



SUDDEN DEATH IN THE YOUNG CASE REGISTRY

Introduction

This Guidance is a reference to assist you in completing the Sudden Death in the Young Autopsy Summary. It will guide you through the steps necessary to conduct a comprehensive autopsy 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 and the US Centers for Disease Control and Prevention. A committee of medical experts representing forensic pathology, cardiac pathology, neuropathology, pediatric medical specialties and death investigators developed this guidance.

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) into 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 the obvious and only 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

General

Sex: 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:

Detailed Review of Specified Organs

Thorax/Lungs

Thorax/Lungs Imaging:

- 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.

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

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.

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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: _____
 - Hypoinflation² 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 only³ 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
 - Cobblestoning⁴ 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 consultant. If consultation is not requested separate the lungs from the heart following 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 edema⁵ Absent Present: _____
- Pulmonary infection Absent Present: _____
- Pulmonary hemorrhage Absent Present: _____
 - If present: Diffuse Focal, location: _____ Aspiration pattern (follows bronchi)
- Pulmonary hypertension⁶ Absent Present
- Other: _____

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

⁵Consider SUDEP

³Probable postmortem change

⁶Muscle layers in subpleural arterioles

⁴Areas of pink hyperinflation and purple hypoinflation

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-ringed 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.

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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:

-Autoimmune disease	-Bacterial infection	-Granulomatous disease (acid-fast bacteria, sarcoidosis, fungi)
	-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 circulation¹⁶ Absent Present

Asthma or other eosinophilic diseases Absent Present

Foreign bodies¹⁷ 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	<input type="checkbox"/> Symmetrical	<input type="checkbox"/> Asymmetrical	Tracheal contents	<input type="checkbox"/> Absent	<input type="checkbox"/> Present
Erythema	<input type="checkbox"/> Absent	<input type="checkbox"/> Present	If present:	_____	
Exudate	<input type="checkbox"/> Absent	<input type="checkbox"/> Present	White foam, pink foam	<input type="checkbox"/> Absent	<input type="checkbox"/> Present
Aryepiglottic folds	<input type="checkbox"/> Symmetrical	<input type="checkbox"/> Asymmetrical	Mucus	<input type="checkbox"/> Absent	<input type="checkbox"/> Present
	<input type="checkbox"/> Flat (normal)	<input type="checkbox"/> Swollen	Necrotic exudate	<input type="checkbox"/> Absent	<input type="checkbox"/> Present
	<input type="checkbox"/> Obstruct the lumen	<input type="checkbox"/> Do not obstruct the lumen	Thin layer of liquid blood along the mucosa	<input type="checkbox"/> Absent	<input type="checkbox"/> Present
Vocal cords	<input type="checkbox"/> Symmetrical	<input type="checkbox"/> Asymmetrical	Pieces of food, vomitus streaking the mucosa	<input type="checkbox"/> Absent	<input type="checkbox"/> Present
Abnormalities	<input type="checkbox"/> Absent	<input type="checkbox"/> Present:	Obstructing blood clots	<input type="checkbox"/> Absent	<input type="checkbox"/> Present
Tracheal mucosa			Obstructing food bolus	<input type="checkbox"/> Absent	<input type="checkbox"/> Present
Erosion	<input type="checkbox"/> Absent	<input type="checkbox"/> Present	Foreign object	<input type="checkbox"/> Absent	<input type="checkbox"/> Present
Erythema	<input type="checkbox"/> Absent	<input type="checkbox"/> Present			
Inflammation	<input type="checkbox"/> Absent	<input type="checkbox"/> Present:			

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

¹⁷Consider polarization to look for talc in granulomata.

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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, random)
 - 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 protocol

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 Right¹⁸ (normal) Left Ambiguous/other: _____

– Venoatrial connections (SVC/IVC) Normal Abnormal: _____

– Coronary sinus os Patent Stenotic Atretic

– Dilation Absent Present: _____ Mild Moderate Severe

– Cavitory thrombus¹⁹ Absent Present: _____

¹⁸Right atrial morphology includes presence of terminal crest, smooth endocardial surface posterior to terminal crest, pectinate muscles anterior to terminal crest and in atrial appendage. ¹⁹Antemortem thrombus; excludes perimortem/postmortem clot.

