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# Emergency care approach to sudden infant death syndrome

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**Abstract**

Sudden infant death syndrome, also called crib or cot death, is the sudden and unexpected death of an infant under one year of age who is thought to be completely healthy, and the cause of death cannot be explained by medical research. It is one of the important causes of mortality in childhood after the neonatal period in developed countries. The exact cause is unknown. The aim of our study is to present an emergency care approach by reviewing the recent studies on sudden infant death syndrome.

**Keywords:** Sudden infant death syndrome, Emergency care, Nursing

## Introduction

Sudden infant death syndrome (SIDS) is defined as the sudden, inexplicable death of an infant under one year of age, even after a comprehensive case examination, including a full autopsy, medical and social history, and crime scene investigation [1]. This concept was first introduced in 1969 to draw attention to the sudden deaths of infants with similar clinical features [2].

Although there are different hypotheses, the inability to fully explain the etiopathogenesis poses a major problem. As in many countries, the question of whether SIDS or a suspicious death could not be answered in many cases in Turkey and these cases were evaluated in the category of "death with unexplained cause" [3].

## Epidemiology

In 2016, approximately 3,600 sudden infant deaths (SID) occurred in the United States of America (USA). Cases occur among infants younger than one year of age with no apparent cause for the event [4]. Another study evaluating 94,038 infant deaths in Turkey between 2007 and 2012 reported that the incidence of SIDS increased by 2.1 times between 2007 and 2014, reaching 4% from 1.9% [5]. According to the 2016 death statistics of the Turkish Statistical Institute [6], babies who die between 1-4 months of age in Turkey constituted 22.6% of infant deaths; however, there is still no comprehensive study on the frequency of SIDS [7].

SIDS cases usually occur while asleep, during the first hours of the morning and at low ambient temperature. Seasonally, it most commonly occurs in the winter between October and April. The month in which infants are born is also related to the frequency of SIDS. While the frequency is the lowest in those born in early spring, it is the highest in those born in early August [8].

The fact that the frequency varies depending on ethnic groups and the higher occurrence in the children of mothers with blood groups (0) and (B), 10 times higher probability of its occurrence in consecutive siblings and 20 times in twins suggest that genetics may also have an etiological role [9].

Situations that cause a rapid and silent clinical picture during sleep have been defined as risks [10]. It has been thought that some chemical stimuli affect the upper respiratory tract or a disturbance in some metabolism or in the secretion of neurotransmitters cause sleep apnea, which, if prolonged, leads to death [9].

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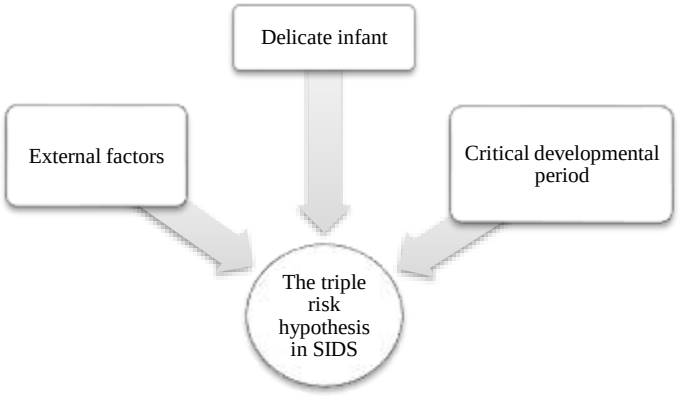
**Risk factors**

In the past centuries, scientists have put forward theories about the risk factors of SIDS. However, the most widely accepted hypothesis was suggested by Filliano and Kinney in 1994 [11] and includes the triple risk model (Figure 1).

Figure 1: The triple risk hypothesis in sudden infant death syndrome (SIDS)




Death occurs when infants with insufficient defense mechanisms and developmental disruptions are exposed to external stressors. SIDS is based on a triple risk model, which demonstrates that three factors that affect the infant occur simultaneously.

A triggering event such as apnea, hypoxia, insufficient cardiorespiratory control, cardiovascular pathologies, infections, anaphylaxis, preterm labor and lying prone, metabolic and congenital diseases, airway obstruction accompanying a brainstem anomaly or an anomaly in serotonin signaling play a role in the pathophysiology [12, 13].

