Defining the Sudden Infant Death Syndrome

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Sudden infant death syndrome (SIDS) is a term that was first proposed in 1969 for a distinctive subgroup of unexpected infant deaths that occur during the postneonatal period with relatively consistent clinical, epidemiological, and pathological features. This term played an important role by focusing attention on a major category of postneonatal infant death, providing support to grieving families, and diminishing the guilt and blame characteristic of these deaths. Unfortunately, the application of this term has become increasingly controversial. Some have applied it too liberally, and others not at all. According to the definition proposed in 1969, despite slight changes suggested in 1989, SIDS remains a diagnosis of exclusion. Although this syndrome has several distinctive features, including age distribution and apparent occurrence during sleep, there has been reluctance to include these features in the definition. The problems created by the lack of an adequate definition are discussed. A 2-tiered approach is suggested, with a more general definition intended primarily for case management and death administration, and a more restrictive one intended primarily for research purposes, which distinguishes those deaths closely fitting the classic SIDS profile from those with one or more less typical features.

A frustrating challenge is presented by diagnostic entities lacking specific laboratory tests or pathognomonic clinical manifestations. Many new diagnostic terms, usually ending in “syndrome,” “complex,” or “disorder,” have been proposed in recent years. Many of these have attracted intense media attention and activist support groups, and pediatricians are constantly faced with the issue of whether a child’s perceived problems justify a label such as attention-deficit/hyperactivity disorder or chronic fatigue syndrome.

Sudden infant death syndrome (SIDS) exemplifies the problems that can arise when a syndrome lacks adequate definitional criteria. Sudden infant death syndrome remains a diagnosis by exclusion, distinguished from other infant deaths only by subjective and permissive variables that can be interpreted according to the whims of the diagnosing pathologist. Current definitional criteria for this diagnosis leave pathologists free to apply this designation either too liberally or too restrictively. Individuals with theories or findings that they believe to be related to SIDS have been able to attach this term to cases that differ drastically from the usual profile. A dramatic illustration of this was a 1972 report of 5 siblings presumed to have died of SIDS.1 One of the victims in that family died at 28 months, and another was said to have died while awake and being fed. Postmortem examinations, performed in only 3 of these deaths, yielded results “consistent with SIDS,” but no details were provided in the report. Historic or laboratory evidence suggesting an apneic disorder preceding the death of some of these infants resulted in massive attention directed toward home apnea monitoring as a means of preventing SIDS. For more than a decade virtually all SIDS research was devoted to the apnea hypothesis to the exclusion of all others, delaying progress in research into other causes or mechanisms. Eventually, the 5 infants in the
original report were proved to have been victims of infanticide.2

When SIDS is diagnosed too restrictively or not at all, the family is often denied the benefits of the SIDS grief support system. Unwillingness to diagnose SIDS can also lead to inappropriate suspicion or blame directed toward parents or caregivers, including unfounded self-accusation.

Some pathologists will not diagnose SIDS if the postmortem examination findings reveal even a slight abnormality.3 Inflammation of the respiratory tract is a commonly certified alternative for SIDS, and is often applied when the microscope reveals only an occasional lymphocyte in the respiratory tract. I have testified in numerous malpractice suits blaming physicians who diagnosed a minor respiratory infection in an infant who not long thereafter had been found dead. The circumstances of death and postmortem findings were typical for SIDS, but the presence of microscopic inflammatory infiltrates in the lungs, no more severe than might be caused by the common cold, led to certification of the cause of death as respiratory infection. This in turn led to malpractice claims against the treating physician.

No less unfortunate are some situations in which the SIDS diagnosis has been applied too liberally. Lethal genetic disorders and infanticide have been misdiagnosed as SIDS. Overuse of the diagnosis has contributed to criticism of the SIDS concept by respected pediatric pathologists who suggest that SIDS is not a true entity, but a “wastebasket” term that interferes with the search for true causes of sudden death in infancy.3-6

In recent years a new factor has further complicated the use of SIDS as a diagnostic term. The “back to sleep” campaign has been associated with a substantial reduction in the number of infant deaths in the United States and elsewhere. This reduction has primarily occurred among those infants who composed the most typical SIDS cases in the past.7 The peak in the age distribution curve between 2 and 4 months has become less prominent. Infants now dying suddenly and unexpectedly include a higher proportion of “atypical” cases, where the index of suspicion is higher than previously. This change in demographics has contributed to an increasing reluctance on the part of many pathologists to use the diagnosis of SIDS. Some pathologists refuse to diagnose SIDS in any infant who dies in a shared sleeping situation or when the infant was found face down, despite lack of evidence that these cases differ in any way from infants found with the face entirely free of obstruction.5-9

The problems cited herein are clearly exacerbated by the lack of an adequate definition for SIDS. This article reviews the history of this definition from the perspective of one who was an active participant in the process, discusses the inadequacy of the currently popular version, and proposes an approach that might prove helpful.

