Improving Forensic Pathologic Investigation of Sudden Death in the Young: Tools, Guidance, and Methods of Cardiovascular Dissection from the Sudden Death in the Young Case Registry

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ABSTRACT

The Sudden Death in the Young (SDY) Case Registry, a prospective, population-based registry active in ten states, has developed tools to aid pathologists and death investigators in the evaluation and autopsy of unexplained, natural sudden deaths in the pediatric population. The tools were developed by a team of experts representing forensic pathology; pediatric-, cardiac-, and neuropathology; cardiology; neurology/epileptology; pediatrics; genetic counseling; and public health. These tools focus on collecting data relevant to determination of cause of death with a focus on dissection of the cardiovascular system. The tools provide an objective checklist format for ease of use and data extraction. By sharing the tools here and highlighting the examination of the cardiovascular system, the SDY Case Registry encourages a standardized approach to death investigation, autopsy, and data collection for sudden, unexpected deaths in the young towards a goal of informing prevention efforts. *Acad Forensic Pathol.* 2018 8(2): 347-391

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INFORMATION

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INTRODUCTION

The sudden death of a child is a devastating event that leaves family members, medical providers, and the community searching for answers. Some causes of sudden death in the young (SDY) are identifiable at autopsy (e.g., infections, motor vehicle accidents, suicide by hanging, or homicide by gunshot wound); but in many cases of sudden death that present to the medical examiner or coroner (ME/C), the history, autopsy, and death scene investigation do not reveal the cause of death (1).

Some medical conditions associated with SDY may be inherited (e.g., long QT syndrome and hypertrophic cardiomyopathy), and surviving family members may not know that they or their other children are at risk. Identifying heritable causes of death in SDY cases is critical to initiate cascade screening of surviving family members and to facilitate access to therapies that may lower their risk of sudden death. In cases of SDY with no identifiable cause at autopsy, there is a critical knowledge gap concerning mechanisms, clinically identifiable features, and epidemiology, fueled partly by a lack of standardized procedures in the United States for investigating, classifying, and reporting SDY. Medical examiners, coroners, and death scene investigators are critical partners in the effort to gather high-quality information to address this knowledge gap and to improve the understanding of SDY.

Most sudden deaths in infancy or childhood will fall under the jurisdiction of ME/C because they occur suddenly, unexpectedly, and in a person in apparently good health. In most states, the determination of whether to perform an autopsy on an SDY case is left to the discretion of the ME/C. Only a small minority of states specifically require an autopsy by law as part of the investigation of SDY (2). When an autopsy is performed, the extent of the autopsy and its ancillary testing may vary by jurisdiction (3, 4). This variation may be due to funding, ME/C preferences, prior experience and training of the ME or coroner's pathologist performing the autopsy, and ME/C workload. Funding for autopsies and ME/C staffing is determined by state or county budgets that may have limited resources

and many competing priorities. In addition, most states are currently experiencing an increase in deaths due to the opioid epidemic, resulting in a significant burden on the death investigation system that reduces the time and resources that ME/C can spend on the investigation of natural deaths. Despite limits on the time and budgets, many ME/C are dedicated to improving the understanding of these unexpected infant and child deaths.

To facilitate improved understanding of SDY and ultimately inform prevention efforts, the Sudden Death in the Young Case Registry created tools and guidance to encourage standardized, comprehensive autopsies and death scene investigations for SDY cases. The tools focus on important elements of the autopsy that are relevant to sudden, unexpected, natural deaths in infants, children, and adolescents. We share these tools herein and provide specific guidance on the performance of the cardiac examination, given the high incidence of cardiac causes of SDY in children over one year of age. We hope that these tools may be useful in the ME/C community and may promote more standardized approaches to characterizing, classifying, understanding, and ultimately preventing sudden, unexpected natural deaths in the young.

DISCUSSION

The Sudden Death in the Young (SDY) Case Registry

The SDY Case Registry is a collaborative effort between the National Institutes of Health (NIH), the Centers for Disease Control and Prevention (CDC), and the Michigan Public Health Institute (MPHI) to address the knowledge gaps in the epidemiology and causes of SDY in the United States (5). The SDY Case Registry includes infants and children from birth through 19 years of age in seven states and specific regions of three additional states. These selected jurisdictions cover 24% of the pediatric population in the United States and approximately 20% of SDY. The SDY Case Registry is one of the largest prospective, population-based cohorts to date of children who have died suddenly in the United States.

The goals of the SDY Case Registry are to: 1) determine the incidence of SDY using population-based surveillance in a large cohort in the United States; 2) compile data from SDY cases to create a resource of phenotypic and genotypic information for research; 3) encourage standardized approaches to investigation, autopsy, and categorization of SDY cases; 4) develop partnerships between local, state, and federal stakeholders towards the common goal of understanding and preventing SDY; and 5) support families who have lost loved ones to SDY by providing resources for bereavement and medical evaluation of surviving family members.

Figure 1 illustrates the flow of cases through the SDY Case Registry. Sudden deaths in infants and children are identified through ME/C offices, where the inclusion and exclusion criteria are applied. Since the SDY Case Registry is population-based, the decedent must have been a resident of the participating jurisdiction or state to be eligible. The registry does not discriminate between witnessed and unwitnessed deaths. Inclusion criteria include: sudden and unexpected death; age less than 20 years; death not explained by a homicide, suicide, drug overdose, or terminal illness; and death due to injury when there may have been an inciting natural cause (e.g., drowning or death of the driver in a motor vehicle accident, which may have been triggered by an underlying cardiac or neurological condition). “Sudden” is defined as death within 24 hours of the first symptom, or death under medical

care after resuscitation from an out-of-hospital cardiac arrest. Although some studies define the window of sudden death as one hour from symptom onset, the SDY Case Registry uses a window of 24 hours due to the high number of infant and childhood deaths that occur during sleep. Comorbid conditions with unrelated symptoms that might overlap this window are expected to have a low incidence, simply because sudden childhood deaths after infancy are so rare. “Unexpected” is defined as the death of an infant or child who was in good health, had either a stable, chronic condition, or had an acute illness that would not be expected to cause death.

Once included, each case is reviewed by a local or statewide child death review team (6), followed by an advanced review by a team of local clinical experts that also categorizes the case using a standardized algorithm to enable an SDY incidence rate to be calculated (5). Review teams are composed of experts in forensic pathology, cardiology, neurology/epileptology, pediatrics, genetic counseling, and public health, among other subject matter experts.

Use of the SDY Case Registry tools for death scene investigation and autopsy is encouraged to promote standardization, as well as to offer the chance of obtaining wide-ranging investigative and autopsy information. During the autopsy, pathologists collect samples of blood and tissue for DNA extraction. Blood in an EDTA tube is the preferred sample type, but

Figure 1: Case flow in the Sudden Death in the Young Case Registry.

fresh frozen liver and/or heart tissue serve as a back-up if the blood does not yield usable DNA. Currently, blood spot cards, formalin fixed tissue, and paraffin fixed tissue do not yield a reliable, adequate quality and quantity of DNA to allow for exome/genome sequencing. Blood and tissue samples are shipped to the SDY Case Registry Biorepository for DNA extraction and storage. The biorepository has a 99% success rate in obtaining adequate quality and quantity of DNA.

For all SDY cases, next of kin consent is sought for diagnostic genetic testing, future research studies that may or may not involve genetic testing, DNA banking, and return of genetic results. Although consent is not required for diagnostic genetic testing, the SDY Case Registry sought advice from ethicists and opted to obtain consent for diagnostic genetic testing as part of a menu of options due to the implications the results may have for the family (7).

The DNA may be used for genetic research by NIH-funded investigators focused on determining the causes of SDY as well as via diagnostic genetic testing. Invitae, a genetic testing company, has partnered with the MPHI and the Biorepository to offer in-kind (donated) diagnostic genetic testing to 900 cases in the SDY Case Registry over a three-year period. Genetic testing is offered on autopsy-negative cases as well as cases suggestive of cardiomyopathy using a panel of arrhythmia and cardiomyopathy genes. Such testing may provide additional information that may aid the ME/C in determining the cause of death and inform the approach to cascade screening of surviving family members. Access to such postmortem genetic testing for the ME/C community has previously been limited due to cost.

Tools to Assist With Autopsy and Death Scene Investigation of SDY

The SDY Case Registry sought guidance from an autopsy protocol task force comprised of ME/C; death investigators; pediatric-, cardiac-, and neuropathologists; genetic counselors; emergency room physicians; and public health experts (**Appendix 1**). After reviewing existing autopsy protocols for SDY from Australia

(8), Ireland (9), Canada (10), and the United Kingdom (11), the task force developed a standardized, comprehensive recommended approach to an SDY Case Registry autopsy and a protocol for sample collection for research and diagnostic genetic testing and DNA banking. The task force also developed tools to facilitate a standardized approach to death investigation. Materials and resources have been created by currently funded SDY states and the SDY technical assistance team for family referral for medical screening and genetic counseling for SDY cases (**Appendix 2 and 3**).

The SDY Field Investigation Guide and Family Interview Summary Sheet

The SDY Field Investigation Guide and Family Interview Tool (**Appendix 4**) was designed for death scene investigators and includes a series of questions, in both checklist and narrative formats, for the family and medical providers to obtain a comprehensive review of the history and circumstances surrounding the child's death. The tool begins with a list of suggested records to collect on the decedent before fatality review, and goes on to include questions ranging from general information about the family to specific information about the events leading up to the child's death. Although experienced investigators will ask many of these questions without the guide, the tool provides a helpful reminder of a comprehensive list of questions about triggers prior to death, symptoms within 24 and 72 hours of death, family history of sudden death, and clues that may help the pathologist identify genetic syndromes. Because the guide offers a standardized list of questions, the same comprehensive review can be performed for every SDY case.

The SDY Case Registry Autopsy Guidance and Summary Tools

The SDY Case Registry autopsy guidance tools focus on medical conditions and other anatomic findings that may be associated with SDY. They include a recommended comprehensive cardiovascular examination due to the high incidence of cardiac causes of SDY in children over age one. The tools are in checklist format to enable rapid and easy use by patholo-

gists. The checklist format also discourages narrative description to facilitate categorization of the cases and easier data mining.

The autopsy guidance tools are available in two forms: 1) the guidance tool and 2) the summary tool. The SDY Autopsy Guidance (**Appendix 5**) is 20 pages long, and although it is less practical for everyday use in the field, it is a valuable resource outlining the elements of a comprehensive SDY-related autopsy. Although the autopsy guidance form may seem at first glance to be time consuming because of its length, with its checklist format and repeated use, it becomes a rapid and easy form to fill out. The SDY Autopsy Summary (**Appendix 6**) is a checklist that is used to guide the pathologist in participating jurisdictions to the critical information that needs to be collected from the SDY Case Registry autopsy. The autopsy summary tool offers the forensic pathologist the chance to ask questions deemed important to the pediatric SDY autopsy by experts on the task force with additional training in several tissue and organ systems.

Many individual ME/C offices have their own existing forms for autopsies, and some offices participating in the SDY Case Registry have added questions from the SDY tools into their own forms. Integration of the SDY Autopsy Summary questions into an existing pediatric autopsy protocol may improve data collection on all pediatric cases for a given office, but states and jurisdictions participating in the SDY Case Registry are strongly encouraged to use the guidance and summary tools to improve data quality for the Registry. After becoming familiar with the tools, pathologists may incorporate elements of the SDY Case Registry summary tool into their routine practice and it may become easier to judge when an organ or system deserves additional expert consultation.

Recommended Practices for SDY Case Registry Autopsies

The following paragraphs describe the recommended practice by the SDY Case Registry to perform a comprehensive autopsy on cases of sudden, unexpected, natural death of a child. This section pays particular

attention to the cardiac examination, given the importance of cardiac causes in such deaths of children, especially over age one. This guidance also highlights some subtle features that deserve specific attention in such cases, and that may help to identify heritable diagnoses with implications for surviving family members.

The SDY Case Registry autopsy tools recommend a complete autopsy. Limited autopsy runs the risk of failing to detect information that may be critical to determining the cause and manner of death, as well as data that may be valuable for research.

After recording basic demographic and identifying information, record the sex, weight, body length, and a general description of body habitus and development. In children three years of age and under, record the head circumference. Use standardized growth charts to plot the anthropometric data and compare it to antemortem measurements, if any are available. These are used to provide a quick glimpse into the child's physical development and to identify a child whose development has been impaired by genetic or acquired chronic illnesses, abuse, neglect, or poor nutrition. The external examination includes the usual descriptions of scars, birthmarks, trauma, medical interventions, and physical stigmata of diseases and syndromes. Significant findings are often most effectively captured with photography as well as narrative; this may include pertinent negative findings.

Radiography is an important adjunct to the autopsy investigation of sudden death. Whole body radiography (so-called "babygrams") may be useful in demonstrating skeletal dysplasias, syndromes, and metabolic disorders (12), but they are technically inadequate for the demonstration of metaphyseal fractures (13). For this reason, a skeletal survey is recommended as part of the postmortem evaluation of infants and small children in cases suspicious for fatal abuse (14). Since the full circumstances of the child's death may not be known before the autopsy is performed, consideration should be given to performing a skeletal survey in all cases of sudden, unexplained death in infants and young children (13). In older children, in addition

to the demonstration of fractures, plain-film radiography of the chest can be useful in individual cases for demonstration of gas embolism, pneumothorax, and pneumomediastinum (15).

