Michigan Department of Community Health

MDCH

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Too Young to Die

An Update on the Impact of Sudden Cardiac Death of the Young in Michigan 1999-2011

"…no important health problem will be solved by clinical care alone, or research alone, or by public health alone- but rather by all public and private sectors working together…"
- JS Marks. Managed Care 2005

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Impact of Sudden Cardiac Death of the Young in Michigan 1999-2011

Introduction

The Michigan Department of Community Health (MDCH) is pleased to present this updated report detailing the impact of sudden cardiac death (SCD) in Michigan young people under age 40—an issue of grave public health concern. SCD is especially devastating when it occurs in children or young adults in the prime of life who were previously thought to be in good health. These deaths are a tremendous loss not only for families, but for entire communities, as evidenced by the often high profile media attention.

MDCH first identified sudden cardiac death of the young (SCDY) as a potentially preventable condition in 2004. A number of surveillance projects to identify SCDY trends occurred from 2005-2011. Analysis of mortality data for the years 1999-2009 has revealed:

- As many as 329 out-of-hospital SCDY occurring each year
- The statewide age-adjusted mortality rate as 5.5 per 100,000 with 9 counties having higher rates than the state average
- Significant disparities with more than two-thirds of the deaths occurring in males, and one-third in blacks
- Dilated cardiomyopathy as the most commonly reported underlying cause for SCDY at 1-29 years of age
- Atherosclerotic cardiovascular disease as the most commonly reported underlying cause for SCDY at 30-39 years of age.

In addition, a telephone survey of 2,856 Michigan adults conducted as part of the Behavioral Risk Factor Surveillance System (BRFSS) in 2007 revealed that about 6.3% have a family history of early SCD in one or more relatives, with a higher rate among black respondents (11.2%).

Based on expert review of selected Michigan SCDY cases from 2006-2008, 21 strategies to prevent SCDY were identified and in 2008, approximately 60 stakeholders from diverse organizations gathered to recommend next steps. From 2009-2011, significant progress has occurred with many of the strategies implemented by MDCH and partners.

The following chapters of this report highlight actual excerpts from reviewed cases and completed actions to date. Investigation of these deaths has led to identification of individual, family, public and provider needs and motivated policy makers to initiate changes to prevent future SCDY. The effectiveness of the human story to convey the burden of SCDY in the state is an instrumental tool to motivate partners and initiate prevention activities, and helps to put a personal face to each of the 3,134 SCDY that have occurred in Michigan. Our hope is that all individuals in Michigan who have experienced a SCDY in their family will know that they are not forgotten and that there are many who care.

We are thankful for the numerous partners who have contributed to this work. The partnership of all concerned individuals and organizations is needed to further increase early identification, treatment and intervention—because victims of SCD are too young to die—and every life lost is one too many.

Chapter 1
Public Health Impact and Significance

Reducing early mortality due to heart disease is an important priority for public health. Heart disease is the second leading cause of premature death as measured by years of potential life lost for males and females of most races, except for black males in whom it is the leading cause of early mortality. Overall, heart disease at any age is the leading cause of mortality in Michigan and has been for over a hundred years.[1,2]

The estimated number of SCDY occurring in Michigan varies depending on the definition. For this report, SCDY is defined as a death that occurred:

- Between 1-39 years of age;
- In the emergency room, en route to the hospital, out of the hospital, or dead on arrival to the emergency room;
- In Michigan to a Michigan resident; and was
- Cardiac-related, due to a congenital cardiac malformation, or had an ill-defined/unexplained underlying cause.

Using this definition, SCDY is estimated to claim the lives of as many as 329 Michigan children and young adults each year. Of these deaths, approximately 10% are in children and teens under age 20. About 40% occur in individuals between 20 and 34, while nearly half occur between 35 and 39 years of age. Moreover, survey data reveal that about 6.3% of all Michigan residents report having at least one biological family member who died suddenly and unexpectedly at a young age.[3]

Although SCDY is a relatively rare occurrence when compared to SCD in older adults, it has a devastating impact not only on the family but on the larger community as well. When they occur, the tragic nature of these deaths frequently attracts media attention. Questions are often asked, especially--

"Could this death have been prevented?"

Concern about the public health impact of SCDY prompted the MDCH Genomics Program and the Cardiovascular Health, Nutrition and Physical Activity Section to identify sudden cardiac death of the young (under age 40) as a potentially preventable condition, and our work in Michigan to address this public health issue began in 2003.

SCDY has critical implications for the victim's family, especially if the death was related to an inherited condition. Advances in understanding the hereditary causes of SCD have led to the realization that individuals with a family history of SCD have an increased risk of suffering a sudden death themselves.[4] New findings have emerged about the benefits of identification, treatment and follow-up to reduce the risk of early cardiac death. Many single genes associated with disorders that may lead to SCDY have been identified.[5] Furthermore, in the past decade, hundreds of new loci associated with polygenic cardiovascular risk factors for SCDY have been discovered. However, the translation of these new gene discoveries to prediction of risk and prevention of disease requires additional research.

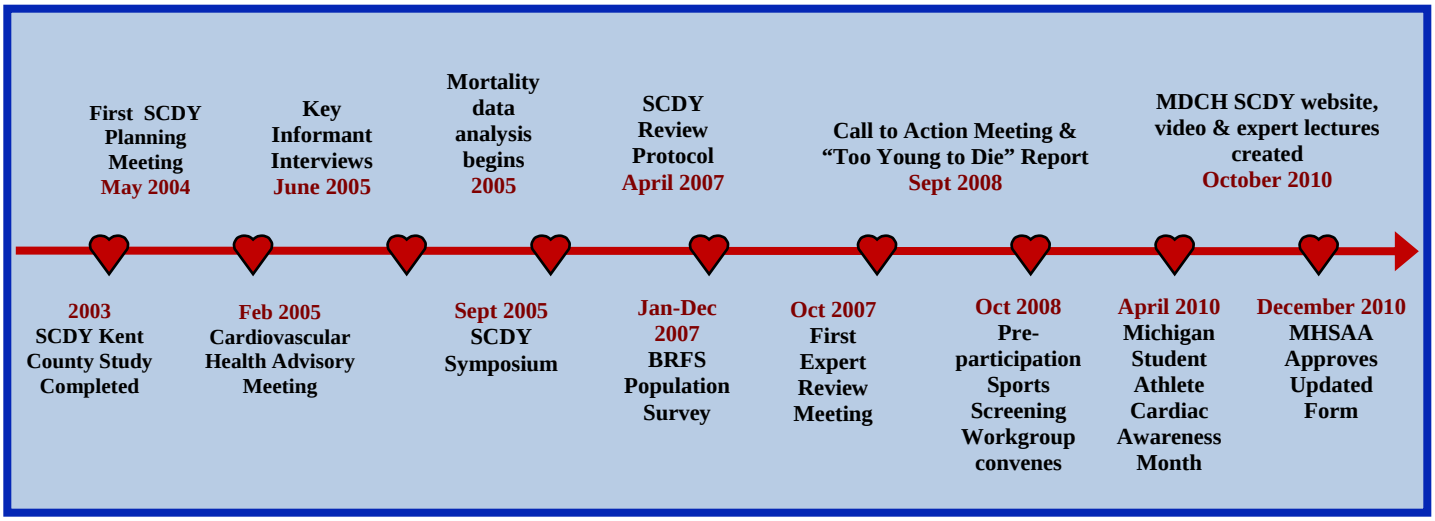
Chapter 2

Overview of the Michigan SCDY Surveillance and Mortality Review System

To address the public health impact of SCDY, MDCH first established a retrospective system for monitoring SCDY based on incoming death certificates filed with the State Registrar. Statewide population-based surveillance and epidemiological review of SCDY is a novel endeavor. To our knowledge, no other state public health agency has attempted to systematically assess and address this cause of early mortality. Several events led to the concept of a state-based SCDY surveillance and mortality review project. As shown in the timeline below, a study by Dr. Michael Lehman and colleagues in 2003 first brought attention to the problem of SCDY in Michigan.[6] The researchers examined records on unexpected deaths to people between 1 and 39 years of age who had received an autopsy in Kent County. The investigators found SCDY occurred mainly in males, 50% of the deaths were due to coronary artery disease, and family history documentation was lacking in medical examiner records.

As shown in the timeline below and in Appendix A, a series of steps were taken between 2004 and 2008 to identify key stakeholders, examine the need for surveillance, and ultimately establish a mortality review system with short-term goals to:

- ♥ Develop and refine a process to collect and review demographic and mortality data regarding SCDY in Michigan residents.
- ♥ Conduct epidemiological assessments of the burden of SCDY in Michigan.



Expert Mortality Review

In 2006, MDCH Genomics Program collaborated with Michigan State University (MSU), Division of Environmental and Occupational Medicine to develop a rapid case investigation and review process as part of a SCDY surveillance system for Michigan. The goals of this pilot mortality review system were to:

- ♥ Implement and refine a process to collect and review medical data and other circumstantial information regarding SCDY cases;
- ♥ Use the expert review process to identify recommendations as a first step toward evidence-based medical system changes and public health prevention efforts that will reduce the occurrence of SCD in Michigan; and
- ♥ Identify unmet needs for family-based interventions including education, support, medical/genetic resources and referrals for relatives who may be at increased risk of SCD themselves.

To our knowledge, this is the first statewide mortality review system that included notification to next-of-kin about possible genetic risks and the need to screen immediate relatives. The mortality review process and outcomes were detailed in a recently published article.[7]

The mortality review process included collecting information from death certificates, medical facilities, medical examiners, emergency responders, and family members. The process of rapidly identifying and reviewing cases is detailed in Appendix B. Deaths occurring to a Michigan resident aged 1-39 years between January 2006 and April 2008 were identified by MDCH Division of Vital Records and Health Statistics on a quarterly basis. Cases were then selected for in-depth review based on the decedent's age, geographic location, and underlying cause of death as coded on the death certificate and likelihood of a true "sudden death." For selected cases, MSU conducted next-of-kin interviews and collected pertinent records including autopsy results and emergency response reports. A summary of each case was prepared with all identifiable information removed.

