Sudden Infant Death Syndrome

STEPHEN M. ADAMS, MD; MATTHEW W. GOOD, MD; and GINA M. DEFRANCO, DO
University of Tennessee College of Medicine Chattanooga, Chattanooga, Tennessee

Sudden infant death syndrome is the leading cause of death among healthy infants, affecting 0.57 per 1,000 live births. The most easily modifiable risk factor for sudden infant death syndrome is sleeping position. To reduce the risk of sudden infant death syndrome, parents should be advised to place infants on their backs to sleep and avoid exposing the infant to cigarette smoke. Other recommendations include use of a firm sleeping surface and avoidance of sleeping with soft objects, bed sharing, and overheating the infant. Pacifier use appears to decrease the risk of sudden infant death syndrome, but should be avoided until one month of age in infants who are breastfed. The occurrence of apparent life-threatening events does not increase the risk of sudden infant death syndrome, and home apnea monitoring does not lower the risk of sudden infant death syndrome. Supine sleeping position has increased the incidence of flattening of the occiput (deformational plagiocephaly), but this condition can be prevented and treated by encouraging supervised “tummy time,” meaning that when awake, infants should spend as much time as possible on their stomachs. All apparent deaths from sudden infant death syndrome should be carefully investigated to exclude other causes of death, including child abuse. Families who have an infant die from sudden infant death syndrome should be offered emotional support and grief counseling. (Am Fam Physician. 2009;79(10):870-874. Copyright © 2009 American Academy of Family Physicians.)

The term sudden infant death syndrome (SIDS) was first used in 1969 at an international conference on the causes of sudden death in infants.1 The current definition of SIDS, developed in 1991, is the sudden death of an infant younger than one year that remains unexplained after a thorough case investigation, including a complete autopsy, examination of the death scene, and review of the clinical history. In the United States, SIDS affects 0.57 per 1,000 live births, totaling more than 2,200 deaths per year. It is the leading cause of death among healthy infants.2

Risk Factors
Infant factors that have been associated with increased SIDS risk include low birth weight, low Apgar scores, recent viral illness, Native American or African American ancestry, and male sex.3 Maternal issues that predict SIDS risk include low socioeconomic status, smoking, illicit drug use, poor prenatal care, and young age.3 Other factors related to poor social and economic status, such as a crowded household, parental unemployment, and single parent status, also increase the risk of SIDS. Pacifier use may decrease the risk of SIDS,4 but sleeping position is the most easily modifiable risk factor. Infants should be placed on their backs to sleep; side sleeping is not recommended.5 A firm sleep surface, such as a firm crib mattress, should be used. Soft objects and loose bedding should be kept out of the crib.6 Parents should avoid overheating the infant during sleep (e.g., keep the room temperature comfortable, do not overdress the infant, use a light blanket).7 Factors that are associated with SIDS are listed in Table 1.4-6,8-11

The relationship between prone sleeping position and SIDS was first noted in 1965, but recommendations supporting supine sleeping were not issued until the early 1990s.12-14 The “Back to Sleep” campaign organized by the National Institutes of Health and the American Academy of Pediatrics (AAP) brought about a decrease in SIDS cases. In spite of recommendations, prone sleeping remains a common presumptive cause of SIDS. Cultural practices regarding sleep position mirror differences in SIDS rates between races. For example, in the United States, black infants are more likely to be placed to sleep in the prone position and have higher rates of SIDS compared with white infants, who are more likely to be placed to sleep in the supine position.15

Pathophysiology
Significant controversy revolves around the pathophysiology of SIDS. Three common...
autopsy findings include unclotted blood in the heart,\(^1\) intrathoracic petechiae,\(^16\) and fluid-filled, heavier organs,\(^17\) but these findings provide little help in understanding the final pathophysiology of SIDS. The now discredited “apnea theory” of SIDS was proposed in the 1970s\(^18\) and led to decades of research and the creation of an apnea monitoring industry. However, in the index case that first prompted investigation of the link between apnea and SIDS, the mother later confessed to killing all five of her children.\(^19\)

Current literature supports a triple-risk model,\(^20\) which suggests that SIDS is the final common pathway of three coinciding factors. This model proposes that an infant must first have an underlying vulnerability and then be stressed by an exogenous source, such as prone sleeping placement. Finally, for SIDS to occur, the stress must occur during a critical developmental period, namely in the first year of life. The last two factors in the triple-risk model have been well researched and defined in the medical literature, but the underlying vulnerability remains to be identified.

