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## **8. Fatal Child Abuse and Sudden Infant Death Syndrome: A Critical Diagnostic Decision**

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## Abstract

Distinguishing between an unexpected infant death due to sudden infant death syndrome (SIDS) and one due to fatal child abuse challenges pediatricians, family physicians, pathologists, and child protection agencies. If child abuse is suspected, the physician must fulfill mandated legal obligations to report the case to the appropriate authorities. Coroners, medical examiners, and pathologists have the added responsibility of rendering a medicolegal opinion as to the cause and manner of death.

## Learning Objectives

- To review historical inquiries into the sudden infant death phenomenon
- To define SIDS
- To describe the clinical presentation of SIDS
- To cite the incidence and epidemiology of SIDS
- To distinguish between SIDS and fatal child abuse
- To describe the role of the autopsy
- To delineate the clinical radiographic study in SIDS
- To identify the importance of death scene investigation to the overall postmortem examination
- To describe criteria for distinguishing SIDS from fatal child abuse and other medical conditions
- To recommend improved practices for determining cause and manner of unexpected infant deaths

## Historical Background

Distinguishing SIDS from death due to inflicted injury is a challenge to professionals. The pediatrician or family physician must know which course to follow in relating to the family. If child abuse is suspected, the physician must fulfill mandated legal obligations to report the case to the appropriate authorities. If the reason for death is SIDS, a sympathetic and supportive role is required. Complicating this decision is the polarization between 2 camps: those who believe that a sympathetic approach to parents losing their baby is the highest priority, and those whose training and experience have convinced them that fatal child abuse is distressingly common. In the final analysis, a non-accusatory approach to the caretakers is best because all parties are treated fairly. Coroners, medical examiners, and pathologists have the added responsibility of rendering a medicolegal opinion as to the cause and manner of death. All

agree that the state of our knowledge in this area is incomplete, and ambiguity exists in some cases.

The subject of crib death was described in the Bible (1 Kings 3:19,22) and was known to happen even before biblical history. For centuries, it was accepted as a natural phenomenon, defying explanation. In the 1950s, spurred by parents suffering the loss of infants to this poorly explained condition, the scientific community entered a period of fruitful research about why these infants were dying suddenly and unexpectedly. At the same time, the medical community, which had previously repressed the abhorrent concept of caretakers harming their children, was being informed by new literature describing this phenomenon. In 1946, Caffey<sup>1</sup> published his seminal paper on multiple fractures and subdural hematomas, and in 1953, Silverman<sup>2</sup> postulated that these injuries were the result of unrecognized trauma. In 1961, Adelson<sup>3</sup> added to the factual information about fatal child abuse. In 1962, Kempe et al<sup>4</sup> coined the phrase the "battered child syndrome" and further raised the consciousness of the medical community about the unpleasant truth that infants and children were being physically abused and killed. The stage was being set for a controversy about death in infancy, its causes, and the possibility of a caretaker's culpability for those deaths.

High-profile cases of serial child homicides focused attention on fatal child abuse as a reason for some sudden, unexpected deaths in infants. Waneta Hoyt of upstate New York was charged with suffocating 5 children—all 5 deaths were initially declared as being due to SIDS. Two of the suffocated children were among 5 whose cases were included in a report by Steinschneider<sup>5</sup> in 1972, who studied those cases and found that SIDS was due to prolonged apnea. Steinschneider's article, heralded as a breakthrough despite the small numbers and questionable data interpretation, elevated prolonged apnea to the top of the list of hypotheses for SIDS. This, in turn, spurred the development of research about apnea and, subsequently, the proliferation of the concept of testing for apnea and home monitoring programs. In their book *The Death of Innocents*, Firstman and Talan<sup>6</sup> skillfully analyze this case, the implications it had for the SIDS research community, and the impact it had on pediatric practitioners and hundreds of thousands of their families. The media accounts of the Hoyt case raised the public awareness about the possibility of infant murders being mistaken for crib death or other

medical illness. For those parents who had lost babies to SIDS and for many health care providers, the suggestion that even some SIDS deaths were actually child murders was painful and unacceptable. They feared that by raising the old specter of infanticide, the 25-year effort of parents and professionals to provide compassion for families losing their babies to SIDS would evaporate. It is apparent that an objective and integrated approach to the ascertainment of death in sudden unexpected infant deaths must occur.

