APNEA SPELLS, SUDDEN DEATH, AND THE ROLE OF THE APNEA MONITOR

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"And this woman's son died in the night because she lay on it." 1 KINGS 3:19

For nearly 3000 years, it has been recognized that apparently healthy infants could die suddenly and unexpectedly, usually during their sleep. Throughout most of history, it was believed that these infants somehow suffocated, either by maternal overlaying or by strangling in bedclothes. Although this is not likely to explain the majority of these infant deaths, 2/1000 infants die suddenly and unexpectedly. Further, other infants may not die but can experience apneic episodes, or other spells, that appear to be life threatening. The causes of these infant deaths and spells remain unknown. The pediatrician, however, is increasingly being asked to evaluate infants for their risk of dying from the sudden infant death syndrome (SIDS) or because they have exhibited an apparent life-threatening event (ALTE). This article addresses the clinical diagnosis and management of infants with apnea spells.

SUDDEN INFANT DEATH SYNDROME

SIDS is defined as the sudden death of an infant, under 1 year of age, that remains unexplained after performance of a complete postmor-
tem investigation, including an autopsy, examination of the death scene, and review of the case history. SIDS is the most common cause of death between the ages of 1 week and 1 year, affecting 1 out of every 500 to 600 live births, accounting for 6000 to 10,000 deaths in the United States per year. The peak incidence is between 2 and 4 months of age, and approximately 95% of SIDS deaths occur before the age of 6 months. SIDS is not a new disorder, as mentioned above. The etiology remains unknown.

A typical clinical presentation for SIDS is that the parents or caregivers place the infant down to go to sleep, either at night or during a daytime nap. They return at some later time to find that the infant has died unexpectedly. Usually, these infants were healthy prior to death, although some had evidence of a mild upper respiratory infection. SIDS deaths have occurred when caregivers have been within hearing distance of the infant and have returned within 20 to 30 minutes to find that their infant had died. Yet, these parents report hearing no signs of a struggle. Thus, SIDS deaths appear to occur swiftly and silently. Although epidemiologic studies have been performed in an attempt to identify risk factors for SIDS, these studies do not point to a specific etiology.

Because SIDS has been referred to as a diagnosis of exclusion, some believe that there may be many causes of sudden death in infants that go unexplained, and thus SIDS may have many causes. There are some unique and characteristic features about SIDS deaths, however, that suggest that most babies dying from SIDS die from the same final common mechanism or final common pathway of death. These features include the unique age distribution, the unique seasonal distribution, and the characteristic findings on postmortem examination of intrathoracic petechiae and minor respiratory tract inflammation. Other natural causes of infant death do not share these unique age and seasonal distributions or have these characteristic autopsy findings.

Postmortem Examination

By definition, the etiology of SIDS is not known. In approximately 15% of infants who die suddenly and unexpectedly, a conventionally accepted cause for the death is found at postmortem examination. These infants are not said to have died from SIDS but rather from the cause of death found. This leaves 85% of infants who died suddenly and unexpectedly in whom no cause of death was found at postmortem examination. These infants comprise the group called SIDS.

By definition, an identifiable cause of death is not found at postmortem examination. Using research techniques, however, Naeyec has found tissue markers in many SIDS victims, suggesting that they may have been chronically hypoxic or hypoxemic prior to death. Some of these findings have been confirmed by other laboratories and some have not. Whether or not SIDS victims were hypoxic prior to death remains controversial and the object of continued study. Gliosis and subtle
changes have been found in brain stem centers responsible for control of breathing and control of sleep/wakefulness cycles.\textsuperscript{25, 26} These findings suggest that the origins of SIDS may lie in abnormal control of breathing or abnormalities in regulation of sleep/wakefulness.\textsuperscript{40}

A common finding in SIDS victims is the presence of intrathoracic petechiae on the thymus, lungs, pleural surfaces, and occasionally the heart.\textsuperscript{5, 27, 28} Extrathoracic portions of the thymus do not show petechiae. These intrathoracic petechiae may have been formed by the generation of highly negative intrathoracic pressures, as might occur when an infant attempts to breathe against upper airway obstruction.\textsuperscript{27} Intrathoracic petechiae suggest to some investigators that obstructive apnea may be involved in the final mechanism of SIDS deaths.\textsuperscript{17}