THE 1969 DEFINITION

In 1969, Abe Bergman, MD, George Ray, MD, and I hosted the Second International Conference on Causes of Sudden Death in Infants near Seattle, Wash.10 Our primary objective was to review the evidence that most infants dying unexpectedly in the postneonatal period had features suggesting a single cause or mechanism of death. Discussions were lively and sometimes heated, but the participants agreed that a large proportion of such deaths shared many features, and that a common cause or mechanism might be involved. To focus attention and research activity on this problem, it seemed important to have a name for this group of cases. It was also necessary to provide a definition that would provide a basis for making this a certifiable cause of death. As chair of a small panel that considered these issues, I suggested SIDS as the most satisfactory designation. This term was accepted by the panel, and subsequently in a plenary session. It was incorporated into the title of the published proceedings of the conference,10 and rapidly achieved wide international acceptance.

More difficult than the name was the problem of definition. This led to lengthy and heated discussion, both in the small group and later in plenary sessions.10 There was agreement the definition should be made as general as possible, pending further research and more widespread recognition of this syndrome. This would encourage use of the diagnostic term without excluding cases that might later be found to be due to the same cause or mechanism.

The result of our discussions was a slightly modified version of the one that we had developed for the Seattle SIDS study. The version adopted at the meeting,10(p18) which will be designated as the 1969 definition, is as follows: “The sudden death of any infant or young child, which is unexpected by history, and in which a thorough post-mortem examination fails to demonstrate an adequate cause for death.” If a prize were offered for the poorest definition of a disease or disorder in the scientific literature, this one would be a strong contender! It contains no limiting criteria, lists none of the features common to most cases, and suggests that this syndrome is only one of exclusion. At the 1969 meeting, I argued for the inclusion of apparent or presumed onset of the lethal event during sleep in the definition, since in our experience those infants observed to die while awake almost always had a demonstrable cause for death. But some participants objected to this wording because one could not be sure that they were asleep when the lethal event began.

I also argued that a narrower age distribution should be part of the definition, based on the consistent age curves available from several large series from around the world. But it was decided that a specific age limitation was premature and that more data were needed. The Seattle group had included in our working definition the term thorough postmortem examination and case study. However, the words emphasized here by italics were excluded from the 1969 definition, not because we were denying the value of careful case studies, but because the term was considered redundant. It was our opinion that careful consideration of the history and circumstances of death is so fundamental a part of every postmortem examination that it does not require mention. This small deletion proved to be unfortunate, as many subsequent critics assumed that our definition ignored the case history,
clinical data, and death scene investigation, and was based solely on dissection of the body.

Despite the vague and permissive nature of the 1969 definition, it was soon recognized that it yielded consistent epidemiological profiles. Large series from several countries and geographic regions reported a similar age distribution curve, sparing the perinatal period, peaking between 2 and 4 months, and declining rapidly thereafter. Virtually all victims were found dead after being put down to sleep. Other common features included a seasonal distribution, tending to spare the summer months in most years, an association with minor viral inflammation, prematurity, or social disadvantage. A similar degree of uniformity was apparent in the autopsy findings, with most babies having prominent intrathoracic petechiae, pulmonary congestion and edema, minor inflammatory infiltrates usually found in the respiratory tract, fluid blood, and empty urinary bladders. Thus the concept of a true syndrome seemed to be justified when applied to collective series of cases. However, disagreement persisted concerning individual cases.

I had agreed to the less restrictive wording of the 1969 definition, since it seemed reasonable to avoid defining an evolving entity too rigidly until consensus is achieved as to its essential features. I was certain that a more specific definition would soon emerge. How wrong I was! For 20 years this definition remained unchanged, and even now has many opponents. Its weaknesses provided a convenient foundation for those who argued that SIDS was a wastebasket term rather than a clinical and pathological entity. It also facilitated the inclusion of inappropriate cases in the medical literature on SIDS.