### **Definition of SIDS**

Sudden infant death syndrome is defined as "the sudden death of an infant under 1 year of age which remains unexplained after the performance of a complete postmortem investigation, including an autopsy, an examination of the scene of death and review of the case history."<sup>7</sup> Because the proportion of SIDS cases having an atypical clinical presentation, potentially unsafe sleeping environment, and/or more "severe" pathological findings at postmortem examination is increasing (*vide infra*), the need to refine and stratify the definition is becoming more important.<sup>8-10</sup>

### **Incidence and Epidemiology of SIDS**

Sudden infant death syndrome occurs most frequently in infants who are between 2 and 4 months of age.<sup>11</sup> Less than 5% of cases occur during the first month of life, and the number of SIDS deaths decreases progressively after the third to fourth months of life. Approximately 90% of SIDS deaths occur by 6 months of age.<sup>11</sup> In most studies, SIDS occurs more often in males (60% to 70%). It occurs more frequently in the winter months in the northern and southern hemispheres, suggesting that temperature alone is not a causative factor. Little and Peterson<sup>12</sup> suggested the possibility that colder outdoor temperatures may encourage overdressing an infant, producing a microclimate of overheating. Because hyperthermia and overheating have been implicated by some authors as etiologic factors,<sup>13</sup> this concept is interesting, but these authors conclude that the "meaning of climate and ambient temperature pattern for risk of SIDS death remains to be elucidated."

The overall incidence of SIDS has declined dramatically in developed countries of the world, yet it remains the most common cause of post-neonatal

infant death.<sup>14</sup> The National Center for Health Statistics reported that there were 5,476 SIDS cases in the United States in 1988, representing an overall rate of 1.4 deaths per 1,000 live births. This means that in 1988, SIDS was the second leading cause of death in infancy, including the neonatal period, accounting for 14.5% of all deaths among white infants and representing a rate of 1.24 per 1,000 live births. In contrast, for African American infants, SIDS was the leading cause of death, accounting for 12.8% of all deaths and representing a rate of 2.26 per 1,000 live births. By 1995, the number of cases declined to approximately 3,000 deaths<sup>15</sup> now yielding a rate of approximately 0.5 deaths per 1,000 live births.

### **Sleep Position**

Sudden infant death syndrome incidence rates in the United States, western Europe, Australia, and New Zealand have declined following the implementation of public awareness initiatives, such as the "Back to Sleep" campaign, in the United States.<sup>16-19</sup> These initiatives recommend the use of the supine, rather than prone or side infant sleep position; that mothers avoid excessive swaddling or wrapping of their babies; and that mothers not smoke cigarettes during their pregnancies or expose their babies to smoke. These campaigns have not reached all segments of the population. Willinger et al<sup>20</sup> have reported that significant predictors of prone placement included maternal African American race, mother's age between 20 and 29 years, residence in the mid-Atlantic or southern states, mothers with a previous child, and infants younger than 8 weeks. In this study, it was encouraging that the prevalence of infants placed in the prone sleep position declined by 66% between 1992 and 1996 and at the same time, SIDS rates declined approximately 38%.

### **Ethnicity**

African American,<sup>21</sup> Alaskan natives,<sup>22</sup> and most, but not all, Native American infants have higher SIDS rates than white and Asian infants.<sup>23,24</sup> In some studies, the excess mortality among African American infants disappears when adjusted for maternal education and income.<sup>25</sup> A recent report indicates a dramatic decrease in the SIDS rates (8.9 to 3.0/1,000 live births) among American Indians and Alaskan natives.<sup>26</sup> Kraus and Bultreys<sup>27</sup> conclude from their studies on the effect of various

factors on the rate of SIDS within certain populations that SIDS rates and socioeconomic status (SES) are inversely related, but SES may act as a confounder, effect modifier, or intermediate variable.

### ***Cigarette Smoking***

Smoking seems to have emerged as the major risk factor of SIDS because supine sleep position has replaced the prone position.<sup>28–30</sup> Maternal smoking prior to and/or during gestation, and after delivery, increase the risk of SIDS. Paternal smoking also increases the risk of SIDS,<sup>28</sup> but this association may be a reflection of a higher rate of passive smoke exposure by a coexisting smoking mother.