The Apnea Hypothesis of SIDS

Currently, most of SIDS research is focused on the respiratory system as a possible cause of SIDS. The infant respiratory system is developmentally immature and rapidly changing. Normal infants, without increased risk for SIDS, can have apneas up to 15-seconds duration during sleep. Infants with unexplained apnea have an increased risk of dying from SIDS.\textsuperscript{7, 24, 36, 45} Tissue markers of chronic hypoxia and hypoxemia have been described in many SIDS victims.\textsuperscript{33, 34} Brain stem lesions have been seen in areas controlling respiration and sleep/wakefulness in many SIDS victims.\textsuperscript{25, 26, 35} Many infants at high risk for SIDS have respiratory control disorders.\textsuperscript{34, 23, 24, 45, 46, 50} Although these findings do not prove that SIDS is due to a respiratory disorder, there is considerable circumstantial evidence suggesting that SIDS may involve central or obstructive apnea during sleep.\textsuperscript{40}

This evidence in favor of respiratory causes for SIDS prompted investigators to formulate the apnea hypothesis of SIDS, which says that SIDS occurs because infants stop breathing during sleep.\textsuperscript{46} It was believed that one might be able to test infants to see if they have apneas. Those with increased apnea during sleep would be at increased risk for SIDS and could be treated.\textsuperscript{24, 48} Home apnea-bradycardia monitoring, which sounds an alarm to summon trained caregivers when an infant has a prolonged apnea or bradycardia, has been advocated as a method to manage these infants.\textsuperscript{24}

The National Institute of Child Health and Human Development collaborative epidemiologic study of SIDS, however, showed no difference in the incidence of neonatal apnea in SIDS victims versus non-SIDS controls.\textsuperscript{40} The study did find an increased incidence of ALTE occurring postterm in SIDS victims (7%) than in non-SIDS controls (3%).\textsuperscript{46} Ninety-three percent of SIDS victims, however, had no apnea observed prior to death. Thus, apnea identified prior to death seems to play a relatively small role in SIDS. Sleep studies performed to detect apneas in infants thought to be at risk for SIDS do not predict SIDS or death.\textsuperscript{7, 44, 53} Home apnea-bradycardia monitoring and infant apnea evaluations have not
substantially decreased the SIDS rates for the general population. SIDS may still be due to a respiratory disorder, but the simplistic approach to treating apnea in infants has not resulted in substantial decreases in SIDS.

**SIDS and Grief**

SIDS deaths have some unique characteristics compared with other infant deaths. SIDS deaths are unexpected, so parents have not had the opportunity to prepare for the death. Because SIDS deaths are unexplained, SIDS parents blame themselves for the death. For most families of SIDS victims, the best resource is a SIDS Parent Support Group. The National Sudden Infant Death Syndrome Alliance is a national SIDS organization, which has parent support counselors who are available to speak with SIDS parents at any time. In contrast, the role of the health professional is to educate the family about SIDS and to reassure them that they did not cause the death of their baby, either by something they did or by something they did not do. It is important to emphasize that SIDS cannot be prevented, and there is nothing they or anyone else could have done to prevent their child's death.

**APPARENT LIFE-THREATENING EVENTS**

The respiratory system is immature and rapidly developing during infancy. Therefore, apnea can result from a variety of systemic disorders that may stress this unstable system. ALTE represent a severe form of apnea, and infants with a history of an ALTE often present to the pediatrician for evaluation and management. Previously healthy infants may present with an ALTE, and the incidence of ALTE in the general population may be as high as 3%. ALTE describes a clinical syndrome that may have many causes, some of which can be identified and some of which cannot. When no treatable cause for the ALTE is found, there is concern that these infants may be at increased risk of subsequently dying from SIDS, compared with the general population. Further, these infants, as a group, have a high incidence of subsequent apneas and require home apnea-bradycardia monitoring. Thus, a diagnostic evaluation and management are required.