Current SIDS research topics include investigation of ion channel abnormalities, autonomic nervous system disturbances, and the effects of nicotine on the developing brain.\(^21\) Multiple ion channel disorders that cause QT interval prolongation have been linked to SIDS\(^22\) and may be a factor in 5 to 10 percent of SIDS cases.\(^23\) In addition, defects in normal arousal mechanisms have long been theorized to cause SIDS,\(^24\) and gene mutations affecting the development of the autonomic nervous system appear in as many as 15 percent of SIDS cases.\(^25\) Finally, pre- and postnatal exposure to cigarette smoke has been a known risk factor for SIDS for more than 30 years.\(^26\) Nicotine exposure has been clearly linked with SIDS, as well as with prematurity, autonomic dysfunction, low birth weight, and spontaneous abortions.\(^27\) These associations and newly identified nicotine metabolizing genes have prompted a search for SIDS causality\(^28\) and further highlight the importance of smoking cessation counseling in SIDS prevention efforts. Women should not smoke during pregnancy, and they should keep the infant’s environment smoke-free.\(^9\)

### Prevention and Counseling Issues

The principles of safe sleeping practices for infants are well established. In a recent case series, 92.2 percent of

<table>
<thead>
<tr>
<th>Table 1. Select Factors Associated with SIDS</th>
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<tr>
<td><strong>Increase SIDS risk</strong></td>
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<tr>
<td>Bed sharing, smoking mother(^4)</td>
</tr>
<tr>
<td>Birth before 37 weeks’ gestation(^7)</td>
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<tr>
<td>Soft bedding material(^6)</td>
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<td>Prone sleeping position (versus back)(^5)</td>
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<tr>
<td>Prone sleeping position (versus any other position)(^5)</td>
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<tr>
<td>Bed sharing, nonsmoking mother(^4)</td>
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<tr>
<td>Cigarette smoking during pregnancy(^9)</td>
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<td>Parental unemployment(^9)</td>
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<tr>
<td>Postnatal exposure to cigarette smoke(^10)</td>
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<tr>
<td>Side sleeping position (versus back)(^5)</td>
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<tr>
<td><strong>Reduce SIDS risk</strong></td>
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<td>Immunizations up to date(^11)</td>
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<tr>
<td>Pacifier use(^4)</td>
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**NOTE:** The above odds ratios were derived from different studies of different populations and, therefore, cannot be directly compared with each other.

\(CI = \) confidence interval; \(SIDS = \) sudden infant death syndrome.

Information from references 4 through 6, and 8 through 11.
deaths occurred in infants who were sleeping in the prone position, bed sharing, or sleeping in a location other than a crib or bassinet.\textsuperscript{29} Patient education efforts that recommended the supine sleeping position in the 1990s resulted in a 50 to 70 percent decrease in SIDS rates.\textsuperscript{3}

**BED SHARING**

Bed sharing remains a controversial topic among health care professionals and families. Possible benefits of bed sharing include increased breastfeeding rates, parent-child bonding, and fewer sleep problems.\textsuperscript{30} The AAP issued a policy statement in 2005 discouraging bed sharing, noting that almost one half of infants who die from SIDS in the United States do so while sleeping with parents.\textsuperscript{31} Bed sharing carries a substantially increased risk in low-birth-weight infants and in children of smokers and persons who use illicit drugs or alcohol.\textsuperscript{8} Recent analyses have suggested that the risks of bed sharing are isolated to infants younger than four months, with no significant increase in risk for older infants.\textsuperscript{30,32} Infants should sleep in a separate, but proximal, sleeping environment, ideally in a bassinet or crib near the mother's bed. Infants should never sleep with other children, or with a parent on a couch or armchair.\textsuperscript{8}

**DEFORMATIONAL PLAGIOCEPHALY**

Deformational plagiocephaly is flattening of the occiput, which affects up to one half of infants who sleep in the supine position, and has become more common since the initiation of recommendations to place infants on their backs to sleep.\textsuperscript{33} To decrease the risk of skull deformities, infants should have supervised “tummy time,” meaning that when awake, infants should spend as much time as possible on their stomachs. The side of the occiput that is placed downward during sleep also should be alternated. Infants should not be placed in car seats when they are not passengers in a vehicle, and the use of other devices that place pressure on the back of the head (e.g., swing, bouncy seat) should be minimized. Once deformational plagiocephaly has developed, parents should avoid placing the flattened side of the occiput down on the mattress during sleep times. Physical therapy is helpful if torticollis is present. A neurosurgical evaluation may be indicated in rare cases when the skull deformity does not improve with changes in head positioning.\textsuperscript{33}

**IMMUNIZATIONS**

Infants who do not receive immunizations may be at greater risk of SIDS.\textsuperscript{11} Some, but not all, case-control studies found a lower rate of SIDS among appropriately immunized children compared with children with incomplete immunizations. However, the apparent protective effect of immunizations against SIDS may in part be because of the relationship between incomplete immunization and lower socioeconomic status and other risk factors for SIDS.\textsuperscript{11}