### ***Substance Abuse During Pregnancy***

Infants of illegal substance-abusing mothers (ISAM) also are reported to be at higher risk of SIDS compared to control subjects.<sup>31–33</sup>

### ***Immunizations***

Diphtheria-tetanus-pertussis vaccine does not increase the risk of SIDS.<sup>34–36</sup>

### ***Co-sleeping***

Co-sleeping as a potential risk factor for SIDS has generated controversy. Mosko and colleagues<sup>37</sup> have hypothesized that the comparatively sensory-rich co-sleeping environment might be protective against SIDS in some contexts. After studying mother-infant pairs, these investigators concluded that bed-sharing promoted infant arousals and reduced stage 3–4 sleep and the mother's responsiveness to infant arousals during bed sharing. From an epidemiologic viewpoint, however, investigators in the New Zealand Cot Death Study found that bed sharing significantly increased the risk of SIDS, particularly among infants of mothers who smoked.<sup>38</sup> A more recent prospective study from this same group led to the conclusion that the risk of SIDS was increased if the mother also smoked.<sup>39</sup> This issue is unresolved and remains intensely controversial.

### ***Twins and SIDS***

In most studies, SIDS is seen to be 2½ times more common in multiple births with twins and triplets than in singleton births. This is probably due to the lower mean birth weight of twins compared with singletons. In a summary of the world's literature

concerning this phenomenon (1956 to 1988), Beal<sup>40</sup> reported that 6 of 625 (1%) of the surviving twins had subsequently died of SIDS. In a large study in 1999, Malloy and Freeman<sup>41</sup> studied twin pairs over a 5-year period in the United States to determine if there was an increased risk of SIDS in this population. In the 767 SIDS twin deaths, there were only 7 sets in which both twins died and in only one of these sets were the deaths on the same day. The authors concluded that, independent of birth weight, twins do not seem to be at greater risk for SIDS compared with singletons.

### ***Recurrence of SIDS Within a Family***

A comparison of SIDS occurrences in siblings of SIDS victims and in non-SIDS siblings in maternal age and birth rank-matched control families revealed no statistically significant difference in SIDS rates or in total infant mortality rates in families with a history of SIDS compared with families in which there was not SIDS. With the exclusion from the SIDS statistics of some of the deaths now thought to be due to inborn errors of metabolism (see subsequent discussion), the chances for recurrent SIDS in families seem even less likely. It should be noted that these studies were reported before implementation of initiatives promoting back sleep position and avoidance of smoke exposure. There is a growing impression among pediatric and forensic pathologists that an increasingly higher proportion of SIDS cases are now of the atypical or non-classical type. In this setting of socioeconomic deprivation, less safe sleep environments, persistence of use of the prone sleep position, and continued exposure to tobacco smoke may increase the recurrence risk. Newer studies are urgently needed to address this issue.

### ***Clinical Presentation of SIDS***

Typically, SIDS is suspected when an apparently previously healthy baby, usually younger than 6 months of age (peaking between 2 and 4 months of age), is found unresponsive in bed in the early morning prompting an urgent call for emergency help. Emergency personnel respond and initiate cardiopulmonary resuscitation in the home and continue it on the way to the hospital, where the baby is ultimately pronounced dead. The infant's medical history is usually unremarkable. In many cases a history of a recent routine pediatric visit is elicited.

The immediate antecedent history indicates that the baby had been fed his or her usual formula or breast milk and had been put to bed. At varying intervals the parents or other caretaker had checked the baby, who seemed to be normal, but later the baby had been discovered lifeless. No crying had been heard, and the baby had been found in the position in which he had been placed at bedtime or naptime. Evidence of terminal motor activity such as clenched fists may be seen and there may be some pinkish serosanguinous, watery, frothy, or mucoid discharge coming from the nose and mouth. The face and dependent portions of the body may have reddish-blue mottling due to postmortem lividity.

In non-classical or atypical SIDS, the infant often lives in lower socioeconomic conditions and frequently has a medical history of recent upper respiratory infection, or gastrointestinal illness accompanied by low-grade fever, poor eating, and diarrhea. The sleep site may not have been completely safe due to an unstable bed, soft sleep surfaces, or the presence of pillows and/or overstuffed toys. The infant may have been in a chaotic co-sleeping situation with adults. The postmortem examination may show some abnormalities, such as pulmonary inflammation, that some, but not all, pathologists would consider sufficiently severe to be the cause of death. These cases often are the subject of considerable controversy at child death review committees.

## Theories of Etiology

Sudden infant death syndrome is an event encompassing several disorders with a final common pathway. In fact, controversy still exists whether SIDS represents a developmentally abnormal infant responding inadequately to some environmental stress that would not be lethal to a normal infant, or the inability of a normal infant to respond to an insurmountable environmental stress. Current evidence suggests that either possibility might exist in any one infant.

