

Sudden Infant Death Syndrome (SIDS)

2.0 Contact Hours

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Sudden Infant Death Syndrome (SIDS)

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Upon completion of this course, the learner will be able to:

1. Define sudden infant death syndrome (SIDS)
2. Explain the pathology of SIDS
3. Explain the pathophysiology of SIDS
4. Discuss the biologic risk factors of SIDS
5. Discuss the environmental risk factors of SIDS
6. Discuss the tests that can be used to identify infants susceptible for SIDS
7. Discuss possible interventions to prevent SIDS

Definition

Sudden Infant Death Syndrome is the most common cause of infant mortality in the United States after congenital anomalies and disorders related to short gestation or low birth weight. It is commonly called “Cot Death” or “Crib Death”. It is defined as the sudden death of an infant that is unexpected by history and unexplained by thorough postmortem examination that includes a complete autopsy, investigation of the scene of death and review of medical history.

An autopsy is essential before the death of an infant can be declared as due to sudden infant death syndrome. This is because the deaths due to cardiac anomalies, brain anomalies and child abuse cannot be ruled out merely by investigating the scene of death and reviewing the medical history.

Sudden Infant Death Syndrome = Unexplained infant death

- ➡ Complete autopsy
- ➡ Investigation of the scene of death
- ➡ Review of medical history

Thus SIDS can be stated as an unexplained death with the conclusion based on the above three modes.

Cases that fail to meet the standards of this definition, including those without a postmortem investigation, should not be classified as SIDS. Cases that are autopsied and carefully investigated but remain unresolved may be designated as undetermined or unexplained.

In most series, SIDS accounts for 35-55% of deaths within the post-neonatal period. SIDS comprises approximately 20-25% of deaths in those younger than 1 year. Despite intensive study and advances in the understanding of associated factors, the specific cause or causes of SIDS remain unknown. SIDS predominates as the single leading cause of unexpected deaths in infancy; however, alternative diagnoses are identified in as many as 15-25% of sudden unexpected deaths in infancy (SUDIs).

Prevalence

The SIDS rate is higher among African American infants and Native American infants compared to white, Hispanic, and Asian American infants. From 1995-2001, SIDS rates among Native American and Alaskan Indian infants were 2.4-2.9 times that seen for white infants. From 1995-2001, SIDS rates in Hispanic infants in the United States were approximately one-third lower than the rates seen in white infants. These racial differences remain unexplained but appear to be independent of other risk factors, such as low birth weight, young maternal age, or high parity.

The prevalence of SIDS has come down dramatically ever since extensive health education began being provided regarding proper sleeping position of babies. SIDS is more common in babies made to sleep in the prone position. By making the infants sleep in the supine position, the incidence of SIDS can be brought down considerably. The NICHD, American Academy of Pediatrics (AAP), Association of SIDS and Infant Mortality Programs (ASIP), SIDS Alliance, and the United States Public Health Service jointly sponsored the 'Back to sleep' campaign in 1994 which has reduced SIDS

considerably. Since 1992, SIDS rates have fallen approximately 58%. In 2002, the National Center for Health Statistics reported a total of 2295 SIDS deaths nationwide, for a SIDS rate in the United States of 0.51 per 1000 live births. Studies conducted by NICHD demonstrated that the rate of prone sleeping for infants decreased from approximately 75% in 1992 to a low of 11.3% in 2002. The observation that the rate of prone sleeping increased to 13% in 2004 is of some concern.

In other countries, the prone sleeping rates have dropped down to 2-5%. 70% of the decline in SIDS in other countries has been attributed to this change in sleeping position. Other factors which have been known to influence SIDS are –

- Avoiding bed sharing, and soft, loose bedding decrease the risk of SIDS. The risk appears to be greatest among infants younger than 3-4 months
- Pacifier use may reduce the risk of SIDS. Pacifier use during sleep modifies cardiac autonomic balance in young infants

The male-to-female ratio for SIDS is 3:2. Approximately two thirds of SIDS deaths occur in infants aged 2-6 months. Ninety percent of deaths occur in children younger than 6 months, and 95% of deaths occur in children younger than 8 months.