There has been much interest in the use of computed tomography (CT) and magnetic resonance imaging (MRI) as an adjunct to or replacement for autopsy, particularly in the setting of trauma. Few studies have been done on the use of so-called “virtual autopsy” specifically in cases of sudden unexplained deaths in infants and small children (16, 17). These studies did not demonstrate an ability to diagnose specific non-traumatic cardiovascular disease, aside from the presence of cardiac hypertrophy. This is inadequate, since a full understanding of the causes of SDY is needed, both in individual cases and across the population. In addition to the limited access most ME/C have to these expensive imaging modalities, interpretation by a radiologist who is experienced in interpreting post-mortem CT/MRI images is required, as postmortem changes may produce artifacts that can mask or simulate pathology (18, 19). Thus, at present, postmortem CT/MRI imaging may be a useful adjunct to the autopsy evaluation of SDY, but should not replace it.

The SDY Case Registry autopsy guidance prompts careful examination of a variety of causes of SDY, including pulmonary etiologies (e.g., embolism, hypertension, pneumothorax), gastrointestinal catastrophe (e.g., volvulus, obstruction, intussusception), infection (e.g., meningitis, myocarditis, sepsis), and neurologic abnormalities (e.g., stroke, hemorrhage, herniation), among others. The SDY Case Registry encourages consultation with experts with additional training in pediatric-, cardiac-, and neuropathology, when feasible, to help identify subtle features of underlying disease.

Cardiovascular Examination

Detailed guides to the examination of the heart have been published elsewhere (20, 21). The following is intended to provide guidance that will suit the examination of most cases of SDY. However, in cases with complex pathology, especially cases involving

congenital heart disease or implanted cardiac devices, consultation with a cardiovascular pathologist is strongly recommended.

Prior to removal of the heart, the pericardium should be assessed for exudates, thickening, or calcification and any effusion or hemorrhage in the pericardial space should be noted and measured. The position and apical direction of the heart should then be documented. The heart may then be removed separately or in a block with the other thoracic organs. En bloc removal is particularly suited to situations in which congenital heart disease is suspected, as it facilitates examination of the arterial and venous connections of the heart, and for disorders involving the proximal portions of the aorta, such as aortic dissection.

After the removal of extraneous extra-cardiac soft tissue, the heart should be weighed. If, during subsequent dissection, it is found that the cardiac chambers contain abundant clotted blood, it is recommended that the heart be re-weighed after dissection and removal of the blood clots.

Prior to opening the heart, the epicardial arteries should then be evaluated by serial sectioning beginning near the aortic root and extending distally for as far as a distinct lumen can be identified. In older children and adolescents, the distance between sections should not exceed 5 mm. In addition, care should be taken not to cut into the ascending aorta and thus disrupt the coronary ostia. Coronary dominance should be noted by determining whether the posterior descending artery arises from the right coronary artery, left circumflex coronary artery, or both. While routine microscopic examination of the coronary arteries is not necessary in most SDY cases, any grossly-observed abnormalities should be submitted for histology.

Multiple specialized methods of cardiac dissection, including methods that reproduce standard echocardiographic views, have been described. However, for most SDY cases, the heart is best examined using the inflow-outflow method (i.e., cutting along the flow of blood), with or without first sectioning the heart along its short axis.

Serial short-axis sections expose the greatest surface area of myocardium and facilitate measurement of the ventricular chambers and walls. It is, therefore, the preferred method for demonstrating myocardial pathology that can result in changes in myocardial color or texture (e.g., infarction, fibrosis, myocarditis), ventricular wall thickness, and chamber size. However, there are circumstances in which it may be desirable to skip short-axis sectioning. This is true of cases of known or suspected congenital heart disease, especially where examination of the muscular ventricular septum for defects is desired (such as a child who was known to have an uninvestigated cardiac murmur during life). Short-axis sections also may be unnecessarily difficult to perform when examining very small hearts, such as those of infants. In these cases, it is reasonable simply to proceed with opening the heart along the flow of blood as described below. When performed, short-axis sections should be made parallel to the base of the heart, visibly identifiable as the atrioventricular groove, beginning just above the apex and continuing superiorly until just below the tips of the left ventricular papillary muscles. These short-axis slices should be no more than about 10 mm in thickness, and will necessarily be much thinner in smaller hearts.

Sectioning the heart along the flow of blood (the so-called “inflow-outflow” method) begins with opening the right atrium from the inferior vena cava orifice to the tip of the right atrial appendage. This permits inspection of the entire right atrium, the atrial septum, and the tricuspid valve. It is not recommended that a further cut be made connecting the inferior and superior vena cavae, since such a cut will pass through the terminal crest of the right atrium, disrupting the sinoatrial node and SA nodal artery and making histologic examination of this portion of the conduction system difficult or impossible in cases where such an examination might be desirable.

The second cut is made through the posterior wall of the right ventricle, just lateral to the septum. This permits examination of the tricuspid valve and the endocardial surface of the right ventricle.

The third cut extends along the anterior wall of the right ventricle and right ventricular outflow tract through the pulmonic valve. This cut affords a view of the outflow portion of the right ventricle and the band of myocardium (the parietal band) that separates the tricuspid and pulmonic valves. The presence of the parietal band confirms that the morphologic right ventricle is correctly sided. The parietal band will be absent in congenital conditions in which the right-sided ventricle is a morphologic left ventricle (e.g., congenitally-corrected transposition of the great arteries).

The fourth cut connects the pulmonary vein orifices, with an additional cut out to the tip of the left atrial appendage. This permits a view of the interior of the left atrium, the left side of the atrial septum, and the mitral valve. This is an ideal time to inspect the atrial septum for septal defects and to determine if there is a patent foramen ovale.

This is also an ideal time to confirm that the heart is demonstrating normal sidedness, which is defined by the morphology of the atria. A morphologic right atrium will have a smooth posterior portion (derived from the sinus venosus) and pectinate muscle in its anterior portion and appendage (derived from the embryologic atrium). The smooth and pectinated parts of the right atrium meet at a ridge of muscle (the terminal crest). In contrast, a morphologic left atrium is entirely smooth-walled (being derived from the confluence of pulmonary veins) except for the appendage, which has pectinate muscles. In normal sidedness (*situs solitus*), the right- and left-sided atria will have the appropriate morphologic features described above. In a mirror-image heart (*situs inversus*), the right-sided atrium will have a left atrial morphology and the left-sided atrium will have right atrial morphology. In heterotaxy syndromes (*situs ambiguus*), both atria may have right atrial morphology (right isomerism, asplenia syndrome) or left atrial morphology (left isomerism, polysplenia syndrome), or one or both atria may have indeterminate morphology. Disorders of cardiac sidedness are typically associated with not only congenital heart disease but also abnormalities in multiple other organ systems. Consultation with a pediatric pathologist or cardiac pathologist is crucial in such cases.

The fifth cut is along the lateral wall of the left ventricle, through the mitral valve, to expose the left ventricular endocardial surface, the mitral valve, and its chordae tendineae and papillary muscles.

The sixth and final cut is along the anterior wall of the left ventricle, turning slightly rightward at the left ventricular outlet to match the direction of takeoff of the ascending aorta. In a properly-sided morphologic left ventricle, it will be apparent at this point that there is direct continuity between the atrioventricular (mitral) valve and the semilunar (aortic) valve, contrasted with the morphologic right ventricle in which these structures are separated by the parietal band. This final cut also affords the ideal opportunity to look for defects in the ventricular septum and to assess the positioning of the coronary ostia.

Properly located coronary ostia will be at, or just below, the sinotubular junction in roughly the midportions of the left and right sinuses of Valsalva. Minor variations of positioning of the ostia are common, as is the co-location of a second small ostium immediately next to that of the right coronary artery (the conus artery which, when not arising from the right sinus, arises as the first branch of the right coronary artery). When major anomalies of the coronary arteries occur, the exact position of the anomalous ostium should be described, along with a statement of the angle at which the artery arises relative to the aortic wall and, if takeoff is at an acute angle, whether or not this results in the presence of an intimal flap that can potentially occlude the ostium. A very acute angle of origin, sometimes nearly parallel to the aortic wall, and an occlusive intimal flap are commonly seen in cases of sudden death caused by anomalous coronary arteries.

Cardiac measurements are useful for detecting and documenting the presence of cardiac pathology. It is therefore recommended in SDY cases that heart weight and ventricular wall thicknesses be routinely collected. Historically, valve circumferences have also been taken but their utility to diagnose pathology has not been proven. Data collected from the SDY Case Registry will be examined to better understand

the utility of valve measurements and thus, taking such measurements could be useful for contributing to a dataset.

Heart weight is the most important of these measurements, as it directly reflects the presence and degree of myocardial loss (atrophy) or excess (hypertrophy). In order to facilitate comparison with published normal heart weights, the heart should only be weighed after the removal of extraneous extracardiac tissue and intracavitary blood clots. As is true with the examination of adult hearts, the hearts of post-pubertal teenagers can reasonably be weighed to the nearest 5 or 10 grams, while the hearts of younger children are best weighed to the nearest gram and those of infants and toddlers to the nearest one-tenth of a gram.

The thicknesses of the left and right ventricular free walls and ventricular septum should be measured at the level of the tips of the left ventricular papillary muscles (20). These measurements should take into account only the compact myocardium and should not include the trabecular muscle or the epicardial fat. In the right ventricle, it is not uncommon for the inferior (posterior) wall to be slightly thicker than that anterior wall, in which case it is recommended that the greater thickness be recorded.

The circumferences of the atrioventricular and semilunar valves could be useful for demonstrating the presence and degree of annular dilation, which can result in regurgitation and stenosis. The recommended method is to use a piece of string or other flexible material that can conform to the curved contours of the valve annulus. The string can then be laid out flat on a ruler to get an accurate measure of the circumference.

These cardiac measurements must be compared with tables of normal values that take into account age, sex, and body size. Body size is typically represented by body weight. However, for situations in which the decedent is very obese or where the body weight has been artifactually reduced (such as after organ and tissue donation) or increased (such as in decedents with anasarca), comparison with normal ranges based on body length is more appropriate (20). Many such

reference tables exist. This author (SG) prefers those from the Mayo Clinic (22), which contains reference ranges for heart weight, wall thickness, and valve circumference for individuals from birth to 19 years old.

The measurements must be interpreted in light of one another and the gross morphology of the heart. For example, a heart may have a left ventricular wall thickness in the published normal range. One might be tempted to interpret this as an absence of left ventricular hypertrophy. However, if the heart weight is significantly increased, a normal LV thickness implies that the left ventricle is both hypertrophied and dilated (the law of Laplace dictates that the wall will thin as the cavity dilates), which should be apparent in the gross appearance of the left ventricle and an increased circumference of the mitral valve.

The presence of structural congenital heart disease may be encountered at autopsy in approximately 1% of the population. Although there is a reported association between sudden death and some more complex forms of congenital heart disease (such as tetralogy of Fallot, transposition of the great arteries, and single ventricle anomalies), many types of congenital heart disease do not typically cause sudden death, and caution should be exercised in attributing cause of death to congenital heart disease in all cases if discovered at autopsy.

Call To Action

Feedback is welcome from the ME/C community on the value and utility of these autopsy and death scene investigation tools. ME/C are encouraged to use these tools and approaches in their routine practice to help improve standardization of investigation and characterization of SDY cases. All tools are available for download from the SDY Case Registry website: www.sdyregistry.org (23). Medical examiners and coroners are also encouraged to participate in child death reviews and/or advanced reviews for SDY Case Registry states/jurisdictions to help improve case review and categorization.

CONCLUSION

The SDY Case Registry tools and guidance documents promote standardization of death investigation and autopsy practices in funded jurisdictions. After informed consent from the next of kin, the SDY Case Registry permits collection of comprehensive information from investigation and autopsy as well as samples for DNA extraction that may inform the cause of death determination, guide cascade screening for families, and create a resource for research. We encourage use of the tools and guidance described herein to foster unified approaches to improving the understanding of sudden, unexpected natural deaths in the young and lead us towards more targeted approaches to prevention.

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APPENDIX 1

The SDY Case Registry Autopsy Protocol Task Force:
Karen Chancellor MD, Beau Clark MD D-ABMDI,
Timothy E. Corden MD, Kim Fallon, Corinne L.
Fligner MD, Sam P. Gulino MD, Wendy M. Gunther
MD, Jennifer L. Hammers DO, Owen L. Middleton
MD, and Michael Murphy DBA D-ABMDI.

APPENDIX 2

INFORMATION FOR FAMILIES

AGENCIES WORKING ON THE REGISTRY

Tennessee Department of Health
West Tennessee Regional Forensic Center
Middle Tennessee Regional Forensic Center
Upper Northeast Regional Forensic Center

THE SUDDEN DEATH IN THE YOUNG CASE REGISTRY
WORKING TO COUNT, UNDERSTAND, AND PREVENT SUDDEN DEATH IN THE YOUNG

SDY
DATA COORDINATING CENTER FOR THE SUDDEN DEATH IN THE YOUNG CASE REGISTRY

Data Coordinating Center for the Sudden Death in the Young (SDY) Registry
Telephone: 800-656-2434
Email: info@SDYregistry.org

RESOURCES

BEREAVEMENT SUPPORT GROUPS

The Compassionate Friends

compassionatefriends.org, 877-969-0010

MISS Foundation

missfoundation.org, 888-455-6477

The Tears Foundation

thetearsfoundation.org

SPECIFIC SUPPORT GROUPS

Sudden Unexplained Infant Death (SUID)/
[Sudden Infant Death Syndrome \(SIDS\)](#)

CJ First Candle Foundation

cjfirstcandle.org

Share Pregnancy and Infant Loss Support

nationalshare.org

[Heart](#)

Hypertrophic Cardiomyopathy Association

4hcm.org

Parent Heart Watch

parentheartwatch.org

Sudden Arrhythmia Death Syndrome (SADS) Foundation

sads.org

[Epilepsy](#)

Epilepsy Foundation SUDEP Institute

epilepsy.com/get-help/about-sudep-institute

The Danny Did Foundation

dannydid.org

Citizens United for Research in Epilepsy (CURE)

cureepilepsy.org/research/sudep.asp

FAMILY MEDICAL FOLLOW-UP

If heart disease is identified as cause of death
or if death remains unexplained

Vanderbilt University Medical Center

The Center for Inherited Heart Disease
Nashville, TN, Phone: 615-322-7602

If family history of epilepsy contact:

Vanderbilt Neurology, Nashville, TN
Phone: 615-936-0060.

