The Nature of the Problem: Sudden Infant Death Syndrome

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The following is an overview of the Sudden Infant Death Syndrome (SIDS) and emphasizes salient features of its epidemiology, pathology, pathophysiology and management. Controversial points are not discussed.

Epidemiology

SIDS is an increasingly common problem with a reported incidence of 2 to 3 per 1,000 live births. In the United States, there are approximately 10,000 deaths per year. For Virginia in 1976, there were 129 reported cases.

In the 1972 King County, Washington study, from one week to 11 months of age the leading cause of death is SIDS (45%), followed by congenital malformations (20%). From one week to 14 years of age SIDS is the third leading specific cause of death behind diseases of early infancy and accidents. SIDS is more common in males than females, 3 : 2. The syndrome is more common in the economically disadvantaged, with blacks having an incidence of 5 per 1,000 live births. Infants of lower birth weight and gestational age have a higher incidence of the problem. For example the risk for babies weighing 3 lbs 8 oz is 13.27 per 1,000 live births. Paradoxically, infants who weighed less than 3 lbs 8 oz at birth have a decreasing incidence, but the lower incidence may reflect the high mortality from respiratory disease in the neonatal period. SIDS is most common at 2 to 4 months of life with few cases being reported before 2 weeks and after 12 months. Nasopharyngitis seems to accompany SIDS. In the northern hemisphere, SIDS tends to occur more frequently in winter; whereas in the southern hemisphere, the syndrome occurs more commonly in summer months. About 74% of all deaths are discovered during sleep in the early morning hours.

It should be remarked that nationally, there is considerable debate as to the true epidemiology of SIDS.

Pathology

During the past ten years pathologists have assisted in bringing about a better understanding of the syndrome. SIDS is defined as, "The sudden death of any infant or young child, which is unexpected by history, and in which a thorough postmortem exam fails to demonstrate an adequate cause of death." * By definition an autopsy is then necessary to rule out SIDS.

Beckwith reported a series of 500 autopsies in infants dying during the first year of life. In 15% a cause of death could be found (infection, trauma, major malformations and others). In 87% of the infants intrathoracic

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* J. B. Beckwith: "The Sudden Infant Death Syndrome." *Pediatrics* 3:29, 1973.

petechiae were noted. A number of observations by investigators from pathological specimens would tend to indicate that infants dying of SIDS are chronically hypoxemic. These findings would include leukomalacia, demyelinization of the brain, intrathoracic petechiae, decreased cytoplasmic granules in carotid chemoreceptors, increased muscle mass in the small pulmonary arterioles, extramedullary hematopoiesis and retention of brown fat. Other autopsy findings in SIDS would include normal nutrition and hydration, perhaps some blood-tinged sputum or frothy fluid at the mouth or nostrils, diapers wet or full of stool, clutched hands, pulmonary congestion or edema, perhaps some interstitial pulmonary lymphocytic infiltrate, normal size adrenal glands, normal thymus and no evidence of aspiration.

Pathophysiology

Why do babies die of SIDS? What is a near-miss SIDS and how does it relate to SIDS? How does the physician identify the infant at risk for SIDS? Answers to these questions would be important to the child and the clinician. As yet there are no clear answers but many clues.

A near-miss infant is defined as a previously well infant who, during sleep, experienced an episode of apnea, limpness, cyanosis or pallor that was terminated by vigorous stimulation or mouth-to-mouth resuscitation. It has now been documented that a number of infants with near-miss SIDS die of SIDS. These near-miss infants need the following: 1) complete physical and neurological examination, 2) complete blood count, 3) urinalysis, 4) serum Ca^{++}, $PO_4^{=}$, Mg^{++}, glucose, Na^{+}, K^{+} and amino acids, 5) capillary blood gases, 6) urine organic and amino acids, 7) chest and skull x-ray, 8) barium swallow, 9) 12-lead electrocardiogram, and 10) sleep and awake electroencephalogram. Included in the differential diagnoses are seizures, esophagopharyngeal reflux, sepsis and aspiration.

The control of ventilation is a very delicately balanced system. The near-miss SIDS and SIDS victim would appear to have abnormal control of ventilation especially during sleep. Study of ventilation in the small infant is difficult and not available except in research laboratories. Sleep and awake electroencephalograms, ventilatory response to various inspired gas mixtures and thoracic and nasal movements are monitored in an integrated system. To date it has been demonstrated that the near-miss SIDS infant has hypoventilation, a depressed ventilatory response to CO_2 breathing, prolonged sleep apnea, frequent short apnea, periodic breathing and apnea clustered in sleep. Apnea in near-miss infants has been demonstrated in REM and Quiet Sleep.

In addition to evidence for abnormal control of ventilation during sleep clinical investigations have hypothesized that obstructive apnea may occur during the terminal event. Tonkin demonstrated the anatomic vulnerability at the oropharyngeal level between the soft palate and the base of the skull in the small infant. In REM sleep with muscle relaxation this anatomic vulnerability may lead to oropharyngeal occlusion. In near-miss SIDS infants abnormalities of muscle tone have been demonstrated particularly shoulder hypotonia. In one case study, loss of electromyogram (EMG) activity of the genioglossus muscle with obstructive sleep apnea has been confirmed. Several family members had died from SIDS.

Therefore, the near-miss SIDS and SIDS patient would appear to have chronic hypoxemia, abnormal control of ventilation during sleep and possibly an obstructive asphyxial event. Despite all of the evidence, the pathophysiology is not completely explained and a simple test to identify the infant at risk has not been forthcoming.

Management

Autopsies should be performed on all infants who are thought to have SIDS. The postmortem will rule out abuse and infanticide and other diagnoses. The parents can then with certainty be told that their infant died from SIDS and that they could not have prevented the death.

A common denominator for the family is guilt which when combined with grief and lack of knowledge can lead to major emotional stress, exacerbation of previous psychiatric conditions, and marital problems. To alleviate the guilt and grief the parents should be referred to a SIDS guild, and should be helped by many counseling visits to their pediatrician or family physician.