In a study investigating the changeable and unchangeable maternal and labor-related risk factors associated with SIDS, it was determined that maternal smoking, chronic hypertension and gestational hypertension in the mother, premature labor, intrauterine growth retardation (IUGR) and being twins were prominent risk factors [14].

Many maternal, infant, and environmental risk factors were identified in the development of SIDS.

**Maternal risk factors:** The mother's low educational and socioeconomic status, single marital status, living alone, overweight, young age (especially 20 years old and below), previous miscarriage or the death of her previous children due to SIDS, consanguineous marriages, racial and ethnic origin are risk factors [15, 16]. Smoking status of mother, particularly during pregnancy, increases the risk of SIDS. Nicotine in the tobacco passes to the fetal circulation through the placenta and binds to the acetylcholine receptors in the fetal brain. In this process, it causes repetitive episodes of hypopnea, thereby bradycardia attacks, and changes the parasympathetic control of the heart, survival of cells and synapse formation. Nicotine also causes chronic hypoxia after labor due to decreased lung capacity of the fetus and increases the risk of respiratory tract infection by increasing the sensitivity of the infant [17, 18]. Since there is a risk of SIDS due to nicotine exposure, the use of electronic cigarettes during pregnancy should also be avoided [19]. An infant's exposure to cigarette smoke is an additional independent risk factor. Maternal use of harmful substances and alcohol are highly associated with SIDS in many studies [3].

**Pregnancy-related risk factors:** Pregnancy-related complications (placenta previa, ablatio placentae, premature rupture of membranes, increased alpha-fetoprotein level in the second trimester) infections, urinary infection, trauma experienced during pregnancy, malnutrition during pregnancy, frequent delivery, low maternal blood pressure in the third trimester of pregnancy, bleeding in the last trimester that causes anemia and inadequate prenatal care also increase the risk of SIDS [9, 15, 19]. Deaths are more common in infants whose mothers use barbiturate or narcotic drugs during pregnancy [20].

**Risk factors regarding labor:** Sedation or anesthesia during labor [9] and oxygenation with resuscitation performed during labor are in the risky group [15].

**Infant-related risk factors:** Premature infants, IUGR and infants with low birth weight are at great risk. In addition, it is more common in intrauterine passive smokers [21]. Studies have found that among infants with low birth weight, the rate of SIDS is 3-4 times higher than infants with normal weight. There is a 5-6-times increase in the risk of SIDS in infants with a history of SIDS in their siblings [22]. Cohort studies on twins have found the mortality rate twice as high as those of single infants [23]. The growth-development and nutritional status of the infant also pose a risk [24]. Among the risk factors, congenital or acquired diseases of the infant, including heart diseases (myocarditis, long Q-T syndrome, congenital heart disease), lung diseases (bronchiolitis, pneumonia), brain diseases (cerebral edema, subdural hemorrhage, meningitis encephalitis, intracranial hemorrhage) and blood diseases (sickle cell anemia) are of great importance [25]. Metabolic diseases constitute 5% of sudden and unexpected infant deaths. A family history of metabolic disease or sibling death with unknown cause, a clinical picture similar to sepsis before death, hyperventilation, feeding intolerance, vomiting, hypoglycemia, lactic acidosis, hyperammonemia suggest a metabolic disease. The most common metabolic disease that causes sudden unexpected infant death is fatty acid oxidation disorder. To exclude metabolic diseases, it is necessary to perform metabolic examinations for all infants with a risk of SIDS. However, since there may be a risk of recurrence in metabolic diseases or abuse, these conditions should be excluded before confirming the diagnosis [10].

**Environmental factors:** There is a strong relationship between the infant's lying position and SIDS and most deaths occur in the prone position [7]. Prone sleeping position is the strongest changeable risk factor for SIDS. In a case-control study, it was shown that the risk of SIDS increased 2.3-13.1 times in the prone sleeping position [26].

The National Institutes of Health (NIH) study suggested that the risk of aspiration was lower among patients in the supine position, since the trachea in infants lying on their back is on top. This proves that infants do not aspirate in case of vomiting during sleep and that lying on their back to breathe more comfortably is a factor that prevents SIDS [27]. In addition, despite the negative effects of the prolonged supine position on musculoskeletal and motor development, it has been reported that it is protective against sudden infant death syndrome due to the decrease in the arousal threshold and more frequent awakenings [28]. The US Food and Drug Administration (FDA)

warned that sleeping the infants on their side is associated with SIDS [29]. According to recent studies, U-shaped pillows have been shown to play a role in infant mortality [30].