**Diagnosis**

The diagnosis of an ALTE is made if an infant has a convincing history of an episode of sudden onset characterized by color change (cyanosis or pallor), tone change (limpness, rarely stiffness), and apnea, which requires significant intervention (vigorous shaking, mouth to mouth breathing, or full cardiopulmonary resuscitation) to revive the
infant and restore normal breathing. ALTE are often frightening to the observer, who may believe that the infant is in the process of dying. ALTE are severe episodes. Mild episodes, which require little or no intervention, probably do not have the same prognostic significance and may not require an aggressive diagnostic and therapeutic approach. The diagnosis is made on the basis of the history of the event, as there are currently no diagnostic tests that accurately confirm the presence of ALTE.

The physician usually has not witnessed the ALTE. Infants often appear entirely normal by the time they reach medical attention following the ALTE. The most important initial diagnostic step is to obtain a careful history from the person witnessing the event. One should specifically ask about the infant's color, tone, apnea, and the need for intervention. Was the infant awake or asleep? How long did the event last? Normal infants without increased risk for SIDS or apnea can have respiratory pauses during sleep lasting up to 15 seconds. Thus, isolated events of this duration or shorter may not be clinically significant. One must make an initial judgment about whether the event represents an ALTE or not. If not, usually no further diagnostic evaluation is required. If the physician believes that the infant did have an ALTE, then an aggressive approach to identifying the etiology of the event and instituting appropriate therapy is necessary.

The respiratory system of infants is immature, and many systemic conditions include apnea as a presenting sign, including seizures, anemia, sepsis, metabolic disorders, pneumonia, lung disease, upper airway obstruction, pertussis, and heart disease. Optimal care of the infant presenting with an ALTE requires a thorough diagnostic evaluation to detect treatable causes of the event. Thus, we recommend hospital admission for protective monitoring, to facilitate the diagnostic evaluation, and for parent training. This diagnostic evaluation should include, but is not limited to, complete blood count, serum electrolytes, blood sugar, calcium, thyroid studies, and serum ammonia; chest radiograph; electrocardiogram; arterial blood gases; electroencephalogram (EEG); and a pneumogram or polysomnogram. Some infants may require a septic workup, laryngoscopy and bronchoscopy, Holter monitoring, or an evaluation for gastroesophageal reflux; in addition, if indicated by history, a physical examination or initial testing may be required. Although not every infant will require all of these tests, most of them are performed before the episodes are termed "unexplained."

Sleep Studies

The pneumogram is a two-channel 12-hour overnight recording of chest wall movement (chest wall impedance respirations) and electrocardiogram. The pneumogram is a useful test to document the ventilatory pattern of infants during sleep as part of the initial diagnostic evaluation. These recordings can be performed in hospital or in the home and are equally accurate and sensitive in either location.

The criteria for normal pneumograms, determined from home re-
cordings on 97 normal infants (aged 5.6 ± 12.0 months, range 0.8 to 10.8, 55% females) in Southern California, are listed in Table 1. It is important to note that the pneumogram is not a diagnostic test for apnea and does not predict the risk of death or subsequent apneas. Some investigators report prolonged apneas, increased frequency of short apneas, and increased periodic breathing in infants with unexplained apnea, whereas other investigators find no such abnormalities.

Even when infants with unexplained apnea have abnormal pneumograms, it bears little correlation with the number or severity of subsequent apneas. Recording the pneumogram following a diptheria-tetanus-pertussis (DTP) immunization does not improve the sensitivity of the pneumogram. Thus, the pneumogram is not a diagnostic test for infant apnea, and infants with normal pneumograms can have multiple severe subsequent apneas or death. It can, however, be a useful and relatively inexpensive test documenting the breathing and heart rate pattern during sleep. Four-channel pneumograms offer the addition of arterial oxygen saturation (pulse oximetry) and esophageal pH or nasal airflow.