WHAT KIND OF INFORMATION IS THE SDY CASE REGISTRY COMPILING?

The Sudden Death in the Young (SDY) Case Registry gathers information to learn more about children who die suddenly and unexpectedly. The project is funded by the National Institutes of Health (NIH) and the Centers for Disease Control and Prevention (CDC). The SDY Data Coordinating Center located in Michigan is working to implement the SDY Case Registry.

The purpose of the SDY Case Registry is to try to understand the cause of these deaths and find ways to prevent these deaths.

You may be contacted in the future to consent to save your child's DNA and/or be re-contacted as part of the SDY Case Registry.

The SDY Case Registry is compiling information from the autopsy and death scene investigation, DNA from samples obtained at the time of autopsy and medical history of the child and the child's family. All of the information gathered for the SDY Case Registry will be kept confidential and private.

WHO DO I CONTACT FOR MORE INFORMATION?

Rachel Heitmann

615-741-0368

THIS PROJECT IS SUPPORTED BY:

Centers for Disease Control and Prevention

National Institutes of Health

FOR MORE INFORMATION ABOUT THE SDY CASE REGISTRY:

www.sdyregistry.org

APPENDIX 3

Date

X
X
X

Dear Mr./Mrs/Ms. BLANK

I would like to offer you my condolences on the loss of your child, NAME. At the time of their death, INSERT STAFF INFO talked with you about the Sudden Death in the Young (SDY) project designed to explore possible genetic causes of unexplained death in children. You consented to have your child's DNA tested. This testing was done to determine if your child had any known genetic abnormalities that might have contributed to their death. I am writing to you about those results.

The DNA sample we collected from your child was sent to INVITAE, a genetic testing lab. A CARDIOMYOPATHY and/or ARRHYTHMIA panel of genes were looked at to see if there were HEART STRUCTURAL/FUNCTIONAL ISSUES or ABNORMAL HEART RHYTHMS. This panel tested for changes (also known as variants) in genes.

Attached is the completed report for your review and for sharing with a genetic counselor, your family physician, family members, and anyone else you choose.

Insert if VUS identified

Genetic testing was inconclusive. The testing found a VARIANT (or variants) OF UNCERTAIN SIGNIFICANCE (VUS) in the [GENE] gene (or genes). This means the laboratory found a unique change in your child's DNA, but based on current knowledge, it is not known if these changes cause heart problems. This gene variant (or these gene variants) could increase the risk for heart problems, or it (or they) could be a harmless change that does not cause any heart problems. Thus, it is not clear if the variant is related to your child's sudden death. Talking with a genetic counselor and gathering more information on your family may help in better understanding the variant(s).

Insert if Mutation Negative

Genetic testing was normal. Testing found NO disease-causing gene changes. This testing cannot rule out a genetic cause of sudden death, but your child's death was most likely NOT related to a change in one of the genes she (or he) was tested for.

I highly recommend you reach out to a genetic counselor, geneticist, or your family physician for further explanation. Genetic counseling may also help you and your family understand which relatives might be at risk for an inherited disease, and what medical follow-up is recommended for your family.

Insert if Mutation Positive

Genetic testing showed a PATHOGENIC VARIANT(S) DETECTED. Thus, NAME's sudden death was most likely related to this gene change.

I highly recommend you reach out to a genetic counselor, geneticist, or your family physician for further explanation. Genetic counseling may also help you and your family understand which relatives might be at risk for an inherited disease, and what medical follow-up is recommended for your family.

For genetic counseling, I suggest you reach out to: INSERT LOCAL GENETICS Resource. Phone-based genetic counseling is also available through BLANK. The Office of BLANK is not affiliated with either of these offices or counselors, but they may be able to assist you further.

Enclosed is a brochure that contains additional information about the SDY registry as well as locally available bereavement resources. If you have any questions about the resources, the testing process, or the project please reach out to me. My phone number is BLANK and my email is BLANK.

Thank you for your time and for participating in the SDY project. We hope that this project has been helpful as we all search for the cause of sudden death in children. Again, please accept our condolences for the loss of your loved one.

Sincerely,

INSERT CONTACT INFO

Enclosure

APPENDIX 4

SDY FIELD INVESTIGATION GUIDE AND FAMILY INTERVIEW TOOL

2.0 | 1.2017

This is as an optional tool to guide the information needed for the Sudden Death in the Young Case Registry Death Investigation and Family Interview. These questions mirror those in the Child Death Review Case Report and should be answered for sudden unexpected deaths in children in ages 0 through 19. It is important that you provide a copy of this tool to your Child Death Review Team, even if you were not able to obtain all the information. If the child whose death you are investigating is an infant (0 to 364 days old) also use the Sudden Unexplained Infant Death Investigation Reporting Form (SUIDIRF). <http://www.cdc.gov/sids/suidrfdownload.htm>

When introducing and using this tool, here are statements to share with families who are providing information: We would like to ask you additional questions about your child's and family's medical history. This information may assist the medical examiner/doctor in understanding why your child died. The questions are similar to what a doctor's office may ask you. This will take approximately 30-45 minutes. I understand that these questions may be difficult, or you may not know the answer, and that is ok. We can take breaks whenever you need. Would this be ok?

Some of the questions may not seem relevant or important since your child was so young. I still need to ask you to answer them the best you can. Even though it may not seem important, every little bit of information helps the medical examiner/doctor find out what happened. Remember it is okay to answer "I don't know."

We appreciate that this is a lot of information and we appreciate your time in helping us.

Name of person being interviewed: _____ Relationship to the deceased: _____

Name of Person conducting this interview: _____ Title: _____

Date/Time of Interview: _____ Signature: _____

Location of interview: _____ Interview Method (phone, in-person): _____

Medical records to collect

- ☐ Pediatric records for well and sick visits (including newborn screening results)
- ☐ If under 1 year of age, include mother's prenatal and obstetric reports
- ☐ Hospital birth records
- ☐ Emergency department records
- ☐ Emergency medical services/first responder records
- ☐ Immunization records
- ☐ Hospital records from day of death and from previous visits, if any
- ☐ Specialty health provider records (including any history of cardiac or neurological conditions)
- ☐ Any cardiac testing including previous electrocardiogram (EKG), echocardiogram, cardiac MRI, stress test, Holter monitors, and chest X-rays
- ☐ If child had epilepsy, records should include history of anti-epileptic drug levels, including frequency of monitoring of levels
- ☐ Any testing/records done as part of organ procurement
- ☐ Comprehensive family history records, if they exist

SDY FIELD INVESTIGATION GUIDE AND FAMILY INTERVIEW TOOL

General Information

OVERLAPS SUIDIRF

Decedent Name: _____ Decedent Date of Birth: _____

Decedent Gender: ☐ Male ☐ Female Decedent Ethnicity: ☐ Hispanic/Latino ☐ Non-Hispanic/Latino

Decedent Race: ☐ Black ☐ Asian, specify: _____ ☐ American Indian, Tribe: _____
☐ Alaskan Native, Tribe: _____ ☐ Pacific Islander, specify: _____ ☐ Native Hawaiian

Decedent Next of Kin's Name: _____ Decedent Next of Kin's Phone No: _____

Decedent Next of Kin's Email: _____

Date of Death: _____ Time of Death: _____

Location of Death: _____ Time Decedent Was Last Seen Alive: _____

Was the incident witnessed? ☐ Yes ☐ No ☐ Unknown If yes, by whom: _____

Was a death scene investigation performed? ☐ Yes ☐ No ☐ Unknown

If yes, check all that apply: ☐ CDC's SUIDI Reporting Form or jurisdictional equivalent ☐ Narrative description of circumstances
☐ Scene Photos ☐ Scene recreation with doll ☐ Scene recreation without doll ☐ Witness interviews

Activities within 24 hours of death

Child's activity at time of incident? Check all that apply.

- ☐ Sleeping ☐ Playing ☐ Working ☐ Eating ☐ Driving/vehicle occupant ☐ Unknown
☐ Other, specify: _____

Did the child experience any of the following stimuli at the time of incident or within 24 hours of incident? Check all that apply.

	at incident	within 24 hrs of incident
Physical activity	☐	☐
If yes to physical activity, specify: _____		
Sleep deprivation	☐	☐
Driving	☐	☐
Visual stimuli	☐	☐
Video game stimuli	☐	☐
Emotional stimuli	☐	☐
Auditory stimuli/startle (loud noises)	☐	☐
Physical trauma (direct blow to chest or head)	☐	☐
☐ Other, specify: _____		
☐ Unknown		

Resuscitation

Resuscitation attempted?

- ☐ N/A ☐ Yes ☐ No ☐ Unknown

If yes, by whom? _____

If yes, type of resuscitation (CPR, Automated External Defibrillator (AED), rescue medications (e.g. atrophine, epinephrine, other), specify: _____

If no AED, was AED available/accessible? ☐ Yes ☐ No

If an Automated External Defibrillator (AED) was used, was a shock administered? ☐ Yes ☐ No

How many shocks? _____

If yes, what rhythm was recorded? (e.g. ventricular fibrillation) _____

Describe the death, including: time lapse between collapse, 911 call, access to CPR and automated external defibrillator, EMS arrival, defibrillation, transit to hospital, death, etc. _____

SDY FIELD INVESTIGATION GUIDE AND FAMILY INTERVIEW TOOL

Symptoms within 72 hours of Death

check all that apply

Cardiac	Yes	No	Unknown
1. Chest pain	☐	☐	☐
2. Dizziness/lightheadedness	☐	☐	☐
3. Fainting	☐	☐	☐
4. Palpitations	☐	☐	☐

Neurologic

5. Concussion	☐	☐	☐
6. Confusion	☐	☐	☐
7. Convulsions/seizure	☐	☐	☐
8. Headache	☐	☐	☐
9. Head injury	☐	☐	☐
10. Psychiatric symptoms	☐	☐	☐
11. Paralysis (acute)	☐	☐	☐

Respiratory

12. Asthma	☐	☐	☐
13. Pneumonia	☐	☐	☐
14. Difficulty breathing	☐	☐	☐

Other Acute Symptoms

15. Fever	☐	☐	☐
16. Heat exhaustion/heat stroke	☐	☐	☐
17. Muscle aches/cramping	☐	☐	☐
18. Slurred speech	☐	☐	☐
19. Vomiting	☐	☐	☐
20. Other, specify: _____			

Previous Serious Injury

check all that apply

	Yes	No	Unknown
1. Near drowning	☐	☐	☐
2. Car accident	☐	☐	☐
3. Brain injury	☐	☐	☐
4. Other, specify: _____			

If yes, describe

Symptoms prior to 72 hours of Death

check all that apply

Cardiac	Yes	No	Unknown
1. Chest pain	☐	☐	☐
2. Dizziness/lightheadedness	☐	☐	☐
3. Fainting	☐	☐	☐
4. Palpitations	☐	☐	☐

Neurologic

5. Concussion	☐	☐	☐
6. Confusion	☐	☐	☐
7. Convulsions/seizure	☐	☐	☐
8. Headache	☐	☐	☐
9. Head injury	☐	☐	☐

Respiratory

10. Difficulty breathing	☐	☐	☐
--------------------------	--------------------------	--------------------------	--------------------------

Other

11. Slurred speech	☐	☐	☐
12. Other, specify: _____			

Exercise

Did the child ever have any of the following uncharacteristic symptoms during or within 24 hours after physical activity?

	Yes	No	Unknown
1. Chest pain	☐	☐	☐
2. Confusion	☐	☐	☐
3. Convulsions/seizure	☐	☐	☐
4. Dizziness/lightheadedness	☐	☐	☐
5. Fainting	☐	☐	☐
6. Headache	☐	☐	☐
7. Palpitations	☐	☐	☐
8. Shortness of breath/ difficulty breathing	☐	☐	☐
9. Other, specify: _____			
10. Unknown	☐	☐	☐

For children age 12 or older, did the child receive a pre-participation physical exam for a sport?

☐ N/A ☐ Yes ☐ No ☐ Unknown

If yes, date: _____

Restrictions? ☐ N/A ☐ Yes ☐ No ☐ Unknown

If yes, specify: _____

SDY FIELD INVESTIGATION GUIDE AND FAMILY INTERVIEW TOOL

Medical History of Decedent - Symptoms/Medical History/Previous Injuries

Had the child ever been diagnosed by a medical professional for the following? Check all that apply.