THE 1989 DEFINITION

In the late 1980s, the National Institute of Child Health and Human Development (NICHD) consulted a variety of individuals, soliciting suggestions for goals for the first 5 years of the next decade. At the top of my list was the need for a better definition of SIDS, primarily for use as a guide to better reporting of research results. Others must have responded similarly, and in June 1989, a group was convened for this purpose. Only 4 of 12 invited participants were pathologists. Some of the panelists had extensive experience with sudden and unexpected infant death, but others were researchers or clinicians with little knowledge of the issues involved in defining cause of death from autopsy examinations.

My goals for the meeting were to incorporate the most consistent epidemiological features of the syndrome into the definition and to establish the distinction between typical and nontypical SIDS cases for purposes of enhancing the quality of research reports. It seemed desirable to retain a somewhat permissive definition in the death administration and case management arena, but a more restrictive distinction of typical from atypical cases would enhance the quality of research and publication. While there seemed to be considerable support for these principles during the meeting, I was disappointed that the version drafted by the organizers rejected the concept of a stratified diagnostic approach and proposed only slight revision of the 1969 definition. This revised version, which will be termed the 1989 definition, was published in 1991: “The sudden death of an infant under one year of age, which remains unexplained after a thorough case investigation, including performance of a complete autopsy, examination of the death scene, and review of the clinical history.” The limitation of age to the first year of life was the only substantive change from the 1969 definition. Review of the history and examination of the death scene were made explicit rather than implicit, but as noted, these were studies assumed to be part of any adequate postmortem examination when we drafted the 1969 definition. Undoubtedly, the explicit mandate in the 1989 version has led to increased investigation of the death scene, and forensic pathologists internationally deserve credit for their support of this mandate.

I was profoundly disappointed that so little improvement in the definition had resulted from 20 years of intensive research. It is my opinion that the organizers of the 1989 meeting attempted to represent too many disciplines, including individuals with scant experience with the problem of diagnosing causes of death in the forensic setting. Only 4 of the participants, and neither of the co-chairs, were pathologists. Sudden infant death syndrome is a diagnosis made by pathologists, based primarily on autopsy findings and death investigation. It therefore seems appropriate that pathologists should play the leading role in establishing definitional criteria, with other disciplines providing advice and consultation.

AFTER 1989

At an international meeting of professionals and parents, sponsored by SIDS International, held in Sydney, Australia, in 1992, I was asked to chair a panel charged with drafting a more specific set of defining criteria. The panelists consisted of 3 pathologists, a pediatrician, and an epidemiologist, all of whom had extensive experience with this problem. The panel easily achieved consensus on a more restrictive definition of SIDS, with criteria for defining subsets of “typical” and “atypical” cases. But when our recommendation was presented at a large plenary session it engendered heated opposition from the large and diverse audience. Many participants favored reverting to the 1969 definition to maximize access of parents to support groups. The 1989 definition, by excluding deaths beyond the first year, and mandating death scene investigation, was considered too restrictive. A good point that did emerge from this session was the suggestion that the words “typical” and “atypical” be avoided, as these terms could lead to problems in dealing with families of cases deemed “atypical.”

At a final session of that meeting, before a smaller and less diverse audience, I presented a modified version of the panel report, replacing the term “typical” with “Category I,” and “atypical” with “Category II.” I also suggested a third category, intended for epidemiological purposes only, in countries or jurisdictions where autopsies were difficult or impossible to obtain. The intended application of this scheme was to develop a 2-tiered definitional approach. A generic designation was suggested for administrative and management purposes, with
more specific criteria identifying Categories I, II, and III SIDS, intended primarily for research and statistical purposes. This proposal was only a recommendation made at a small session and had no effect on the general use of the 1969 and 1989 definitions.

A revised version of my 1992 proposal is presented in the Table. This approach uses a generic definition intended for death administration purposes and a list of more specific criteria that could enhance the quality of research and publication concerning sudden unexpected death in infancy. This scheme is presented only to show one way in which more specific criteria might be inserted into the definition of SIDS. This approach should not severely reduce the number of infants eligible for this diagnosis and would enhance the quality of research by clearly distinguishing the more typical, or classic, SIDS case from the remainder. The practicability of distinguishing classic from nonclassic SIDS was nicely described in the study by Haas et al.15 These authors demonstrated, among other things, that risk factors for SIDS are more prevalent in equivocal cases than in the more classic ones.