Pathology

An autopsy is essential before declaring an infant's death is due to SIDS. The autopsy findings are generally supportive rather than conclusive. In most of the cases, lesions are seen in the pulmonary tissue and the central nervous system. Though these lesions are found during autopsy, if they are not sufficient enough to explain the sudden and unexpected death, only then should the death be declared as due to SIDS.

Pulmonary findings –

- Signs of chronic asphyxia are seen in nearly two thirds of infants who die of SIDS.
- Mild pulmonary edema and petechiae may be seen

Central nervous system findings –

- Brain stem abnormalities especially in the areas related to respiratory neuro-regulation
- Anatomical and functional abnormalities of the arcuate nucleus which is related to cardio-respiratory control.

Other abnormalities-

- Growth retardation
- Elevated blood cortisol levels
- Elevated hypoxanthine in vitreous humor – which indicates there is a long period of hypoxia before death

Pathophysiology

As mentioned earlier, no specific etiology has been identified for SIDS. But the autopsy findings in infants dying of SIDS point towards a brain stem abnormality as a causative factor for SIDS. The brain stem functions, like cardio-respiratory control, including arousal responsiveness, and other autonomic controls like blood pressure and sleep regulation, are said to be abnormal in infants dying of SIDS. The clinical abnormalities in high risk infants who later died of SIDS confirmed this hypothesis.

They showed features like abnormalities in the following functions –

- Respiratory pattern
- Chemoreceptor sensitivity
- Heart rate
- Asphyxic arousal responsiveness

There are two categories of risk factors thought to be responsible for SIDS in susceptible infants.

- Biologic risk factors
- Environmental risk factors

Interaction of these two groups of risk factors is very complex and is thought to be very important in causing SIDS.

Biologic risk factors

All these risk factors are found to be associated with SIDS in high risk infants. The following are the various biologic factors thought to be interacting with the environmental risk factors causing SIDS –

- Family history of SIDS
- Idiopathic apparent life threatening events (IALTE)
- Prematurity
- Deficient brain stem function
 - Arousal / gasping
 - Ventilatory responsiveness
 - Respiratory pattern
 - Cardiac control
 - Temperature regulation
 - Other autonomic deficits – eg. Vagal tone, blood pressure
- Hypothetical
 - Metabolic
 - Infectious / inflammatory
 - Immune

Abnormal respiratory pattern –

As mentioned above, abnormalities of the respiration is one of the biologic factors responsible for SIDS. The various abnormalities of respiration that can occur include –

- Prolonged apnea
- Excess brief apneas
- Periodic breathing
- Restricted breath to breath variability at lower respiratory rates

Abnormalities of chemoreceptor sensitivity –

Many infants who later die of SIDS showed poor ventilatory responsiveness to hypercarbia or hypoxia

Defective arousal responsiveness –

Abnormal responsiveness predisposes an infant to SIDS, especially those who are preterm, and those showing idiopathic apparent life threatening events. But it may be insufficient to cause SIDS in the absence of other biologic or environmental factors. Infants in whom the arousal responsiveness is absent are incapable of responding sleep related asphyxia. These infants also show deficit in gasping. Gasping is a form of auto-resuscitation which acts as a complement to the asphyxic arousal response deficit. Absence of gasping is considered the final step in the pathogenesis of SIDS.

Poor temperature regulation –

Hyperthermia can result in SIDS. An increased body temperature or environmental temperature is hazardous to the infant. The body temperature is closely related to cardio-respiratory control. This interaction is a very complex phenomenon. Overheating will affect the cardio-respiratory control which increases the risk of SIDS.

Abnormal cardiac control –

In normal infants, the QT interval shortens as the heart rate increases. This is a physiological response. If the QT interval does not shorten, it predisposes to ventricular arrhythmias. In infants who die of SIDS, this physiological response does not happen and they subsequently develop ventricular arrhythmias which results in SIDS. The abnormalities of cardiac control are thought to be due to decreased vagal tone. The decreased vagal tone may be due to vagal neuropathy or due to the damage of the parasympathetic control center in the brain stem.