Case summaries were reviewed by a panel of experts representing the multiple disciplines of emergency medicine, pediatric and adult cardiology, medical genetics, cardiac pharmacology, public health, pathology, primary care, sports medicine, nursing, and health insurance. For each case, the panel was asked to:

- Confirm the cause of death listed on the death certificate or suggest an alternative diagnosis;
- Describe all significant factors that may have contributed to the death, including health system or community issues such as access to an automated external defibrillator (AED) in the location where the death occurred;
- Determine the likelihood that additional family members could be at risk and attempt to determine whether appropriate medical interventions for family members were identified or had been recommended; and
- Suggest recommendations related to the patient, provider, and system levels that could have prevented the reviewed death or future deaths in the family.

If deemed appropriate by the expert panel, the next-of-kin were notified by MDCH to provide information about potential heritable risks and the importance of screening immediate family members.

Medical records, including autopsy reports, were obtained and reviewed on 23 SCDY cases that occurred between 2006 and 2008 in Michigan. The two most common causes of death in these cases were attributed to cardiomyopathy (7 cases) and fatal arrhythmia (6 cases). A possible heritable cause was attributed to 17 of the 23 cases (73.9%). Sixteen next-of-kin interviews were conducted; most of these next-of-kin were not aware of the possible heritable cause of SCDY and were also not aware of the importance of family screening. All next-of-kin received introductory and follow-up letters from MDCH regarding the expert case review. If appropriate, the next-of-kin follow-up letters included information about the importance of screening immediate family members. The next of kin, family members and/or their providers were encouraged to contact MDCH with any questions. Some families and their providers did contact MDCH for additional information and specifics about appropriate screening for family members. Follow-up evaluations of immediate family members, even in the absence of a known SCDY cause of death, can lead to potential prevention of sudden death in the surviving family members.

Based on review of these SCDY cases, the expert panel identified 21 prevention measures grouped into five themes (also see Table 5):

- Provider education and public awareness of SCDY risk factors
- Enhanced pre-participation sports screening, physical examination and follow-up
- Public awareness of cardiac symptoms and training in the use of cardiopulmonary resuscitation (CPR) and AED for coaches and the general public
- Creation and dissemination of emergency response protocols
- Creation and dissemination of medical examiner protocols for possible SCDY cases

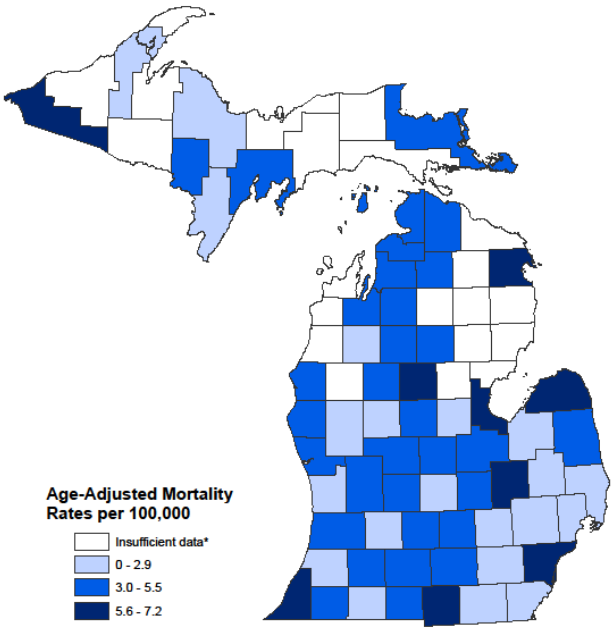
Chapter 3

Epidemiology of SCDY in Michigan

Who is affected by SCDY?
Where do most deaths occur?
Why do these deaths happen?

To understand the burden of sudden death in Michigan young people, mortality data spanning the time period from 1999-2009 were examined for residents between 1 and 39 years of age whose underlying cause of death was reported on the death certificate using specific codes based on the International Classification of Diseases-Version 10 (ICD-10) system (Appendix C). All inpatient hospital deaths were excluded. A retrospective methodology was used to examine death certificates similar to previously published studies.[8] Cases were assigned to one of three groups according to eligible codes that included cardiac (I00-I51 except I12-13 and I26-28), congenital cardiac malformation (Q20-Q24 and Q87.4) or ill-defined (R96-R99) etiologies. Analysis of death certificate data alone does not provide potential explanations for reasons for death—for instance, the impact of environmental or behavioral factors such as smoking, drug or alcohol use, obesity or level of physical activity. Although there is ambiguity regarding some of the codes used to report underlying causes of death, the MDCH Division for Vital Records and Health Statistics classifies reported deaths to Michigan residents in accord with national coding requirements. This retrospective code-based method may result in an overestimation of true SCD cases. Further assessments of SCDY using *multiple* sources of case ascertainment (i.e., death certificates, emergency medical response, medical examiners, hospitals), as previously described in our expert mortality process, enhance the accuracy of the data.

Figure 1
Age-Adjusted Mortality Rates of SCDY among Michigan Residents, Ages 1-39 by County of Residence, 1999-2009



*Counties with fewer than five sudden cardiac deaths from 1999-2009
Michigan's age-adjusted mortality rate is 5.5 per 100,000
Source: MDCH Vital Statistics
Age-adjusted to the 2000 U.S. standard population

From 1999-2009, there were 3,134 deaths that met Michigan's case definition for SCDY based on death certificates filed with the State Registrar. The statewide age-adjusted mortality rate for children and young adults aged 1-39 was 5.5 per 100,000 residents. There were significant differences between counties of residence, as depicted in Figure 1. Nine counties had mortality rates for residents that were higher than the statewide rate, including Alpena (7.2), Claire (7.1) and Genesee (6.7) counties. Wayne County had the largest number of deaths (1,166), followed by Oakland (251), Macomb (203), Genesee (164), and Kent (146) counties. The numbers and rates for all Michigan counties are included in Appendix D.

Data were also analyzed by sex, race, place of death, autopsy status, and cause of death (Table 1). Significant disparities were observed for sex and race, with more than two-thirds of the deaths occurring in males, and one-third in blacks. The greatest numbers of SCDY were reported in adults ages 35-39 years. Nearly 47% of individuals died in the emergency room or were dead on arrival, whereas 43% died at home. An autopsy was performed in more than three-quarters of the cases (79%).

Overall, the rate for males was 7.6 per 100,000 and for females, 3.4 per 100,000. The rate for blacks (12.2 per 100,000) was significantly higher than the rate for whites (4.3 per 100,000). The single highest age-adjusted mortality rate was for black males at 16.5 per 100,000 which is nearly two-and-a-half times the rate for white males (6.1 per 100,000). The rate for black females was 8.3 per 100,000, more than triple the rate for white females (2.4 per 100,000) [Data not shown].

The lowest mortality rate was for the 1-9 year old group at 1.0, followed by the 10-19 year old group at 1.2 and the 20-29 year old group at 4.1 per 100,000. There was a dramatic increase between those in their 20s and those in their 30s, with the highest mortality rate for the 30-39 year old age grouping at 14.5 per 100,000.

The significantly increased mortality rate in the 30-39 year old group requires further investigation into possible modifiable risk factors associated with these deaths.

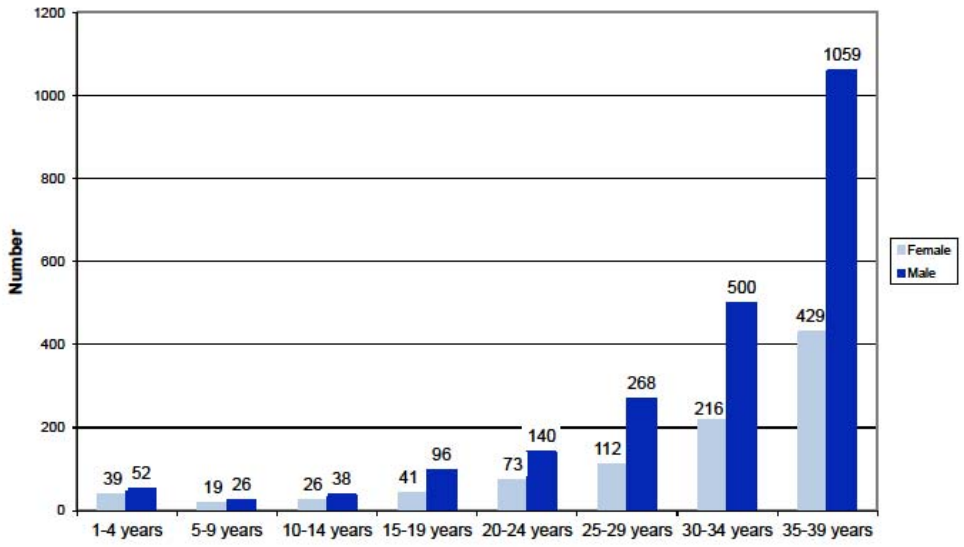
Table 1
Sudden cardiac deaths* of Michigan residents aged 1 - 39 years, 1999 - 2009

		Number	Percent
Total		**3,134**	
Sex			
	Male	2,179	69.5
	Female	955	30.5
Race			
	White	1,961	62.6
	Black	1,089	34.7
	Other	84	2.7
Age			
	1-4 years	91	2.9
	5-9 years	45	1.4
	10-14 years	64	2.0
	15-19 years	137	4.4
	20-24 years	213	6.8
	25-29 years	380	12.1
	30-34 years	716	22.8
	35-39 years	1,488	47.5
Place of death			
	Home	1,339	42.7
	Nursing home, extended care	28	0.9
	Hospital: emergency room / outpatient	1,462	46.6
	Ambulance	34	1.1
	Other / unknown	271	8.6
Autopsy			
	Yes	2,474	78.9
	No	658	21.0
	Unknown	2	0.1

* Includes decedents who died out of the hospital, or in an emergency department, or were dead on arrival to an emergency department, and had one of the identified ICD-10 codes reported as the underlying cause of death on the death certificate

Significant disparities were also observed for age and sex (Figure 2). For all age groups analyzed, males were more commonly affected with SCDY than females. The largest disparity between sexes was observed in the 35-39 year old age group, with males comprising approximately 70% of cases.