## Neurologic Hypotheses

There are 2 principle hypotheses centering on neurophysiology and neuropathology in SIDS. The first focuses on an abnormality in the brain's regulation of respiration during sleep and/or stimulation of normal protective arousal during life-threatening events that occur in all infants. The second is known

as the developmental hypothesis in which case SIDS victims are neuroanatomically and neurophysiologically immature. Both conditions may be present simultaneously.

Investigators propose that the maturational regulation of the arcuate nucleus is abnormal in SIDS brains, such that there is a developmental deficiency of neurons with an associated deficiency of neurotransmitter receptor binding. It has been suggested that there may be a lag in neurotransmitter maturation, regardless of a deficiency of neuron number. This may explain the apparently asymptomatic behavior of most SIDS victims and why eliminating an external stressor (eg, prone sleeping position) may reduce the incidence of SIDS. The model also explains why most infants who sleep prone do not die of SIDS, because they are "normal" and do not have an underlying vulnerability.

The concept that an external stressor precipitates sudden death is derived from epidemiologic studies indicating that minor respiratory or gastrointestinal illness occurs around the time of death in some SIDS victims, as well as symptoms of more severe illness in the 2 days before death.<sup>42,43</sup> The prone sleeping position is associated with the spontaneous face-down sleeping position in infants.<sup>15</sup> The face-down position is associated with re-breathing exhaled gases and increased end-tidal CO<sub>2</sub> in normal infants, particularly those sleeping on soft bedding where pockets of exhaled gas form and trap CO<sub>2</sub>.<sup>35</sup> The prone position also may lead to partial or complete upper airway obstruction by repositioning the mandible and occluding the pharynx, compressing the nose directly, or closing the larynx reflexively as part of the "diving" response. At least 26% of SIDS victims are found in the face-down position and 71% are found in the prone position.<sup>17</sup>

It is reasonable to propose that the underlying vulnerability in at least a subset of SIDS victims is an abnormality in the arcuate nucleus cell populations of the ventral medulla. It is possible that the external stressor is hypoxia or hypercapnia that results from upper airway obstruction and/or asphyxial re-breathing in the face-down (prone) sleeping position. A normal infant's nervous system detects progressive hypercapnia and hypoxia and responds by arousal and a series of protective reflexes and behaviors to ensure airway patency. Due to the underlying abnormality in the ventral medulla, the vulnerable SIDS infant fails to arouse,

cry, increase ventilation, and move his or her head in response to the hypercapnia and hypoxia, and death results.

### **Upper Airway Obstruction**

Several investigators have proposed the theory of upper airway obstruction.<sup>44, 45</sup> In 1971, Cross and Lewis<sup>46</sup> suggested a common "march of events" taking place, commencing with a "mild nasal infection in a baby who is an obligate nose breather." Because of the failure to respond to rising CO<sub>2</sub> levels, they argue, the baby finally succumbs. The defect in this theory is that obligate nasal breathing diminishes around 4 to 6 weeks of age, which is before the high SIDS incidence period of 2 to 4 months. Pharyngeal obstruction, because of backward falling of the tongue; pharyngeal collapse during sleep; or neck flexion leading to obstruction have all been suggested as instrumental in producing upper airway obstruction.

### **Gastroesophageal Reflux**

Gastroesophageal reflux with reflex apnea from laryngospasm is posed as another mechanism and has led to preventive medical measures to reduce the number and amount of refluxes, and in extreme cases, to surgical fundoplication. The efficacy of this surgical procedure in preventing any cases of SIDS is unproven, and its use for this purpose now has few proponents.

### **The Apnea Hypothesis**

In 1972, Steinschneider<sup>5</sup> described 5 infants who had been referred to him for cyanotic spells or apnea. Two of those patients (siblings) died, and in both, the diagnosis was initially SIDS. All had been documented as having had episodes of prolonged apnea, made worse by upper respiratory tract infections. However, the 2 infants whose deaths were attributed to SIDS had no prolonged apnea observed in the hospital, and later the mother, Waneta Hoyt, confessed to having smothered them.<sup>6</sup> However, this flawed research paper began a period of vigorous research about apnea as the etiology for SIDS. Guntheroth<sup>47</sup> postulated that a lack of an arousal response to apnea might be instrumental in SIDS. Shannon and Kelly<sup>48</sup> implicated sleep apnea as the mechanism for some "near miss SIDS" based on home monitoring results. Belief in the apnea hypothesis began to erode when Southall

and colleagues<sup>49</sup> presented data on continuous electrocardiogram (ECG) and pneumogram monitoring of more than 9,000 infants, of whom 29 subsequently died of SIDS. Among those SIDS cases, no infant had shown abnormalities in the pneumograms, whereas those with abnormal pneumograms did not die. In 1991, Schechtman et al<sup>50</sup> reported that the 24-hour pneumograms taken between 40 and 65 days of life in SIDS victims showed significantly fewer respiratory pauses than age-matched control infants in quiet sleep and REM sleep; however, during the first month of life, the SIDS victims did not differ significantly from controls. This finding supported the view that some as yet undefined abnormality makes the future SIDS victim more vulnerable between 1 and 4 months. Current consensus is that SIDS victims seldom have a history of apnea as manifested either clinically or by pneumography.