The various abnormalities of cardiac control which makes the infant prone for SIDS are –

- Absence of shortening of QT interval in response to an increasing heart rate
- Higher heart rates during sleep/waking states
- Diminished heart rate variability during wakefulness

These abnormalities of the cardiac control can be recorded with home cardio respiratory monitors. These may show abnormalities in the rhythm or sudden rapid progression to bradycardia. Home cardio respiratory monitoring is one of the options available for monitoring high risk infants who are prone to SIDS. But this is not completely sensitive and various other factors like poor compliance by the parents make this option not a worthy one.

Prematurity –

Prematurity and low birth weight are significant risk factors for SIDS. There is an inverse relationship between risk for SIDS and birth weight and gestational age. Approximately 12-20% of infants with SIDS are born prematurely (<37 weeks' gestation) or at low birth weight (<2500 g). The post natal age of preterm infants dying of SIDS is about 5-7 weeks older and the post conceptional age is 4-6 weeks younger than for full term infants.

Idiopathic Apparent Life Threatening Events (IALTE) –

An apparent life-threatening event (ALTE) is defined as an episode that is frightening to the observer and is characterized by some combination of apnea (central or obstructive), change in muscle tone (usually diminished), and choking or gagging. In some cases, the observer fears that the infant has died. The estimated frequency of ALTE among healthy term infants is 1-3%. More than 50% of ALTE is idiopathic (IALTE). Infants with IALTE are at an increased risk for SIDS. The risk of SIDS appears to be increased in infants with two or more IALTE.

Metabolic disorder –

The recurrent risk for SIDS and for infant mortality from other causes is increased in subsequent siblings. A familial metabolic disorder should be considered in families with more than one unexplained infant death especially when the history is atypical for SIDS.

Epidemiological/ environmental factors

There are numerous risk factors with varying sensitivity and specificity which are associated with SIDS. The various risk factors are:

Maternal & antenatal risk factors –

- Intrauterine hypoxia
- Fetal growth retardation
- Urinary tract infection
- Smoking
- Anemia
- Drug exposure (cocaine, heroin)
- Nutritional deficiency
- Less prenatal care
- Low socioeconomic status
- Decreased age, education
- Increased placental weight
- Increased parity
- Shorter inter-pregnancy interval

Infant risk factors

- Age (peak 2-6 months)
- Asphyxia
- Bottle feeding
- Growth failure
- Male gender
- No pacifier
- Prone sleep position
- Recent febrile illness
- Smoking exposure
- Soft sleeping surface, soft bedding

- Thermal stress

Other risk factors –

- Cold season & climate
- Race / ethnicity – African, Native American, Gypsy, Maori, Hawaiian, Filipino)

There are numerous obstetric risk factors which contribute to SIDS which infers that the intrauterine environment is not ideal for the infants which later die of SIDS. Maternal smoking is an important risk factor. It causes –

- dysfunction of the pulmonary neuro-endocrine cells
- decreased ventilatory and arousal responsiveness to hypoxia

As prone sleeping prevalence has decreased, prenatal and postnatal exposure to tobacco smoke has emerged as an important risk factor for SIDS. Elimination of prenatal exposure to smoking can reduce the risk of SIDS by 30%. The other important factors include sub-optimal post natal care causing relative growth failure and prone sleeping position.

Prone sleeping position –

Prone sleeping is considered one of the significant risk factor for SIDS. There has been a decrease of 50% or more in rates of SIDS after dramatic declines in the prevalence of the prone sleep position to 10% or less. The ‘back to sleep’ campaign was first initiated in 1994. The recommendation calls for the supine position for sleeping in all infants without medical contraindications (e.g., micrognathia, obstructive sleep apnea).

Preterm infants were initially excluded from this campaign. This was based on the data that the ventilation was optimal when sleeping prone, especially in the presence of lung disease. But it was later confirmed that even preterm infants are at increased risk of SIDS in the prone position. The current recommendation is that supine should be the recommended sleeping position for all preterm infants and that supine sleeping position should begin in the hospital before discharge from NICU.