Figure 2
Number of SCDY in Michigan residents aged 1-39 years, by age and sex, 1999-2009



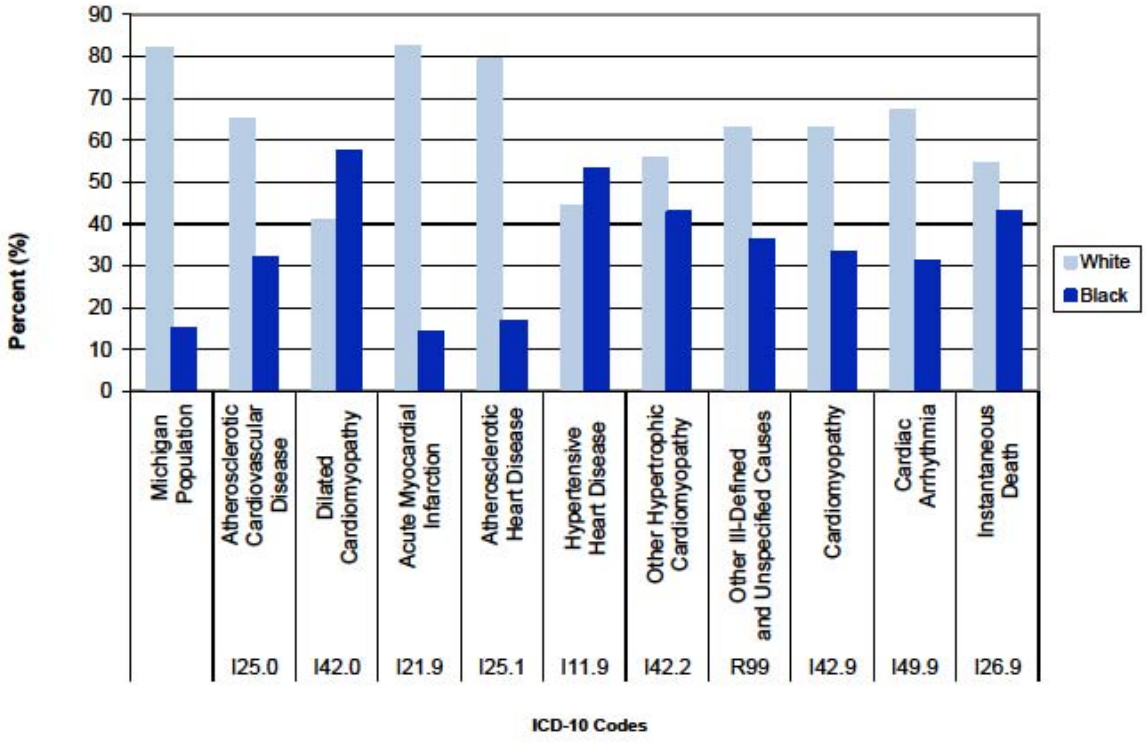
The overall age-adjusted rate for deaths due to cardiac causes was 4.8 per 100,000, significantly higher than rates for ill-defined causes (0.4) and for congenital cardiac malformations (0.3). The single most frequently reported cause of death between 1-39 years of age was atherosclerotic cardiovascular disease (14.8%), followed by dilated cardiomyopathy (14.2%) and acute myocardial infarction (10.6%), as noted in Table 2. In contrast, the single most frequently reported cause of death between 1-29 years of age was dilated cardiomyopathy (41.1%; n=122) [Data not shown]. Significant genetic components are known to be associated with early onset atherosclerosis, cardiomyopathies and many of the other identified causes of death which include anatomical and arrhythmogenic cardiac disorders. For a list of common causes of SCDY, please see Appendix E.

Table 2
Ten most frequent underlying causes of death of Michigan SCD victims, 1-39 years, 1999-2009 (n=3,134)

ICD 10 Code	Cause of death	Number	Percent
I25.0	Atherosclerotic cardiovascular disease	464	14.8
I42.0	Dilated cardiomyopathy	444	14.2
I21.9	Acute myocardial infarction	331	10.6
I25.1	Atherosclerotic heart disease	303	9.7
I11.9	Hypertensive heart disease without heart failure	221	7.1
I42.2	Other hypertrophic cardiomyopathy	180	5.7
R99	Other ill-defined and unspecified causes of mortality	124	4.0
I42.9	Cardiomyopathy	121	3.9
I49.9	Cardiac arrhythmia	109	3.5
I26.9	Instantaneous death	86	2.7

The relative frequency of different SCDY etiologies also varies by sex, race and age. For the top ten causes of SCDY, there were notable differences by race, with **blacks disproportionately represented among deaths relating to any cause except acute myocardial infarction and atherosclerotic heart disease**. In the 1-39 year old age group, the most commonly reported underlying cause in blacks was dilated cardiomyopathy (n=255) while in whites it was atherosclerotic cardiovascular disease (n=301) (**Figure 3**). In the 1-29 year old age group, the most commonly reported cause of death was dilated cardiomyopathy for blacks and whites. Among deaths due to dilated cardiomyopathy in the 1-29 year old age group, approximately 60% (n=72) were among blacks [data not shown]. **Among deaths in the 1-39 year old age group, dilated cardiomyopathy and hypertensive heart disease were notably higher among blacks compared to whites and the deaths in blacks comprised more than half of all deaths due to those two etiologies** (Figure 3).

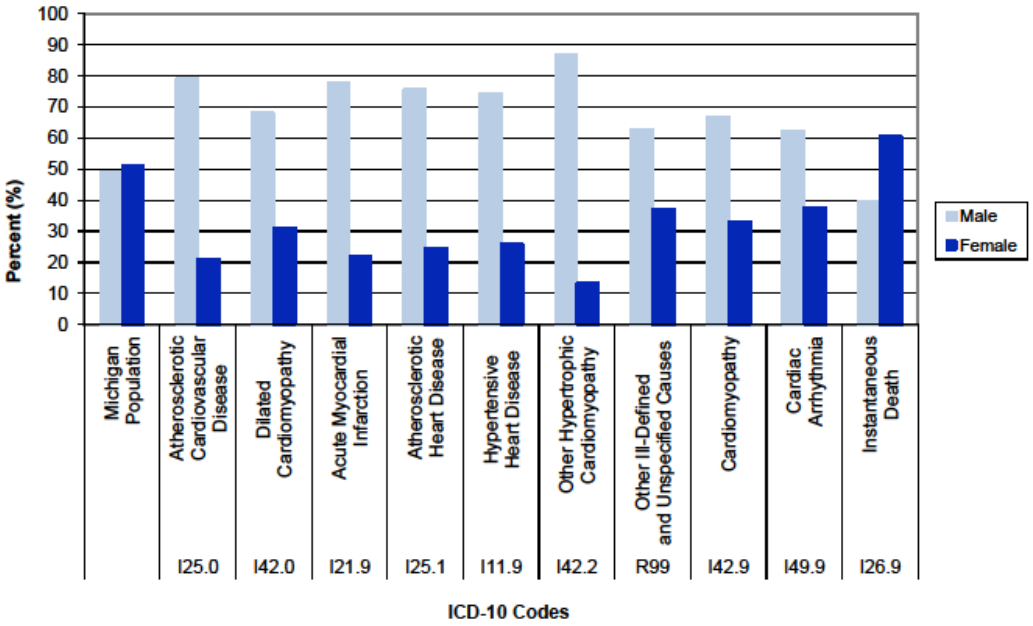
Figure 3
Top ten causes of SCDY by race, Michigan residents aged 1-39 years, 1999-2009



There were also notable differences between males and females, with males more likely than females to die of SCDY for all age groups and most of the ten top SCDY causes. However, the one exception was for SCDY between 1-39 years of age, in which females were more likely to die of instantaneous death. The greatest difference between males and females was in the "other hypertrophic cardiomyopathy" category where females comprised just less than 14%, while males represented 86% of all deaths (**Figure 4**).

Figure 4

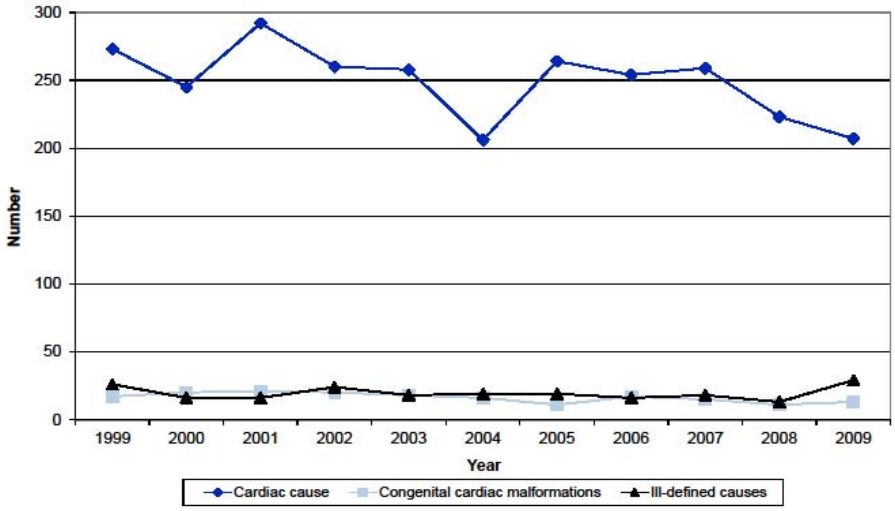
Top ten causes of SCDY by sex, Michigan residents aged 1-39 years, 1999-2009



There was little year to year fluctuation in the number of SCDY cases due to congenital and ill-defined causes. A slight decrease in cardiac cases was observed in 2008 and 2009 (Figure 5), but it is unclear whether this is a new trend or normal variation. MDCH will continue to gather data in coming years.

