Sudden Infant Death Syndrome: An Update

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Abstract:

The sudden infant death syndrome (SIDS) accounts for most of the deaths in infancy. The precise etiology of this condition, despite extensive research, remains unknown. Each year additional studies advance newer explanations. In this paper an attempt is made to review the old and the new theories explaining why an apparently healthy infant dies. The pathological investigations in the past have not contributed substantially in solving the mystery of the SIDS etiology. The clinical studies also have not provided complete answers but they have helped recognize the entity of near-SIDS paving the way to possible prevention of the SIDS. Current thinking on this subject is presented for the primary care physicians and the researchers.

Introduction

One of us (A.F.) co-authored an article in Virginia Medical Monthly in 1968 in which an analysis of 199 cases of sudden deaths in infancy and the etiological factors were presented. Since that time a considerable amount of new and useful information has emerged on the subject of SIDS and near-SIDS. An attempt is made to update that information in this article principally for the primary care physician.

The sudden unexpected death in infancy, also known as "crib death," "cot death," or "sudden infant death syndrome (SIDS)" is a condition characterized by the sudden death of an infant who had previously been well or affected with a mild illness such as coryza, cough or a mild gastro-intestinal upset which was in no way felt to be of sufficient severity to have caused death. The entity is also defined as a sudden death of any infant or young child, which is unexpected by history and in which a thorough postmortem examination fails to demonstrate an adequate cause for death. A popular hypothesis in recent years is that the infants who die suddenly and unexpectedly are thought to have an abnormality in the autonomic regulation of cardiovascular and respiratory activity sometimes exaggerated by a stimulus such as inflammation of the respiratory tract. It is also important to define near-SIDS. Sudden onset of apnea in infancy with cyanosis or pallor is a high-risk state sometimes causing death. When it is non-fatal, it is called "near-SIDS."

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The SIDS is a common condition. There is no doubt that every family physician, pediatrician, and emergency room physician will be directly or indirectly involved during his practice with this syndrome.

Epidemiology

The SIDS is responsible for more deaths in infancy after the first week of life than any other condition. The reported incident is two to three deaths of infants per thousand live births.² The survey of the research indicates that the following are recognized as important risk factors:

Risk factors related to the mother are important. The risk of SIDS is increased if the mother

- a. is less than twenty years of age;
- b. is unmarried:
- c. is economically deprived;
- d. has delayed or failed to seek prenatal care;
- e. has had a short interval between pregnancies;
- f. has been ill during pregnancy;
- g. has had previous abortion or miscarriage;
- h. has been a smoker especially during pregnancy;
- i. has abused narcotics,3

The risk is also increased if the father is less than twenty years old and comes from a low socio-economic bracket.4

The important factors relating to the infant are:

- a. sex there is a 55 to 83 percent male preponderance in the incidence of the SIDS.⁵
- age most cases fall in the age group of
 weeks to 8 months with the extremes being
 days and 3½ years. The peak age of infant deaths is between 2 and 4 months;
- c. race in the U.S., the Asians have the lowest rate (0.51 per thousand), while the Blacks have the highest rate (5.04 per thousand);

- d. low socio-economic status of the infant;
- e. prematurity;
- f. low birth weight;
- g. infants not first born;
- infants who are products of multiple birth pregnancies; triplets are a greater risk than twins;
- infants with mild gastro-intestinal or respiratory symptoms such as cold, sniffles, cough, vomiting or diarrhea;
 - j. infants discharged after post-birth intensive care unit treatment;
 - infants who are less active and less responsive and have an unusual cry;⁶
 - 1. illegitimacy;
 - m. infants from large families;
 - increased risk of SIDS in the siblings of the SIDS victims.

The following additional general factors are worth noting:

- a. infants tend to die during their sleep at night;
- the majority of infant deaths occur in the early hours of the morning, approximately 60% occurring between 3:00 A.M. and 9:00 A.M.;
- the majority of infant deaths occur in the colder months of fall, winter and spring with the peak in December, January and February.

Pathology of the SIDS

The majority of the autopsies on the cases of SIDS are, in a conventional sense, unrevealing. The following autopsy findings, both positive and negative, are sometimes observed:

- a. normal or subnormal development of the body and normal or subnormal nutrition;
- clear, frothy or blood tinged secretions in the nostrils;
- petechial hemorrhages in the surfaces of the thymus, heart, lungs and pleura;
- d. increased weight of the lungs, patchy or uniform purplish discoloration of the surface feeling slightly firm, and congestion and edema of the tissue. Microscopically, the lungs reveal congestion and patchy edema. The alveolar walls are thickened with an infiltrate of lymphocytes, occasional neutrophils and monocytes. In some areas, alveolar atelectasis is seen and occasional alveoli are found to contain macrophages and alveolar lining cells;7
- thin frothy secretions and aspirated gastric contents in the larynx, trachea, and bronchi;
- f. enlarged thymus gland;
- g. focal necrosis of the vocal chords;
- thickening of the smooth muscle in small pulmonary arteries;
- i. brain stem gliosis, astroglial proliferation

- around the nucleus of the vagus and the nucleus ambiguous;8
- j. smaller glomus cells in carotid body with fewer chemoreceptor granules.9

Etiology of SIDS

The etiology of the SIDS is still unknown in spite of a great deal of investigative work. There is a great diversity of thought on the subject, and the thinking has changed with the times.

Older literature discussed the following theories to explain the SIDS.

