Sudden infant death syndrome (SIDS) or cot death: A review

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Summary

Sudden death of an infant is not an uncommon event and when there is no adequate cause to explain the death it poses a diagnostic dilemma for the clinician, and makes discussion of the cause of death with parents quite difficult and unsatisfactory. Sudden infant death syndrome (SIDS) or cot death is a diagnostic entity which can be applied in such a case. It is one of the most common causes of infant deaths world-wide in the postneonatal period, from 1 month to 1 year of age. Features which may assist the clinician to arrive at such a diagnosis are discussed in this paper.

Since the first sign of SIDS is death, leaving the clinician with no opportunity to intervene, attention has been focussed on whether or not the infants prone to suffer from this tragic demise can be identified and preventive measures instituted to save them. In this respect, intensive and extensive research efforts over the last two decades have led to what is now commonly referred to as the Apnee Hypothesis for SIDS, and the resultant current intervention method of home apnea monitoring. The apnea hypothesis and other aetiological factors that may help predict infants at risk are discussed.

Finally, current management of infants at risk is discussed for completeness since the facilities for such management are not yet in common use in Nigeria and the West African sub-region, and the level of literacy may severely limit the size of the population that could benefit from such management even if the facilities were available.

Keywords: Sudden, Infant, Death, Syndrome, SIDS, Crib.

Résumé

La mort subite du nourrisson n’est pas un événement rare, mais, elle constitue un dilemme diagnostique pour le clinicien et read très difficile et peu satisfaisante toute discussion avec les parents sur ce qui aurait pu engendrer ce décès, dans la mesure ou personne ne peut en établir une cause précise. Le syndrome de la mort subite du nourrisson représente une entité diagnostique applicable dans ce cas. Car, il représente une des plus communes des causes de la mortalité infantile pendant la période postneonatale, c’est-à-dire, entre 4 et 52 semaines de naissance dans le monde entier. Les traits pouvant aider le clinicien a aboutir a un tel diagnostic font l’objet de discussion dans cette communication.

Puisque le tout premier signe du syndrome ce la mort subite du nourrisson, c’est la mort immédiate, ce qui n’accorde pas au clinicien l’opportunité d’intervention, on a mis l’accent, dans cette communication, sur la possibilité d’identifier au préalable tout enfant predispose a ce deces tragique, aussi bien que sur la possibilité d’apporter des mesures preventives pour sauver la vie. A cet egard, des efforts de recherches intensifs et extensifs ont abouti au cours de ces derniersse annees, a ce que l’on appelle aujourd’hui l’Hypothese Apnee par le controle de la mort subite du nourrisson. 11 en resulte aussi une methode moderne d’intervention qui comporte la surveillance d’Apnee dans la famille meme. L’hypothese Apnee et d autres facteurs etiologiques capables d aider a prevoir les nourrissons future victimes de cette horrible maladie enfant l’object de discussion ici.

Enfin, la gestion courante des nourrissons en risque est discutée pour arriver a une therapie complete, etant donne que les facilites pour telle gestion ne sont pas en usage libre au Nigeria aussi bien que dans la sous region de l’Afrique de l’Ouest, ou le niveve de savior et ecriture limite le nombre de ges capables de profiter de cette gestion au sein de la faille, meme si les facilites existent.

Introduction

In Nigeria 85 out of every 1000 live births die within the first year of life1. Though there may be no accurate data categorizing the exact causes of death in these infants, it is reasonable to assume that a good number of them will have no identifiable cause of death even after very thorough investigations, including post-mortem examinations, have been carried out. Diagnosis in such cases poses a problem to the clinician and the anticipated informed and professional discussion of the cause of death with the victim’s parents becomes very difficult and unsatisfactory. Lack of diagnosis may lead to accusations and finger pointing as to who killed the infant by witch-craft. It is for these reasons that the diagnosis of sudden infant death syndrome (SIDS) (also called Cot or Crib Death) becomes very important as it would be helpful in explaining the death to the parents. SIDS is responsible for 40–50% infant deaths between 1 month and 1 year of age in developed countries2.