Figure 5

Number of SCDYs in Michigan residents aged 1-39 years, by year and underlying cause of death, 1999-2009



Michigan BRFS and Family History of SCDY

Although age, sex, and race/ethnicity are risk factors for SCDY, the greatest risk factor is family history.[9] In fact, as much as 40% of families with a victim of SCDY have been identified as having a heritable disease. Collection of family history of SCDY is a vital first step to identify, screen, and manage individuals at greatest risk for SCDY.

Despite increased risks for family members, it appears that family history of SCDY is rarely documented in the clinical setting.[10] There is a paucity of reports in the medical literature on the psychosocial, emotional or clinical follow-up of family members of SCDY victims. Therefore, in 2007, using the Michigan BRFSS (www.michigan.gov/brfs), we surveyed 2,856 Michigan adults about their family history of SCDY.

Overall, the proportion of adults in Michigan reporting a family history of early sudden cardiac or unexplained death was 6.3% (Table 3). Of adults reporting a family history, 35.5% had an immediate relative (parent, sibling, or child) and 26.2% had multiple family members that had SCDY. The most commonly reported family member who had SCDY was a sibling. This is significant since individuals with multiple affected family members and/or an affected first degree relative are known to have the greatest risk based on their family history.

Table 3
Family History of Sudden Cardiac Death of the Young[a]
2007 Michigan Behavioral Risk Factor Survey

	%	95% Confidence Interval
Total	**6.3**	**(5.2 - 7.7)**
Age		
18 – 24	3.8	(1.6 - 8.7)
25 – 34	8.6	(4.9 - 14.6)
35 – 44	4.2	(2.4 - 7.1)
45 – 54	7.7	(5.4 - 10.9)
55 – 64	5.9	(4.1 - 8.5)
65 – 74	8.5	(5.4 - 13.3)
75 +	5.4	(3.5 - 8.2)
Gender		
Male	5.4	(3.9 - 7.4)
Female	7.7	(6.1 - 9.6)
Race/Ethnicity		
White non-Hispanic	5.4	(4.3 - 6.8)
Black non-Hispanic	11.2	(7.7 - 16.0)
Other non-Hispanic	9.4	(3.8 - 21.3)
Hispanic	--[b]	
Education		
Less than high school	10.8	(5.8 - 19.3)
High school graduate	8.8	(6.6 - 11.7)
Some college	4.7	(3.3 - 6.8)
College graduate	4.4	(2.8 - 6.8)
Household Income		
< $20,000	7.8	(5.1 - 11.7)
$20,000 - $34,999	8.4	(5.9 - 11.8)
$35,000 - $49,999	8.8	(5.5 - 13.8)
$50,000 - $74,999	4.1	(2.1 - 7.9)
$75,000 +	3.2	(1.9 - 5.2)

[a] Among all respondents (n = 2,856), the proportion who reported having at least one biological family member that had a sudden cardiac death, or sudden unexplained death, between the ages of 1 and 39.

Note: Interviewers were instructed not to include spouses of the respondent, infants less than one year of age, as well as drug-related deaths, traumatic deaths (such as car crashes), suicides, homicides, or individuals who had a long illness.

[b] The denominator in this subgroup is less than 50.

Table 4

Prevalence of health-related characteristics among Michigan adults by family history of SCDY

2007 Michigan Behavioral Risk Factor Survey

Health-Related Characteristic	Has Family History of SCDY: Yes % (95% CI)	Has Family History of SCDY: No % (95% CI)
No health insurance	17.8 (11.0-27.5)	10.6 (9.0-12.5)
On Medicaid insurance	23.1 (15.4-33.1)	10.6 (9.0-12.4)
No personal doctor	13.0 (7.9-20.5)	15.0 (12.9-17.4)
No routine checkup in past year	29.3 (20.1-40.5)	31.3 (28.7-34.0)
No blood cholesterol test in past 5 years	27.4 (17.9-39.6)	20.1 (17.6-23.0)
Health Status		
Fair to poor general health	16.1 (11.1-22.7)	14.3 (12.6-16.2)
Rarely-never receive needed emotional support	12.3 (7.1-20.4)	6.2 (5.1-7.6)
Has a disability	26.4 (19.6-34.6)	21.8 (19.8-23.9)
Obese (BMI ≥ 30)	34.0 (25.0-44.4)	27.6 (25.2-30.2)
Chronic Conditions		
Ever diagnosed with high blood pressure	39.5 (30.8-49.1)	27.9 (25.8-30.2)
Ever diagnosed with high cholesterol (among tested)	42.4 (33.0-52.4)	40.8 (38.1-43.5)
Ever diagnosed with diabetes	13.1 (8.9-19.1)	8.6 (7.5-9.9)
Ever diagnosed with cardiovascular disease	10.0 (6.2-15.8)	9.5 (8.3-10.9)
Behaviors		
Current smoking	32.2 (23.3-42.6)	20.1 (17.9-22.6)
No leisure-time physical activity	20.2 (13.3-29.5)	19.3 (17.2-21.5)
Inadequate physical activity	48.3 (38.2-58.5)	47.3 (44.5-50.0)
Inadequate fruit and vegetable consumption	82.4 (75.6-87.6)	78.1 (75.7-80.2)

Consistent with the health disparities found among SCDY victims (Table 1), there are considerable differences in the proportion of respondents with a family history of SCDY by racial/ethnic background (Table 3). Blacks had the highest reported prevalence of SCDY family history at 11.2%, more than double the prevalence of white non-Hispanic adults, 5.4%. Adults with a household income of less than $50,000 also reported family history of SCDY more frequently than those with a household income of $50,000 or more. Adults with a high school degree or less education more often reported a family history of SCDY than those with some college education. Among those with a family history of SCDY, there was also a significantly higher proportion who reported having Medicaid insurance compared to those without a family history (23.1% vs.10.6%).

Furthermore, those with a family history of SCDY also had a significantly higher proportion of people who had ever been diagnosed with high blood pressure (39.5% vs. 27.9%) and a higher proportion who were current smokers compared to those without such a family history (32.2% vs. 20.1%) (Table 4). These data highlight the need to collect family history of SCDY on all individuals, identify high-risk individuals and initiate public health and clinical interventions to reduce the burden of SCDY.

Chapter 4
Strategies and Opportunities for Prevention

Certain causes of SCD in younger adults and children are more likely to have genetic determinants than similar conditions in older persons. These include inherited arrhythmias, hypertrophic cardiomyopathy, undetected congenital heart defects and early coronary artery disease/atherosclerotic disease. Over the last decade, there has been significant progress in identifying inherited disorders that increase the chance of SCDY.[11,12,13] There have also been major strides in uncovering new genes, mechanisms and syndromes that have significantly advanced the diagnosis and treatment of genetic SCDY disorders.[14] These advances can enhance presymptomatic diagnosis and help establish definitive molecular diagnosis for symptomatic patients.[15] Furthermore, recent discoveries about genotype-phenotype correlations can now assist providers in predicting risk factors and selecting treatment for patients with inherited arrhythmic disorders.[16] Despite the major advances in cardiovascular genetics, a careful review of personal and family health history remains the current standard for assessing SCDY risk.[17] Throughout this chapter, each section includes blue boxes quoting excerpts from our expert mortality review which highlight the need for action to prevent SCDY in Michigan.

Provider Education on SCDY Risk Assessment

Collection of personal and family history is an effective and simple method for providers to screen and identify individuals at risk for SCDY. From 2005-2007, MDCH worked with a major Michigan health plan to conduct a review of 668 charts from Michigan primary care providers (i.e. pediatric, family medicine, internal medicine). Disturbingly, less than 1% of these charts indicated that the provider had inquired and documented the presence or absence of a family history of SCDY.[18]

It is critical for providers to be aware of personal and family risk factors for SCDY. Cardiovascular risk assessment should be administered at routine intervals starting in early childhood. There are specific personal and family history questions[19] that can indicate a potential risk for SCDY including:

- ⇒ Unexplained syncope/fainting particularly during exercise, emotion or startle
- ⇒ Chest pain or discomfort during exercise
- ⇒ Excessive, unexpected and unexplained shortness of breath or fatigue associated with exercise
- ⇒ Past detection of heart murmur, high cholesterol, myocarditis, or increased systemic blood pressure
- ⇒ Family history of premature death in close relatives younger than 50 years old, including unexplained accidents, drowning, SIDS
- ⇒ Family history of unexplained syncope
- ⇒ Family history of congenital deafness
- ⇒ Family history of individuals with pacemakers or automatic implantable cardioverter-defibrillators (ICDs)
- ⇒ Specific knowledge of the occurrence of certain familial conditions associated with SCDY [see Appendix F]

After their son died in his 20s, the parents learned that he had been complaining of chest pain and shortness of breath for a couple weeks but did not feel he needed medical attention.

An example of a risk assessment form for use in the clinical setting was published by Campbell and Berger in 2006.[20] For those identified with a significant personal or family history of SCD and/or related heart conditions, referral to a cardiologist and/or clinical geneticist who specializes in conditions known to be associated with SCD should be considered.

Pre-participation Sports Screening

One opportunity for primary prevention of SCDY that has received substantial national and international attention is to identify high risk athletes through pre-participation screening. Importantly, SCD is the leading cause of death in young athletes.[21] Hypertrophic cardiomyopathy (HCM) is the single most common cause of death among U.S. athletes, occurring most commonly in those playing football and basketball. Male athletes are more likely to die suddenly than female athletes (male:female ratio for SCDY is up to 9:1). Sudden Cardiac Death of the Young is also disproportionately more likely to occur in African American athletes.[22] Identification of athletes at high risk for SCDY

is critical in order to prevent adverse events. However, since athletes with unsuspected cardiovascular disease can appear healthy, it can be difficult for providers to detect 'silent' cardiovascular abnormalities than can lead to sudden death without a careful history and examination.