- a. suffocation;
- b. teething;
- c. smothering;
- d. status thymolymphaticus;
- e. interstitial pneumonitis without any cause;
- f. bacterial infection;
- g. viral infection;
- h. hypersensitivity to cow's milk antigens;
- i. neurogenic cardiac arrest from strong vagal impulses;
- j. hypogammaglobulinemia;
- k. anaphylactic shock following intra-uterine infection;
- congenital abnormalities.

More **recent literature** stresses the importance of the following three factors:

- a. infection mild respiratory tract infection of viral origin is thought to potentiate the risk of the SIDS and is present in 40 to 75 percent of the cases of SIDS. Infection with clostridium botulinum and amnioitis in preterm births are associated with SIDS.³
- b. neurological abnormalities the research demonstrates that SIDS infants are physiologically and neurologically abnormal prior to death, they tend to have Apgar scores less than 6 at one minute, a greater need for resuscitation, more feeding problems and jitteriness or abnormal Moro reflex.²
- arrhythmias caused by abnormal cardiac conduction, alteration in control of the heart rate and cardiovascular failure account for a small number of cases of SIDS.

Apnea in Infancy: Near-SIDS

The American Academy of Pediatrics defines prolonged apnea as the cessation of breathing for 20 seconds or longer, or as a briefer episode associated with bradycardia, cyanosis or pallor. The occurrence of periodic apnea during periodic breathing is commonly observed in preterm infants. Some investigators consider periodic apnea as a benign form of respiratory instability, but several reports associate it with high risk for SIDS. A great deal of modern research deals with the relationship between apnea and near-miss SIDS infants (near-SIDS). The apnea is probably caused by abnormal cardiac conduction and

abnormal regulation of breathing.

Near-SIDS identified by the detection of a sudden onset of apnea with cyanosis or pallor can be helped by prompt resuscitative efforts. This fact is very important because of its significance in the prevention of SIDS.

Physician's Role in Advising the Parents of the Victims of SIDS

Every primary care physician should be acutely aware of the parents' feelings after the death of an infant. He should realize that most parents know very little about SIDS. Therefore, a sudden and unexpected loss instantly creates intense guilt feelings. A parent blames himself or herself for causing the death of the infant. Such a reaction is due to ignorance of the facts of the SIDS. A primary care physician is perhaps in the best position to alleviate the crisis situation with a full discussion of the SIDS. To be able to do this he should also recognize the intensity of grief and stress brought on by the sudden unexpected death of an infant. He should be prepared to sit down with the parents of the SIDS victim and discuss the SIDS with a great deal of compassion, at any time of day or night, soon after the death. In particular, during such a session the physician should stress the following important facts about the SIDS:

- a. it is a natural death:
- it is a condition not associated with any definite signs and symptoms sufficient to forecast the impending death;
- the death from this syndrome is not unequivocally predictable;
- d. the death is not definitely preventable;
- e. the death is not from suffocation or aspiration;
- f. the death is painless;
- g. the condition is not contagious;
- h. it is not hereditary;
- the death from this syndrome can occur in spite of the best of care;
- i. the cause of this condition is not known.

Such a presentation of the facts will greatly help the parents by reducing their shock and minimizing their guilt feelings. After this initial discussion, the parents should be gently told that an autopsy will probably be done because in most states a sudden, unexpected death requires an investigation by a medical examiner or coroner. They should be encouraged to cooperate in such an investigation because the autopsy would help confirm the cause of death.

Identification of Near-SIDS and Possible Prevention of SIDS

After the initial shock following the sudden death of an infant subsides, some parents accept the fact of the loss. Then they start raising questions about the siblings, future births and possible approaches to prevent the SIDS. The physician could assist the families with the knowledge of more recent work in the field of near-SIDS. This work relates to the identification of the infants at risk, intervention through monitoring and possible prevention of the SIDS by homemonitoring programs.

The following three groups of infants are considered to be at risk for the SIDS:

- a. siblings of an infant dying of the SIDS.11
- b. infants experiencing a "near miss."12
- c. infants with prolonged apnea and/or bradycardia in the neonatal period.^{13, 14}

The parents of an infant considered to be at risk of SIDS may be advised to bring the infant for evaluation and assessment by infant monitoring equipment. Some such devices available are Electronic Monitoring Inc., Model RE134C or D, Fort Worth, Texas; Airshield, Model AM-46-1, Hatboro, Pennsylvania for chest impedance monitoring and Parks Manufacturing Inc., Model 510-A, Beaverton, Oregon for heart-rate monitoring. After the high risk infant is assessed with history, physical examination and laboratory investigations such as urinalysis, complete blood count, blood glucose, serum calcium, electrolytes, chest X-ray, EKG and EEG, to rule out any identifiable causes of apnea, it is selected for continuous monitoring of respiratory patterns and cardiac activity. The monitors are set to sound an alarm after a non-breathing period of twenty seconds or at heart rate less than eighty beats per minute. For infants with abnormal recordings, follow-up monitoring is recommended every six to eight weeks until two consecutive normal recordings are obtained. Most times, when an abnormal recording is obtained a tactile stimulation of the infant is sufficient to reverse the breathing to normal. In some instances mouth-tomouth resuscitation may be required. Doctors Kelly and Shannon of Massachusetts General Hospital have extensively used the monitoring devices and have been helpful in developing programs of evaluation and monitoring in several parts of the country. Many hospitals and private organizations in different states also offer this service. With this program of monitoring, several anxious parents have gained reassurance and are better prepared to face up to the aftermath of the tragedy they once faced.

Finally, in order to educate such families further, they should be encouraged to join one of the several organizations that operate to increase the understanding of the SIDS and near-SIDS.

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