Polysomnography can be performed overnight or during a nap. Polysomnography records multiple channels of physiologic information. It is important to include the measurement of respiratory parameters, such as arterial oxygen saturation (pulse oximetry); end-tidal CO2 tension; chest wall and abdominal movement (inductance plethysmography); and EEG, electrooculogram, and electromyogram channels for sleep staging. Polysomnography is particularly useful in the evaluation of infants with atypical presentation, a prolonged clinical course, or severe events. These may be due to hypoxemia or hypoventilation that can be diagnosed using polysomnography. Like the pneumogram, polysomnography is not a diagnostic test for ALTE or unexplained apnea nor does it predict the risk of recurrent apneic episodes. There are no diag-

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<th>Table 1. NORMAL PEDIATRIC PNEUMOGRAM VALUES FOR TERM INFANTS</th>
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<td>Mean ± SEM</td>
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<tr>
<td>Quiet (sleep) time (min)*</td>
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<td>Longest apnea (s)</td>
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<td>Sustained bradycardia†</td>
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<td>Periodic breathing‡ (%) sleep time</td>
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<td>Apnea density (apnea ≥6 s)</td>
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*At least 300 minutes of quiet time (sleep) is the accepted minimum standard as a sufficient sample to adequately evaluate the ventilatory pattern during sleep.
†Sustained bradycardia is defined as bradycardia lasting at least 5 seconds, which are less than 80 in the first month of life, less than 70 between 1 and 3 months of life, and less than 60 between 3 and 12 months of life, and less than 50 thereafter.
‡Periodic breathing is defined as three or more respiratory pauses, each at least 3 seconds in duration, no two separated by more than 20 seconds of normal breathing. Periodic breathing is quantitated as the total time from the beginning of the first pause to the end of the last pause in a run of periodic breathing.
nostic tests that are consistently abnormal in, or diagnostic of, ALTE or unexplained apnea.14 16 18 22 44 46

Management

In our experience, treatable etiologies are found in approximately 30% of infants presenting with ALTE to our referral center.25 The proportion of infants with treatable causes of ALTE presenting to primary care physicians may be higher. Episodes that have occurred during wakefulness are more likely to be secondary to a treatable etiology, such as a seizure disorder or gastroesophageal reflux. Infants in whom a treatable cause of ALTE can be identified are best managed by treating the specific etiology of the event. Occasionally, some may require the addition of home apnea-bradycardia monitoring if ALTE cannot be controlled despite specific treatment.

The diagnosis of apnea of infancy (AOI) is made when an identifiable cause for the ALTE cannot be found. There are presently no specific treatments for AOI; thus, home apnea-bradycardia monitoring is recommended for these infants.21 24 28 Although scientific studies have not been performed to prove the efficacy or lack of efficacy of home apnea-bradycardia monitoring in saving the lives of these infants, they have a high risk for subsequent apneas, and home monitoring is used to detect these episodes.21 24 39

Parents or caregivers are instructed to use home monitors whenever the infant is not being otherwise observed. They are most commonly used during sleep and in automobiles. Alarms are set to sound for central apneas greater than 20 seconds and/or bradycardias less than 80 beats/min in the first month of life, less than 70 beats/min from 1 to 3 months, less than 60 beats/min from 3 to 12 months, and less than 50 beats/min thereafter. Home monitors should sound an alarm for both prolonged apnea and bradycardia. These monitors do not sound an alarm for obstructive apneas, unless an accompanying bradycardia sounds an alarm. We have not found tachycardia alarms to be useful in identifying clinically significant events. We require that home apnea-bradycardia monitors be powered by a rechargeable battery system for use in automobiles and while outside the home.

Home monitors only alert the caregiver that a potential episode is occurring. The caregiver must then respond to evaluate and/or terminate the episode. Parents and caregivers must be trained in the proper operation of the monitor, a graded response to monitor alarms, and infant cardiopulmonary resuscitation. Thorough education of the parents and psychosocial support of monitoring families are important for successful home apnea-bradycardia monitoring.4

Documented Monitoring (Event Recordings)

It may be difficult for parents or caregivers to distinguish true apnea or bradycardia alarms from loose lead alarms or alarms for nonsignifi-
Documented monitoring, with event recorders built into the monitors, provide objective recordings of apnea and bradycardia alarms and may be helpful in making these distinctions. Only 8% of 14,131 alarms, validated by event recordings, were caused by true apnea or bradycardia. Of these true events, 70% were triggered by apnea and 30% by bradycardia. These true events occurred in 48% of the patients. Loose lead alarms accounted for 69% of all events and false alarms for 23%. The most common reason for a false alarm was a low amplitude signal. Therefore, in many instances, alarms viewed as real by the caregivers can be shown to be false, and the time of monitoring may be decreased. Documented monitoring, however, may also confirm the caregiver's observations regarding the clinical significance of episodes. In addition, it provides information regarding compliance with monitor use, because the length of time the monitor was turned on each day is recorded. Compliance with monitoring may be enhanced with documented monitoring because the physician has access to data on monitor use.