International Guidelines for ECG Pre-Participation Sports Screening

Guidelines for screening athletes differ widely on an international, national and state basis.[23,24,25,26,27,28,29,30] Many professional sports leagues, the International Olympic Committee and some countries routinely incorporate the use of ECGs with the physical and health assessment for early detection of cardiovascular abnormalities in athletes. A national screening program in Italy led to a 10-fold reduction in the incidence of SCDY in competitive athletes (with a 7% false-positive rate) over a 25 year period, while a study conducted in Israel did not find the rate of SCDY to be affected by mandatory mass ECG screening over a 12 year period.[31,32]

Mass Universal ECG Pre-Participation Sports Screening Not Currently Recommended in the United States

There has been considerable debate in the United States about the appropriate strategies, effectiveness and logistics of various pre-participation screening methods.[33,34] Evidence-based guidelines for pre-participation screening of non-professional athletes currently do not exist, and national research on the best method of conducting pre-participation screening for athletes has led to inconsistent conclusions.[35,36] A recent study of 400 healthy children (5-19 years of age) in Philadelphia who received history and physical examination, ECG and echocardiograms detected 5.8% with previously undiagnosed cardiac abnormalities.[37] The authors concluded that ECG was three times more likely than history and physical examination alone to identify true abnormalities.

The American Heart Association (AHA) supports pre-participation cardiovascular screening for student-athletes in organized competitive sports as "justifiable, necessary, and compelling on the basis of ethical, legal and medical grounds." However, the AHA Council on Nutrition, Physical Activity and Metabolism did not believe it to be…"prudent or practical to recommend the routine use of tests such as 12-lead ECG or echocardiography in the context of mass, universal screening" it concedes that …"such a complex initiative would have benefit in terms of detecting greater numbers of athletes with important heart diseases." The Council also did not …"arbitrarily oppose volunteer-based athlete screening programs with noninvasive testing performed selectively on a smaller scale in local communities if well designed and prudently implemented." [38]

> As reported during his pre-participation sports screening, he had been experiencing vague symptoms for the past several months. His heart skipped beats, he was dizzy when getting up from a chair, his legs hurt and he was tired all the time. The teen thought the symptoms meant he was out of shape so he would practice harder. A few days before his death, he told his mother "I'm going to die... my heart's going to stop."

Studies in support of ECG screening are evolving rapidly with more rigorous ECG criteria leading to lower total positive rate and reduced levels of false positives.[39] If SCDY is found to be more common than previously suspected and the accuracy of ECG screening continues to increase, mass ECG screening may become more feasible in the United States. In 2010, a group from Stanford University using computer simulation concluded that screening young athletes with 12-lead ECG plus cardiovascular focused history and physical examination may be cost effective.[40] In 2011, a National Heart, Lung and Blood Institute (NHLBI) working group published support for development of a research agenda to further clarify the national incidence of SCDY and determine the best approach to reduce SCDY.[41] The research would evaluate if screening with ECG effectively reduces SCDY and adds overall healthcare value.

Current Consensus on Pre-participation Sports Screening in the United States

Despite differing opinions among cardiology experts on the value and feasibility of including additional elements such as routine ECG screening, there is general agreement nationally on the basic elements of cardiovascular screening. The AHA and American College of Sports Medicine recommend that an athlete pre-participation evaluation include detailed family history and physical examination. The recommended 12-point screening protocol for young competitive athletes includes:

1) A personal history to assess palpitations, exertional chest pain/discomfort, unexplained syncope/near syncope (fainting), exertional unexplained dyspnea/fatigue, and elevated systemic blood pressure and heart murmur;

2) A family history to assess premature death, disability from heart disease in a close relative younger than 50 years old, and known cardiovascular genetic conditions; and

3) A physical exam to assess heart murmur, femoral pulses, physical stigmata of Marfan syndrome and brachial artery blood pressure.

If there is a significant finding in the history or physical, a 12-lead ECG, echocardiogram, exercise testing and/or cardiovascular consultation should be considered. Parental verification of any responses provided by a youth is essential for secondary education students.[42,43]

The mother voiced concern over the fact that the sports physical form was not seen, filled out, or signed by her. After his death, she found a letter her son had received clearing him to play sports.

In 2010, the American Academy of Family Physicians, American Academy of Pediatrics, American College of Sports Medicine, American Medical Society for Sports Medicine, American Orthopaedic Society for Sports Medicine, and American Osteopathic Academy of Sports Medicine published the fourth edition of a pre-participation physical evaluation monograph that includes a recommended pre-participation history and physical form, (Appendix F).[44] The physical exam includes a focus on detecting the heart murmur of left ventricular outflow tract obstruction and physical findings suggestive of Marfan syndrome. Patients suspected or identified to be at risk for SCDY by the history or exam should be referred to a cardiovascular specialist for further evaluation. In addition, providers should be aware that guidelines for athletes with known genetic conditions, such as hypertrophic cardiomyopathy (HCM), and known coronary artery disease have been published.[45,46,47]

Public Awareness of Cardiac Symptoms and AED/CPR Training

Sudden cardiac arrest (SCA) most often results from a sudden, unexpected heart arrhythmia such as ventricular fibrillation during which the heart's electrical impulses become chaotic and ineffective. Blood flow to the brain is lost and death usually follows unless a normal heart rhythm is quickly restored. Defibrillation with a device such as an AED provides an electrical shock to re-establish the heart's normal rhythm and is the only known treatment for ventricular fibrillation.[48]

An AED was not available during the practice and the boy's coach did not have training on how to use one.

Availability of an AED provides the opportunity for secondary prevention of early deaths. However, symptoms of possible cardiac arrest must first be recognized by bystanders, and an AED must be accessible by trained users with a target goal of three to five minutes from time of collapse to first shock. Currently the chance of surviving SCA is less than five percent. The AHA has identified a "Chain of Survival" that increases the likelihood of survival. The sequence of four steps includes early access (recognizing an emergency exists and calling 911), early CPR, early defibrillation and early advanced care.[49,50]

According to the AHA, early CPR and defibrillation within the first three to five minutes after collapse, plus early advanced care can result in greater than 50% long-term survival for witnessed ventricular fibrillation events.[51]

Emergency Response Protocols

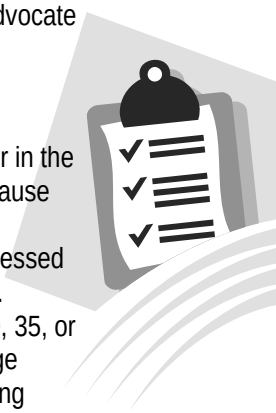
EMS response time in most communities in the United States has been reported to range from 12-15 minutes.[52] The time delay is related to many factors, including the time to activate EMS by telephone, arrival of EMS at the site, parking of the rescue vehicle, and initiation of first defibrillation shock. The AHA advocates for equipping all EMS first-response vehicles and ambulances with an AED or other defibrillation device. It also supports "Public Access to Defibrillation" (PAD), which means making AEDs available in targeted public and/or private places where large numbers of people gather or where people who are at high risk for cardiac arrest live—venues such as sports

arenas, gated communities, office complexes, shopping malls, etc. When AEDS are placed in the community, they need to be part of a defibrillation program that includes notification of the local EMS office, oversight by a licensed medical authority to ensure quality control, and training in both CPR and use of the defibrillator for persons responsible for using the AED.[53]

> The deceased's sports team was not affiliated with the school district or the city's recreation program. The coach told the family there was an AED in the building where the deceased had collapsed but it was locked in an office and not available.

The National Athletic Trainers' Association has convened a task force to ensure an efficient response to SCA.[54,55] The task force states that it is critical for schools to be prepared for cardiac emergencies. They recommend that structured emergency action plans should be developed through discussion with local EMS, school safety officials, first responders and school administrators. The task force further notes that the emergency plan should be practiced annually; the first responders should be trained and certified in CPR and defibrillation; the collapse-to-EMS call time and CPR initiation should be less than one minute; and the access to early defibrillation must have a target time of less than three to five minutes from time of collapse. They also state that SCA should be suspected for any collapsed and unresponsive athlete. Finally, the task force developed uniform recommendations for management of SCA in athletes, similar to the "chain of survival" recommendations- early activation of EMS, early CPR, early defibrillation, and rapid transition to advanced cardiac life support.[56,57] These guidelines are comparable to statements that have been published by AHA taskforces and other organizations on this issue.[58]

While the effectiveness of widespread AED availability in the secondary prevention of SCDY is unknown, a number of individuals and families who have survived SCA or lost a loved one to SCDY have started foundations to advocate for placement of AEDs in schools and other public areas.[59,60,61]



Medical Examiner Protocols

Sudden Cardiac Death describes an unexpected, non-violent, non-traumatic death occurring out of hospital, or in the emergency room, or being dead on arrival. Sudden cardiac death is usually defined as death from a cardiac cause occurring within one hour from onset of symptoms; however, the time frame for onset of symptoms varies in definitions from 1 to 24 hours.[62,63] Some definitions further specify whether the death was witnessed or unwitnessed and the amount of time since the victim was last seen without acute symptoms in his/her usual state of health. *Sudden Cardiac Death of the Young* has been variably defined as occurring in people under the age of 25, 30, 35, or 40 years.[64] Most often the term is used for deaths to individuals between the ages of 1 and 39, which is the age range used for identifying cases in Michigan's SCDY surveillance program. Sudden Cardiac Death of the Young generally occurs in the emergency room or at home with the underlying cause of death reported as cardiac disease (ICD-10 codes: I00-I51). However, the selection of a specific ICD-10 code as the underlying cause of death is often ambiguous; for instance, the rationale for selecting "I26.9 instantaneous death" over the myriad of other available codes for SCDY remains unclear. One important limitation of the SCDY surveillance project identified by the expert panel is an inconsistency in coding the cause of death (ICD codes) on death certificates. Therefore, there is an urgent need to standardize reporting by medical examiners and others in order to improve data accuracy, completeness and comparability across the state and nationally. The advent of electronic medical records and data reporting systems may present an opportunity for achieving more consistency.