**Sleep environment:** The use of a soft sleeping surface was found to increase the risk of SIDS in a case-control study. In another study, it was found that using a soft mattress in the prone position increased the risk 21 times [31]. The prone sleeping position increases the $CO_2$ level with re-breathing of expiratory air, particularly in infants who sleep in soft mattresses [32]. This position may also cause a reflexive displacement of the lower jawbone, obstruction of the pharynx, or closure of the nose, thereby an obstruction of the upper airway [33].

Couch or reclining chair, beads, polystyrene foam mattresses or mattresses with natural fiber, face cover, non-stretched bed sheet, blanket or quilt, pillows, toys and any other soft objects on the bed that cover the face while sleeping can increase the risk of SIDS by airway obstruction. Infants who are overdressed while asleep, swaddled tightly or sleep-in rooms that are overheated are also at higher risk. It has been recommended that car seats or strollers not be routinely used for sleeping [34, 35]. Otherwise, the risk of SIDS increases if infants do not have sufficient neck control to support their airways when they fall asleep. In addition, it should be kept in mind that aspiration due to increased gastroesophageal reflux (GER) may develop with the use of these equipment [36]. Although GER is physiological in the first months, recent studies have shown that lying in a supine position does not increase the risk of asphyxiation and aspiration in infants suffering from GER [37].

Again, an increase in the risk of SIDS has been observed in infants sharing the same bed with their parents [35, 38]. It has been shown in many studies that sleeping together and bed sharing increase the risk, particularly in infants younger than three months of age [26]. Parents being very tired, parental smoking, use of alcohol and sedative drugs and overweight increase the risk of SIDS among those who share their beds with the infant. A low birth weight of the infant, preterm labor and the infant being younger than 8-14 weeks are also risky conditions [39].

**White noise**

White noise is a continuous monotonous sound in the form of buzz that has a calming feature suppressing disturbing sounds in the environment [40, 41]. It is thought that while the infant is still in the womb, it is affected by the heartbeat of the mother and that re-finding this familiar sound and rhythm after birth has a relaxing effect on the infant [40, 42]. With the use of "sleep aids" (such as pacifiers, white noise, rocking cradles and swaddling), lonely and long-term infant sleep is targeted from an early age (deep sleep with minimal warnings). Inhibition of the stimulation response is a characteristic of vulnerable infants and increases with exposure to external stress [8]. Despite the potential benefits, white noise doesn't always offer risk-free peace and silence. In 2014, AAP tested 14 white noise devices designed for infants. They found that they all exceeded the recommended noise limits set at 50 decibels. In addition to increasing hearing problems, the study found the use of white noise to be risky in speech development. Based on the results of the AAP, pediatricians recommend placing any white noise device at least 2 m from the crib. In addition, it is recommended to lower the noise level of the device [43].

**Typical characteristics of SIDS**

In a well-developed infant, foamy blood around the nose, cyanosis in the lips and nail fold can be identified. Pulmonary edema is usually present and can be significant. Subacute inflammation in the upper respiratory tract, minimal stress effects on the thymus and adrenal glands and empty bladder have been found in most.

In infants died of SIDS, differences are found in the lung and other organs, brain stem and functions. SIDS mostly occurs in a healthy infant approximately 20 minutes after being put to sleep by his/her mother or his/her babysitter during the day or night. This was also associated with laryngospasm [8].

**Diagnosis**

All possible diseases and causes of death should be excluded for a diagnosis. For a complete investigation, standard protocols including detailed autopsy, scene investigation should be performed, and infant and family history should be obtained thoroughly [44].

In some cases, fatal child abuse can be confused with sudden infant death syndrome. In autopsy, it is difficult to differentiate between SIDS and incidental or intentional asphyxiation with a soft object [45]. Cases of infant deaths older than six months, simultaneous twin deaths, recurrent cyanosis, apnea, mouth and nose bleedings and unexplained sibling deaths may suggest abuse [7].

Especially in recurrent infant deaths, “Munchausen syndrome by proxy”, fatal child abuse and asphyxiation should be kept in mind. Intrathoracic petechial and intraalveolar hemorrhages seen in asphyxia deaths are also seen in SIDS cases [45].