There may be interaction between the prone sleeping position and impaired cardio-respiratory control. Face down or nearly face down sleeping does occasionally occur in prone sleeping infants and can result in episodes of airway obstruction and asphyxia even in healthy full term infants. Infants with insufficient arousal responsiveness to asphyxia would be at risk for fatal asphyxia. Prone sleeping also increases the thermal stress which increases the risk of SIDS especially in infants with poor cardio-respiratory control. Sleeping on a very soft surface would further increase the risk of life threatening asphyxia in the face down or nearly down sleeping position. Thus, there may be links between risk factors such as soft bedding, prone sleeping and thermal stress and biological risk factors such as cardio-respiratory control deficits.

Identifying the babies at risk of SIDS

Even though 18.5% of SIDS victims are premature and the risk of SIDS progressively increases as birth weight decreases, those preterm infants destined to die of SIDS cannot be accurately identified prospectively. Methods to identify the infants at risk of sudden infant death syndrome are still under research. Research is going on to develop a screening test that is capable of accurately identifying the high risk infants which will allow the caregivers to give extra care to prevent SIDS. The ideal screening test should have negligible false –negative rates and acceptable false positive rates.

A few tests have been tried as screening tests for SIDS. These include pneumogram and polysomnogram. These tests essentially recorded the respiratory pattern or cardiac abnormalities and were not sensitive or specific enough to identify all the high risk babies. Moreover, the extent to which the cardio-respiratory control contributes to the risk of SIDS itself is not clearly defined. Hence, they were not considered as suitable screening tests for identifying infants at risk of SIDS.

New technologies using event recordings now permit home memory monitoring that includes respiratory pattern, heart rate, electrocardiography and oxygenation. Thus it is possible now to obtain ongoing home assessment of cardio-respiratory pattern. But as

mentioned above, it is still not possible to identify any specific cardio-respiratory pattern associated with an increased risk of SIDS.

Intervention

Though respiratory pattern is not a critical component in the pathogenesis of SIDS, home monitoring could still be effective if bradycardia or desaturation representative of life threatening conditions were occurring sufficiently early to be amenable to intervention.

But the drawbacks include uncertainty about the monitor use and compliance as suggested by the following reports –

- In more than 50% of the cases of SIDS the monitor was not used at the time of death
- Less than 10% of the monitor alarms were due to physiological events
- Parental difficulties with movements, loose lead or other non-significant alarms lead to frustration and non compliance

But these problems can be overcome by using memory monitors which identify and minimize problems with frequent false alarms. This will improve the family compliance.

There are a few other options which are available for intervention. Drugs like caffeine and theophylline which are meant to treat apnea of prematurity can be tried. These improve the respiratory pattern and reduce the frequency and severity of the clinical symptoms. But there are no well authenticated reports regarding the effectiveness of these drugs.

Conclusion: Strategies to prevent SIDS

Although, the peak incidence of SIDS occurs after the newborn period, parents frequently express concern about their child's risk. Although SIDS occurs most frequently in premature or low birth weight babies, the apnea of prematurity commonly

seen in them do not increase the risk of SIDS. The following strategies may be tried to decrease the risk of sudden infant death syndrome.

Sleeping position –

Sudden infant death syndrome is common in babies made to sleep in the prone position. Sleeping on the back or on the sides reduces this risk. But there are a few exceptions for this. These exceptions include preterm infants with respiratory disease, infants with gastro esophageal reflux and infants with craniofacial abnormalities or evidence of upper airway obstruction. For these infants soft bedding should be avoided.

Avoiding smoking –

Infants exposed to maternal smoking during pregnancy and post-natally have a higher risk of SIDS. Smoking should be avoided by parents and infants should not be exposed to smoke.

Avoiding overheating –

Infants exposed to excessively high room temperatures or overheating from excess wrapping have an increase of SIDS. Caregivers should avoid practices that result in overheating.

Breast – feeding –

Infants who were never breast-fed have an increased risk of SIDS than those who were breast-fed. So breast-feeding should be encouraged to prevent sudden infant death syndrome.

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