The definitional approach proposed in the Table would by no means represent a final solution to the problem of defining SIDS. More specific criteria for certain subsets of SIDS should evolve over time. However, the present needs of research and case management demand better tools than the 1969 and 1989 definitions.

**WHERE DO WE GO FROM HERE?**

I suggest the appointment of a relatively small ad hoc panel, composed mainly of pediatric and forensic pathologists with extensive experience in dealing with sudden infant death. This panel should represent major pediatric and forensic pathology organizations and would be enhanced by international representation. Representatives of other disciplines, especially statistics and epidemiology, could participate as consultants, but the final wording of the definition would be the responsibility of pathologists. Such a group, if not too large or disparate in nature, should be able to achieve this goal in a relatively short time. The ability of such a panel to achieve consensus was demonstrated by a study involving 5 experienced forensic and pediatric pathologists who reviewed 838 candidate SIDS cases that were included in a large NICHD collaborative study of epidemiological factors in SIDS.13 A high degree of consensus was achieved by this panel. Their work resulted in a histopathology atlas illustrating the typical features of SIDS and of some of the entities that exclude that diagnosis. Perhaps the most useful feature of this atlas is the inclusion of a variety of microscopic abnormalities that were deemed to

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**Proposed Definitional Approach of Sudden Infant Death Syndrome (SIDS)**

### Generic Definition*

The sudden and unexpected death of an infant younger than 1 year and usually beyond the immediate perinatal period, which remains unexplained after a thorough case investigation, including performance of a complete autopsy and review of the circumstances of death and of the clinical history. Onset of the lethal episode was presumably during sleep (ie, the infant was not known to be awake). Minor inflammatory infiltrates or other abnormalities insufficient to explain the death are acceptable.

### Subset Definitions†

#### Category I SIDS

An infant death that meets the generic criteria and also meets all of the following standards:
- Age between 3 weeks and 8 months
- No similar deaths in siblings, close genetic relatives, or other infants in custody of same caregiver‡
- No evidence indicative of significant trauma, abuse, neglect, or accident
- No evidence of unexplained moderate or severe stress in thymus, adrenals, or other organs and tissues§
- Intrathoracic petechial hemorrhages are a supportive but not an obligatory or diagnostic finding

#### Category II SIDS

An infant death that meets the criteria for Category I SIDS except for 1 or more of the following features:
- Age younger than 1 year but outside the 3-week to 8-month range
- Similar deaths in siblings or other close genetic relatives that are not considered suspicious for infanticide (genetic consultation indicated)
- Inflammatory changes or other abnormalities somewhat greater than usual for Category I but not sufficient to be an unequivocal cause of death
- Cases in which accidental asphyxia is considered possible but not certain:
  - Depending on specific features of each case and the preference of the certifying pathologist, such cases can be designated as Category I or II SIDS, or as an undetermined cause. A diagnosis of suffocation or asphyxia in a case that would otherwise fit Category I SIDS should be made only with strong supporting evidence. Sometimes infants may, during a death struggle, get into situations that falsely suggest mechanical asphyxia.

#### Category III SIDS

While performance of a complete autopsy is a mandatory prerequisite to a diagnosis of SIDS, in some developing nations, religious groups, or economic settings, the performance of autopsies is difficult or impossible. Category III SIDS is suggested solely for purposes of developing statistical data from such situations and is intended to apply to those cases that seem to fit the generic criteria for SIDS but in which no autopsy is performed. It should not be considered an acceptable alternative to autopsy in most developed societies.

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*For purposes of death certification and administration.
†For research and statistical purposes.
‡While true SIDS can on rare occasions recur in the same sibship or in custody of the same individual, such cases should be earmarked for careful consideration of genetic, environmental, or inflicted causation.
§For infants found in extremis who are partially resuscitated and die later and who seem otherwise to fit the criteria for Category I SIDS, the designation temporarily interrupted SIDS is appropriate. In these cases inflammatory changes, stress effects, and other pathologic features related to postevent alterations need not disqualify consideration for Category I SIDS.
be consistent with SIDS. The scheme suggested in the Table might be useful as a starting point for the deliberations of the proposed SIDS Definition Panel.

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