**Outpatient Management**

After discharge, ARI infants should be followed on an outpatient basis every 1 to 2 months. Parents are requested to keep a log or diary of all apnea and bradycardia alarms that were thought to be real, especially those requiring intervention. During outpatient visits, the clinician reviews these logs with the family and compares them with event recordings, if available. The usual clinical pattern is that true alarms will decrease in both frequency and severity with time. Infants whose alarms become more frequent or severe, those infants with multiple alarms requiring intervention, and those infants who continue to have true alarms after 6 to 8 months of monitoring require further diagnostic evaluation, including overnight polysomnography. With severe episodes, these infants may require hospitalization for observation and further diagnostic evaluation. Sometimes the character of the events may change, suggesting the presence or development of other clinical problems, such as a seizure disorder, which also require specific evaluation.

Outpatient visits are used to reinforce parent education and compliance. Parent response to monitor alarms, including infant cardiopulmonary resuscitation, should be reviewed at each outpatient visit. It is important to emphasize compulsive use of the home monitor at all times when the infant is not being otherwise observed. Because parents often find it difficult to stop home monitoring once their infant no longer requires it, they are taught the criteria for monitor discontinuation. Therefore, parents can begin psychologic preparation for the time when home monitoring can be safely discontinued.

**Discontinuing Home Apnea-Bradycardia Monitoring**

Home apnea-bradycardia monitoring can be discontinued after 3 months of no apnea or bradycardia alarms that require intervention.
Tolerating a physical stress (upper respiratory infection or other intercurrent illness) without an apnea or bradycardia is reassuring information but not required to discontinue home monitoring. Most infants with ALTE require 4 to 6 months of home monitoring, indicating that they had subsequent apneas for 1 to 3 months after the initial ALTE. Some infants may continue to have lengthened breathing pauses, documented by polysomnogram or event recording, after these criteria for discontinuation have been met. If these episodes no longer require intervention, however, and are therefore not life threatening, they do not necessarily require the continuation of home monitoring.

Outcome

It has been suggested 40% to 60% of infants with unexplained apnea have subsequent apneas detected by the home monitor.1, 27, 24, 36, 39, 51 AOI infants are more likely to have apneas when they are physically stressed, with an upper respiratory infection or with fatigue. In most AOI infants, apnea resolves before 1 year of age, but a few still have apneas after their first birthday.

Even with the above diagnostic evaluation and home monitoring, AOI infants have twice the risk of dying from SIDS than the general population.7 Sleep studies are not predictive of subsequent apneas, SIDS, or death, and a normal sleep study does not eliminate the possibility that a child will die from SIDS.7, 41, 53 AOI infants have died when home monitor function and response to the alarms appeared to be appropriate.7, 36 Infants with AOI who have received full cardiopulmonary resuscitation on more than one occasion are at high risk of dying.7, 36 and some of these infants have died even with appropriate home monitoring. Some infants continue to die in temporal association with non-compliance or with errors in home monitoring technique.7 This reemphasizes the importance of parental teaching and reinforcement of monitoring skills.