### **Cardiac Hypotheses**

Prolonged QT interval, possibly resulting from a developmental abnormality in cardiac sympathetic innervation, has been advocated as a cause of SIDS.<sup>51</sup> This study has received intense criticism.<sup>52-55</sup> Although the study is strengthened by the large number of cases and a single group analyzing the ECGs, it is weakened in part by the lack of blinded analyses, unknown aware (activity) or sleep states, and failure to list the other causes of death. There is strong consensus that screening newborns for this abnormality would yield results too non-specific to be useful. Further, treatment of infants with powerful medications for an unproven goal (prevention of SIDS) would be inadvisable.

### **Environmental Factors**

Overheating and hyperthermia have been suggested as factors in SIDS after descriptions of such infants being overdressed, over-wrapped, and hot and sweaty when found.<sup>56,57</sup> The mechanism suggested is that of "febrile apnea," the younger infant equivalent of a febrile seizure. Apnea has been associated with elevated ambient temperatures in premature babies,<sup>58</sup> and this occurrence is cited as reason to believe the existence of the phenomenon in older infants. Hyperthermia as a factor contributing to SIDS deaths is gaining deserved attention, but more carefully crafted studies need to be conducted to know the relative importance of this factor. One

such study,<sup>13</sup> using a case-control method, examined this issue in 41 infants who died of SIDS by measuring thermal conditions at the death scene and at the scene of last sleep for control infants. A questionnaire also was administered to all the parents. The results showed that those who died of SIDS had more excess thermal insulation for their given room temperature than the matched controls.

Byard et al<sup>59</sup> reported on 30 cases of unexpected death by asphyxia caused by unsafe sleeping conditions. Deaths occurred from hanging from loose retainers, clothing, or curtain cords; positional asphyxia (wedging); and suffocation from plastic bed covers.

### **Other Disease States**

In 1989, Burchell et al<sup>60</sup> reported finding raised hepatic glycogen levels in the livers of 10 infants who had died of SIDS. Eight of those infants had glucose-6-phosphatase deficiency (type 1a glycogen storage disease), and 2 infants had transport protein T2 deficiency (type 1c glycogen storage disease). Another metabolic abnormality, medium chain acyl-CoA dehydrogenase deficiency (MCAD), allegedly accounts for death in 2% to 3% of infants in whom the diagnosis is SIDS. This disease is an autosomal recessive abnormality in fatty acid oxidation and usually presents with recurrent episodes of hypoglycemia and lethargy mimicking Reye syndrome, or with features clinically indistinguishable from SIDS. Mortality in MCAD is 60% in the first 2 years of life, but when recognized, the disease can be managed effectively.

### **Distinguishing Between SIDS and Child Abuse**

In 1961, Adelson<sup>3</sup> reported on 46 child homicides occurring between 1944 and 1961. Ten children were younger than 1 year. Of those, 5 drowned and 3 died of starvation. The causes of death of the other 2 are not described. In 1991, Adelson<sup>61</sup> reported 194 child homicides: 28 occurred in infants younger than 1 year, 16 occurred in infants between ages 1 month and 1 year, and 7 occurred in infants between ages 1 month and 6 months. All were fatally and obviously battered. Therefore, in this series, there were no cases likely to be confused with SIDS.

Emery and Taylor<sup>62</sup> described a 24-year period in Sheffield, England (1960 to 1984), during which postperinatal deaths (birth to 2 years) were investigated by gathering information about the death scene, obstetric and pediatric care, reviewing autopsy findings, and conducting extensive home visits. As a result of this process, accidental suffocation was thought to be the cause of death in 10% of these cases, and the possibility of active intervention on the part of one or both parents was raised in another 10%, a rate consistently double that of overt child abuse in this age group. Specific data on infants between ages 1 month and 1 year were not reported.

