0.3
White (other)	2	0.7
Mixed White/Black Caribbean	2	0.7
Mixed White/Black African	18	5.9
Indian	1	0.3
Unknown	4	1.3
Missing	152	49.7
Total	306	100.0

Co-existing illnesses

Table 10
Co-existing illnesses*

	n
None	40
Pregnancy	20
Epilepsy	7
Schizophrenia	2
Major depressive illness	1
Asthma	21
Liver disease alcohol related	1
Congenital heart disease	11
Liver disease other	2
Sickle Cell	1
Post partum	13
Other	72
Neuromuscular disease	3
Unknown	83
Missing	39

* More than one entry per patient in some cases

Family history

Table 11
Family history

	n
None	74
Premature sudden death	12
Cardiomyopathy	2
Death age 35-60	3
Arrhythmia	1
Death age > 60	1
Diabetes, Other	1
Other	10
Unknown	159
Missing	43
Total	306

History of illegal drug and alcohol use

Table 12
Alcohol use

	n	%
No	169	55.2
Yes - unspecified	38	12.4
Yes - degree unknown	5	1.6
Yes - light use	1	0.3
Yes - heavy use	7	2.3
Yes - recent withdrawal	1	0.3
Unknown	53	17.3
Missing	32	10.5
Total	306	100.0

Table 13
Illegal drug use

	n	%
No	261	85.3
Yes	21	6.9
Unknown	15	4.9
Missing	9	2.9
Total	306	100.0

Heart weight

Table 14
Heart weight

	n	%
<400g	122	43.4
400 - 449g	47	16.7
450 - 499g	29	10.3
>500g	83	29.5
Total	281	100.0

The distribution of results for cause of death in adult patients (table 15) largely reflects the distribution of diagnoses from other published national studies. There is a high proportion of sudden arrhythmic death syndrome cases together with a number of cardiomyopathies.

Cause of death

Table 15
Final diagnosis - ICD10 codes

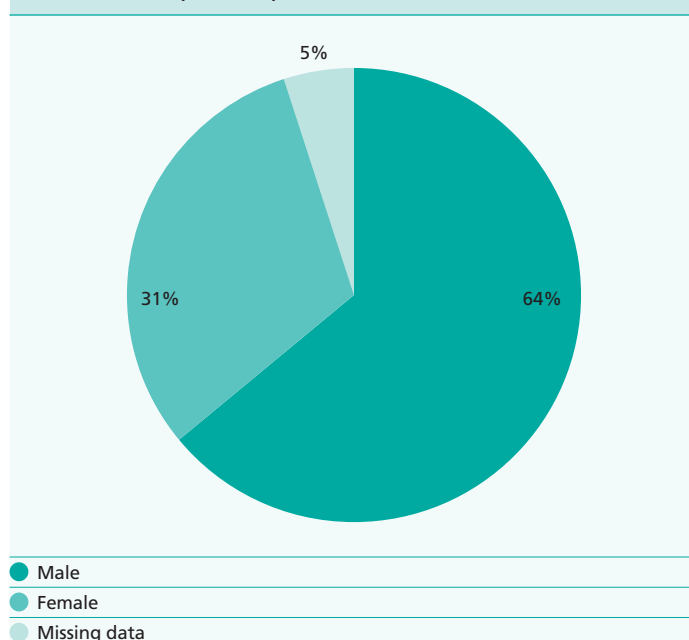
	n
I011. Acute rheumatic endocarditis	1
I050. Mitral stenosis	1
I078. Other tricuspid valve diseases	1
I083. Combined disorders of mitral- aortic and tricuspid valves	1
I11. Hypertensive heart disease	2
I110. Hypertensive heart disease with (congestive) heart failure	1
I152. Hypertension secondary to endocrine disorders	1
I21. Acute myocardial infarction	2
I219. Acute myocardial infarction- unspecified	2
I249. Acute ischaemic heart disease- unspecified	1
I269. Pulmonary embolism without mention of acute cor pulmonale	1
I27. Other pulmonary heart diseases	2
I278. Other specified pulmonary heart diseases	1
I279. Pulmonary heart disease- unspecified	2
I312. Haemopericardium- not elsewhere classified	1
I328. Pericarditis in other diseases classified elsewhere	1
I340. Mitral (valve) insufficiency	1
I341. Mitral (valve) prolapse	1
I358. Other aortic valve disorders	1
I371. Pulmonary valve insufficiency	1
I40. Acute myocarditis	2
I400. Infective myocarditis	1
I401. Isolated myocarditis	2
I411. Myocarditis in viral diseases classified elsewhere	1
I418. Myocarditis in other diseases classified elsewhere	1
I42. Cardiomyopathy	7
I420. Dilated cardiomyopathy	13
I421. Obstructive hypertrophic cardiomyopathy	10
I422. Other hypertrophic cardiomyopathy	3
I424. Endocardial fibroelastosis	1
I426. Alcoholic cardiomyopathy	4
I427. Cardiomyopathy due to drugs and other external agents	1
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	1
I456. Pre-excitation syndrome	1
I458. Other specified conduction disorders	1
I46. Cardiac arrest	26
I461. Sudden cardiac death- so described	78
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	68
Total	238

Children and adolescents

58 cases were less than 16 years of age at the time of death.

Figure 6 shows the sex distribution of this group, with ethnicity, co-existing illness, family history and cause of death presented in tables 16, 17, 18 and 19.