Other Conditions That Can Cause ALTE

Inborn Errors of Beta-Oxidation of Fatty Acids

ALTE and SIDS-like deaths are rarely associated with inborn errors of fat oxidation.15, 43 An inborn error of metabolism is more likely to be associated with an ALTE if there is a family history of ALTE, consanguinity, seizure disorders, or SIDS. Inborn errors of fat oxidation may only be apparent during times of metabolic stress, such as fasting, when the infant is forced to utilize fatty acids as substrates for energy production rather than carbohydrates. Medium chain acyl-CoA dehydrogenase deficiency (MCADD) is the most common of these disorders and the most likely to present as an ALTE.15

Each infant with an ALTE should have blood NH₄ measured. A
more thorough diagnostic evaluation is recommended for ALTE infants with (1) a positive family history for AOL, seizure disorders, or SIDS; (2) laboratory evidence of hypoglycemia, hyperammonemia, metabolic acidosis, elevated liver enzymes, or abnormal hemostasis; and/or (3) a positive patient history of unexplained failure to thrive, developmental delay, or seizures. These include blood for the DNA diagnostic test for MCADD, blood and urine carnitine, urine nonvolatile organic acids, and urine acylglycine and acylcarnitine. Treatment of MCADD involves the avoidance of fasting, L-carnitine supplementation, a low-fat/high-carbohydrate diet, and home apnea-bradycardia monitoring for infants with ALTE.35

Munchausen Syndrome by Proxy

SIDS and AOL represent true organic disease; they are not a form of child abuse. Child abuse exists, however, and may masquerade as SIDS and/or AOL. Munchausen syndrome by proxy is a disorder of parenting and a form of child abuse, where the parent creates a factitious illness in the child.32,38 Most parents afflicted with this syndrome do not harm their children, but rather the factitious illness is created by providing a fabricated history. Munchausen syndrome by proxy, however, may involve inflicted physical injury to the child with suffocation used to induce "apneic spells" or even death, which can mimic SIDS.

Confirmation of the diagnosis of Munchausen syndrome by proxy is difficult. Physicians are trained to believe the parents' history and to treat illness, which makes recognizing factitious illness an uncomfortable task. Techniques that have been used include documenting induced "apneic spells" by hidden video camera or by separating the infant from the mother and documenting the resolution of the symptoms.32,38 Treatment of Munchausen syndrome by proxy involves a team approach utilizing psychiatry and social and protective services.32

APNEA OF PREMATURETY

As a group, preterm infants are at statistically increased risk for SIDS.7 At present, however, there is no way to accurately identify which preterm infants will die from SIDS.7-41 Apnea of prematurity is defined as a respiratory pause 20 seconds in duration or longer or any respiratory pause associated with bradycardia or cyanosis in an infant less than 37 weeks postconception. A large epidemiologic study of SIDS indicated that apnea of prematurity was not, in and of itself, a precursor or predictor of subsequent SIDS death.40 Therefore, the optimal management of preterm infants to prevent SIDS is unclear. Apnea of prematurity is a natural consequence of immaturity. It is expected that infants will outgrow apnea of prematurity as they mature.
Management

Preterm infants who continue to exhibit symptomatic apnea when they would otherwise be ready for hospital discharge should have their oxygenation carefully evaluated, because hypoxia can cause apnea in preterm infants, and relieving it may resolve the problem.\textsuperscript{11} Bronchopulmonary dysplasia is frequently associated with apnea in preterm infants. In our experience, treating the chronic lung disease optimally may be effective treatment for apnea in preterm infants with lung disease who are past term. In the absence of hypoxia or chronic lung disease, preterm infants who are still having clinically apparent episodes of apnea can be discharged on home apnea-bradycardia monitoring. If theophylline or caffeine reduces the frequency of these episodes, then these infants can be treated in addition to the home apnea-bradycardia monitor. If theophylline or caffeine has no clinical effect, however, these infants should be discharged on home apnea-bradycardia monitoring alone. Theophylline, if used, may be stopped after 40 weeks postconceptional age. If there is no recurrence of apnea, home monitoring can be discontinued 1 month later. If theophylline is not used, home monitoring is continued until 1 month postterm and discontinued if the child has had no real apnea or bradycardia alarms. If the child has had real apnea alarms, then the infant is managed in the same way as AOI.