Suggested etiologies for unexpected infant deaths that have been reported include accidental strangulation, intentional suffocation, and Munchausen by proxy. Deaths in infant twins also have been studied extensively, from the standpoint of the possible increased risk of death in the twin survivor of a SIDS death and to ascertain whether there is increased risk of being abused because of twin status. In 1982, Groothuis and colleagues<sup>63</sup> reported on this latter phenomenon after studying 48 families with twins and 124 single-birth families, matched for hospital of delivery, birth date, maternal age, race, and socioeconomic status. Three control (2.4%) and 9 twin (18.7%) families had been reported for maltreatment, with one fatality. Siblings of these twins were abused more frequently than the twins themselves, and abuse was limited to the twins in only 3 families. When analyzing the variables in the families studied, the authors concluded that twin status had the greatest impact on the risk of subsequent child abuse, suggesting that the stress of rearing twins, added to the other elements of childrearing in already marginally functioning families, was a significant determinant for subsequent abuse.

Beal,<sup>40</sup> in her summary of the world's literature concerning the phenomenon of SIDS in twins (1956 to 1988), reported that 6 (1%) of 625 of the surviving twins had subsequently died of SIDS. Data concerning the rate of simultaneous twin SIDS are difficult to interpret. Beal's estimate, based on published series, is that SIDS occurs in 12 of 637 twin infant pairs, or 2% of all twin pairs.

In 1985, Christoffel et al<sup>64</sup> examined 43 unexpected deaths in children brought to Children's Memorial Hospital in Chicago during 1980 to 1981. Nine were

due to child abuse and in 3 the correct diagnosis was established only by postmortem examination. In the same journal issue, Kirschner and Stein<sup>65</sup> described 10 cases in which the diagnosis of child abuse was made based on incomplete or erroneous medical observation. Five of those cases were proved by autopsy as cases of SIDS. The recording of the clinical physical examinations had described conditions that were postmortem changes (eg, lividity and sphincter dilation), misinterpreted skin markings (mongoloid pigmentation), or a physical finding often seen in SIDS deaths (serosanguinous discharge from nose and mouth). These reports emphasize the need for appropriate evaluation before and after death, including thorough physical examinations, autopsies, and death scene investigation.

There are convincing data that at least in some cases, postpartum depression and other psychiatric disturbances, particularly in mothers who had histories of maltreatment themselves, have led to infanticide.

### **Death Investigation: Scene Investigation and Medical History**

By delaying the death scene investigation one may lose accurate documentation in terms of risk factors associated with the environment and sleeping conditions. Prompt interviewing of the discovering caretaker is needed to ascertain details of the infant's situation when first found lifeless. The gathering of information from physicians familiar with the baby and the family and from local child protection agencies also should be accomplished. Scene investigation is integral to the definition and accurate diagnosis of SIDS. It is the forensic pathologist's "medical history" and is analogous to the importance of the clinical history to the pathologist performing postmortem examinations in the hospital. Because the diagnosis of SIDS is one of exclusion, the postmortem examination does not reveal findings of sufficient severity to assign death to another cause. In this regard, it is vital that the scene where the infant was found lifeless be evaluated for environmental conditions, noxious gases, safety of the crib or bed and its sleep surface, and the presence of harmful substances and medications. To facilitate the scene investigation, the Centers for Disease Control and Prevention has produced and published "Guidelines for the Scene Investigation of Sudden Unexpected

Infant Death."<sup>66,67</sup> These guidelines have been endorsed by the SIDS Global Strategy Task Force.

### **Role of the Autopsy**

Although the autopsy has not elucidated the etiology of SIDS, and despite the presence of minor pathologic findings in many cases, it is still considered *sine qua non* in determining the cause of sudden and unexpected death in infancy. In a recent study, Kumar et al<sup>68</sup> reviewed the autopsy findings in 107 cases of post-neonatal patients who had died during a 10-year period at a large suburban medical center. Sixty of those patients were less than a year old. In 34%, a new diagnosis was made at autopsy, while complete concordance was seen in 66%. This is a convincing argument about the value of conducting pediatric autopsies in general. A standardized protocol has been developed for pathologists performing postmortem examinations on infants whose deaths were sudden and unexpected.<sup>69</sup> Endorsed by the Society for Pediatric Pathology, National Association of Medical Examiners, and the SIDS Global Strategy Task Force, this protocol prompts recording of positive and negative observations important to reaching a diagnosis in sudden unexpected infant death by use of a checklist. The protocol encourages narrative descriptions of abnormalities supplemented by microscopic, microbiologic, and toxicologic analyses, as well as the use of radiographic and photographic images when indicated.