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Heart – Gross Examination (continued)

Left atrium

- Morphology Right²⁰ (normal) Left Ambiguous/other: _____
- Venoatrial connections (SVC/IVC) Normal Abnormal: _____
- 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:²¹ _____

Atrioventricular valves Two valves (right and left) Common valve (atrioventricular canal²²)

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 Right²³ (normal) Left Ambiguous/other: _____
- Wall thickness²⁴ Anterior: _____ cm Posterior: _____ cm
- Fat infiltration²⁵ Absent Present If present, which wall: Anterior Posterior
- Maximum % thickness of wall involved: _____
- Right ventricular thinning²⁶ Absent Present Location: _____
- Dilation Absent Present: _____ Mild Moderate Severe
- Cavitory thrombus Absent Present: _____
- Endocardium Thin, translucent (normal) Abnormal: _____

Left ventricle

- Morphology Left²⁷(normal) Right Ambiguous/Other: _____
- Wall thickness²⁸ 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 scar²⁹ Absent Present: _____
- Myocardial discoloration Absent Present: _____

²⁰Left atrial morphology includes absence of terminal crest and smooth endocardial surface throughout atrium except for pectinate muscles in atrial appendage.

²¹Description should include location, size, and any intervention.

²²Describe morphology and pathology under “Left atrioventricular valve” section.

²³Right ventricular morphology includes coarse endomyocardial trabeculations and presence of a moderator band.

²⁴Measurements 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).

²⁵Concerning for arrhythmogenic right ventricular cardiomyopathy

²⁶Concerning for arrhythmogenic right ventricular cardiomyopathy

²⁷Left ventricular morphology includes fine endomyocardial trabeculations and absence of a moderator band.

²⁸Measurements should be taken at the level of the tips of the left ventricular papillary muscles and should include only the compact myocardium (not epicardial fat or papillary/trabecular muscle).

²⁹Includes remote myocardial infarctions

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Heart – Gross Examination (continued)

Ventricular septum

Septal thickness³⁰ _____ cm Intact Ventricular septal defect³¹ _____

Semilunar valves Two valves Single valve³² (truncus arteriosus, pulmonary or aortic atresia)

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

D-malposed³³ 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: _____

– Aorta³⁴ 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 Normal³⁵ Abnormal: (e.g., stenosis) _____

– Distribution Normal, right dominant Normal, left dominant³⁶ 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

³⁰Measurement should be taken at the level of the tips of the left ventricular papillary muscles.

³¹Description 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.

³²Describe morphology/pathology in “Left semilunar valve” section.

³³D-malposition is commonly referred to as “complete transposition” (i.e.,

aorta is anterior and rightward of the pulmonary artery).

³⁴The aorta is the vessel that gives rise to the coronary arteries.

³⁵“Normal” includes origin of the conus artery adjacent to right coronary ostium (normal variant).

³⁶The 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).

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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 Ventricular septal thickness³⁷ _____ cm
- Dilated cardiomyopathy Absent Present
- Left ventricular noncompaction Absent Present
- Restrictive cardiomyopathy Absent Present
- Congenital heart disease^{38,39} Absent Present: (type) _____
- Valve disease
- Mitral valve prolapse Absent Present
- Valve stenosis Absent Present: (location, severity) _____
- Cardiovascular interventions present at autopsy⁴⁰ 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: (location) _____
- Other: _____

³⁷“Normal” includes origin of the conus artery adjacent to right coronary ostium (normal variant).

³⁸Probe patent foramen ovale is considered a normal variant and should not be included under congenital heart disease.

³⁹Surgical status will be recorded under evidence of cardiovascular interventions

⁴⁰With the exception of valve prostheses, which should be described in the valve sections above.

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Heart – Microscopic Examination

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)

Brain – Gross Examination

- 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

- 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)
- Intramycardial 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 system⁴¹ 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.

⁴¹A 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.

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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 removed⁴² No Yes: By pathologist By pathology resident By technician
 Brain weight (unfixed)⁴³ _____ g

- Fix brain in 10 – 20% buffered formalin for 2 weeks or longer.^{44,45}
- 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: _____

Gyral 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 Chronic⁴⁶ Acute

⁴²Removal by forensic pathologist is recommended. This decreases the chances of artifacts, such as tearing of cranial nerves.

⁴⁵In some jurisdictions the family must be notified if the brain is retained for fixation.

⁴³Skip this step if the brain is very fragile and the brain can be fixed.

⁴⁶As in a malformation such as Arnold Chiari

⁴⁴Except 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.*⁴⁷

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.*⁴⁸

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

⁴⁷Other techniques may be useful (e.g., sagittal sectioning of brainstem if pontomedullary tear suspected; sagittal sectioning of brainstem with cerebellum if Arnold Chiari suspected)

⁴⁸Photographs 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

- Take sections of any abnormal areas⁴⁹
- Also take sections of:
 - Dura⁵⁰
 - 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.⁵¹

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: _____

⁴⁹Sections should include borders between normal and abnormal areas.

⁵⁰If subdural hemorrhage/neomembrane present, include interface with the normal dura.