Previous Diagnoses

Blood Disease

	Yes	No	Unknown
1. Sickle cell disease	☐	☐	☐
2. Sickle cell trait	☐	☐	☐
3. Thrombophilia (clotting disorder)	☐	☐	☐

Cardiac

4. Abnormal electrocardiogram (EKG or ECG)	☐	☐	☐
5. Aneurysm or aortic dilatation	☐	☐	☐
6. Arrhythmia/arrhythmia syndrome (irregular heart rhythm, palpitations) (DD: long QT, Brugada, CPVT, WPW)	☐	☐	☐
7. Cardiomyopathy (hypertrophic, dilated, arrhythmogenic right ventricular, left ventricular noncompaction)	☐	☐	☐
8. Commotio cordis (blow to chest causing cardiac arrest or death)	☐	☐	☐
9. Congenital heart disease	☐	☐	☐
10. Coronary artery abnormality	☐	☐	☐
11. Coronary artery disease (atherosclerosis)	☐	☐	☐
12. Endocarditis	☐	☐	☐
13. Heart failure	☐	☐	☐
14. Heart murmur	☐	☐	☐
15. High cholesterol	☐	☐	☐
16. Hypertension	☐	☐	☐
17. Myocarditis (heart infection)	☐	☐	☐
18. Pulmonary hypertension	☐	☐	☐
19. Sudden cardiac arrest	☐	☐	☐

Neurologic

20. Anoxic brain Injury (injury caused by lack of oxygen to the brain)	☐	☐	☐
21. Traumatic brain injury/ head injury/concussion	☐	☐	☐
22. Brain tumor	☐	☐	☐
23. Brain aneurysm	☐	☐	☐
24. Brain hemorrhage	☐	☐	☐
25. Developmental brain disorder (cerebral palsy, structural brain malformation)	☐	☐	☐
26. Epilepsy/seizure disorder	☐	☐	☐
27. Febrile seizure	☐	☐	☐
28. Mesial temporal sclerosis	☐	☐	☐
29. Neurodegenerative disease	☐	☐	☐

30. Stroke/mini stroke/ TIA- Transient Ischemic Attack	Yes ☐	No ☐	Unknown ☐
31. Central nervous system infection (meningitis or encephalitis)	☐	☐	☐

Respiratory

32. Apnea	☐	☐	☐
33. Asthma	☐	☐	☐
34. Pulmonary embolism	☐	☐	☐
35. Pulmonary hemorrhage	☐	☐	☐
36. Respiratory arrest	☐	☐	☐

Other

37. Connective tissue disease (Ehlers Danlos, Marfan syndrome, bicuspid aortic valve with aortic root dilation and/or cystic medial necrosis)	☐	☐	☐
38. Diabetes	☐	☐	☐
39. Endocrine disorder, other: thyroid, adrenal, pituitary	☐	☐	☐
40. Hearing problems or deafness	☐	☐	☐
41. Kidney disease	☐	☐	☐
42. Mental illness/ psychiatric disease	☐	☐	☐
43. Metabolic disease	☐	☐	☐
44. Muscle disorder or muscular dystrophy	☐	☐	☐
45. Oncologic disease treated by chemotherapy or radiation	☐	☐	☐
46. Prematurity	☐	☐	☐
47. Congenital disorder/ genetic syndrome	☐	☐	☐
48. Other, specify: _____			

Follow-up Testing and Evaluation for Diagnosis

(e.g. echo, EKG, neuro eval)

If a more specific diagnosis is known, provide any additional information:

Describe:

Routine treatment for diagnosis?

SDY FIELD INVESTIGATION GUIDE AND FAMILY INTERVIEW TOOL

Medical History of Decedent - Symptoms/Medical History/Previous Injuries (circle all that apply)

Medications

In the 72 hours prior to death was the child taking any prescribed medication(s)?

☐ Yes ☐ No ☐ Unknown

If yes, describe:

Medication History

Was the child compliant with their prescribed medications?

If not, describe why and how often:

Recent medication changes? (extra doses, missed doses, change of medication)

☐ Unknown for all

Other Substances within 24 hours of Death

1. Over the counter (OTC) medicine
2. Recent/short term prescriptions (antibiotics)
3. Energy drinks
4. Caffeine
5. Performance enhancers
6. Diet assisting medications
7. Supplements
8. Tobacco (cigarettes, chewing, electronic/nicotine)
9. Alcohol
10. Illegal drugs (cocaine, heroin)
11. Legalized marijuana
12. Other, specify

If yes to any, describe:

Family History

Include information on 1st and 2nd degree relatives: *siblings, parents, grandparents, aunts, uncles and first cousins of the deceased as well as if they were older children if they had their own children.*

Family can be referred for genetic counseling at this center:

☐ Unknown for all

Sudden Death

1. Sudden, unexpected death before age 50, describe (SIDS, drowning, relative who died in single and/or unexplained motor vehicle accident (driver of car))

Heart Disease

1. Heart condition/heart attack or stroke before age 50
2. Aortic aneurysm or aortic rupture
3. Arrhythmia (fast or irregular heart rhythm)
4. Cardiomyopathy
5. Congenital heart disease

Neurologic Disease

1. Epilepsy or convulsions/seizure
2. Other neurologic disease

Symptoms

3. Febrile seizures
4. Unexplained fainting

Other

1. Congenital deafness
2. Connective tissue disease (Ehlers Danlos Syndrome, Marfan syndrome)
3. Mitochondrial disease
4. Muscle disorder or muscular dystrophy
5. Thrombophilia (clotting disorder)
6. Other diseases that are genetic or run in families

Genetic Testing

Has any blood relative (siblings, parents, aunts, uncles, cousins, grandparents) had genetic testing?

☐ Yes ☐ No ☐ Unknown

If yes, describe the results (disease, gene, mutation)

SDY FIELD INVESTIGATION GUIDE AND FAMILY INTERVIEW TOOL

Epilepsy/Seizure Disorder

Answer only if child was diagnosed with an epilepsy/seizure disorder.

How old was the child when diagnosed with epilepsy/seizure disorder?

What was the underlying cause of the child's seizures?

Check all that apply.

	Yes	No	Unknown
Brain injury/trauma	☐	☐	☐
Brain tumor	☐	☐	☐
Cerebrovascular	☐	☐	☐
Central nervous system infection	☐	☐	☐
Degenerative process	☐	☐	☐
Developmental brain disorder	☐	☐	☐
Inborn error of metabolism	☐	☐	☐
Genetic/chromosomal	☐	☐	☐
Mesial temporal sclerosis	☐	☐	☐
Idiopathic or cryptogenic	☐	☐	☐
Other acute illness or injury other than epilepsy, other, specify:	☐	☐	☐

What type(s) of seizures did the child have?

Check all that apply.

	Yes	No	Unknown
Non-convulsive	☐	☐	☐
Convulsive (grand mal seizure or generalized tonic-clonic seizure)	☐	☐	☐
Occur when exposed to strobelights, video game, or flickering light (reflex seizure)	☐	☐	☐

Describe the child's epilepsy/seizures.

Check all that apply.

	Yes	No	Unknown
Last less than 30 minutes	☐	☐	☐
Last more than 30 minutes (status epilepticus)	☐	☐	☐
Occur in the presence of fever (febrile seizure)	☐	☐	☐
Occur in the absence of fever	☐	☐	☐
Occur when exposed to strobe lights, video game, or flickering light (reflex seizure)	☐	☐	☐

How many seizures did the child have in the year preceding death?

Did treatment for seizures include anti-epileptic drugs?

If yes, how many different types of anti-epilepsy drugs (AED) did the child take?

Was night surveillance used?

DATA COORDINATING CENTER FOR THE
SUDDEN DEATH IN THE YOUNG
CASE REGISTRY

Sudden Death in the Young Case Registry Data Coordinating Center
c/o Michigan Public Health Institute, 2479 Woodlake Circle, Okemos, MI 48864

Telephone: 800-656-2434 Email: info@SDYregistry.org
Web: www.sdyregistry.org Fax: 844-816-9662

APPENDIX 5

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE SUBJECT ID:

Introduction

This Guidance is a reference to assist you in completing the Sudden Death in the Young (SDY) Autopsy Summary. This summary sheet of autopsy results should be completed following your investigation of a sudden and unexpected death in a child or youth under age 20. It includes instructions for specific components of the autopsy.

The SDY Autopsy Guidance was developed as part of the SDY Case Registry, an initiative of the National Institutes of Health (NIH) and the US Centers for Disease Control and Prevention (CDC). This summary, the guidance and instructions were developed by the SDY Autopsy Protocol Committee composed of medical examiners with experience in pediatric, cardiac and/or neuro pathology; physician coroners, death investigators, and other medical professionals with experience in cardiology, neurology, emergency medicine, public health and genetics.

Your jurisdiction is participating in the Sudden Death in the Young Case Registry with funding from the NIH and CDC. The autopsy findings will be summarized with other case review information and biospecimen data (upon family consent) in the SDY Case Registry. This Registry of de-identified data will be used to better understand the etiologies and risk factors for sudden death in the young so that improved prevention strategies may be developed.

Additional instructions and information are provided throughout this document in italics and footnotes.

SDY Definitions and Inclusion/Exclusion Criteria for the SDY Case Registry

"Sudden" implies death within 24 hours of the first symptom, or those resuscitated from cardiac arrest and dying during the same hospital admission.

"Unexpected" refers to a death in someone who dies from an accidental injury or someone who was believed to have been in good health, or had a stable chronic condition or had an illness but death was not expected. Examples could include hypertrophic or dilated cardiomyopathy, congenital heart disease, epilepsy, asthma and pneumonia.

Inclusion and Exclusion Criteria

This autopsy results summary sheet is a key component of the SDY Case Registry and should be used for all cases that meet all of the following inclusion criteria and none of the following exclusion criteria:

Inclusion Criteria

Is the child under 20 years old?	☐ Yes, Include	☐ No, Exclude
Was the death sudden and unexpected and/or unwitnessed?	☐ Yes, Include	☐ No, Exclude

Exclusion Criteria

Was the death caused by an accident in which the external cause was <u>the obvious and only</u> reason* for the death?	☐ Yes, Exclude	☐ No, Include
*Exception: All infants under 1 year of age whose death was caused by suffocation		☐ Include
Was the death an obvious homicide?	☐ Yes, Exclude	☐ No, Include
Was the death an obvious suicide?	☐ Yes, Exclude	☐ No, Include
Was the death caused by an accidental or intentional overdose of drugs even if this caused cardiac or respiratory arrest?	☐ Yes, Exclude	☐ No, Include
Was the death caused by a terminal illness in which the death was reasonably expected to occur within 6 months?	☐ Yes, Exclude	☐ No, Include

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

General

Sex: ☐ Male ☐ Female

Body weight: _____ kg Body length: _____ cm

Head circumference: _____ cm

External Exam: If abnormalities suggest trauma, disease/syndrome, or medical intervention, please describe:

Photography (external): ☐ Yes ☐ No

Imaging

(Circle all that were performed and describe the location)

X-Ray, single:

X-Ray, multiple views:

CT scan:

MRI:

Describe any abnormalities found on imaging:

Detailed Review of Specified Organs

Thorax/Lungs

Thorax/Lungs Imaging:

Radiographs of chest ☐ Prior to death (hospital, emergency room, other) ☐ Postmortem

- *If there is a question about the possibility of extra lobar or intra-lobar sequestration, or congenital pulmonary adenomatoid malformation (CPAM; old name CCAM), remove the heart, lungs, central diaphragm, inferior vena cava, and descending aorta as a block, and send for pediatric pathology consultation.*

Thorax/Lungs – External Gross Examination

Chest

Contour ☐ Normal ☐ Abnormal

If abnormal: ☐ Increased anteroposterior diameter ☐ Asymmetry

Costal margin flaring ☐ Other: _____

Injuries ☐ Absent ☐ Present: _____

Axillary lymphadenopathy ☐ Absent ☐ Present

Other: _____

Nasal choanae (infants)¹ ☐ Patent ☐ Obstructed

¹Testing to see if the nasal choanae are patent may be performed by sounding each nostril with a flexible probe. This can be performed with the nasopharyngeal swab for viral culture.

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Thorax/Lungs – Internal Gross Examination

- *Photography: (optional)* ☐ *In situ* ☐ *On cutting board* • *Testing: sampling for viral and bacterial cultures (as indicated)*

Tracheal deviation	☐ Absent	☐ Present:	☐ Left	☐ Right
Lungs				
Pneumothorax	☐ Absent	☐ Present:	☐ Left	☐ Right ☐ Bilateral
If present, diagnosed by:	☐ X-ray	☐ Other means:		
Hypoinflation ²	☐ Absent	☐ Present:	☐ Left	☐ Right ☐ Bilateral
Lung(s) sunken towards the back	☐ Absent	☐ Present:	☐ Left	☐ Right ☐ Bilateral
Hyperinflation	☐ Lungs do not approach midline	☐ Approach midline	☐ Meet in midline	
Color	☐ Pink	☐ Dark red	☐ Alternating pink and purple	☐ Fibrinous/purulent exudate
	☐ Dark red in all lobes, posterior only ³	☐ Other: _____		
Pleural effusion	☐ Absent	☐ Present:	☐ Left	☐ Right ☐ Bilateral
If present, appearance:	☐ Clear	☐ Bloody	☐ Straw	☐ Purulent ☐ Other: _____
Amount:	_____ ml			
Hemidiaphragm elevation:	☐ Absent	☐ Present:	☐ Left	☐ Right ☐ Bilateral

Thorax/Lungs – Gross Dissection

- *Take heart and lungs out as a block after inspecting aorta for vascular ring around trachea, and inspecting pulmonary arteries and veins (see heart section).*
- *The trachea / upper respiratory tract should be removed as a block with the lungs.*

Vascular ring (aorta around trachea) ☐ Absent ☐ Present

Lungs				
Blood on the pleural surface (adherent hemothorax)	☐ Absent	☐ Present:	☐ Acute	☐ Chronic
Blood beneath the pleura	☐ Absent	☐ Present:	☐ Petechiae	☐ Confluent/Large hemorrhages
Necrotic exudate on the pleural surface	☐ Absent	☐ Present		
Prominent/discolored/dilated lymphatics visible through the pleura	☐ Absent	☐ Present		
Cobblestoning ⁴	☐ Absent	☐ Present		
Rib markings on the pleura	☐ Absent	☐ Present		
Other:	_____			

- *Perform the initial examination of the heart/lung block. If a cardiovascular pathology or pediatric pathology consultation is requested, send the heart/lung block to the consultant. If consultation is not requested, separate the lungs from the heart following the initial examination.*

Lung weights within normal range for age	☐ Yes	☐ No:	☐ Increased	☐ Decreased
Right lung approximately 1/3 heavier than the left lung	☐ Yes	☐ No:		
Resuscitation-related changes	☐ Absent	☐ Present:		
Pulmonary edema, NOS	☐ Absent	☐ Present:		
Neurogenic pulmonary edema ⁵	☐ Absent	☐ Present:		
Pulmonary infection	☐ Absent	☐ Present:		
Pulmonary hemorrhage	☐ Absent	☐ Present:		
If present:	☐ Diffuse	☐ Focal, location: _____	☐ Aspiration pattern (follows bronchi)	
Pulmonary hypertension ⁶	☐ Absent	☐ Present		
Other:	_____			

²Do the lungs approach each other or meet in the midline?