Actual autopsy findings in SIDS have been described extensively. The bodies of SIDS victims seem well nourished and well developed, but their weights are typically below the 50th percentile expected for age. A mucoid discharge, occasionally foamy and often tinged with blood, often is present around the nares and mouth. Fibers from bedclothes may be found in the hands, which are often clenched. Signs of resuscitation and postmortem changes must be distinguished from non-accidental trauma. Reddish blue mottling of the skin indicative of postmortem lividity may alter dependent portions of the body. The blood in the heart is liquid and often oozes from venipuncture sites. The bladder and rectum are empty. Intrathoracic petechiae, the most common abnormality seen with the naked eye, are identified in about 80% of SIDS cases.<sup>70,71</sup> Despite opinion to the contrary,<sup>43</sup> facial and conjunctival petechiae are not seen in SIDS, and their presence should provoke a search for another cause of death. Experimental evidence and observations in human

postmortem examinations suggest petechiae limited to the thorax can result by breathing against an obstructed upper airway during the moments preceding death.<sup>42,70,71</sup> Alternatively, it also has been suggested that bronchiolar obstruction could cause the same finding.

The lungs are congested and variably edematous, but not consolidated. It is unlikely that pneumonia that cannot be seen with the naked eye is lethal. Given that infants may have considerable clinical and radiographic evidence of pneumonia and, even if ill, are nevertheless alive, it seems all the more improbable that microscopic pulmonary inflammatory infiltrates are lethal.

Microscopic examination of the lungs, heart, and leptomeninges may show mild, focal interstitial lymphocytic infiltrates in SIDS cases. These should not be interpreted as a lethal finding. These infiltrates in the heart are not accompanied by myocardial necrosis, thus precluding a diagnosis of myocarditis using the Dallas criteria, and are seen with about equal frequency in other causes of sudden, unexpected infant death. Trivial microscopic inflammatory infiltrates also may be seen in the meninges but are not accompanied by brain swelling, encephalitis, or hemorrhage, and are not therefore a lethal finding.

Accumulation of small lipid droplets collectively in the liver, renal tubular epithelium, and smooth, skeletal, and cardiac muscle does not occur in SIDS and warrants evaluation for metabolic disorders such as MCAD deficiency. Death during the first week of life is rare in SIDS and, if microscopic lipid accumulation is found in these tissues, then MCAD deficiency should be considered.

To date, a pathognomonic marker for SIDS has not been found. It is unlikely that such a marker will be found given the likelihood that SIDS consists of more than one entity and has more than one cause.

### **Radiographic Studies**

The use of radiographs as an ancillary study in postmortem examinations is routine in most jurisdictions. In most cases where radiographs are used, however, the "babygram" is the standard. A skeletal survey should be the choice of examination, and properly informed and motivated technologists should be able to obtain high-quality postmortem skeletal

surveys in most medical examiners offices.<sup>72</sup> The widespread failure to obtain such studies is probably due more to inertia than to actual technical or economic factors. The clinical radiographic study commonly referred to as the "skeletal survey" has been outlined by Kleinman.<sup>72</sup> It consists of numerous projections and is clearly superior to the "babygram" done in most autopsy settings.

### **Criteria for Distinguishing SIDS From Fatal Child Abuse and Other Medical Conditions**

See Table 1-1 on page 11.

### **Child Death Review Teams**

If child abuse or neglect is a contributory factor in a substantial proportion of unexpected infant deaths, what should be done to minimize mistakes in the ascertainment of the cause and manner of death? The following are the 1999 recommendations of the American Academy of Pediatrics<sup>73</sup>:

1. Accurate history taking by emergency responders and medical personnel at the time of death and made available to the medical examiner or coroner
2. Prompt death scene investigation where the infant was found lifeless and careful interviews of household members by knowledgeable individuals (potentially including a pediatrician)
3. Postmortem examination following established protocol within 24 hours of death, including radiographic skeletal survey and toxicologic and metabolic screening
4. Collection of medical history through interviews of caretakers, interviews of key medical providers, and review of previous medical records
5. Maintenance of a supportive approach to parents during the death review process
6. Consideration of intentional asphyxia in cases of unexpected death with a history of recurrent cyanosis, apnea, or apparent life-threatening event (ALTE) witnessed only by a single caretaker or in a family with previous unexplained infant death(s)
7. Use of accepted diagnostic categories on death certificates as soon as possible after review

**TABLE 1-1. CRITERIA FOR DISTINGUISHING SIDS FROM FATAL CHILD ABUSE AND OTHER MEDICAL CONDITIONS**

	Consistent With SIDS	Less Consistent With SIDS	Highly Suggestive or Diagnostic of Child Abuse
History surrounding death	Apparently healthy infant fed, put to bed. Found lifeless. Silent death. EMS resuscitation unsuccessful.	Infant found apneic. EMS transports to hospital. Infant lives hours to days. Substance abuse, family illness.	History atypical for SIDS. Discrepant history. Unclear history. Prolonged interval between bedtime and death.
Age at death	Peak 2–4 mo. 90% <7 mo. Range 1–12 mo.	8–12 mo.	>12 mo.
PE and laboratory studies at time of death	Serosanguinous watery, frothy, or mucoid nasal discharge. PM lividity in dependent areas. Possible marks on pressure points of body. No skin trauma. Well-cared-for baby.	Organomegaly of viscera. Stigmata of disease process (PE, laboratory, x-ray).	Cutaneous injuries. Traumatic lesions of body parts (conjunctiva, fundi, scalp, intraoral, ears, neck, trunk, anogenital extremities, malnutrition, neglect. Fractures.
History of pregnancy, delivery, and infancy	Prenatal care—minimal to maximal. Frequent history of cigarette use during pregnancy. Some future SIDS victims are premature or LBW. Subtle defects in state, feeding, cry, neurological status (hypotonia, lethargy, irritability). Less postneonatal height and weight gain. Twins, triplets. Spitting, GE reflux. Thrush, pneumonia, illnesses requiring hospitalization, tachypnea, tachycardia, cyanosis. Usually: No signs of antecedent difficulty.	Prenatal care—minimal to maximal. History of recurrent illnesses and/or multiple hospitalizations. "Sickly" or "weak" baby. Specific diagnosis of organ system disease.	Unwanted pregnancy. Little or no prenatal care. Late arrival for delivery. Birth outside of hospital. Few or no well baby care. No immunizations. Use of cigarettes, drugs/alcohol during and after pregnancy. Baby described as hard to care for or to "discipline." Deviant feeding practices.
Death scene investigation	Crib, bed in good repair. No dangerous bedclothes, toys, plastic sheets, pacifier strings, pellet pillows. No cords, bands for possible entanglement. Accurate description of position with attention to possible head/neck entrapment. Normal room temperature. No toxins, insecticides. Good ventilation, furnace equipment.	Defective crib/bed. Use of inappropriate sheets, pillows, sleeping clothes. Presence of dangerous toys, plastic sheets, pacifier cords, pellet pillows. Cosleeping. Poor ventilation, heat control. Presence of toxins, insecticides. Unsanitary conditions.	Chaotic unsanitary crowded living conditions. Evidence of drugs/alcohol. Signs of terminal struggle in crib, bed, bedclothes or other equipment. Discovery of blood-stained bedclothes. Evidence of hostility by caretakers. Discord between caretakers. Display of violence between caretakers. Admission of harm. Accusations.
Previous infant deaths in family	First unexplained and unexpected infant death.	One previous unexpected or unexplained infant death.	More than one previous unexplained or unexpected infant death.
Autopsy findings	No adequate cause of death at PM. Normal: skeletal survey, toxicology, chemistry studies (blood sugar may be high, normal, or low), microscopic examination, metabolic screen. Presence of: large numbers of intrathoracic petechiae; dysmorphic, dysplastic, or anomalous lesions; gliosis of brainstem; sphincter dilation. Occasionally subtle changes in liver, including fatty change and extramedullary hematopoiesis.	Subtle changes in liver, adrenal, myocardium. Few or no intrathoracic petechiae.	Traumatic cause of death (IC or visceral bleeding). External bruises, abrasions, or burns. No intrathoracic petechiae. Malnutrition. Fractures. Subgaleal hematoma. Abnormal body chemistry values (Na, Cl, K, BUN, sugar; liver, pancreatic enzymes; CPK). Abnormal toxicology.
Previous CPS or LE involvement	None.	One.	Two or more. One or more family member arrested for violent behavior.

\* Abbreviations: SIDS, sudden infant death syndrome; EMS, emergency medical services; PM, postmortem; PE, physical examination; LBW, low birth weight; GE, gastroesophageal; WBC, well baby care; IC, intracranial; BUN, blood urea nitrogen; CPK, creatine phosphokinase; CPS, children's protective services; LE, law enforcement.

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8. Prompt informing sessions with parents when results indicate SIDS or medical causation of death

9. Locally based infant death review teams to review collected data with participation of the medical examiner or coroner in the review

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