Figure 6
Sex distribution for paediatric patients



Ethnicity

Table 16
Ethnic group of children

	n	%
White (British)	32	55.2
Black African	3	5.2
Other Ethnic Group	1	1.7
White (other)	1	1.7
Mixed White / Black African	1	1.7
Pakistani	3	5.2
Other	2	3.4
Missing	15	25.9
Total	58	100.0

Co-existing illnesses

Table 17
Co-existing illnesses in children

	n
None	19
Epilepsy, Asthma	1
Asthma	1
Asthma, Congenital heart disease	1
Congenital heart disease	13
Other	7
Unknown	14
Missing	2
Total	58

Family history

Table 18
Family History of Sudden Death in children

	n
None	10
Yes - age < 35	2
Yes - age 35-60	1
Other	1
Unknown	43
Missing	1
Total	58

Cause of death

Table 19
Final Diagnosis - ICD10 codes

	n
I058. Other mitral valve diseases	1
I230. Haemopericardium as curr comp follow acute myocard infarct	1
I24. Other acute ischemic heart diseases	1
I249. Acute ischaemic heart disease- unspecified	1
I270. Primary pulmonary hypertension	1
I34. Nonrheumatic mitral valve disorders	1
I342. Nonrheumatic mitral (valve) stenosis	1
I348. Other nonrheumatic mitral valve disorders	1
I350. Aortic (valve) stenosis	1
I37. Pulmonary valve disorders	4
I370. Pulmonary valve stenosis	1
I372. Pulmonary valve stenosis with insufficiency	1
I40. Acute myocarditis	4
I42. Cardiomyopathy	4
I420. Dilated cardiomyopathy	2
I421. Obstructive hypertrophic cardiomyopathy	2
I422. Other hypertrophic cardiomyopathy	3
I429. Cardiomyopathy- unspecified	1
I431. Cardiomyopathy in metabolic diseases	1
I46. Cardiac arrest	2
I461. Sudden cardiac death- so described	10
I490. Ventricular fibrillation and flutter	1
I50. Heart failure	1
I514. Myocarditis- unspecified	2
I516. Cardiovascular disease- unspecified	1
I517. Cardiomegaly	1
Missing	8
Total	58

The child and adolescent group shows a higher number of specific diagnoses reflecting the prevalence of congenital heart disease, although these are widely distributed. However, even in this group sudden death with a normal heart (I46 and I461) accounted for 21 per cent of deaths.

Summary

The number of cases held on the database is increasing and the patterns in the data reflect those seen in other data collections.

Participation and case submission rates remain lower than would be hoped for. Dr Elliott has previously written to his cardiology colleagues to emphasise the importance of maintaining the database. More recently the UKCPN has written to all the heads of the Regional College Councils asking them to identify the lead histopathologist in the mortuary in all hospitals. Unfortunately, this has occurred just at the time the College is disbanding its network of Regional Councils and resulted in a muted response.

The UKCPN has offered to send representatives to regional groups to demonstrate the database, discuss the issues of tissue retention and provide an update on the pathology of sudden cardiac death. Other than in Wales, which has retained a regional council, there has been to date no uptake. The UKCPN will write to all the Designated Individuals (DIs) identified through the HTA to ask for a lead pathologist in each Trust. If they can be arranged, regional educational meetings will be undertaken. UKCPN would ideally like to secure longer-term funding for the audit to help encourage greater participation and case submission.

Data quality and completeness could be improved. Key information is either missing or inaccurately recorded in significant numbers of cases. HSCIC intend to transfer the SADS data collection system to their new Clinical Audit Platform and this transition would allow improvement in data quality by the introduction of automated validation rules in the data entry process and the designation of appropriate data items as required fields.

There may also be opportunities to reduce the burden of data entry and improve data quality by replacing some free text fields with pre-defined pick lists. It may also be that the dataset is more comprehensive than necessary and provides too many options for inputting data. Migration to a new platform would provide the opportunity to review the dataset in the light of a number of years' experience of data collection and reporting. This process is currently under consideration by UKCPN and should make sure that the dataset is still appropriate for the project's purposes, that all the data items are still necessary, and that we are collecting the right data items to support the project's current and future aims. Reducing the burden of data entry by modification of the dataset might be expected to have a positive effect on participation, case submission rates and data quality.

UKCPN plan to develop the audit to support quality monitoring and improvement in the cardiac description in autopsy practice. There should be minimum standards for the description of the heart, and in cases where blocks and slides are sent, a copy of the autopsy description could be audited against those standards. This development should complement the review of autopsy guidelines currently being undertaken by the Royal College of Pathologists.

Future aims

Aims for the near future include:

1. Secure long-term funding for the database
2. Improve engagement by identifying leads in all hospital trusts/mortuaries and encouraging them to register with the audit.
3. Improve awareness of the database and UKCPN amongst both pathologists and coroners by use of educational updates.
4. Develop the database to support audit of agreed standards of macroscopic description of the heart in referred cases where only slides and blocks are submitted for an opinion.
5. Transfer the database to the HSCIC's Clinical Audit Platform.

References

1. Sheppard M. Saving families from sudden death. The Bulletin of the Royal College of Pathologists 2012 October; 160: 236-241
<http://www.rcpath.org/Resources/RCPPath/Migrated%20Resources/Documents/O/October2012Bulletinweb.pdf>
2. National Audit of Sudden Arrhythmic Death Syndrome 2011. Second annual report: Key findings. Health and Social Care Information Centre 2012.
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