**Autopsy findings**

The purpose of the autopsy protocol is to provide additional information to standardize autopsy practices, increase diagnostic accuracy and support the information obtained from clinical history and death events [2]. Autopsy is appropriate in cases of sudden and unexpected infant deaths, since all known causes cannot be excluded by history and imaging techniques [46].

**The diagnostic criteria for SIDS:**

- Term pregnancy,
- Normal clinical history, normal growth and development,
- Safe sleeping bed with inspection of the place of death,
- Other causes of death such as severe infections like sepsis, fluid-electrolyte imbalance, severe congenital anomalies, congenital metabolic diseases and poisoning should be excluded,
- Autopsy findings including cranium and cranial structures should support SIDS,
- There should be no macroscopic or microscopic findings suggestive of trauma, accident and/or serious disease,
- There should be no evidence of trauma on skeletal radiographs,
- There should be no evidence of intoxication with substances such as alcohol, medicine, etc.,
- There should be no findings suggestive of specific etiology in history and imaging,
- Based on the information obtained, the cause of death should be inexplicable [45].

**External Examination**

External examination begins by determining the infant's height and weight. In the meantime, changes specific to newborns such as vernix caseosa and lanugo covering the body are observed. In order to obtain information about the term of the child, it is necessary to carefully examine and photograph the head circumference, shoulder width, sole length, the length of the finger and toe nails, the umbilical cord and anogenital area [47]. The infant's body surfaces, oral and nasal cavities should be carefully examined, post-traumatic injuries or lesions on the skin, conjunctiva and mucosa should be checked and if any, petechiae should be recorded [2].

After the initial external examination is completed, a radiograph of the body should be taken to exclude bone injuries in the body. Infants who die of SIDS usually do not have external abnormalities. In other words, they give the impression of a well-groomed infant. Cyanosis on the lips and nasal wings and the presence of foamy, sometimes bloody mucus around the mouth and nose are among important findings. Dead examination findings are completely negative. There is no trauma [2].

**Approach to SIDS in the Emergency Department**

In deaths from SIDS, healthy infants are generally fed at night and put in their cribs. The next morning, the parents find their infant cold, purple and unresponsive. The emergency team is called and the infant is either transferred to the emergency room or stays at home depending on the emergency protocol. If the infant stays at his/her home and his/her room should not be touched until the forensic team reaches the scene [48]. Parents are asked questions to obtain information about the condition of the infant before death (health status, last feeding time, whether he/she was checked at night, sleeping position, his/her status when the infant was found dead) [49, 50].

If the infant is brought to the emergency room after he/she was found dead, the relevant units are called for a forensic medical examination. Scene investigation is important in determining the factors influencing the death of the infant. After the scene investigation is completed, the information gathered is reported to the physician who will perform the external examination and then the autopsy. In all cases, autopsy including detailed examination and toxicological examination are necessary to determine the cause of death [48, 49].

The parents may have difficulties accepting sudden death. Stress, anger and guilt are the most common reactions. Interviewing the family about death and waiting for autopsy results for a long time increases emotional stress [48]. All procedures should be explained and appropriate language should be used for interview techniques in traumatic cases. Healthcare professionals should explain to the family that it is routine to identify possible mechanisms leading to death. It should be specified that the questions in the interviews are asked to determine the risk factors and gather information and are not intended to accuse the family. Open-ended questions should be asked while taking history from the family in shock due to death and it should be emphasized that the collected information will be used by the forensic medicine team. It should be explained that autopsy performed to determine the mechanisms and etiology leading to sudden death is medically and legally necessary [50]. While allowing families to express their feelings, the necessary support and counseling should be provided to facilitate their coping with the trauma. They can be directed to group therapies, and informed that SIDS can be prevented to a great extent with the measures taken for preventable risk factors. Due to the lack of standard practice and a multidisciplinary approach, the number of studies in Turkey are very limited.

**Conclusion**

Emergency room personnel should be trained in the management of SIDS cases and develop appropriate approaches by working in coordination with the forensic team members. To reduce SIDS, the health personnel should be educated according to known etiologies, and parents should be informed as a preventive public health service. Families' awareness of the prevention of SIDS should be raised, standard procedures for case management and reporting should be developed and the family should be supported during the mourning process.

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