SIDS is defined as “the sudden death of an infant which is unexpected by history and in which a thorough post-mortem examination fails to demonstrate adequate cause of death”3. The phrase “unexpected by history” means that there was no history of any antecedent illness prior to the event and the infant has been apparently quite healthy with normal growth and development.

This type of unanticipated death of an apparently healthy infant is quite devastating to parents, and in an effort to understand or explain the tragedy, accusations and counter-accusations, finger-pointing at their parents, relatives, neighbours and friends may arise regarding the cause of death particularly in the West African sub-region. The clinician should understand the phenomenon of SIDS and be able to explain it as a possible cause of death in such cases to the bereaved parents to assist them to understand the nature of the death and perhaps to minimise or even eliminate any social problems it may create for them by suspecting someone of foul play or witch-craft in the death of their infants. Parental grief reactions to sudden and unexpected deaths have been reported to include shock, disbelief, denial, negativism, hostility, anger, self-reproach and
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guilt. A clinician who is well informed on SIDS will be able to provide a warm, caring professional counselling which may ameliorate the psychological burden of guilt or self-reproach of either or both parents arising from their presumption that it may be something they did or did not do that caused or contributed to the death of their infant. The cause of the guilt reactions is lack of information, and prevention of guilt is possible through informed and compassionate counselling by a clinician who understands the phenomenon of SIDS. The first objective of this paper is to review basic information about SIDS to give the clinician a better understanding of the phenomenon.

One of the frustrating and unique features of SIDS is that it leaves the clinician with no opportunity to intervene except to make diagnosis of cause of death. This uniqueness has led to intensive search for factors that could identify infants at risk for SIDS thereby allowing preventive measures to be instituted. This search has resulted in the hypothesis that prolonged sleep apnea is an important etiological factor in the pathogenesis of SIDS. Subsequently home apnea monitoring was instituted as a preventive measure and has become a multi-million dollar industry in the USA, Europe and other advanced countries.

The second objective of this paper is to review the information on the factors associated with high risk for SIDS and the basis for the hypothesis that prolonged sleep apnea should be considered a significant factor in the etiology of SIDS. The current intervention measures are also discussed for completeness.

Epidemiology

SIDS is a world-wide problem and constitutes one of the most common causes of infant death in the post-natal period from 23 days to 12 months of age. Incidence rate varies in different countries and racial groups.

Practically all cases of SIDS deaths occur within the first year of life with peak incidence at 2–3 months of age. Death due to SIDS is quite rare under 2 weeks and after 6 months of age. In other words most infants who will die of SIDS will do so within the age range of 1–6 months. Subsequent siblings of SIDS victims have a higher incidence that is at least 5 times that of all live births. However it has been shown that if the effect of maternal age and birth rank are removed the incidence in siblings will not be significantly different from that in the general population. The incidence in twins is not higher than in non-twins, but nine pairs of twins have been reported to have died from SIDS simultaneously. Eight pairs were reported in the USA and one pair from Belgium.

Diagnosis

By definition SIDS is a diagnosis of exclusion, that is, a thorough investigation and post-mortem examination must have excluded all possible causes of sudden death before the diagnosis of SIDS can be made. Extensive post-mortem examination have shown that there are certain pathological findings that are seen in infants who died of SIDS. These abnormal pathological findings include:

- Increased pulmonary arterial smooth muscle
- Increased right ventricular muscle mass
- Increased extra-medullary hematopoiesis
- Brain stem gliosis
- Retention of adrenal brown fat
- Abnormal carotid body size
- Intrathoracic petechiae
- Hyperplastic adrenal chromaffin tissue.