Need for Autopsy and Implications for Family Members

When a young person dies suddenly, it is imperative to investigate the underlying cause in order to identify possible risks for other family members. Routine autopsy can detect some structural conditions that predispose to SCDY but ideally the autopsy examination should be performed by a pathologist with cardiac expertise and appropriate samples be taken for histological and DNA analysis.[65] To our knowledge, there is no published evidence-based protocol or universal procedure that medical examiners follow when investigating SCDY in the United States. However, there are advocacy groups that recommend a mandatory autopsy for all cases of SCDY, and have issued recommendations for the postmortem process.[66] Other countries have also established SCDY uniform protocols for medical examiners, including recent guidelines by the Association of European Cardiovascular Pathology.[67]

It is critical to review autopsy information in light of possible genetic implications for surviving family members, and consider DNA testing or banking as part of the post-mortem work-up. Both health care providers and families need to

be aware that the presence of SCDY in a biological relative may increase the risk for many other family members due to numerous underlying genetic causes. It is the potential impact on the entire family that makes a thorough investigation, including autopsy and careful medical consideration of genetic causes, vital in every case of SCDY. In the case of familial hypertrophic cardiomyopathy (HCM), a structural cause of SCD often identified on autopsy, there is a one in two chance that first degree relatives (parent, sibling, or child) also inherited the gene for this condition. Clinical recommendations for screening high risk individuals (based on history of an affected family member) include clinical assessment with electrocardiogram (ECG) and echocardiogram at regular intervals starting in childhood. Genetic testing for HCM is also commercially available, and allows for early detection prior to the expression of typical clinical findings.

In contrast to "single gene" disorders such as HCM, the etiology of coronary artery disease is typically more complex, based on both genetic and behavioral/environmental causes (multifactorial). Certain lifestyle choices, such as smoking, high-fat diet, and lack of physical activity, are known risk factors. However, when a young person dies of coronary artery disease, there is likely a strong underlying genetic predisposition. Family members should, therefore, be carefully screened for risk factors related to early coronary artery disease so they can modify other preventable risk factors and monitor for development of coronary artery disease.

A white female in her 20s was found unresponsive and lying in bed. After a full autopsy, examination of histologic tissue sections and full toxicology, the cause and manner of death were undetermined. There was no documentation that DNA testing or banking was considered.

Molecular Autopsy

About 30% of young sudden deaths have negative autopsies.[68] Because electrical and other conditions will not be detected, it is not unusual for a routine autopsy to conclude that the cause of death was indeterminate. In such cases, heritable conditions such as cardiac ion channel disorders, are suspect and genetic analysis could help to elucidate the cause of death and prevent future deaths in families at risk.[69,70] Post-mortem DNA testing is commonly referred to as a "molecular autopsy". Studies have found that molecular autopsy leads to a specific diagnosis with a known deleterious mutation in about 30-40% of cases.[71,72] Detecting a mutation has also been shown in some studies to be more likely when there is more than one SCDY in a family[73] and in SCDY cases with a syncope history.[74] When a routine autopsy is inconclusive or negative, continued investigation through cardiology and genetic evaluation of first or second degree relatives or a molecular autopsy may help to determine the underlying cause of the SCDY.[75] Laboratories conducting DNA testing for various causes of SCDY can be found at: www.genetests.org. DNA testing can be quite costly; however, funding assistance may be available in some situations.

Since current genetic technology can detect less than half of SCDY cases with known deleterious mutations, DNA testing for families is most informative when it includes testing the affected individual's DNA first; otherwise, 'negative' DNA test results on family members will be uninformative.[76] An appropriate assessment of family members is highly dependent on determining the cause of the SCDY. Once the specific cause of death has been determined, the best approach is to use a stepwise assessment or cascade screening through the family to identify key individuals most likely to benefit from screening.[77] Another approach to consider is combining molecular autopsy with family cardiac screening.[78] Newer technologies are emerging that will make DNA testing even more effective.[79]

DNA Banking

DNA banking is another important option to consider when SCDY occurs. DNA banking is offered through a number of clinical laboratories that can be found at: www.genetests.org . DNA banking by a clinical laboratory is typically not covered by insurance. However, some medical examiners in Michigan routinely store a dried blood spot from autopsies. Also, if DNA banking or autopsy were not considered at the time of death, another approach to obtaining DNA from a deceased individual is through stored newborn screening dried blood spots. In fact, a study in New Zealand of 21 cases of SCDY utilized stored dried blood spots to conduct molecular autopsies up to 13 years after death.[80] In Michigan, there exists an important resource of archived newborn screening dried blood spots that are stored by the MDCH. With permission from a parent, guardian or next-of-kin, these archived blood spots could be used for molecular testing to further investigate causes of death in those born in Michigan since 1984.[81,82,83]

Chapter 5

Michigan's Progress in Preventing SCDY, 2008-2011

In September 2008, a "Call to Action" meeting to address SCDY was attended by approximately 60 individuals representing a diverse group of organizations that included professional medical societies, hospitals, health plans, industry, faith-based groups, academia, and foundations. The attendees were asked to identify the most appropriate and feasible immediate next steps to prevent SCDY in Michigan. Since 2008, over 50 additional individuals have joined our efforts to prevent SCDY. Table 5 provides an overview and status report on key action steps identified as a result of the original mortality review process, with progress in specific areas highlighted below.

> "I thought we were forgotten.... I thought no one cared..."
>
> — Mother of 18 year old victim, upon being asked for a next-of-kin interview

Provider Education on SCDY Risk Assessment and Public Awareness Of Risk Factors

From 2009-2011, a multitude of educational resources were created —including a website, video, and expert presentations—to increase provider and public awareness of personal and family history risk factors. In addition, several press releases and newspaper articles drew public attention to the issue of SCDY. Many partners held educational events to increase public and provider awareness and prevention of SCDY; and there have been numerous presentations at national and state conferences about Michigan's efforts to prevent SCDY. For example, in September 2011, educational initiatives were implemented by three key partners. The University of Michigan held an event to increase provider knowledge regarding SCDY in Ann Arbor (view at: http://www.uofmhealth.org/protectinghearts). Spectrum Health conducted public service announcements including local radio and television interviews and held an educational event in Grand Rapids to increase public awareness about SCDY risk factors. Genessee County Health Department and Community Based Organization Partners (CBOP) held an event in Flint to increase public awareness about SCDY risk factors. It is estimated that over 10,000 individuals were reached regarding SCDY with these three activities alone. Information regarding educational resources and upcoming events is available at www.michigan.gov/scdy.

Pre-participation Sports Screening

The Michigan High School Athletic Association (MHSAA) is a private, not-for-profit corporation with over 1,500 middle and high school members that agree to abide by MHSAA policies.[84] These schools have approximately 315,000 high school and middle school students who participate in school athletics per year. The MHSAA Handbook includes a regulation that 'no student shall be eligible to represent a high school for whom there is not on file in the offices of the superintendent or principal or athletic director of that school, statements for the current school year certifying that the student has passed a physical examination'.[85] From 2008-2010, MDCH convened a workgroup to make formal recommendations to MHSAA regarding updates to align its pre-participation sports screening history and exam forms with national recommendations.

In 2010, the workgroup's recommended form was provided to MHSAA with letters of support from over 20 Michigan professional organizations and advocacy groups. MHSAA subsequently approved this recommendation and updated its medical history form template.[86,87] The history and exam portions of the form can now be accessed at: www.michigan.gov/scdy or www.MHSAA.com. The history form, including 12 questions to assess SCDY risk factors, must be completed by an adult parent or guardian unless the student-athlete is 18 years of age or older. If the personal, family history and/or exam reveal significant findings, a referral to cardiology for consideration of appropriate evaluations, such as an ECG and/or echocardiogram, are suggested prior to clearance of the athlete.

MHSAA is now one of the first state athletic organizations in the United States to be aligned with current national recommendations. In 2011, MHSAA created an awareness campaign to disseminate health messages regarding heart, head, heat and medical history for their member organizations.[88] With this important policy change, it is hoped that public awareness about SCDY risk factors and provider practice regarding collection and documentation of SCDY risk factors in Michigan will increase.

Public Awareness of Cardiac Symptoms and AED/CPR Training

Public awareness of SCDY and the need to recognize cardiac symptoms appears to be increasing in Michigan. For instance, April 2010 and April 2011 were declared Michigan Student Athlete Awareness Month by the Michigan House of Representatives and Senate as a statewide effort to increase awareness and prevention of sudden cardiac events in students.[89]

In 2011, the MHSAA conducted a survey that revealed 93% of schools who responded (n=346) have AEDs; however, only 73% indicated that an AED is readily available at all athletic venues.[90] Furthermore, 67% of respondents reported that there is no requirement by their school for coaches to be certified to provide CPR/AED or first aid assistance. Several Michigan foundations, created by families whose child died as a result of SCDY at a school athletic event, have continued to donate AEDs to schools. Additionally, the first Michigan chapter of the Sudden Cardiac Arrest Association (SCAA) was formed in 2011 by Beaumont Hospital. SCAA is an organization that works to increase awareness of SCA and public access to defibrillation. Additional information on public awareness is available at www.michigan.gov/scdy

There is currently no state law that mandates placement of AEDs in schools; however in 2011 and 2012, bills were introduced in the state legislature that would require schools to have AEDs in place, emergency response plans, and required AED training for personnel. Michigan already has a Good Samaritan Act for CPR and AED that protects individuals who in good faith voluntarily render CPR and/or AED to another individual from civil action damages.[91] There is currently no federal or state funding to implement CPR and/or AED training and placement in schools. MDCH and Michigan pediatric cardiologists are therefore investigating the feasibility of becoming an affiliate of Project ADAM to provide CPR and AED trainings and resources to schools at a voluntary community level.

Medical Examiner Protocols

In 2011, MDCH worked with University of Michigan cardiologists and the Washtenaw County Medical Examiner to create a standardized checklist for SCDY cases for integration into a new electronic death certificate reporting system. MDCH has also been working with the University of Michigan and interested Michigan pathologists to further investigate causes of SCDY through autopsy reports. Activities regarding medical examiner protocols will be a primary focus and further explored in coming years.