³Consider SUDEP

³Probable postmortem change

⁴Muscle layers in subpleural arterioles

⁴Areas of pink hyperinflation and purple hypoinflation

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Thorax/Lungs – Gross Dissection (continued)

Abnormalities/disease processes visible at the hilum of either lung:

- Pulmonary artery thromboemboli⁷ ☐ Absent ☐ Present, location: _____
- Bronchial mucus/purulence ☐ Absent ☐ Present: _____
- Bronchial aspirated food, foreign object ☐ Absent ☐ Present: _____
- Other: _____

Is the right lung anatomically right-sided, and is the left lung left-sided?⁸ ☐ Yes ☐ No

If no, partially divided lobes: ☐ Absent ☐ Present: _____

Relationship of mainstem bronchus to mainstem pulmonary artery:⁹

- ☐ Normal: left hyperarterial bronchus and right eparterial bronchus
- ☐ Left side is normal but right side is not (two left lungs)¹⁰
- ☐ Right side is normal but left side is not (two right lungs)¹¹
- ☐ Neither side is normal¹²

• Section through all lobes, central and peripheral, including mainstem bronchi.

- Hilar lymph nodes ☐ Normal ☐ Abnormal
- If abnormal: ☐ Enlarged ☐ Anthracotic ☐ Granulomatous disease ☐ Hemorrhagic
- ☐ Gross infection ☐ Tumor deposits ☐ Other: _____
- Aspiration ☐ Absent ☐ Present: _____
- Atelectasis ☐ Absent ☐ Present: _____
- Hyperinflation with/without mucus plugs¹³ ☐ Absent ☐ Present: _____
- Rib markings on pleura ☐ Absent ☐ Present: _____
- Cobblestoning ☐ Absent ☐ Present: _____
- Copious clear fluid ☐ Absent ☐ Present: _____
- Copious blood-tinged fluid (from bronchi and/or parenchyma on sectioning) ☐ Absent ☐ Present
- Hemorrhage ☐ Absent ☐ Present:
- If present: ☐ Diffuse ☐ Focal, location: _____ ☐ Aspiration pattern (follows bronchi)
- Pneumonia/consolidation, exudate in bronchi, abscesses, or other signs of infection ☐ Absent ☐ Present
- Cavitation ☐ Absent ☐ Present: _____
- Granulomatous process¹⁴ ☐ Absent ☐ Present: _____
- Infarction/thromboemboli¹⁵ ☐ Absent ☐ Present: _____
- Tumor or suspected benign or neoplastic process ☐ Absent ☐ Present: _____
- Congenital anomaly ☐ Absent ☐ Present: _____
- Other: _____

⁷If there is any question whether blood clots in the mainstem pulmonary artery branches are antemortem thromboemboli or postmortem clot, histology is definitive.

⁸Three lobes on the right and two lobes on the left

⁹Does the main bronchus enter the hilum above, or approximately level with, the mainstem pulmonary artery branch on the right side (normal right eparterial bronchus), and below the mainstem pulmonary artery branch on the left side (normal left hyperarterial bronchus)? If abnormal, consider pediatric pathology consultation.

¹⁰Look for polysplenia.

¹¹Look for asplenia.

¹²Look for Kartagener syndrome.

¹³Consider asthma.

¹⁴Consider infection or sarcoidosis

¹⁵Propagation of thromboemboli causes red-purple "sausages" to exude from cross-sectioned pulmonary artery branches.

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Thorax/Lungs – Microscopic Examination

- Central
- Peripheral: including pleura and subpleural pulmonary artery branches and medium-sized bronchi
- Through areas of grossly evident or suspected disease processes
- There is no definitive number of lung sections supported by research that can be stated as required in every case. Peripheral and central lung samples each yield different diagnoses, and both should be sampled. Sampling from multiple areas may detect patchy diseases. Grossly suspicious areas are likely to reward sampling. Storage of multiple lung segments allows further sampling if disease processes are detected that require it. If in doubt, consult a pediatric pathologist.

- Obtain special stains as indicated for:
 - Bacterial infection
 - Granulomatous disease (acid-fast bacteria, sarcoidosis, fungi)
 - Autoimmune disease
 - Neoplasia
 - Resolving hemorrhage (iron)

Aspiration	☐ Absent	☐ Present:	☐ Food	☐ Blood	☐ Other: _____
Pulmonary edema	☐ Absent	☐ Present:	_____		
Alveolar hemorrhage	☐ Absent	☐ Present:	_____		
Hemorrhage in bronchial lumens	☐ Absent	☐ Present:	_____		
Red cell morphology	☐ Normal	☐ Typical postmortem	☐ Sickle cells on formalin-exposed tissue		
Inflammation	☐ Absent	☐ Present			
If present, location:	☐ Bronchi/bronchioles	☐ Alveoli	☐ Alveolar walls		
Bronchus-associated lymphoid tissue	☐ Normal	☐ Abnormal:	_____		
Pulmonary thromboemboli	☐ Absent	☐ Present:	_____		
Secondary pneumonia around obstructed bronchi/infarcted lung parenchyma	☐ Absent	☐ Present			
Chronic lung disease following prematurity	☐ Absent	☐ Present			
Pulmonary hypertension or evidence of persistent fetal circulation ¹⁶	☐ Absent	☐ Present			
Asthma or other eosinophilic diseases	☐ Absent	☐ Present			
Foreign bodies	☐ Absent	☐ Present			

Trachea:

- Methods of examining the trachea should include opening along the long axis and cross-sectioning.
- The neck contents (proximal esophagus, trachea, thyroid gland, and overlying strap muscles) may be cross-sectioned together in one piece for histologic examination, particularly if tracheal/bronchial infection or narrowing of the lumen are of concern.
- Areas not cross-sectioned may be opened along the long axis. The epiglottis is easily sectioned. The aryepiglottic folds may be sectioned to look for eosinophils.
- The carina lends itself to a triangle-shaped sagittal cross-section that includes the carinal nodes.
- Trachea should be removed, including the hyoid bone, epiglottis, aryepiglottic folds, arytenoid cartilage, thyroid cartilage, trachea, and carina.

Epiglottis	☐ Symmetrical	☐ Asymmetrical	Tracheal contents	☐ Absent	☐ Present
Erythema	☐ Absent	☐ Present	If present:		
Exudate	☐ Absent	☐ Present	White foam, pink foam	☐ Absent	☐ Present
Aryepiglottic folds	☐ Symmetrical	☐ Asymmetrical	Mucus	☐ Absent	☐ Present
	☐ Flat (normal)	☐ Swollen	Necrotic exudate	☐ Absent	☐ Present
	☐ Obstruct the lumen	☐ Do not obstruct the lumen	Thin layer of liquid blood along the mucosa	☐ Absent	☐ Present
Vocal cords	☐ Symmetrical	☐ Asymmetrical	Pieces of food, vomitus streaking the mucosa	☐ Absent	☐ Present
Abnormalities	☐ Absent	☐ Present: _____	Obstructing blood clots	☐ Absent	☐ Present
Tracheal mucosa			Obstructing food bolus	☐ Absent	☐ Present
Erosion	☐ Absent	☐ Present	Foreign object	☐ Absent	☐ Present
Erythema	☐ Absent	☐ Present			
Inflammation	☐ Absent	☐ Present			

¹⁶Including muscle layers in subpleural arterioles; other abnormalities of pulmonary artery branches

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Heart – Gross Dissection

- Weigh the heart
- Make note of epicardial adhesions, exudate, or discoloration
 - Make note of amount and distribution of epicardial fat
- Section the epicardial coronary arteries at 3-5 mm intervals, avoiding cutting into great arteries and cardiac chambers
 - Note arterial dominance (right/left/shared) and locations and degrees of obstructions
- Make transverse (short axis) slices through the ventricles beginning 1 cm above the apex and at 1 cm intervals; do not section above the level of the tips of the left ventricular papillary muscles
 - Note all gross lesions in the myocardial sections including scars, discolorations, and softening
 - Lesions should be described by the usual descriptors (e.g., size, color, firmness) as well as:
- Vertical location (e.g., basal, midventricular, apical)
- Lateral location (e.g., anteroseptal, inferolateral)
- Distribution (e.g., subendocardial, transmural, subepicardial)
 - Take measurements of left ventricular thickness, right ventricular thickness, and septal thickness in the uppermost (most basal) slice
- When taking measurements, include only the compact myocardium; do not include trabecular muscle or papillary muscles
 - Examine the right ventricular wall for fat infiltration
 - It is recommended that the myocardial slices be photographed, especially if there are grossly visible lesions
- Open the heart in the direction of blood flow:
 - Open the right atrium from the inferior vena cava orifice to the tip of the atrial appendage

- Do not open through the superior vena cava orifice; doing so may cut through the SA node, hampering dissection of the conduction system if that is desired later
 - Open from the right atrium to the right ventricle along the posterior or lateral wall
 - Open the right ventricular outflow tract anteriorly
 - Open the left atrium by connecting all of the pulmonary veins and cutting to the tip of the atrial appendage
 - Open from the left atrium to the left ventricle along the lateral wall
 - Open the left ventricular outflow tract anteriorly
- Remove postmortem clot from all chambers
 - If large amount of postmortem clot is present, consider re-weighing heart after the clot is removed
- Describe degree of dilation of chambers, if any, and document presence/absence of mural thrombi
- Document presence/absence of patent foramen ovale, atrial septal defect, or ventricular septal defect (describe size and location if present)
- Examine the valves, noting number of leaflets/cusps of each and presence of any abnormalities (e.g., myxoid change, calcification, vegetations)
- Examine the coronary ostia
 - If ectopic origin is present, note acuity of the origin (e.g., sharp angle of origin), course of the proximal segment of the artery (e.g., within aortic adventitia), and presence/absence of an occlusive ostial flap
- If any of the above findings are present, it is recommended that they be photographed in addition to being described in the autopsy guidance

Heart – Gross Examination

Heart weight _____ g	☐ Unfixed	☐ Fixed	
Thoracic position	☐ Left (normal)	☐ Right	☐ Midline ☐ Ectopic: _____
Apex	☐ Leftward (normal)	☐ Rightward	☐ Other: _____
Spleen	☐ Single	☐ Accessory	☐ Polysplenia ☐ Asplenia
Liver	☐ Right (normal)	☐ Left	☐ Midline/ambiguous
Pericardial effusion	☐ Absent	☐ Present	If present: Amount: _____ ml
– Appearance	☐ Clear	☐ Straw	☐ Purulent ☐ Other: _____
Hemopericardium	☐ Absent	☐ Present	
Vascular Ring	☐ Absent	☐ Present	
Epicardium – Exudate	☐ Absent	☐ Present: _____	
– Adhesions	☐ Absent	☐ Present: _____	
– Fat	☐ Present, normal amount	☐ Increased ☐ Decreased	
Right atrium – Morphology	☐ Right ¹⁷ (normal)	☐ Left	☐ Ambiguous/other: _____
– Venoatrial connections (SVC/IVC)	☐ Normal	☐ Abnormal: _____	
– Coronary sinus OS	☐ Patent	☐ Stenotic ☐ Atretic	
– Dilation	☐ Absent ☐ Present: ☐ Mild ☐ Moderate ☐ Severe		
– Cavitory thrombus ¹⁸	☐ Absent ☐ Present: _____		

¹⁷Right atrial morphology includes presence of terminal crest, smooth endocardial surface posterior to terminal crest, pectinate muscles anterior to terminal crest and in atrial appendage.

¹⁸Antemortem thrombus; excludes perimortem/postmortem clot.