⁵¹If 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.*

Organ weights

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.*

Evaluate grossly for:

- | | |
|--|---|
| Pancreatitis <input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ | Volvulus <input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ |
| Adhesions/sequelae of surgery
<input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ | Toxic megacolon
<input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ |
| Bleeding <input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ | Prolapse (rectal or other)
<input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ |
| Thrombosis <input type="checkbox"/> Absent <input type="checkbox"/> Present, vessel: _____ | Reflux <input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ |
| Obstruction <input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ | Inflammation <input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ |
| Dilatation <input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ | Diarrhea <input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ |
| Stenosis <input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ | Constipation <input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ |
| Fistulas <input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ | Sequelae of necrotizing enterocolitis
<input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ |
| Foreign objects
<input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ | Sequelae of G.I. diseases/infections ⁵²
<input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ |
| Masses - wall, including reduplications
<input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ | Congenital abnormalities
<input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ |
| Masses in the lumen
<input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ | |
| Intussusception
<input type="checkbox"/> Absent <input type="checkbox"/> Present: _____ | |

⁵²In neonates, systemic Herpes infection may include hepatitis.

SUDDEN DEATH IN THE YOUNG AUTOPSY GUIDANCE

Gastrointestinal Tract – Microscopic Examination

- 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)
 - Jejunum 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

- Encephalitis Absent Present
- Meningitis Absent Present

Respiratory

- Pharyngitis Absent Present
- Epiglottitis Absent Present
- Bronchitis/bronchiolitis Absent Present
- Pneumonia Absent Present

Cardiac

- Myocarditis Absent Present
- Endocarditis Absent Present

Gastrointestinal

- Enterocolitis Absent Present

Other

- Diffuse rash Absent Present
- Soft tissue lesion Absent Present
- Lymphadenitis Absent Present
- Sepsis syndrome (e.g., disseminated intravascular coagulopathy)
 - Absent Present
- Urinary tract infection Absent Present
- Other: _____

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: _____

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 ⁵³ ?
Brain (including leptomeninges)				<input type="checkbox"/> Normal	<input type="checkbox"/> Yes <input type="checkbox"/> No
Neck structures ⁵⁴		Thyroid gland ⁵⁵ Thymus	<input type="checkbox"/> Fresh <input type="checkbox"/> Fixed	<input type="checkbox"/> Normal	<input type="checkbox"/> Yes <input type="checkbox"/> No
Body cavities ⁵⁶			<input type="checkbox"/> Fresh <input type="checkbox"/> Fixed	<input type="checkbox"/> Normal	<input type="checkbox"/> Yes <input type="checkbox"/> No
Heart			<input type="checkbox"/> Fresh <input type="checkbox"/> Fixed	<input type="checkbox"/> Normal	<input type="checkbox"/> Yes <input type="checkbox"/> No
Kidneys			<input type="checkbox"/> Fresh <input type="checkbox"/> Fixed	<input type="checkbox"/> Normal	<input type="checkbox"/> Yes <input type="checkbox"/> No
Liver			<input type="checkbox"/> Fresh <input type="checkbox"/> Fixed	<input type="checkbox"/> Normal	<input type="checkbox"/> Yes <input type="checkbox"/> No
Lungs			<input type="checkbox"/> Fresh <input type="checkbox"/> Fixed	<input type="checkbox"/> Normal	<input type="checkbox"/> Yes <input type="checkbox"/> No
Pancreas			<input type="checkbox"/> Fresh <input type="checkbox"/> Fixed	<input type="checkbox"/> Normal	<input type="checkbox"/> Yes <input type="checkbox"/> No
Spleen			<input type="checkbox"/> Fresh <input type="checkbox"/> Fixed	<input type="checkbox"/> Normal	<input type="checkbox"/> Yes <input type="checkbox"/> No
GI tract			<input type="checkbox"/> Fresh <input type="checkbox"/> Fixed	<input type="checkbox"/> Normal	<input type="checkbox"/> Yes <input type="checkbox"/> No

⁵³Small tissue samples in formalin.

⁵⁴Neck 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.

⁵⁵In infants the thyroid may be too small to weigh.

⁵⁶Body cavities include the pleural, peritoneal and pericardial cavities and pelvis.

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 ⁵⁷		
Gastrointestinal tract		

⁵⁷Endocrine organs include: adrenal glands, pituitary gland, and the thyroid gland. The testes/ovaries can also be included.

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		<input type="checkbox"/> Normal <input type="checkbox"/> Abnormal If abnormal, describe:
Postmortem metabolic screen		<input type="checkbox"/> Normal <input type="checkbox"/> Abnormal If abnormal, describe:
Toxicology		<input type="checkbox"/> Normal <input type="checkbox"/> Abnormal If abnormal, describe:
Vitreous testing		<input type="checkbox"/> Normal <input type="checkbox"/> Abnormal If abnormal, describe:
Genetic testing		<input type="checkbox"/> Normal <input type="checkbox"/> Abnormal If abnormal, describe:
Other, specify:		<input type="checkbox"/> Normal <input type="checkbox"/> 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:



SUDDEN DEATH IN THE YOUNG
CASE REGISTRY

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