Future Steps for Prevention of SCDY

In 2010, MDCH and partners were pleased to learn that the NHLBI convened a Working Group to develop a national research agenda and to identify resources to evaluate whether screening for SCDY would effectively reduce SCD and add health care value. The Working Group was unanimous in stating the importance of preventing SCD, but decided that more research is required to determine the best national approach to achieve this goal. Specific areas to be addressed were strikingly similar to our Michigan work, including prospectively defining the incidence and epidemiology of SCDY; the development and evaluation of protocols for screening populations for SCDY; and, protocols for management of individuals with a known condition at risk for SCDY.[91,92] The NHLBI research agenda includes development of a SCDY registry, with resources leveraged through existing infrastructure of the National Center for Child Death Review using state-based child death review teams to assist with this activity.[91] The National Center for Child Death Review is administered by the Michigan Public Health Institute, and has recently become a new partner involved with our efforts. For instance, MDCH recently authored a SCDY factsheet to be utilized by state-based child death review teams which can be found at: http://www.childdeathreview.org/causesNOO.htm. MDCH will continue to work with the National Center for Child Death Review, NHLBI and our partners to determine the best approach to prevention of SCDY. For further updates or to become involved in our efforts, please visit www.michigan.gov/scdy

Table 5
Overview of Action Steps from Expert Panel Review & Current Progress to Prevent SCDY

Themes	Action Steps Identified by Expert Panel	2009-2011 Progress
Provider Education and Public Awareness of SCDY Risk Factors	Increase public awareness and provider assessment of SCDY risk factors & family history, through various means and multiple different health system contact points	Events for providers and public in 2009-2011
	Identify clinical screening protocols for those with family history of SCDY	Completed in 2010
	Consider developing continuing medical education regarding SCDY risk factors, assessment, referral and treatment for health providers	Provider education events in 2009-2011
	Emphasize the need for case management for young adults with chronic disorders (such as diabetes, hypertension, high cholesterol, heart defects, & obesity)	Ongoing
	Identify options for obtaining medications for uninsured and underinsured individuals at risk for SCDY, and disseminate to providers	No progress
	Increase awareness and access to appropriate medical and/or genetic services for individuals with a family history of SCDY	Ongoing
Pre-Participation Sports Screening/ Physical and Follow-up	Make recommendations to MHSAA to revise sports participation form based on mortality case findings and national guidelines	Completed in 2010
	Identify and disseminate clinical guidelines for evaluation and management of students with known conditions at risk for SCDY	Expert presentations in 2010-2011
	Identify and/or disseminate recommendations to coaches about symptoms and/or conditions that may place athletes at higher risk	Events held for coaches in 2011
Public Awareness of Cardiac Symptoms & CPR/AED Training	Explore existing state mandates for CPR and AED training	Ongoing
	Increase appropriate availability of AEDs and trained users	Ongoing
	Increase awareness of cardiac symptoms and appropriate actions	Ongoing
	Increase public and community knowledge of CPR	Ongoing
Emergency Response Protocols	Explore appropriateness of response time and initiation of immediate defibrillation for SCA cases; provide feedback to EMTs	Ongoing
	Investigate availability of AEDs for all responders	Ongoing
	Make formal clinical recommendations regarding appropriate use of medications in SCDY situations to appropriate EMS organizations	No progress
Medical Examiner Protocols	Suggest consideration of DNA banking for SCDY cases; explore expense and insurance reimbursement options for DNA banking	Ongoing
	Increase awareness of familial risks among medical examiners	Ongoing
	Develop mechanism to ensure autopsy results and recommended follow-up are conveyed to families and primary providers	Ongoing
	Develop suggested protocols for autopsy of SCDY cases	Ongoing
	Consider pilot to independently assess pacemakers function after SCDY	No progress

Appendix A

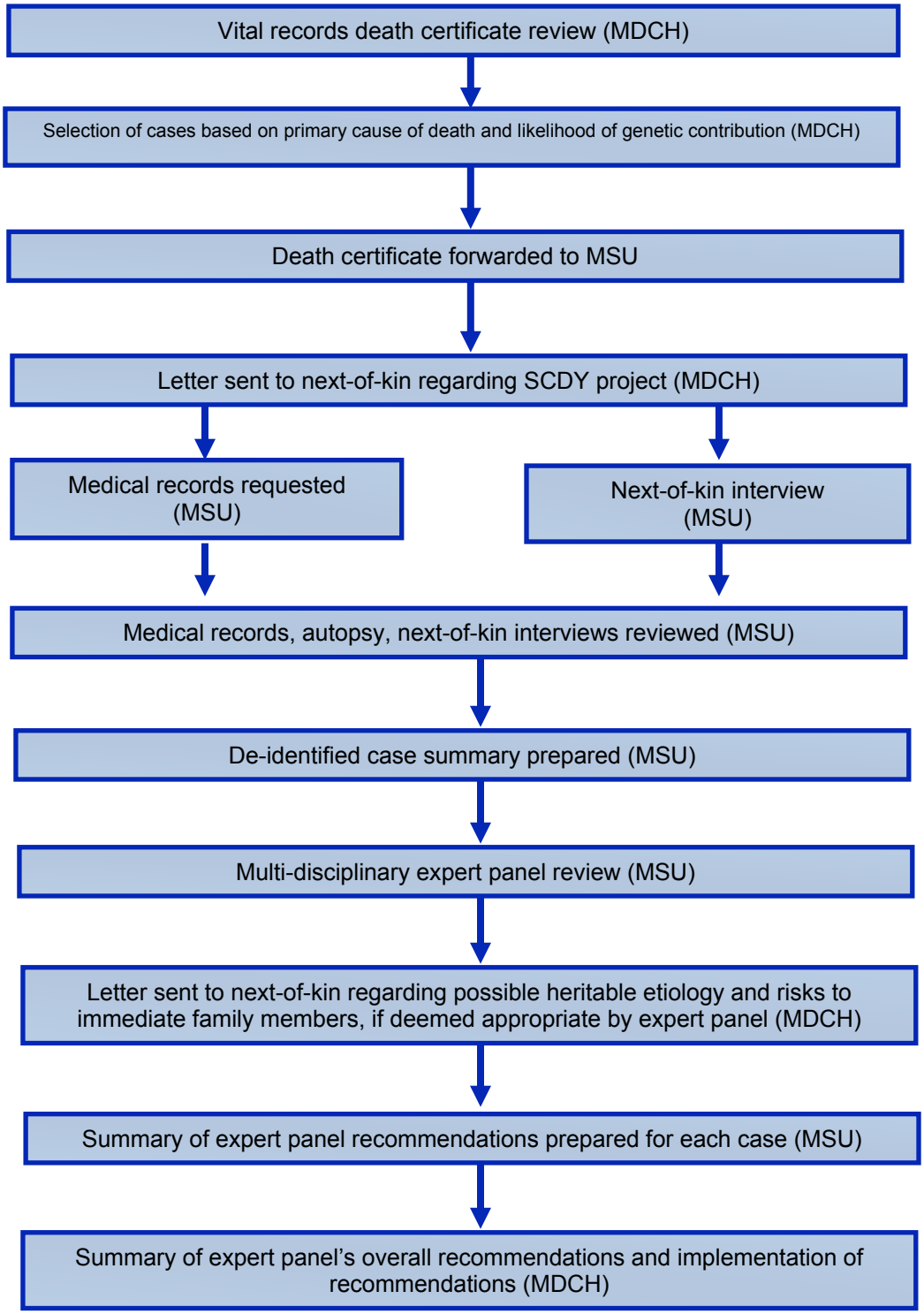
Development of SCDY Data to Action in Michigan

Highlights of Key Steps

- **Interviews with key medical experts**—provided important information about current practice and procedures following unexpected deaths, revealing considerable variation and lack of consistency in medical examiner post-mortem protocols from county to county.
- **Presentation to the state cardiovascular health advisory committee**—confirmed interest in developing a mortality review system.
- **Symposium on SCDY**—invited national and state experts to review causes of SCDY and set priorities; stakeholders (including family members and advocacy groups) identified the need for a state surveillance system and awareness campaign.
- **Analysis of existing mortality records**—reviewed death certificate data from 1999-2009 retrospectively to estimate burden of SCDY.
- **Population survey**—provided estimates of how many Michigan families are affected by SCDY based on answers to a question on the 2007 BRFS (a BRFS random-digit-dialed telephone survey) about family history of SCD in young relatives.
- **SCDY investigation protocol and data collection instruments**—reviewed by MDCH Institutional Review Board (IRB), and project deemed to be of public health surveillance.
- **MDCH Medical Research Designation**—issued by MDCH Chief Medical Executive as allowed by the Public Health Code to assure that all information collected during the project is kept confidential and used only for public health purposes.
- **Michigan SCDY Expert Review Panel**—met for the first time in October, 2007 to review four SCDY cases. The panel met twice in 2008 to review nineteen SCDY cases. Sixteen next-of-kin were interviewed. Twenty-one action items to primary and secondary prevention of SCDY were identified. Published in Journal of Community Health in 2010 (9).
- **Call to Action Event –** held on September 18, 2008 with 60 participants to discuss and identify key next steps to prevent SCDY in Michigan.
- **Listserv Formed**- listserv was created in September 2008 and includes individuals interested in next steps to prevent SCDY in Michigan.
- **Workgroups Formed**- three workgroups were formed in October 2008 to address the next steps to prevent SCDY in Michigan; one is the pre-participation sports screening workgroup which has over 55 members.
- **Consensus Based Recommendations for Pre-Participation Sports Screening adopted by MHSAA-** pre-participation sports screening workgroup reviewed published literature and existing forms in all 50 states. Workgroup and over 20 Michigan organizations approved form based on the 2010 consensus form from the American Academy of Pediatrics, American Academy of Family Physicians, et al and the 2007 12-point American Heart Association recommendation. The workgroup's form was approved by MHSAA in December 2010 and began dissemination in 2011.
- **MDCH SCDY Website & Other Resources Created-** website created at www.michigan.gov/scdy; video created to increase awareness of SCDY and steps to prevent; expert web-based presentations on a variety of SCDY topics created.