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Heart – Gross Examination (continued)

Left atrium

- Morphology ☐ Left (normal)¹⁹ ☐ Right ☐ Ambiguous/other: _____
- Coronary sinus OS ☐ Patent ☐ Stenotic ☐ Atretic
- Dilation ☐ Absent ☐ Present: ☐ Mild ☐ Moderate ☐ Severe
- Cavitory thrombus ☐ Absent ☐ Present: _____

Atrial septum

- ☐ Intact ☐ Probe-patent foramen ovale ☐ Atrial septal defect:²⁰ _____

Atrioventricular valves

- ☐ Two valves (right and left) ☐ Common valve (atrioventricular canal²¹)

Right atrioventricular valve

- Morphology ☐ Tricuspid (normal) ☐ Prosthetic: (type) _____ ☐ Other: _____
- Abnormalities ☐ Absent ☐ Present: If present, circle/describe all that apply:
 - Vegetations _____ – Leaflet thickening _____
 - Prolapse/ballooning _____ – Leaflet perforation _____
 - Commissural fusion _____ – Apical displacement of septal leaflet (Ebstein's anomaly) _____
 - Other: _____

Left atrioventricular valve

- Morphology ☐ Mitral (bicuspid, normal) ☐ Prosthetic: (type) _____ ☐ Other: _____
- Abnormalities ☐ Absent ☐ Present: If present, circle/describe all that apply:
 - Vegetations _____ – Leaflet thickening _____
 - Prolapse/ballooning _____ – Leaflet perforation _____
 - Commissural fusion _____ – Chordal thickening _____
 - Chordal stretching/rupture _____ – Other: _____

Right ventricle

- Morphology ☐ Right²² (normal) ☐ Left ☐ Ambiguous/other: _____
- Wall thickness²³ ☐ Anterior: _____ cm ☐ Posterior: _____ cm
- Fat infiltration²⁴ ☐ Absent ☐ Present ☐ If present, which wall: ☐ Anterior ☐ Posterior
 - ☐ Maximum % thickness of wall involved: _____
- Right ventricular thinning²⁴ ☐ Absent ☐ Present, location: _____
- Dilation ☐ Absent ☐ Present: ☐ Mild ☐ Moderate ☐ Severe
- Cavitory thrombus ☐ Absent ☐ Present: _____
- Endocardium ☐ Thin, translucent (normal) ☐ Abnormal: _____

Left ventricle

- Morphology ☐ Left²⁵ (normal) ☐ Right ☐ Ambiguous/Other
- Wall thickness²³ ☐ Anterior: _____ cm ☐ Lateral: _____ cm ☐ Inferior/posterior: _____ cm
- Dilation ☐ Absent ☐ Present: ☐ Mild ☐ Moderate ☐ Severe
 - If present, chamber diameter (at same level as wall thicknesses): _____ cm
- Cavitory thrombus ☐ Absent ☐ Present: _____
- Endocardium ☐ Thin, translucent (normal) ☐ Abnormal: _____
- Myocardial infarction (acute/recent) ☐ Absent ☐ Present: _____
- Myocardial scar²⁶ ☐ Absent ☐ Present: _____
- Myocardial discoloration ☐ Absent ☐ Present: _____

¹⁹Left atrial morphology includes absence of terminal crest and smooth endocardial surface throughout atrium except for pectinate muscles in atrial appendage.

²⁰Description should include location, size, and any intervention.

²¹Describe morphology and pathology under "Left atrioventricular valve" section.

²²Right ventricular morphology includes coarse endomyocardial trabeculations and presence of a moderator band.

²³Measurements should be taken at the level of the tips of the ventricular papillary muscles and should include only the compact myocardium (not epicardial fat or papillary/trabecular muscle).

²⁴Concerning for arrhythmogenic right ventricular cardiomyopathy

²⁵Left ventricular morphology includes fine endomyocardial trabeculations and absence of a moderator band.

²⁶Includes remote myocardial infarctions

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Heart – Gross Examination (continued)

Ventricular septum

– Septal thickness²⁷ _____ cm ☐ Intact ☐ Ventricular septal defect²⁸ _____

Semilunar valves

☐ Two valves ☐ Single valve²⁹ (truncus arteriosus, pulmonary or aortic atresia)

– If two valves: ☐ Aorta posterior and rightward of the pulmonary valve (normal)

☐ D-malposed³⁰ ☐ Other arrangement: _____

Right semilunar valve

– Number of cusps ☐ 3 (normal) ☐ Other: _____ ☐ Prosthetic (type) _____

– Abnormalities ☐ Absent ☐ Present: If present, circle/describe all that apply:

– Vegetations _____

– Thickening _____

– Calcification _____

– Perforation _____

– Commissural fusion _____

– Other: _____

Left semilunar valve

– Number of cusps ☐ 3 (normal) ☐ 2 (bicuspid) ☐ Other: _____ ☐ Prosthetic (type) _____

– Abnormalities ☐ Absent ☐ Present: If present, circle/describe all that apply:

– Vegetations _____

– Thickening _____

– Calcification _____

– Perforation _____

– Commissural fusion _____

– Other: _____

Great vessels

– Pulmonary artery ☐ Normal ☐ Dilated ☐ Hypoplastic

– Discontinuous branch pulmonary arteries ☐ Absent ☐ Present

– Supravalvar pulmonary stenosis ☐ Absent ☐ Present: ☐ Mild ☐ Moderate ☐ Severe

– Thromboemboli ☐ Absent ☐ Present: _____

– Aorta³¹ ☐ Leftward arch (normal) ☐ Rightward arch

– Other arch anomaly (e.g., vascular ring) ☐ Absent ☐ Present: _____

– Root dilatation ☐ Absent ☐ Present: _____ cm (circumference)

– Dissection ☐ Absent ☐ Present: (type) _____ Ruptured? ☐ Yes ☐ No

– Coarctation/Interruption ☐ Absent ☐ Present

– Supravalvar aortic stenosis ☐ Absent ☐ Present: ☐ Mild ☐ Moderate ☐ Severe

– Ductus arteriosus ☐ Ligamentous (ligamentum arteriosum) ☐ Present, closed

☐ Probe patent ☐ Visibly patent: _____ mm (diameter)

Coronary arteries

– Ostia ☐ Normal³² ☐ Abnormal: (e.g., stenosis) _____

– Distribution ☐ Normal, right dominant ☐ Normal, left dominant³³ ☐ Abnormal

If abnormal ☐ Single ☐ Left anterior descending from right ☐ Circumflex from right

☐ Other: _____

– Aneurysm ☐ Absent ☐ Present: _____

– Dissection ☐ Absent ☐ Present: _____

– Narrowing ☐ Absent ☐ Present: _____ ☐ Atherosclerotic ☐ Non-atherosclerotic

²⁷Measurement should be taken at the level of the tips of the left ventricular papillary muscles.

²⁸Description should include location, size, and any intervention. If malalignment is present (e.g., as in tetralogy of Fallot), describe extent and direction – anterior or posterior.

²⁹Describe morphology/pathology in "Left semilunar valve" section.

³⁰D-malposition is commonly referred to as "complete transposition" (i.e., aorta is anterior and rightward of the pulmonary artery).

³¹The aorta is the vessel that gives rise to the coronary arteries.

³²"Normal" includes origin of the conus artery adjacent to right coronary ostium (normal variant).

³³The right coronary artery may be small in left-dominant hearts. Describe in further detail in "Other" section if absent/hypoplastic or if downstream sequelae exist (e.g., myocardial infarction).

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

If atherosclerosis is present, fill out the following table:

Coronary Artery	Greatest % obstruction	Proximal ✓	Mid ✓	Distal ✓	Thrombus +/-	Calcification +/-
Left main						
Left anterior descending						
Diagonal						
Left circumflex						
Obtuse marginal						
Right						
Posterior descending						
Other						

Hypertrophic cardiomyopathy	☐ Absent	☐ Present
Dilated cardiomyopathy	☐ Absent	☐ Present
Left ventricular noncompaction	☐ Absent	☐ Present
Restrictive cardiomyopathy	☐ Absent	☐ Present
Congenital heart disease ^{34,35}	☐ Absent	☐ Present: (type) _____
Valve disease		
– Mitral valve prolapse	☐ Absent	☐ Present
– Valve stenosis	☐ Absent	☐ Present: (location, severity) _____
Cardiovascular interventions present at autopsy ³⁶		
– Pacemaker: (make, model, type) _____	☐ Absent	☐ Present
Interrogated?	☐ Yes ☐ No	Results: _____
– Implantable cardioverter defibrillator: (make, model) _____		
Interrogated?	☐ Yes ☐ No	Results: _____
– Implanted loop recorder: (make, model) _____		
Interrogated?	☐ Yes ☐ No	Results: _____
– Ventricular assist device: (type, location) _____		
– Evidence of congenital heart surgery: (type, location) _____		
– Stents/coils/plugs/occluder devices: (location) _____		
– Other: _____		

³⁴Probe patent foramen ovale is considered a normal variant and should not be included under congenital heart disease.

³⁵Surgical status will be recorded under evidence of cardiovascular interventions

³⁶With the exception of valve prostheses, which should be described in the valve sections above.

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Heart – Microscopic Examination (Describe findings on page 17)

The extent of microscopic examination is guided by the available history and the gross findings.

For a grossly normal heart, at a minimum:

- 2 sections of left ventricle that include the anterolateral and posteromedial papillary muscles
- 1 section of basilar ventricular septum
- 1 section of right ventricle
- An additional 4-6 sections of myocardium taken from a variety of locations in the ventricles and septum (to look for myocarditis, which can be patchy; if there is recent history of viral illness, it is advisable to take more)

Myocardium:

- Take sections of any areas of discoloration, softening, or mass.
- Taking sections of old myocardial infarction scars is usually uninformative, but areas of myocardium with randomly dispersed interstitial scars should be sampled.
- In cases of suspected hypertrophic cardiomyopathy, the ventricular septum should be carefully sampled to look for myocyte disarray.
- In cases of suspected arrhythmogenic right ventricular cardiomyopathy, multiple sections of the anterior and posterior walls of the right ventricle should be taken.
- Make note of:
 - Hypertrophy
 - Myocyte disarray
 - Necrosis (coagulative vs. contraction-band; focal vs. geographic; specific distribution)

- Fibrosis (replacement vs. interstitial; specific distribution)
- Inflammation (prominent cell type(s); presence/absence of myocyte necrosis)
- Infiltrate (e.g., fat, amyloid)
- Epicardial surface (e.g., presence/absence of inflammation and exudate)
- Epicardial arteries (atherosclerosis)
- Intramyocardial arteries (thrombi, fibromuscular dysplasia)

Coronary arteries:

- Take sections of the greatest area of obstruction of each artery.
- Take sections of any other grossly visible lesion (e.g., aneurysm, dissection); consider including elastic stain.

Valves:

- Take sections of any vegetations (consider including Brown & Brenn tissue gram stain).
- Take a section of a mitral leaflet if it appears to have myxoid degeneration (include an Alcian Blue (AB)-Periodic acid-Schiff (PAS) stain).

Conduction system:

- Examination of the conduction system³⁷ should be done in all cases where:
 - There is documented history of heart block, OR
 - The decedent is an infant/small child and there is a known history of maternal lupus, OR
 - Myxoid valvular disease is present.
- If number of histology blocks is not a financial consideration, doing microscopic examination of the conduction system should be considered in any apparent sudden cardiac death case.

Brain – Gross Examination (Describe findings on page 17)

- Photographs should be taken with the brain in place and cranial vault removed. This is helpful for evaluation of brain swelling. All photographs should be made with a ruler.

-Photographs: ☐ Vertex view ☐ Right view ☐ Left view ☐ Base View

- Photographs
 - Epidural surface of dura mater
 - Subdural surface of dura mater
 - Dorsal brain
 - Ventral brain
 - Right side of brain
 - Left side of brain
 - Evidence of surgical intervention

Evidence of surgical intervention ☐ Absent ☐ Present: If present, circle/describe all that apply:

- Craniotomy: _____
- Craniectomy: _____
- Hardware in skull: _____
- Dural grafts: _____
- Tubes, drains: _____

Dural sinus thrombosis ☐ Absent ☐ Present: ☐ Sagittal ☐ Transverse

Subdural hemorrhage ☐ Absent ☐ Present: ☐ Left ☐ Right ☐ Bilateral

- If present:
 - Amount _____ ml
 - Color _____
 - Appearance: ☐ Clotted ☐ Liquid ☐ Shiny surface

³⁷A stepwise description of the technique can be found in Gulino SP. Examination of the cardiac conduction system: forensic application in cases of sudden cardiac death. Am J Forensic Med Pathol 2003;24(3):227-38.

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Brain – Gross Examination (continued)

Purulent material in subdural space ☐ Absent ☐ Present

– If present, bacterial culture obtained ☐ Yes, results: _____ ☐ No

Subarachnoid hemorrhage ☐ Absent ☐ Present

– If present Pattern: ☐ Diffuse ☐ Scattered ☐ Focal, location: _____
Severity: ☐ Mild ☐ Moderate ☐ Severe _____

Leptomeninges

– Clear ☐ Yes ☐ No:

If no:

– Purulent material ☐ Absent ☐ Present

If present, bacterial culture obtained ☐ Yes, results: _____ ☐ No

– Clouding ☐ Absent ☐ Present

If present, bacterial and viral culture obtained ☐ Yes, results: _____ ☐ No

– Congestion ☐ Absent ☐ Present

Brain removed³⁸ ☐ No ☐ Yes: ☐ By pathologist ☐ By pathology resident ☐ By technician

Brain weight (unfixed)³⁹ _____ g

• Fix brain in 10 – 20% buffered formalin for 2 weeks or longer.^{40,41}

• Suspend brain so that is not deformed by container. This can be done by suspension with a thread under the basilar artery or by using concentrated formalin until the brain floats

• Request antemortem imaging reports if available for review prior to cutting.

Brain weight (fixed): _____ g

Photographs: -Epidural surface of dura mater -Subdural surface of dura mater -Dorsal brain -Ventral brain
-Right side of brain -Left side of brain -Evidence of surgical intervention

Intradural hemorrhage ☐ Absent ☐ Present

– If present Location: _____
Severity: ☐ Mild ☐ Moderate ☐ Severe

Subdural neomembrane ☐ Absent ☐ Present

– If present Location: ☐ Right cerebral ☐ Left cerebral ☐ Superior tentorium ☐ Inferior tentorium ☐ Posterior fossa
Color: _____

Gyrus pattern ☐ Normal ☐ Aberrant: _____

– Polymicrogyria ☐ Absent ☐ Present, location(s): _____

Circle of Willis:

– Distribution ☐ Normal ☐ Abnormal: _____

– Obstruction ☐ Absent ☐ Present

– Size ☐ Normal ☐ Small ☐ Large Vessel(s): _____

– Aneurysm ☐ Absent ☐ Present

If present: Size _____ mm

Location: _____

Cranial nerves All present: ☐ Yes ☐ No: _____

Symmetric: ☐ Yes ☐ No: _____

Cingulate herniation ☐ Absent ☐ Present: ☐ Right ☐ Left

Uncal herniation ☐ Absent ☐ Present: ☐ Right ☐ Left ☐ Bilateral

Tonsillar herniation ☐ Absent ☐ Present: ☐ Right ☐ Left ☐ Bilateral ☐ Chronic⁴² ☐ Acute

³⁸Removal by forensic pathologist is recommended. This decreases the chances of artifacts, such as tearing of cranial nerves.

