Sudden Unexplained Death in Children: A Selected Annotated Bibliography

Sudden death in toddlers associated with developmental abnormalities of the hippocampus: a report of five cases.

Sudden unexplained death in childhood (SUDC) is the sudden death of a child older than 1 year of age that remains unexplained after review of the clinical history, circumstances of death, and autopsy with appropriate ancillary testing. We report here 5 cases of SUDC in toddlers that we believe define a new entity associated with hippocampal anomalies at autopsy. All of the toddlers died unexpectedly during the night, apparently during sleep. Within 48 hours before death, 2 toddlers had fever, 3 had a minor upper respiratory tract infection, and 3 experienced minor head trauma. There was a history of febrile seizures in 2 (40%) and a family history of febrile seizures in 2 (40%). Hippocampal findings included external asymmetry and 2 or more microdysgenetic features. The incidence of certain microdysgenetic features was substantially increased in the temporal lobes of these 5 cases compared with the temporal lobes of 39 (control) toddlers with the causes of death established at autopsy (P < 0.01). We propose that these 5 cases define a potential subset of SUDC whose sudden death is caused by an unwitnessed seizure arising during sleep in the anomalous hippocampus and producing cardiopulmonary arrest. Precipitating factors may be fever, infection, and/or minor head trauma. Suggested risk factors are a history of febrile seizures and/or a family history of febrile seizures. Future studies are needed to confirm these initial findings and to define the putative links between sudden death, hippocampal anomalies, and febrile seizures in toddlers.

Full-text available at: www.pedpath.org/ (not a U.S. government site)

Hemoglobin F in sudden infant death syndrome: A San Diego SIDS/SUDC research project report.

Whether levels of fetal hemoglobin (HbF), a possible marker of antecedent hypoxemia, are increased in Sudden Infant Death Syndrome (SIDS) compared to controls is unresolved. Our aims are to: (1) Compare percent fetal hemoglobin (%HbF) levels in SIDS and control cases, and (2) compare our findings with those reported in previous studies. Using Triton-acid-urea gel electrophoresis and quantitative densitometry, %HbF was determined in whole blood specimens obtained at autopsy from SIDS and control
cases accessioned into the San Diego SIDS/SUDC Research Project database. The SIDS and control cases were not different with respect to mean age, gender, gestational age, method of delivery, birth weight, or mean autopsy interval; %HbF levels in SIDS and control cases were not significantly different. Given that our results were obtained using optimal methods in well-defined SIDS and control cases, we concur with others that %HbF is not elevated in SIDS.

Full-text available at www.sciencedirect.com (not a U.S. Government site)


We report a 9-year-old, previously healthy girl who died suddenly and unexpectedly and was found at postmortem examination to have a cardiac rhabdomyoma, megalencephaly, and an involuting adrenal ganglioneuroma. Her death was possibly caused by a fatal cardiac arrhythmia resulting from interference of the ventricular septal rhabdomyoma with the cardiac conduction fibers. Her extended family history included a variety of disorders, including cleft lip and palate and ill-defined cardiac and neurologic diseases. The constellation of her autopsy findings suggested a diagnosis of tuberous sclerosis, for which there are gene defects that can be identified in surviving family members.

Full-text available at: www.pedpath.org/ (not a U.S. government site)


Sudden unexplained death in childhood (SUDC) is rare, with a reported incidence in the United States of 1.5 deaths per 100,000 live births compared with 56 deaths per 100,000 live births for sudden infant death syndrome in 2001. The objectives of this study include a proposal for a general definition for SUDC and presentation of 36 cases of SUDC and 14 cases of sudden unexpected death in childhood. Cases were accrued through referrals or unsolicited via our Web page (http://www.sudc.org ). Our analyses tentatively suggest a SUDC profile characterized by cases being 1 to 3 years in age, predominantly male, and frequently having a personal and family history of seizures that are often associated with a fever. A history of recent minor head trauma is not uncommon. They are usually born at term as singletons and occasionally have a family history of sudden infant death syndrome or SUDC. Most are found prone, often with their face straight down into the sleep surface. Minor findings are commonly seen at postmortem examination but do not explain their deaths. Comprehensive review of the medical history and circumstances of death and performance of a complete postmortem examination including ancillary studies and extensive histologic sampling of the brain are critical in determining the cause of death in these cases of sudden unexpected childhood death. Legislation enabling research...
and formation of a multicenter research team is recommended to unravel the mystery of SUDC.


The aim of the present study was to determine the incidence of various causes of sudden unexpected child deaths (SUCD) and to assess the importance of an autopsy in predicting the likelihood of finding a cause of death. A retrospective analysis of autopsy findings in 97 cases of SUCD between the ages of 0--11 years was undertaken at the Council of Forensic Medicine, Ankara during a 5-year period (1995--2000). Cases were classified as explained causes (80.42 per cent) and sudden infant death syndrome (SIDS) (19.58 per cent). A total of 25.77 per cent of the deaths occurred in the neonatal period, 45.31 per cent of them in the first year of life and the remaining 28.86 per cent after 1 year of life. The causes of neonatal deaths were respiratory pathology (five cases), birth complications (four cases), gastrointestinal pathology (one case), homicide (10 cases), and SIDS (five cases). The incidence of SIDS in the newborn period was 33 per cent. The incidence of unexplained causes of deaths in the postneonatal period was 31 per cent and the causes of deaths were respiratory pathology (15 cases), aspiration (five cases), gastrointestinal pathology (four cases), SIDS (14 cases), and other causes (four cases). The study of an entire population provides more reliable data regarding causes of sudden unexpected child deaths than does the study of small groups and it is also recommended that in addition to a through evaluation, a detailed autopsy must be performed for each case in experienced centers.


Tumors are rare causes of sudden death in infancy and early childhood. The goals of this study were to determine the types and frequency of the tumors associated with sudden death occurring in cases between birth and age 3 years. The San Diego Sudden Infant Death Syndrome/sudden unexplained death in childhood (SUDC) Research Project database and the literature were reviewed retrospectively. Sixty-eight cases, with the most (84%) affecting the heart and brain, were identified. Tumors are a rare but significant cause of sudden death in infancy and early childhood, and their diagnosis may have significant genetic implications for planning future pregnancies. The diagnosis of these lesions can be established only after thorough postmortem examination.

Sudden unexplained death (SUD) claims over 4000 persons between the age of 1 and 22 each year in the United States. Nearly half of all pediatric SUD cases have a normal structural autopsy evaluation and are dismissed without a diagnosis. With the discovery of the genetic basis for potentially fatal arrhythmias associated with the inherited long QT syndrome (LQTS), postmortem molecular diagnosis of this disorder is possible. The authors describe the results of a molecular autopsy performed on a 17-year-old boy found dead in bed. A novel clinical test involving an epinephrine challenge in the decedent's mother implicated a potential defect in the phase 3 potassium current encoded by the gene KVLQT1. Exon-specific amplification by polymerase chain reaction and direct DNA sequencing of KVLQT1 revealed a 5-base pair deletion in the genetic material recovered from the decedent's paraffin-embedded heart tissue. The ability to perform molecular autopsies on archived necropsy material undoubtedly will transform the forensic evaluation of SUD. The combination of catecholamine provocation testing in survivors and a postmortem LQTS gene analysis may unmask families with "concealed" LQTS and establish the cause and manner of death in SUDS.

Full-text available at: www.amjforensicmedicine.com (not a U.S. government site)