**OTHER TOPICS**

A history of one or more apneic episodes does not increase the risk of SIDS, and the use of home apnea monitors does not lower the risk of SIDS.\textsuperscript{34} Pacifier use appears to decrease the risk of SIDS, and the 2005 AAP position statement on SIDS prevention suggests offering the infant a pacifier at nap or bedtime.\textsuperscript{7} However, the potential SIDS risk reduction from pacifier use must be weighed against the possibility that early pacifier use may shorten the duration of breastfeeding.\textsuperscript{35} Use of a pacifier should be delayed until one month of age in infants who are breastfed.\textsuperscript{4}

**Apparent Life-Threatening Events**

Apparent life-threatening events (ALTEs) are apneic episodes associated with changes in skin color (e.g., cyanosis, pallor, erythema), changes in muscle tone, and choking or gasping. Common diagnoses and evaluation of infants with ALTEs are listed in Table 2.\textsuperscript{36} ALTEs are relatively common, occurring in approximately one in 400 infants.\textsuperscript{37} In the past, ALTEs were referred to as “near-miss SIDS,” but this characterization does not appear to be accurate. The education campaigns that have reduced the incidence of SIDS have not affected the rate of ALTEs, and the only important SIDS risk factor that is associated with ALTEs is maternal smoking during pregnancy.\textsuperscript{37} Risk factors for ALTEs (in order of most to least important) include history of apnea, cyanosis, or pallor; feeding difficulties; single parenthood; family history of infant death; and smoking during pregnancy.\textsuperscript{36}

An etiology for ALTEs is determined in only one half of cases.\textsuperscript{37} Gastrointestinal etiologies (usually gastroesophageal reflux) account for most cases in which a cause is identified. Respiratory infections are another important cause. Cardiac disorders, such as valvular disease, arrhythmia, and cardiomyopathy, are responsible for a minority of cases. Neurologic causes, such as malignancy, structural abnormalities of the brain, and seizures, are uncommon, but should be considered. The history and physical examination are the most important factors in determining an etiology.\textsuperscript{39} In a case series of 243 patients admitted to a single academic medical center, fewer than 6 percent of all tests ordered were useful in making the diagnosis.\textsuperscript{39} The only tests found to be of value in detecting causes of ALTEs that were not evident
from the history and physical examination were testing for gastroesophageal reflux, urinalysis, neuroimaging, chest radiography, and white blood cell count. \(^{39}\)

### Differential Diagnosis of SIDS

Other causes of death, such as infection, electrolyte abnormalities, inborn errors of metabolism, and child abuse, must be eliminated before a death can be attributed to SIDS (Table 3\(^ {40}\)). Investigation of a potential SIDS death should include a thorough death scene evaluation and a complete autopsy, with a radiographic skeletal survey and toxicology studies. Evaluation of the death scene should include notation of the infant’s position, bedding, type of bed or crib, body temperature, presence or absence of rigor mortis, room temperature, type of heating or cooling used, and caregiver response.

Intentional suffocation is difficult to distinguish from SIDS, and the incidence of infanticide among deaths labeled as SIDS is estimated to be 1 to 5 percent. \(^{40}\) Small studies of children with recurrent ALTEs showed a high incidence of Munchausen syndrome by proxy. Factors that raise the possibility of intentional suffocation are a history of ALTEs occurring with a lone caretaker, death of infants older than six months, history of unexplained death of siblings, simultaneous death of twins, a previous death of a child under the care of the same person, or evidence of previous pulmonary hemorrhages at autopsy. \(^{40}\)

### Post-SIDS Support and Counseling

Families who have had an infant die from SIDS should be treated with compassion and empathy. They should be supported through the process of the death investigation and guided through problems, such as ending lactation and funeral planning. Grief counseling and referral to a SIDS support group should be offered. Parents should be counseled that the risk of future children dying from SIDS is not increased. \(^{40}\)

### The Authors

STEPHEN M. ADAMS, MD, is an associate professor and the program director of the Department of Family Medicine at the University of Tennessee College of Medicine Chattanooga. He received his medical degree from the University of Tennessee College of Medicine, Memphis, and completed a family medicine residency at the Huntsville Regional Medical Campus of the University of Alabama School of Medicine.

MATTHEW W. GOOD, MD, FAAP, is an assistant professor in the Department of Pediatrics and a pediatric hospitalist at the University of Tennessee College of Medicine Chattanooga. He received his medical degree from the Medical College of Virginia, Richmond, and completed a pediatric residency at Emory University, Atlanta, Ga.

GINA M. DEFRANCO, DO, is an assistant professor in the Department of Family Medicine at the University of Tennessee College of Medicine Chattanooga. She received her degree in osteopathic medicine from Kansas City (Mo.) University of Medicine and Biosciences, and completed a family medicine residency at the University of Tennessee College of Medicine Chattanooga.
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