These findings which are also tissue markers for hypoxia are seen in infants who died of hypoxic diseases such as cyanotic congenital heart disease. These became the first pathological support for the hypothesis that recurrent sleep apnea may play an important role in the pathogenesis of SIDS. Ideally, diagnosis of SIDS should be made by post-mortem examination by a forensic pathologist. However, where the services of a pathologist is not always available, the clinician who has adequate knowledge of the syndrome can still have very high index of suspicion and make a provisional diagnosis of SIDS based on history and post-mortem external physical examination of the victim's body. A typical presentation is a 6-month old infant who appears apparently healthy, shows normal development but is found dead in the Cot or Crib some hours after being put to sleep. At the time of going to bed there was nothing whatsoever to raise any suspicion of an impending death. An infant who presents in this manner, and in whom external physical examination of the body and thorough history show no possible cause of death, may rightly be suspected to have died of SIDS. When this diagnosis is made it is easier to discuss the death intelligently and professionally with the family and to counsel them. It minimises their misery and grief for the death, and decreases the profound suspicion of foul play or witchcraft that such deaths can elicit in the African society.

Infants at risk

SIDS has perplexed the medical community throughout the world for ages and at various times different etiological factors have been considered as being responsible for these tragic deaths. These factors include: over laying and smothering by the mother; infantilicide and child abuse; gastrointestinal reflux; allergic reactions; neurological damage at birth; infection, cardiorespiratory abnormalities, and in the Nigerian context, witchcraft. While the theoretical basis of some of these etiological factors may be more plausible than others, there has not been established any direct cause and effect relationship between SIDS and any of these factors. It is possible that SIDS may have multiple etiologies which acting in concert or separately may result in SIDS death. This possibility is reflected in its being regarded as a syndrome.

SIDS gives the clinician no opportunity to intervene in favour of the victim and therefore the only desirable and effective approach for the clinician is to identify infants at risk of dying from SIDS and to institute preventive measures to abort it. Unfortunately there are no physical examinations or reliable tests on the basis of which the clinician can predict an infant who may or may not die of SIDS.

The recognition of these facts has led to a flurry of research efforts throughout the world in the past two decades to identify the infant at risk. In prospective studies, a host of factors have been identified as being associated with or predictive of infants who are at risk for and/or later died of SIDS. These factors include young maternal age, birth rank, poor social conditions, prematurity, low birth weight, asphyxia and neurological damage at birth, subsequent sibling of a SIDS victim, and cardiorespiratory abnormalities such as prolonged QT interval, excessive periodic breathing and prolonged sleep apnea. Two groups of researchers have even gone further to develop predictive scoring systems based upon epidemiological, physical, and physiological characteristics of a larger series of neonates who later died of SIDS. However, these scoring system has been found to have high false-positivity, and low specificity to be truly predictive.

One of the predictive factors mentioned above, respiratory abnormalities, has received very intensive and extensive research activities throughout the world resulting in numerous
publications in the past two decades. This resulted from the so-called 'Apnea Hypothesis' for SIDS which states that infants who die of SIDS have recurrent prolonged sleep apnea, and hypoxic episodes during these events. The implication of prolonged sleep apnea in the pathogenesis of SIDS is derived from two important studies: The first was by steinshneider in 1972 in which he recorded recurrent prolonged apnea during sleep studies of some infants who later died of SIDS. He then theorized that repeated prolonged sleep apnea in these infants produced chronic hypoxia and that during these vulnerable period one of these episodes will become terminal from which the infant never recovers but goes on to die. The second group of studies were by Naeye and others where in an effort to find pathological support for the chronic hypoxia theory they carried out post-mortem examinations on SIDS victims. From these examinations they identified eight tissue markers of chronic or recurrent hypoxia in these victims (listed earlier in this paper). These findings lent strong support to the hypothesis that recurrent prolonged sleep apnea may play an important role in the etiology of SIDS.