INFANTS OF SUBSTANCE-ABUSING MOTHERS

Infants of substance-abusing mothers (ISAM) have an increased risk of SIDS\textsuperscript{6,10,37} and abnormalities of the ventilatory pattern during sleep.\textsuperscript{8} In a large, population-based, retrospective study of SIDS risk in ISAM, nearly one fourth of the ISAM who died from SIDS had a history of symptomatic apnea prior to their death.\textsuperscript{6}

Many of the health practices and social, economic, and demographic characteristics of substance-abusing mothers are independent risk factors for SIDS.\textsuperscript{2} Thus, the SIDS risk in ISAM may be largely or partly explained by other independent risk factors. Programs providing adequate prenatal care to pregnant opiate addicts result in reduced obstetric complications.\textsuperscript{49} Therefore, it is possible that the SIDS risk in ISAM could be decreased by pre- and postnatal programs that improve the general health and socioeconomic status of pregnant substance-abusing women and their infants, even in the face of continuing drug abuse.

ISAM may have a number of medical problems that are best managed by comprehensive pediatric care. Home-apnea bradycardia monitoring is only indicated for ISAM who are symptomatic with apnea. There is no evidence to suggest, however, that home apnea-bradycardia monitoring is useful or indicated in asymptomatic ISAM.\textsuperscript{35} Diagnostic tests of respiratory control for asymptomatic ISAM, such as pneumograms or sleep studies, have not been found to be predictive of subsequent clinical outcome and are not recommended. In 27 ISAM evaluated
by pneumograms, a significant percentage of infants had abnormal pneumogram recordings, yet none of the infants with abnormal recordings developed symptomatic apnea or died of SIDS.  

**BREATHE-HELDING SPELLS**

A classic breath-holding spell is triggered by anger or frustration leading to crying with subsequent breath-holding after a prolonged exhalation. Pallor or cyanosis results. The spells may terminate with loss of consciousness and, in particularly severe spells, tonic-clonic movements. The loss of consciousness and tonic-clonic movements are related to hypoxia and do not represent a primary seizure disorder. Breath-holding spells are commonly accompanied by extreme parental anxiety and are occasionally confused with seizure disorders. Breath-holding spells may affect up to 5% of all children. The etiology is unknown. The age of onset of breath-holding spells is usually between 6 and 15 months, although occasionally they may begin in the neonatal period. They tend to peak in severity during the third year of life and subside during the fourth and fifth years. There is a familial tendency.

There are no specific physical findings associated with breath-holding spells. No diagnostic evaluation is required with a classic history of breath-holding spells. Many patients will have an EEG, however, to eliminate the possibility of a seizure disorder. The resting EEG in breath-holding spells is normal.  

Increased vagal tone has been proposed in the pathogenesis of breath-holding spells. About 40% of children with breath-holding spells will have prolonged asystole (>2 seconds) with ocular compression, which supports this concept. The cyanosis of breath-holding spells occurs much sooner than with voluntary breath-holding. This is due, in part, to the breath being held in expiration, to a decreased venous return, and decreased cardiac output (as seen in a Valsalva maneuver). Recordings of physiologic parameters during breath-holding spells have documented asystole, electrical silence of the EEG, and active muscular contraction of the abdominal muscles of respiration with the breath held in expiration.

Polysomnographic recordings to evaluate sleep architecture, ventilatory patterns, and gas exchange during sleep have been performed in children with breath-holding spells. These failed to demonstrate sleep-disordered breathing in children with breath-holding spells. The outcome of children with breath-holding spells is generally excellent, although some may go on to have syncopal episodes in later life. No specific treatment other than parental reassurance is warranted. Atropine has been utilized in some severe cases but with limited success. Home apnea-bradycardia monitors are not helpful for breath-holding spells because the episodes are obvious and they occur during wakefulness when infants would be unlikely to be using their home monitors.
SUMMARY

SIDS is the most common cause of death between the ages of 1 week and 1 year. It affects 1 out of every 500 to 600 live births. The etiology of SIDS is unknown. There are no tests currently available to predict the infant who will die from SIDS prior to death. SIDS cannot be prevented. Many infants experience serious apneic spells, however, that require diagnostic evaluation and treatment. Even if the treatment of these infants does not have a large impact on the SIDS rate for the general population, thorough diagnostic evaluations and appropriate use of home apnea-bradycardia monitoring is indicated for this population and may reduce their risk of morbidity or mortality.

References


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