Appendix B

SCDY Expert Mortality Review Process and Roles of
Michigan Department of Community Health (MDCH) & Michigan State University (MSU)



Appendix C

International Classification of Diseases and Related Health Problems (ICD-10) Codes Utilized for SCDY Case Definition

Code	Disease
I00-I09	Rheumatic heart disease
I11	Hypertensive heart disease
I20-I25	Myocardial infarction; Atherosclerotic disease
I30-I31	Pericardium disease
I33	Endocardium disease
I34-I38	Valve disorders
I40	Myocarditis
I42	Cardiomyopathy
I44-I45	Conduction disorders
I46	Cardiac arrest
I47-I49	Cardiac dysrhythmias
I50	Heart failure
I51	Complications and ill-defined heart disease
Q20-Q24	Congenital abnormalities of the heart
Q87.4	Marfan syndrome
R96-R99	Ill-defined causes of death

Appendix D

Age-Adjusted Mortality Rates by County of Residence

Ten-year age-adjusted mortality rate per 100,000 persons aged 1-39 years of age, 1999-2009

County	Rate	95% Confidence Interval	Number of SCDs
Alcona	-	-	-
Alger	-	-	-
Allegan	3.6	(2.1-5.0)	22
Alpena	7.2	(2.7-11.7)	10
Antrim	3.9	(0.5-7.4)	5
Arenac	-	-	-
Baraga	-	-	-
Barry	2.5	(0.8-4.3)	8
Bay	6.1	(4.0-8.1)	34
Benzie	.	-	-
Berrien	6.4	(4.7-8.2)	53
Branch	5.4	(2.6-8.2)	14
Calhoun	5.1	(3.5-6.7)	38
Cass	3.5	(1.2-5.7)	9
Charlevoix	3.8	(0.5-7.1)	5
Cheboygan	4.3	(0.8-7.7)	6
Chippewa	3.5	(1.0-6.0)	8
Clare	7.1	(2.9-11.3)	11
Clinton	2.9	(1.2-4.7)	11
Crawford	-	-	-
Delta	4.4	(1.3-7.5)	8
Dickinson	4.8	(1.0-8.6)	6
Eaton	3.2	(1.7-4.7)	18
Emmet	5.0	(1.5-8.4)	8
Genesee	6.7	(5.7-7.7)	164
Gladwin	-	-	-
Gogebic	6.3	(0.8-11.7)	5
Grand Traverse	4.7	(2.7-6.6)	21
Gratiot	4.6	(1.8-7.3)	11
Hillsdale	6.2	(3.0-9.4)	15
Houghton	2.6	(0.1-5.1)	5
Huron	5.7	(2.0-9.4)	9
Ingham	4.6	(3.7-5.9)	78
Ionia	3.1	(1.3-4.8)	12
Iosco	-	-	-
Iron	-	-	-
Isabella	4.2	(1.9-6.5)	15
Jackson	5.5	(3.9-7.0)	50
Kalamazoo	3.9	(2.8-5.0)	53
Kalkaska	5.4	(0.7-10.1)	5
Kent	4.1	(3.4-4.8)	146
Keweenaw	-	-	-
Lake	-	-	-
Lapeer	1.5	(0.7-2.3)	14

County	Rate	95% Confidence Interval	Number of SCDs
Leelanau	.	.	.
Lenawee	1.7	(0.9-2.5)	17
Livingston	1.5	(0.9-2.1)	27
Luce	-	-	-
Mackinac	-	-	-
Macomb	2.3	(2.0-2.6)	203
Manistee	-	-	-
Marquette	1.0	(0.2-1.9)	6
Mason	5.0	(2.3-7.8)	13
Mecosta	2.5	(0.9-4.2)	10
Menominee	2.2	(0.3-4.2)	5
Midland	1.7	(0.8-2.6)	14
Missaukee	4.4	(0.9-7.9)	6
Monroe	2.9	(2.0-3.8)	44
Montcalm	5.4	(3.6-7.1)	35
Montmorency	-	-	-
Muskegon	4.0	(3.0-4.9)	68
Newaygo	2.9	(1.3-4.4)	13
Oakland	2.0	(1.7-2.2)	251
Oceana	3.0	(0.9-5.1)	8
Ogemaw	-	-	-
Ontonagon	-	-	
Osceola	3.4	(0.9-6.0)	7
Oscoda	-	-	-
Otsego	4.6	(1.9-7.3)	11
Ottawa	1.2	(0.8-1.6)	32
Presque Isle	-	-	-
Roscommon	5.4	(2.0-8.8)	10
Saginaw	3.2	(2.4-4.0)	62
St. Clair	2.4	(1.7-3.2)	42
St. Joseph	2.3	(1.1-3.5)	14
Sanilac	4.1	(2.2-6.1)	17
Schoolcraft	-	-	-
Shiawassee	3.1	(1.8-4.4)	22
Tuscola	2.5	(1.1-3.8)	13
Van Buren	1.8	(0.8-2.8)	13
Washtenaw	1.6	(1.2-2.0)	63
Wayne	5.6	(5.2-5.9)	1166
Wexford	2.3	(0.5-4.1)	7

Source: MDCH Vital Statistics
- Insufficient data, less than five deaths in the county

Appendix E

Common Causes of Sudden Cardiac Death

*single or multiple genes play a role in determining susceptibility to most of these conditions

Anatomical Abnormalities	
Aortic root aneurysm/ dissection	Some aortic root aneurysms can be caused by genetic connective tissue diseases that weaken the aortic wall causing aortic aneurysm and subsequent death due to aortic dissection. Examples include Marfan syndrome, familial thoracic aortic aneurysms and dissections, and vascular type Ehlers-Danlos syndrome.
Aortic stenosis	An abnormal narrowing of the aortic valve that impairs blood flow to the arteries and can lead to heart failure, this is three times more common in men than women.
Dilated cardiomyopathy (DCM)	Left ventricular enlargement and systolic dysfunction, which usually results in heart failure with symptoms of congestion and/or reduced cardiac output, arrhythmias, and thromboembolic disease, including stroke. Genetic and acquired (non-genetic/idiopathic) forms exist.
Hypertrophic cardiomyopathy (HCM)	Unexplained left ventricular wall thickening in the presence of a non-dilated left ventricular cavity. HCM can often be genetic. It occurs in 1 in 500 individuals, and is reportedly the most common cause of SCDY in U.S. athletes.
Mitral valve prolapse	A "floppy" heart valve that can lead to irregular or rapid heartbeats and shortness of breath. Some may be caused by Marfan syndrome or Ehlers-Danlos syndrome.
Myocarditis/ endocarditis	An inflammation of the heart muscle, usually caused by viral infection but may occur as a complication of other medical conditions or exposure to drugs.
Arrhythmias	
Arrhythmogenic right ventricular cardiomyopathy (ARVC)	The heart muscle becomes thin due to an abnormal amount of fat and scar tissue in its walls. Mainly affects the right side of the heart. Most common cause of SCDY in Italian athletes.
Brugada syndrome	An electrocardiographic pattern of right bundle branch block, ST segment elevation in leads V1 to V3, and sudden death. More common in young males and in Southeast Asians.
Catacholaminergic polymorphic ventricular tachycardia (CPVT)	Arrhythmogenic disorder characterized by exercise, stress- or emotional-induced ventricular tachycardia, syncope or sudden death in the absence of detectable structural heart disease.
Long QT syndrome (LQTS)	A disorder of the heart's electrical rhythm leading to an increase in the QT interval that is often asymptomatic. Symptoms may include fainting and an abnormal rate and/or rhythm of the heartbeat (arrhythmia). Deafness is also associated with one type of inherited LQTS.
Short QT syndrome	A disorder of the heart's electrical rhythm leading to a decrease in the QT interval. If untreated, irregular heartbeats can lead to symptoms including dizziness, fainting, cardiac arrest and sudden death.
Wolff-Parkinson-White syndrome (WPW)	A condition that results in an extra conduction pathway for electrical signals within the heart. Symptoms include dizziness, chest palpitations, fainting and rarely cardiac arrest.
Vascular Abnormalities	
Coronary artery abnormality	A malformation of an artery, present at birth but often undetected.
Coronary artery disease (CAD)/Atherosclerosis	Plaque build-up narrows the coronary arteries and reduces blood flow to the heart muscle. Blood clots are more likely to form, blocking blood flow. Over time, CAD can weaken the heart muscle, leading to heart failure or arrhythmias.

Appendix F

Suggested history questions from the American Academy of Pediatrics et al., 2010

1. Have you ever passed out or nearly passed out DURING or AFTER exercise?
2. Have you ever had discomfort, pain, tightness, or pressure in your chest during exercise?
3. Does your heart ever race or skip beats (irregular beats) during exercise?
4. Has a doctor ever told you that you have any heart problems (high blood pressure, high cholesterol, a heart murmur, a heart infection, Kawasaki disease, or other)?
5. Has a doctor ever ordered a test for your heart (for example, ECG/EKG or echocardiogram)?
6. Do you get lightheaded or feel more short of breath than expected during exercise?
7. Have you ever had an unexplained seizure?
8. Do you get more tired or short of breath more quickly than your friends during exercise?
9. Has any family member or relative died of heart problems or had any unexpected or unexplained sudden death before age 50 (including drowning, unexplained car accident, or SIDS)?
10. Does anyone in your family have hypertrophic cardiomyopathy, Marfan syndrome, arrhythmogenic right ventricular cardiomyopathy, long QT syndrome, short QT syndrome, Brugada syndrome, or cateholaminergic polymorphic ventricular tachycardia?
11. Does anyone in your family have a heart problem, pacemaker or implanted defibrillator?
12. Has anyone in your family had unexplained fainting, unexplained seizures, or near drowning?

Appendix G
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The Michigan Department of Community Health is committed to the vision that "Michigan will be a safe and healthy state where all people realize their fullest health potential and live enriched and productive lives". Every life lost to SCD of the young distances us from achieving this vision. Together with our partners, MDCH will continue efforts to prevent SCDY in Michigan.

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