In other word, any infant who has significant apneic episodes during sleep is considered to be at high risk for SIDS, and the infant at the highest risk is the "Near-Miss" SIDS infant. A "Near-Miss" SIDS infant is defined as an infant who suffered prolonged sleep apnea leading to cyanosis and limpness, and requiring vigorous stimulation and/or mouth-to-mouth resuscitation too resume breathing again and in whom no underlying cause is found after thorough investigation. In other words the infant seemed to have died suddenly and unexpectedly but was brought back to life by timely intervention. Currently infants with near-miss SIDS or aborted SIDS are referred to as having infantile apnea.

Following the apparent acceptance of recurrent prolonged sleep apnea as important in the pathogenesis of SIDS, infants who were deemed at risk for SIDS were studied for respiratory abnormalities with pediatric sleep pneumographs, or polygraphic recording of heart rate and respirations during sleep at various SIDS/Apnea Monitoring Programme Centres in the US, UK and other countries. Those infants who were found to have abnormal recordings, namely excessive periodic breathing or prolonged apnea with or without cyanosis and/or bradycardia, were sent home on home apnea impedance monitors equipped with an alarm device as a preventive measure; after thorough investigation in the hospital to rule out treatable causes of the abnormalities. Periodic breathing is defined as an event of 3 episodes of apnea of 3 or more seconds in duration interrupted by respirations lasting 20 seconds or less. It is abnormal if an infant spends more than 3.5% of pneumograph recording or sleep time in this pattern of breathing. It is well known that normal full term infants rarely have apneic episodes lasting up to 15 seconds. An infant with apnea of greater than 15 seconds duration with or without bradycardia was regarded as abnormal. Apnea of any duration accompanied by significant bradycardia for age was also regarded as abnormal. Any infant who showed any of the above abnormal respiratory patterns and/or bradycardia was regarded as at high risk for SIDS and was a candidate for home apnea monitoring. Siblings of SIDS victims and premature infants with respiratory instability at the time of discharge were also candidates for home monitors. Bradycardia was defined for age as equal to or less than 80 beats per minute for age 0–1 month, 70 beats per minute for 1–3 months olds and 60 beats per minute for ages greater than 3 months for a duration of 10 seconds in each insurance.

### Management of infants at risk

After evaluating an infant at risk to ensure that there are no treatable or untreatable pathological conditions that could be responsible for the respiratory abnormalities, a home apnea monitoring is prescribed for the infant. Prior to discharge from the hospital the parents and caretakers receive detailed and extensive instructions on (a) how to use the monitor properly including response to monitor alarms, (b) Mouth-to-mouth resuscitation techniques and (c) Record keeping of events to aid the clinician in evaluating the progress of the infant. Full explanations are also given to them as to why the monitor is being prescribed and the criteria for discontinuing it. Sometimes when the respiratory abnormalities are quite severe, the infant is placed on some respiratory stimulants such as theophylline at a dose of 5–7.5mg/kg/day with serum theophylline level maintained around 11 micrograms/dL.

It is generally agreed that the use of some apnea monitors may abort prolonged sleep apnea but it has remained controversial as to whether or not it prevents SIDS. SIDS deaths are not uncommon amongst infants placed on home monitors in many of the Apnea Monitoring Programmes in the USA and other countries.

### Conclusion

It is sad to note that after decades of intensive and extensive research and millions of dollars spent, the etiology of SIDS remains elusive and the prediction of the infant at risk remains unsatisfactory. The medical community world-wide remains challenged to solve the riddle of what causes SIDS and which infants are at risk.

But until further studies convincingly prove otherwise; infants who have significant cardio-respiratory abnormalities should be regarded as at high risk and appropriate preventive measures including home apnea monitoring, where available, should be instituted. Similarly, even in the absence of post-mortem examination for definitive diagnosis, provisional diagnosis of SIDS should be made based on the characteristics enumerated earlier in this paper. This will assist the clinician in explaining the death to the parents and to eliminate or at least minimize wrongful accusation of someone for the death of the infant.

Sudden infant death syndrome or Cot death is a diagnostic entity listed as No. R-95 in the international statistical classification of disease (ICD) 10. It is recommended for use when appropriate.

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### References


