Sudden Unexplained Death in Childhood. An Audit of the Quality of Autopsy Reporting

Abstract:
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Introduction
Sudden unexplained death in childhood (SUDC) is defined as sudden death of a child over one year of age which remains unexplained after a thorough case investigation, including review of the clinical history and circumstances of death, and performance of a complete autopsy with appropriate ancillary testing. In contrast to sudden infant death syndrome (SIDS), referring to infants less than one year of age, SUDC is not widely studied. Our aim was to audit the quality of autopsy reporting in Ireland in cases of SUDC, and to compare it with the quality in cases of SIDS.

Methods
The database of the National Sudden Infant Death Register (NSIDR) in Ireland was interrogated and all cases registered as SUDC were reviewed covering a fifteen-year period were limited to those aged <5 years. It is common practice both nationally and internationally for statistics relating to paediatric deaths to be divided into the following categories: <1 year, 1-4 years, 5-9 years, etc, the rationale being to separate preschoolers from older children. Cases of sudden death in older children are currently not notified to the register, hence our limitation to those <5 years of age. The SUDC cases from this period were compared with SIDS cases matched for year of death and age at death. The two groups of SIDS and SUDC cases were reviewed and a modified Rushton score (MR score) was calculated for each case. The objective scoring system for autopsy quality was employed by Rushton, to assess neonatal pathology services in the UK in the 1980s with a maximum MR score of 700, and an arbitrary minimumscore of 300 (43%).

Results
There were 48,530 live births in Ireland in 1995, increasing to 74,728 live births in 2009. The total number of live births between 1995 and 2009 was 899,226. During this period there were 45 deaths registered as SUDC (age range 52-152 weeks) with no autopsies performed by paediatric pathologists. Autopsy reports were available in 43/45 (95%) of SUDC cases. The MR scores for these SUDC cases were inadequate using CESDI guidelines. Overall MR scores were higher in the SIDS cases, with 29/43 (67%) cases obtaining the minimum arbitrary score (MAS) of >300 compared to 25/43 (58%) of SUDC cases. Paediatric pathologists carried out examinations. Rushton’s comment that “the inadequate or badly performed autopsy has shown that autopsy quality, assessed using MR scores, was higher in SIDS cases than in SUDC cases, was better if performed by paediatric pathologists, and improved after the publication of the Royal College of Pathologists Guidelines on autopsy procedure. The results of this study on autopsy quality reporting is comparable to that of other studies and in the Northern region of the UK. Porter and Keeling reported an improved yield of new information when paediatric pathologists carried out examinations. Rushton comment that the inadequate or badly performed autopsy is probably worse than no autopsy at all since it may lead to a false sense of security for the parents” is still applicable.

Discussion
The aim of this study was to audit the quality of autopsy examination in SUDC cases as compared to SIDS cases as it occurs in children over 1 year of age. SUDC cases were reviewed covering a fifteen-year period were limited to those aged <5 years. Cases of sudden death in older children are currently not notified to the register, hence our limitation to those <5 years of age. The SUDC cases from this period were compared with SIDS cases matched for year of death and age at death. The two groups of SIDS and SUDC cases were reviewed and a modified Rushton score (MR score) was calculated for each case. The objective scoring system for autopsy quality was employed by Rushton, to assess neonatal pathology services in the UK in the 1980s with a maximum MR score of 700, and an arbitrary minimum score of 300 (43%).

Figure 1: Breakdown of SUDC Autopsy Investigations

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Discussion
The aim of this study was to audit the quality of autopsy examination in SUDC cases in Ireland and compare with SIDS cases over a 15 year period. In contrast to adult deaths the cause of death in young children and infants is frequently elusive and additional investigations are often required to exclude disease and clarify the cause of death. Our study has shown that autopsy quality, assessed using MR scores, was higher in SIDS cases than in SUDC cases, was better if performed by paediatric pathologists, and improved after the publication of the Royal College of Pathologists Guidelines on autopsy procedure. The results of this study on autopsy quality reporting is comparable to that of other studies.

SUDC is rare. The incidence in the USA is 1.3/100,000 live births. We, the Irish incidence of SUDC has varied between...
1995 and 2009 with an extrapolated mean rate of 5.1/100,000 live births. The differences in these figures might be explained by different reporting and recording practices. Our recently published paper on SUDC epidemiology suggests an increase in rates of SUDC. The definition of SUDC by Krous et al states that a complete autopsy with appropriate ancillary testing must be performed. However our study has illustrated in many cases that the autopsy is not sufficiently thorough, and ancillary testing is not always performed. The range of ancillary tests available is growing rapidly. In cases of SUDC the Royal College of Pathologists recommends routine autopsy organ weights and measurements along with wide range of molecular testing (congenital lqng GT syndrome, catecholaminergic polymorphic ventricular tachycardia, Brugada syndrome, short QT syndrome). Tester et al suggests that up to a third of sudden death after the 1st year of life may stem from a lethal cardiac channelopathy in addition to 10% of SIDS cases, a figure which is higher in SIDS cases >6-months. Since the epidemiological profile, risk factors and pathogenesis of SUDC are not fully understood, standardisation of autopsy procedure will be crucial in adding to our knowledge and understanding of this entity as well as providing important information for future ancillary and genetic testing. This audit of the quality of autopsy reporting has shown that autopsies performed by paediatric pathologists are superior and guidelines issued by the Royal College of Pathologists contribute to improving quality. We aim to raise the profile of SUDC as a diagnosis, and recommend that all paediatric autopsies be referred to specialist centres where RCPath Guidelines are adhered to. Centralisation of the paediatric autopsy service will lead to an increased workload in regional and tertiary referral centres and may require more trained personnel but will ultimately result in improved service provision.

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References

3. National Sudden Infant Death Register Ireland 2009