⁴¹In some jurisdictions the family must be notified if the brain is retained for fixation.

³⁹Skip this step if the brain is very fragile and the brain can be fixed.

⁴²As in a malformation such as Arnold Chiari

⁴⁰Except in jurisdictions in which this is not allowed.

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Brain – Gross Examination (continued)

Pontomedullary tear ☐ Absent ☐ Present: _____ Depth _____ mm

Cerebral hemispheres ☐ Symmetric ☐ Asymmetric: ☐ Right larger ☐ Left larger

Cerebellar hemispheres ☐ Symmetric ☐ Asymmetric: ☐ Right larger ☐ Left larger

Cerebellar folial sclerosis ☐ Absent ☐ Present, location: _____

Areas of softening ☐ Absent ☐ Present, location: _____

Areas of firmness ☐ Absent ☐ Present, location: _____

Surgical drains or other materials ☐ Absent ☐ Present

– If present: Location: _____ Type of material: _____

Drains patent ☐ Yes ☐ No ☐ N/A

Shunts patent ☐ Yes ☐ No ☐ N/A

- Separate brainstem/cerebellum by horizontal cut through the midbrain.⁴³

Aqueduct: ☐ Normal ☐ Obstructed ☐ Dilated

- Cut the cerebrum in the coronal plane at 1.5 – 2.0 cm intervals.
- Separate the brainstem from the cerebellum by cutting the cerebellar peduncles.
- Divide the cerebellum in midline; slice each hemisphere with sagittal cuts at 0.5 cm intervals.
- Section the brainstem at 0.3 cm intervals.
- Photograph the cut brain sections.⁴⁴

Brain ☐ Symmetric ☐ Asymmetric: _____

Lateral ventricles ☐ Symmetric ☐ Asymmetric: ☐ Right larger ☐ Left larger

☐ Not Dilated ☐ Dilated: ☐ Mild ☐ Moderate ☐ Severe

Mass ☐ Absent ☐ Present: _____

Third ventricle ☐ Normal ☐ Dilated ☐ Obstructed

Fourth ventricle ☐ Normal ☐ Dilated ☐ Obstructed

Cortical ribbon

– Size ☐ Normal ☐ Narrow: _____ ☐ Diffuse ☐ Focal, location(s): _____

– Discoloration ☐ Absent ☐ Present: _____ ☐ Diffuse ☐ Focal, location(s): _____

White matter

– Distribution ☐ Symmetric ☐ Asymmetric: _____

– Discoloration ☐ Absent ☐ Present: _____ ☐ Diffuse ☐ Focal, location(s): _____

Myelination ☐ Normal for age ☐ Abnormal for age: _____

Hippocampi ☐ Symmetric ☐ Asymmetric: ☐ Right smaller ☐ Left smaller

Deep nuclei

– Distribution: ☐ Symmetric ☐ Asymmetric: _____

– Discoloration: ☐ Absent ☐ Present: _____ ☐ Diffuse ☐ Focal, location(s): _____

Pituitary

– Size ☐ Normal ☐ Small ☐ Large

– Necrosis ☐ Absent ☐ Present: _____

– Mass ☐ Absent ☐ Present: _____

– Areas of softness ☐ Absent ☐ Present

If present: Location(s): _____ Size: _____ mm

⁴³Other techniques may be useful (e.g., sagittal sectioning of brainstem if pontomedullary tear suspected; sagittal sectioning of brainstem with cerebellum if Arnold Chiari suspected)

⁴⁴Photographs of cut brain can be done in 2 to 6 photos with multiple sections in each. If abnormalities are found, photograph the involved brain section(s) with possible close-up views of the abnormalities.

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Brain – Gross Examination (continued)

– Areas of firmness ☐ Absent ☐ Present
 If present: Location(s): _____ Size: _____ mm

– Areas of discoloration ☐ Absent ☐ Present
 If present: Location(s): _____ Size: _____ mm
 Color: _____

– Hemorrhage ☐ Absent ☐ Present
 If present: Location(s): _____ Size: _____ mm

– Encephomalacia ☐ Absent ☐ Present
 If present: Location(s): _____ Size: _____ mm

Stroke ☐ Absent ☐ Present, location: _____

Heterotopia ☐ Absent ☐ Present, location: _____

Arterio-venous malformation ☐ Absent ☐ Present, location: _____

Compression of cerebral hemisphere ☐ Absent ☐ Present

Anoxic ischemic encephalopathy ☐ Absent ☐ Present

Other congenital anomalies of the brain ☐ Absent ☐ Present, describe: _____

Brain – Microscopic Examination (Describe findings on page 17)

- Take sections of any abnormal areas⁴⁵
- Also take sections of:
 - Dura⁴⁶
 - Frontal cortex including subcortical white matter
 - Parietal cortex including subcortical white matter
 - Temporal cortex including subcortical white matter and ependymal surface
 - Right hippocampus at level of lateral geniculate nucleus
 - Left hippocampus at level of lateral geniculate nucleus
- Amygdala
- Hypothalamus
- Cerebellum including dentate nucleus and folia
- Midbrain
- Pons
- Medulla
- Keep sectioned brain in formalin until histologic examination is complete.
- Retain brainstem and hippocampi.⁴⁷

Gastrointestinal Tract – Gross Examination

External Examination

Abdominal distention ☐ Absent ☐ Present
 -If present: ☐ Postmortem gas ☐ Asymmetry ☐ Fluid wave

Scar(s) from previous abdominal surgery ☐ Absent ☐ Present: _____

External feeding tube ☐ Absent ☐ Present: _____

Internal Examination

- Photography: optional *In situ* *On cutting board*
- Testing: sampling for viral and bacterial cultures (as indicated)

Peritoneal Cavity

– Evidence of peritonitis ☐ Absent ☐ Present: _____

– Ruptured abdominal organ ☐ Absent ☐ Present: _____

– Fluid accumulation ☐ Absent ☐ Present: _____

– Injury from resuscitation ☐ Absent ☐ Present: _____

⁴⁵Sections should include borders between normal and abnormal areas.

⁴⁶If subdural hemorrhage/neomembrane present, include interface with the normal dura.

⁴⁷If jurisdiction allows.

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Gastrointestinal Tract – Gross Examination (continued)

- Adhesions ☐ Absent ☐ Present: _____
- Previous surgery ☐ Absent ☐ Present: _____
- Hernia ☐ Absent ☐ Present: _____
- If present: Incarceration: ☐ Absent ☐ Present: _____
- Volvulus ☐ Absent ☐ Present: _____
- Intussusception ☐ Absent ☐ Present: _____
- Appendicitis ☐ Absent ☐ Present: _____
- Foreign object in the peritoneum ☐ Absent ☐ Present: _____

- *Examine the tongue. During examination look for tongue bites if the child has teeth; examine the area of the foramen cecum for a visible or microscopic trace of the origin of the thyroid gland.*

Liver weight within normal range for age ☐ Yes ☐ No: ☐ Larger ☐ Smaller

If the liver is enlarged, does it appear to be a sequela of right heart failure (not a primary liver problem)? ☐ Yes ☐ No

- *Look at the epiglottis (may fall under respiratory/trachea).*
- *Open the esophagus, stomach, and duodenum, and consider opening the jejunum and ileum (strongly recommended).*
- *Open the large bowel.*
- *Use dissection or the squeeze test to evaluate whether the biliary tree passes bile.*
- *Open the gallbladder; optional, obtain bile for later evaluation.*
- *Section the liver and the pancreas.*
- *The pancreas may be sectioned with the duodenum and ampulla (preferred), or after separation from the duodenum.*

- Pancreatitis ☐ Absent ☐ Present: _____
- Adhesions/sequelae of surgery ☐ Absent ☐ Present: _____
- Bleeding ☐ Absent ☐ Present: _____
- Thrombosis ☐ Absent ☐ Present, vessel: _____
- Obstruction ☐ Absent ☐ Present: _____
- Dilatation ☐ Absent ☐ Present: _____
- Stenosis ☐ Absent ☐ Present: _____
- Fistulas ☐ Absent ☐ Present: _____
- Foreign objects ☐ Absent ☐ Present: _____
- Masses - wall, including reduplications ☐ Absent ☐ Present: _____
- Masses in the lumen ☐ Absent ☐ Present: _____
- Intussusception ☐ Absent ☐ Present: _____

- Volvulus ☐ Absent ☐ Present: _____
- Toxic megacolon ☐ Absent ☐ Present: _____
- Prolapse (rectal or other) ☐ Absent ☐ Present: _____
- Reflux ☐ Absent ☐ Present: _____
- Inflammation ☐ Absent ☐ Present: _____
- Diarrhea ☐ Absent ☐ Present: _____
- Constipation ☐ Absent ☐ Present: _____
- Sequelae of necrotizing enterocolitis ☐ Absent ☐ Present: _____
- Sequelae of G.I. diseases/infections⁴⁸ ☐ Absent ☐ Present: _____
- Congenital abnormalities ☐ Absent ☐ Present: _____

⁴⁸In neonates, systemic Herpes infection may include hepatitis.

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Gastrointestinal Tract – Microscopic Examination (Describe findings on page 17)

- Take sections of any abnormal areas.
- Also take sections of:
 - Tongue at foramen cecum (optional)
 - Epiglottis (optional)
 - Proximal esophagus (optional)
 - Gastroesophageal junction, for reflux (required in infants; optional in children/young adults)
 - Gastric wall (optional)
 - Pyloroduodenal junction (recommended in infants; optional in children and young adults)
 - Proximal duodenum (if evaluating for villous atrophy, some immunodeficiency syndromes, or parasites; optional otherwise)
 - Ampulla of Vater with adjacent duodenum and head of the pancreas (optional)
 - Tail of the pancreas (optional)
 - Liver
 - Gallbladder, biliary tree (optional)
 - Jejunal and ileal sections (if evaluating for villous atrophy, enteritis, or parasites; optional otherwise)
 - Ileocecal junction (recommended in infants; optional in children and young adults)
 - Appendix tip or base (optional)
 - Ascending or transverse colon (optional)
 - Descending or rectosigmoid colon (recommended in infants and children; optional in children and young adults)
 - Anorectum (optional)

Infectious Diseases

Neurologic			Gastrointestinal		
- Encephalitis	☐ Absent	☐ Present	- Enterocolitis	☐ Absent	☐ Present
- Meningitis	☐ Absent	☐ Present	Other		
Respiratory			- Diffuse rash	☐ Absent	☐ Present
- Pharyngitis	☐ Absent	☐ Present	- Soft tissue lesion	☐ Absent	☐ Present
- Epiglottitis	☐ Absent	☐ Present	- Lymphadenitis	☐ Absent	☐ Present
- Bronchitis/bronchiolitis	☐ Absent	☐ Present	- Sepsis syndrome (e.g., disseminated intravascular coagulopathy)	☐ Absent	☐ Present
- Pneumonia	☐ Absent	☐ Present	- Urinary tract infection	☐ Absent	☐ Present
Cardiac			- Other: _____		
- Myocarditis	☐ Absent	☐ Present			
- Endocarditis	☐ Absent	☐ Present			

Specimens

The following should not be construed as requiring every sample for every examination, but should guide the autopsy physician's selection of specimens recovered based upon antemortem signs and symptoms and postmortem anatomic findings.

- ☐ Nasopharyngeal swab for viral culture
- ☐ Cerebrospinal fluid
- Blood cultures ☐ Aerobic ☐ Anaerobic
- Trachea ☐ Culturette ☐ Fresh tissue (obtained in a sterile fashion)
- Bronchus ☐ Culturette ☐ Fresh tissue (obtained in a sterile fashion)
- Lung culturette(s)
 - ☐ Right upper lobe ☐ Right middle lobe ☐ Right lower lobe
 - ☐ Left upper lobe ☐ Left lower lobe
- Sterilely obtained fresh lung tissue
 - ☐ Right upper lobe ☐ Right middle lobe ☐ Right lower lobe
 - ☐ Left upper lobe ☐ Left lower lobe
- ☐ Stool sample

Were additional specialists consulted on this autopsy (e.g., cardiac pathologist, neuropathologist)? ☐ Yes ☐ No

If yes, specify: _____

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Gross Examination of Organs Summary Table

Organ	In situ exam	Gross weight of organ	Fixed or fresh (check)	Gross inspection (check box if normal; if not, describe abnormalities)	Sections retained? ⁴⁹
Brain (including leptomeninges)				☐ Normal	☐ Yes ☐ No
Neck structures ⁵⁰		Thyroid gland ⁵¹ Thymus	☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Body cavities ⁵²			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Heart			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Kidneys			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Liver			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Lungs			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Pancreas			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Spleen			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Gastrointestinal tract			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No

⁴⁹Small tissue samples in formalin.

⁵⁰Neck structures include: epiglottis, aryepiglottic folds, arytenoid and thyroid cartilage to include the vocal cords, cricothyroid membrane, the cricoid cartilage and the tracheal rings, thyroid gland, strap muscles, and the vessels and nerves including those within the carotid sheath and tongue. Under 1 year old include the subglottic musculature.

⁵¹In infants the thyroid may be too small to weigh.

⁵²Body cavities include the pleural, peritoneal and pericardial cavities and pelvis.

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Tissue Sampling and Histology

Sampled Tissue	Number of Sections	Describe Abnormalities
Airways		
Brain (including leptomeninges)		
Heart		
Kidneys		
Liver		
Lungs		
Pancreas		
Spleen		
Thymus		
Bone or costochondral tissue		Location: Abnormalities:
Endocrine organs ⁵³		
Gastrointestinal tract		

⁵³Endocrine organs include: adrenal glands, pituitary gland, and the thyroid gland. The testes/ovaries can also be included.

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Ancillary Testing

Testing	Describe Testing Performed	Results
	Lab name and type of testing (toxicology panel or genetic testing for Long QT, etc.)	
Microbiology/cultures for infectious disease		☐ Normal ☐ Abnormal If abnormal, describe:
Postmortem metabolic screen		☐ Normal ☐ Abnormal If abnormal, describe:
Toxicology		☐ Normal ☐ Abnormal If abnormal, describe:
Vitreous testing		☐ Normal ☐ Abnormal If abnormal, describe:
Genetic testing		☐ Normal ☐ Abnormal If abnormal, describe:
Other, specify:		☐ Normal ☐ Abnormal If abnormal, describe:

Final Pathologic Diagnosis

Was the family referred to a tertiary care center with subspecialty expertise relevant to the cause of death (e.g., cardiology, neurology) for screening of at-risk relatives and genetic counseling?

☐ Yes ☐ No ☐ N/A Where:

APPENDIX 6

SDY AUTOPSY SUMMARY

SUBJECT ID:

Instructions

This summary sheet of autopsy results should be completed following your investigation of a sudden and unexpected death in a child or youth under age 20. Once completed, please share this summary with your local or state Child Death Review Team for its SDY case review process. We hope you are able to participate in the reviews. Your jurisdiction is participating in the Sudden Death in the Young Case Registry, funded by the National Institutes of Health and the Centers for Disease Control and Prevention. The autopsy findings will be summarized with other case review information and biospecimen data (upon family consent) into the SDY Case Registry. Analysis of this comprehensive data will help us better understand the etiologies and risk factors for sudden death in the young.

Limited guidance is provided throughout this summary sheet in italics and footnotes. Please try to complete all elements by circling the appropriate responses and most importantly describing any abnormalities.

A longer and more detailed document called the SDY Autopsy Guidance is also available to you for further direction and instructions. If you do not have a copy you can request one from your SDY State Coordinator.

"Sudden" implies death within 24 hours of the first symptom, or those resuscitated from cardiac arrest and dying during the same hospital admission.

"Unexpected" refers to a death in someone who dies from an accidental injury or someone who was believed to have been in good health, or had a stable chronic condition or had an illness but death was not expected. Examples could include hypertrophic or dilated cardiomyopathy, congenital heart disease, epilepsy, asthma and pneumonia.

This summary, the guidance and instructions were developed by the SDY Autopsy Protocol Committee composed of medical examiners with experience in pediatric, cardiac and neuropathology, physician coroners, death investigators, and other medical professionals with experience in cardiology, neurology, emergency medicine, public health and genetics.

Inclusion and Exclusion Criteria

This autopsy results summary sheet is a key component of the SDY Case Registry and should be used for all cases that meet all of the following inclusion criteria and none of the following exclusion criteria:

Inclusion Criteria

Is the child under 20 years old?	☐ Yes, Include	☐ No, Exclude
Was the death sudden and unexpected and/or unwitnessed?	☐ Yes, Include	☐ No, Exclude

Exclusion Criteria

Was the death caused by an accident in which the external cause was <u>the obvious and only</u> reason* for the death?	☐ Yes, Exclude	☐ No, Include
*Exception: All infants under 1 year of age whose death was caused by suffocation		☐ Include
Was the death an obvious homicide?	☐ Yes, Exclude	☐ No, Include
Was the death an obvious suicide?	☐ Yes, Exclude	☐ No, Include
Was the death caused by an accidental or intentional overdose of drugs even if this caused cardiac or respiratory arrest?	☐ Yes, Exclude	☐ No, Include
Was the death caused by a terminal illness in which the death was reasonably expected to occur within 6 months?	☐ Yes, Exclude	☐ No, Include

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SDY AUTOPSY SUMMARY

SUBJECT ID: _____

SDY Autopsy Guidance and Instructions consulted?☐ Yes ☐ No**Were additional specialists consulted on this autopsy** (e.g., cardiac pathologist, neuropathologist)?☐ Yes ☐ No

If yes, specify:

GeneralSex: ☐ Male ☐ Female

Body weight: _____ kg Body length: _____ cm

Head circumference: _____ cm

External Exam: If abnormalities suggestive of trauma, disease/syndrome, or medical intervention, please describe:

Photography (external) ☐ yes ☐ no**Imaging**

(Circle all that were performed and describe the location)

X-Ray, single:

X-Ray, multiple views:

CT scan:

MRI:

Describe any abnormalities found on imaging:

SDY AUTOPSY SUMMARY

SUBJECT ID: _____

Gross Examination of Organs

Organ	In situ exam	Gross weight of organ	Fixed or fresh (check)	Gross inspection (check box if normal; If not, describe abnormalities)	Sections retained ¹ ?
Brain (including leptomeninges)				☐ Normal	☐ Yes ☐ No
Neck structures ²		Thyroid gland ³ Thymus	☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Body cavities ⁴			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Heart			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Kidneys			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Liver			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Lungs			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Pancreas			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
Spleen			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No
GI tract			☐ Fresh ☐ Fixed	☐ Normal	☐ Yes ☐ No

¹Small tissue samples in formalin.

²Neck structures include: epiglottis, aryepiglottic folds, arytenoid and thyroid cartilage to include the vocal cords, cricothyroid membrane, the cricoid cartilage and the tracheal rings, thyroid gland, strap muscles, and the vessels and nerves including those within the carotid sheath and tongue. Under 1 y.o. include the subglottic musculature.

³In infants the thyroid may be too small to weigh.

⁴Body cavities include the pleural, peritoneal and pericardial cavities and pelvis.

SDY AUTOPSY SUMMARY

SUBJECT ID: _____

Detailed Review of Specified Organs

Thorax/Lungs

Aspiration

Pneumonia/consolidation ☐ Absent ☐ Present
Pulmonary artery thromboemboli⁵ ☐ Absent ☐ Present (location): _____
Hemorrhage ☐ Absent ☐ Present
If present: Diffuse _____ Focal, location: _____ Aspiration pattern (follows bronchi)
Pulmonary hypertension⁶ ☐ Absent ☐ Present

Heart

Hemopericardium ☐ Absent ☐ Present
Vascular ring ☐ Absent ☐ Present
Right ventricular fat infiltration⁷ ☐ Absent ☐ Present
If present, circle which wall ☐ Anterior ☐ Posterior Maximum % thickness of wall involved: _____
Right ventricular thinning⁸ ☐ Anterior ☐ Posterior Location: _____
Hypertrophic cardiomyopathy ☐ Absent ☐ Present
Ventricular septal thickness⁹: _____ cm
Dilated cardiomyopathy ☐ Absent ☐ Present
Left ventricular noncompaction ☐ Absent ☐ Present
Restrictive cardiomyopathy ☐ Absent ☐ Present
Congenital heart disease^{10,11} ☐ Absent ☐ Present (type) _____
Valve disease:
Mitral valve prolapse ☐ Absent ☐ Present
Valve stenosis ☐ Absent ☐ Present (location, severity) _____
Myocardial infarction (recent) ☐ Absent ☐ Present (location) _____
Coronary arteries: ☐ Ostia: ☐ Normal¹² ☐ Abnormal: (location) _____
Distribution: ☐ Normal, right dominant ☐ Normal, left dominant¹³ ☐ Co-dominant
☐ Single ☐ Left anterior descending from right ☐ Circumflex from right
☐ Other: _____
Aneurysm: ☐ Absent ☐ Present
Dissection: ☐ Absent ☐ Present
Atherosclerosis: ☐ Absent ☐ Present

⁵If there is any question whether blood clots in the mainstem pulmonary artery branches are antemortem thromboemboli or postmortem clot, histology is definitive.

⁶Muscle layers in subpleural arterioles

⁷Concerning for arrhythmogenic right ventricular cardiomyopathy

⁸Concerning for arrhythmogenic right ventricular cardiomyopathy

⁹"Normal" includes origin of the conus artery adjacent to right coronary ostium (normal variant).

¹⁰Probe patent foramen ovale is considered a normal variant and should not be included under congenital heart disease.

¹¹Surgical status will be recorded under evidence of cardiovascular interventions.

¹²"Normal" includes origin of the coronary artery adjacent to right coronary ostium (normal variant).

¹³The right coronary artery may be small in left-dominant hearts. Describe in further detail in "Other" section if absent/hypoplastic or if downstream sequelae exist (e.g., myocardial infarction).

SDY AUTOPSY SUMMARY

SUBJECT ID: _____

Heart (continued)

Evidence of cardiovascular interventions ☐ Absent ☐ Present

Pacemaker (make, model, type) _____

Interrogated? ☐ Yes ☐ No

Results: _____

Implanted defibrillator (make, model) _____

Interrogated? ☐ Yes ☐ No

Results: _____

Implanted loop recorder (make, model) _____

Interrogated? ☐ Yes ☐ No

Results: _____

Ventricular assist device (type, location) _____

Evidence of congenital heart surgery (type, location) _____

Stents/coils/plugs/occluder devices (type, location) _____

Prosthetic valves (type, location) _____

Other: _____

Brain

Dural sinus thrombosis

☐ Absent

☐ Present

☐ Sagittal

☐ Transverse

Epidural hemorrhage

☐ Absent

☐ Present

Subdural hemorrhage

☐ Absent

☐ Present

☐ Left

☐ Right

☐ Bilateral

If present:

Amount: _____ ml

Color: _____

Appearance:

☐ Clotted

☐ Liquid

☐ Shiny surface

Subarachnoid hemorrhage

☐ Absent

☐ Present

If present:

Pattern:

☐ Diffuse

☐ Scattered

☐ Focal, location: _____

Severity:

☐ Mild

☐ Moderate

☐ Severe

Circle of Willis

Distribution:

☐ Normal

☐ Abnormal: _____

Obstruction:

☐ Absent

☐ Present

Size:

☐ Normal

☐ Small

☐ Large

☐ Vessel(s): _____

Aneurysm:

☐ Absent

☐ Present

If present:

Size: _____ mm

Location: _____

Cingulate herniation

☐ Absent

☐ Present

☐ Right

☐ Left

Uncal herniation

☐ Absent

☐ Present

☐ Right

☐ Left

☐ Bilateral

Tonsillar herniation

☐ Absent

☐ Present

☐ Right

☐ Left

☐ Bilateral

☐ Chronic¹⁴

☐ Acute

Stroke

☐ Absent

☐ Present

☐ Location: _____

Heterotopia

☐ Absent

☐ Present

☐ Location: _____

Arterio-venous malformation

☐ Absent

☐ Present

☐ Location: _____

Compression of cerebral hemisphere

☐ Absent

☐ Present

Anoxic ischemic encephalopathy

☐ Absent

☐ Present

Other congenital anomalies of the brain

☐ Absent

☐ Present

Describe: _____

¹⁴As in a malformation such as Arnold Chiari

SDY AUTOPSY SUMMARY

SUBJECT ID: _____

Detailed Review of Specified Organs

Gastrointestinal Tract

Intussusception	☐ Absent	☐ Present
Obstruction	☐ Absent	☐ Present
Ruptured abdominal organ	☐ Absent	☐ Present
Volvulus	☐ Absent	☐ Present

Infectious Diseases

Epiglottitis	☐ Absent	☐ Present
Encephalitis	☐ Absent	☐ Present
Meningitis	☐ Absent	☐ Present
Myocarditis	☐ Absent	☐ Present
Endocarditis	☐ Absent	☐ Present
Pneumonia	☐ Absent	☐ Present
Urinary tract infection	☐ Absent	☐ Present

Tissue Sampling and Histology

Sampled Tissue	Number of Sections	Describe Abnormalities
Airways		
Brain including leptomeninges		
Heart		
Kidney		
Liver		
Lungs		
Pancreas		
Spleen		
Thymus		
Bone or costochondral tissue		Location: _____ Abnormalities: _____
Endocrine organs ¹⁵		
Gastrointestinal tract		

¹⁵Endocrine organs include: adrenal glands, pituitary gland, and the thyroid gland. The testes/ovaries can also be included.

SDY AUTOPSY SUMMARY

SUBJECT ID: _____

Ancillary Testing

Testing	Describe Testing Performed	Results
	E.g. lab name and type of testing (toxicology panel or genetic testing for Long QT, etc...)	Circle Normal or Abnormal If Abnormal, Describe
Microbiology/cultures for infectious disease		☐ Normal ☐ Abnormal If abnormal, describe:
Postmortem metabolic screen		☐ Normal ☐ Abnormal If abnormal, describe:
Toxicology		☐ Normal ☐ Abnormal If abnormal, describe:
Vitreous testing		☐ Normal ☐ Abnormal If abnormal, describe:
Genetic testing		☐ Normal ☐ Abnormal If abnormal, describe:
Other, specify:		☐ Normal ☐ Abnormal If abnormal, describe:

Final Pathologic Diagnosis

Was the family referred to a tertiary care center with subspecialty expertise relevant to the cause of death (e.g. cardiology, neurology) for screening of at-risk relatives and genetic counseling?

☐ Yes ☐ No ☐ N/A

Where: