Part I

Introduction

1

Sudden pediatric death: an overview

Introduction

There have been considerable developments in our understanding of pediatric natural diseases, with many complex genetic links being discovered. More than 900 disease genes have been documented, each of which has from one to many hundreds of mutations. This now places considerable responsibility on pediatric and forensic pathologists to correctly identify diseases that may present as sudden and unexpected childhood deaths, as family screening and genetic counseling may be required (Dietz & Pyeritz, 1994; Goodwin, 1997; Gregersen, Andresen & Bross, 2000).

The pathologist also has to continually evaluate non-inflicted and inflicted trauma, liaising with coroners, product safety experts, and community groups dealing with childhood injury prevention. Preventive pathology refers to this type of activity, with information being taken from the morgue back to the community for use in educational and intervention programs (Byard, 1999; Byard, 2000; Rivara, Grossman & Cummings, 1997a; Rivara, Grossman & Cummings, 1997b). Pathological evaluation of cases of sudden childhood death involves, therefore, considerably more than simple dissection and slide examination.

A question that sometimes arises concerns the definition of terms such as "sudden" and "unexpected," but as long as it is stated clearly how the words are being used then there really should not be any problems in terminology.

How sudden is "sudden?"

The definition of sudden death is quite variable, with different authors setting limits of zero, one, six and 24 hours from the time of onset of symptoms and signs to the time of death. There has again been considerable flexibility in the inclusion of cases in this edition, both from the literature and from personal files, as too rigid an adherence to definitions is impractical and may result in the exclusion of important disease entities. However, generally victims have either been completely well or have been suffering from only an apparently minor illness. If they did have a major illness, then they had been thought to be stable. The common theme uniting all of these cases is that rapid deterioration occurred, culminating in death, although the unavoidable reliance on the presenting history to make this assessment does leave scope for inaccuracy. Quite often the children described in the text were found dead in bed, or suffered a cardiorespiratory arrest while they were engaged in usual activities.

How unexpected is "unexpected?"

It can be argued that the term "unexpected" might not be appropriate to describe death in children who have diseases that are known to be lethal. If this philosophy is applied too rigidly, however, it would be impossible to call a death unexpected in any child suffering from a number of conditions such as Marfan syndrome, tetralogy of Fallot, sickle cell

disease, aortic stenosis, myocarditis, and bronchopulmonary dysplasia, to name but a few. The only way to avoid the problems associated with this form of semantic tangle was to avoid the term "unexpected" and to concentrate instead on deaths that occurred suddenly. In this way, cases of sudden death due to fungal thromboemboli in immunocompromised leukemic patients, or due to massive intracranial hemorrhage in patients with brain tumors, could be discussed without having to go into an excessive analysis of whether the case was truly "unexpected." The common feature of these cases is that death occurred before it was anticipated.

What constitutes being "well?"

A child has been accepted as being well, or only mildly unwell, if neither the parents nor the attending physician felt that there was any evidence of serious disease of a type that would require immediate hospitalization or emergency treatment. This will not always be accurate, particularly in cases of parental neglect and in cases of inflicted injury.

Should conditions be described if sudden death has been reported only in adults, or if sudden death in childhood is only a theoretical possibility?

If the clinicopathological features are otherwise similar in all age groups, then it appears reasonable to accept that these entities may be potential, albeit rare, causes of sudden pediatric death.

Overview

Cases fall into three broad groups:

1 Apparently completely well infants and children who suffered an unexpected cardiac arrest/collapse and died within hours, or who were found dead in bed. Examples include certain types of congenital cardiac defects, cerebral hemorrhage, trauma, and sudden infant death syndrome (SIDS).
2 Infants and children who were considered to be mildly unwell and who presented in a similar manner to the first group. Examples include a variety of diverse infectious diseases, such as viral myocarditis, bacterial meningitis, and epiglottitis. A number of infants who succumb to SIDS would also be included in this group, as a history of low-grade fever, upper respiratory tract infection, and non-specific malaise is often elicited. This group also includes (a) children who had minor illnesses completely coincidental to the underlying lethal process, (b) infants who exhibited only relatively minor symptoms and signs of a serious disease, and (c) those in whom major symptoms and signs were either missed or deliberately ignored, as in cases of inflicted injury and neglect. On occasion there may be an additive effect of an acquired disease to a previously established abnormality with lethal consequences, for example an infant with an anomalous coronary artery circulation who decompensates when the added stress of anemia exacerbates myocardial hypoxia.
3 Children with a known serious but stable condition who suddenly die. Examples include children with asthma and epilepsy.

Detailed tables in the following chapters list conditions that are associated with sudden death in the pediatric age group. Some of these conditions are unique to early childhood, usually causing death before the second decade is reached, while others are found more often in later adolescence. Examples of this age-related variability in mortality include certain congenital cardiovascular anomalies, which may manifest within the first few months of life, compared with sudden death in asthma, which tends to occur in later childhood.

Frequency

The incidence of pediatric sudden death is difficult to determine precisely, since there is variability from community to community, and from year to year in the same population (Denfield & Garson, 1990). In addition, death certificate diagnoses are known to be inaccurate, particularly in cases that do not come

to autopsy. It has been estimated that sudden natural death in the age range of 1–20 years ranges from 1.1 to 13.8 per 100,000 of the pediatric population per year (Denfield & Garson, 1990; Morentin *et al.*, 2000), accounting for 2–5% of deaths in that group (Berger, Dhala & Friedberg, 1999; Driscoll & Edwards, 1985; Molander, 1982). As for trauma, a study from Melbourne, Australia, reported an overall mortality rate from injury in children aged from 0 to 14 years of 10.6 per 100,000 per year (Nolan & Penny, 1992). This compares with a lethal injury rate of 30.3 per 100,000 per year that has been quoted in the USA for the age range 0–19 years (Guyer & Gallagher, 1985). The sudden death rate is higher in boys than in girls (Gillette & Garson, 1992).

Causes of sudden death

SIDS remains the most common cause of sudden and unexpected death in infancy (one week to one year of age) despite dramatic falls in rates over the past decade (Byard, 1991; Côté, Russo & Michaud, 1999). Over one year of age, the major causes of sudden natural death are malignancies, congenital anomalies, and infections (Corey Handy & Buchino, 1998; Denfield & Garson, 1990; Vetter, 1985).

Infectious disorders that are most commonly associated with sudden pediatric death include myocarditis, meningitis, epiglottitis, bronchopneumonia, bronchiolitis, tracheobronchitis, septic shock, gastroenteritis, and peritonitis. Acute infections were responsible for the greatest number of deaths in a study of 207 sudden deaths in the age range 1–21 years, the median age of death being 4.3 years (Neuspiel & Kuller, 1985).

Cardiovascular anomalies or diseases rank along with infections as major causes of sudden death in childhood, although the age of death tends to be older than for infections, the median age being 16.2 years in the study of Neuspiel & Kuller (1985). The usual cardiovascular disorders are hypertrophic cardiomyopathy, dilated cardiomyopathy, aortic stenosis, congenital coronary artery anomalies, tetralogy of Fallot, Ebstein anomaly, pulmonary hypertension, mitral valve prolapse, conduction disturbances, and Eisenmenger syndrome (Klitzner, 1990). The relative percentages of these entities depend on the particular series. For example, while Topaz & Edwards (1985) found mitral valve prolapse to be equal in frequency to myocarditis (24%), with only 4% of cases having aortic stenosis, Lambert and colleagues (1974) found an incidence of 18% for aortic stenosis with only one case of "myxomatous" mitral valve disease. Sudden death following surgery in children with congenital heart defects, particularly tetralogy of Fallot and transposition of the great vessels, also accounts for a significant number of cases (Vetter, 1985).

The most likely etiology for sudden death in young athletes or children engaged in strenuous physical activity is cardiovascular. For example, Maron and colleagues (1980) found a structural cardiovascular anomaly in 28 of 29 competitive athletes aged between 13 and 30 years who died suddenly. The most common abnormality in that series was hypertrophic cardiomyopathy (48%), contrasting with a similar study by Corrado and colleagues (1990), in which the most common abnormality found was right ventricular dysplasia (27%). Whether this variability in results reflects a difference in the populations studied, a difference in diagnostic practice, or a difference in the pattern of referral of autopsy cases for review is difficult to ascertain.

Other "common" causes of sudden pediatric death include epilepsy, intracranial hemorrhage, and asthma (Kitada, Nakagawa & Yamaguchi, 1990; Neuspiel & Kuller, 1985; Norman, Taylor & Clarke, 1990). Less usual causes involve hematologic, gastrointestinal, genitourinary, metabolic, endocrine, genetic, and immunologic disorders.

Use of the term "accident" has been discouraged in recent years by some clinicians as it is argued that most injuries are predictable and preventable (Davis & Pless, 2001). Certainly there is merit in this view, but not all injuries are preventable and most people have a clear concept of what is meant when the term "accident" is used. Additionally, standard categories for the manner of unnatural death in forensic terminology are homicide, suicide, accident, and

undetermined. For these reasons, the term "accidental death" has continued to be used in this edition until a comparable and generally accepted term has been devised.

Deaths due to accidents (i.e. traumatic episodes arising from non-inflicted injury) in children are most often caused by traffic accidents and drowning; other causes of injury in the pediatric age range include burns, scalds, falls, poisoning, choking, and non-accidental trauma (Nolan & Penny, 1992; Norton, 1983).

The percentage of cases of sudden death in childhood that remain unexplained even after an autopsy has been conducted will vary greatly depending on the rigor with which the postmortem examination has been conducted and the significance that is subsequently attached to the findings. The cases that are left serve to highlight the inadequacy of investigations into childhood death and the insensitivity of standard pathological techniques in ascertaining the cause of certain pediatric fatalities.

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Part II

Unintentional trauma

Gin Alley, a nineteenth-century etching by Hogarth, which demonstrates vividly the dangers to infants from trauma associated with poverty and poor living conditions.

2

Accidents

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Introduction

Accidents account for nearly half of all deaths of individuals aged from one to 24 years old in the USA – 25,814 of 54,239 in 1987 (*Accident Facts*, 1990). Accidents cause only 3% of deaths in infants less than one year of age, but cause 71% of deaths in those aged 15–19 years. At all ages motor vehicle crashes account for a significant percentage of non-intentional deaths (Johnston, Rivara & Soderberg, 1994), from 23% in infants under one year to 80% of deaths in the 15–19-year group. These results are summarized in Table 2.1. Generally, the term "adolescence" refers to the 13–19-year-old age group and "young adulthood" after this.

In the *Accident Facts* survey, suffocation was the second leading cause of death in infants under one year of age. In older age groups, drowning was next to motor vehicle accidents (MVAs). Firearm deaths were the third major cause of accidental deaths in 15–19-year-olds in the USA. About half of drownings and motor vehicle-related deaths involve alcohol (Rosenberg, Rodriguez & Chorba, 1990).

Injury mortality for those aged between one and 19 years from 1984 to 1986 in the USA exceeded that of other countries, with an overall mortality rate of 30.5/100,000 of the population, compared with 26.1/100,000 for Canada, 22.3 for Norway, 21.5 for France, 15.6 for England and Wales, and 13.1 for the Netherlands (Williams & Kotch, 1990). In Australia more recent data on accidental death show a mortality rate of 10.8/100,000 for children under four years, 5.3/100,000 in five- to nine-year-olds, 6.4/100,000 in 10–14-year-olds, and 29.2 in 15–24-year-olds (National Injury Surveillance Unit, 1995). Again, drowning and motor vehicle deaths account for the majority (65%) of cases (Pitt, Balanda & Nixon, 1994).

In North Carolina Runyan and colleagues (1985) found fires and burns to be the leading cause of death due to non-inflicted injuries in the under four years age group, followed by motor vehicle accidents, pedestrian injuries, suffocation, and drowning. In the 15–19 years age group, motor vehicle fatalities and drowning were the major causes of death, with suicides and homicides also accounting for many deaths in that age group. Of 267 children with life-threatening injuries treated at a regional trauma center, 55% were injured in motor vehicle accidents, 27% were hurt in falls, 10% were due to sports and other injuries, and 8% were assault victims. In 75% of the motor vehicle accidents, the child was a pedestrian. Fifty one percent of those injured had involvement of one organ system, 29% two organ systems, and 20% three or more organ systems. Fourteen of the 17 deaths resulted from head injury, primarily accounting for the mortality of 6.7% (Colombani *et al.*, 1985).

In Peclet and colleagues' study (1990) of 3472 children up to 14 years of age admitted to an urban trauma center, traffic-related injuries accounted for 29% of all patients. School-age children (5–10 years) tended to have falls and traffic-related injuries. Child abuse, drowning, and penetrating injuries accounted for 5% of all injuries but 40% of all deaths. Seventy five percent of the deaths in this study involved head injury.

Table 2.1. Types of accidental deaths by age (percentages) (derived from *Accident Facts*, 1990)

	Age (years)				
	<1	1–4	5–9	10–14	15–19
Percentage of deaths due to accidents	3	39	48	48	55
Motor vehicle accidents	23	34	55	59	80
Drownings	13	23	15	13	6
Fires, burns	11	24	13	5	1
Firearms	<1	1	3	7	3
Poisoning	2	1	1	<1	1
Falls	4	2	1	1	1
Mechanical suffocation	19	2	2	3	1
Other accidents	28	13	9	11	7

There is marked variability between communities in the rates of childhood gunshot deaths. For example, in 1992 firearms were responsible for 72% of homicides in the 10–14-years age group in the USA (O'Donnell, 1995), but in Australia such deaths are rare. A significant, although small, number of childhood firearm homicides in a study from South Australia were perpetrated by parents in murder/suicide situations (Byard *et al.*, 1999b).

Accidental deaths occurring during the first year of life have been described by Corey and colleagues (1992) and also in a study produced by the New York State Department of Health (1991). Corey and colleagues reported 36 unintentional deaths in this age group over an 11-year period. Nineteen (53%) died from asphyxia: eight were in mechanically unsafe sleeping environments, six were overlaid, three suffocated in plastic bags, and two asphyxiated on foreign objects. Four (11%) drowned, three (8%) each died in house fires, of scald burns, and in motor vehicle accidents, two (6%) died of hypothermia, one (3%) fell from a height, and one (3%) died of alcohol toxicity. The New York study found that injuries were the third leading cause of death after perinatal conditions and congenital anomalies. Between 1984 and 1988 there were 343 deaths due to injury in this series. Homicide accounted for 116 (34%) deaths. Of the remaining 227 deaths, the intention was unknown in 71, and these were not described further. Of the remaining 156 deaths, 39 (25%) were caused by fires and 60 (38%) were caused by asphyxia: 27 by suffocation, 22 by choking on food, and 11 by choking on a foreign object. Nineteen (12%) died in motor vehicle accidents, 15 (10%) drowned, and 23 (15%) died of miscellaneous causes. Infants and young children are particularly vulnerable to injury or death due to their high dependency, and may be at particular risk if a carer dies or becomes incapacitated. For example, three deaths occurred in South Australia in children aged one, two and a half, and three years, respectively, due to dehydration when children became trapped in houses with carers who had died (Byard, 2002) (Figure 2.1).

Figure 2.1 Handprints left on a closed bedroom door by a 2.5-year-old boy who was unable to reach the handle. Both the boy and his one-year-old brother died of dehydration in the room after their mother had died in bed of epilepsy.

Table 2.2. Details of 369 cases of accidental death of children aged from 0 to 16 years in South Australia from 1963 to 1996

Category	*n*	Male	Female	Age range	Mean
Motor vehicle accident					
Unspecified	66	35	31	1–14 years	6 years 5 months
Passenger	27	13	14	1 month–13 years	4 years 9 months
Pedestrian	69	47	22	6 months–15 years 11 months	7 years 5 months
Cyclist	24	17	7	5 years 7 months–15 years 8 months	10 years 2 months
Motorcyclist	3	2	1	6 years 6 months–13 years	9 years 10 months
Drowning	63	46	17	3 months–12 years 8 months	3 years 6 months
Cot/bed asphyxia	40	25	15	1 month–4 years	10 months
Fire/flame/scald	24	11	13	1 year 1 month–14 years 2 months	5 years 1 month
Foreign body aspiration	14	12	2	3 months–8 years	2 years 2 months
Poisoning	10	5	5	1 year 1 month–6 years	2 years 9 months
Farm-related*	10	6	4	2 years 6 months–11 years 6 months	4 years 10 months
Electrocution	6	4	2	2 years–12 years	5 years 8 months
Other asphyxia	5	5	0	3–9 years	5 years 5 months
Sporting	4	4	0	3 years 9 months–14 years 10 months	9 years 3 months
Falls	2	1	1	3 years 2 months–14 years	8 years 7 months
Industrial	1	0	1	9 years 6 months	9 years 6 months
Train	1	1	0	12 years 4 months	12 years 4 months
Total	369	234	135	1 month–15 years 11 months	5 years 2 months

*In addition, there were five other deaths on farms included in other categories: two asphyxias, one drowning, one incineration, and one motorcycle accident.

As a background to this chapter, two studies were undertaken. In the first, pediatric autopsies (age 16 years and younger) performed in Grand Rapids, Michigan, USA, from 1983 to September 1991 were reviewed. A total of 568 children were autopsied, including 330 natural deaths, 168 accidents, 42 homicides, and 15 suicides. The manner of death in 13 cases was indeterminable. The 168 accidental deaths serve as the focus for this chapter. Under the Michigan Medical Examiner Law, all sudden and unexpected deaths and all unnatural deaths (accidents, suicides, and homicides) must be investigated by a physician medical examiner, who, as part of his or her investigation, may order an autopsy. These 168 autopsies were thus ordered by the investigating medical examiner. There were 44 cases of craniocerebral trauma (defined as fatal injury to the head or neck), 38 cases of multiple injuries (defined as serious or fatal injuries involving at least two different areas, e.g. head and chest), 30 cases of asphyxia, 26 drownings, 22 fire deaths, and eight miscellaneous causes of death.

In the second study, a review of 369 cases of non-intentional childhood deaths occurring in South Australia over a 34-year period from 1963 to 1996 was undertaken (Byard, 2000; Byard, 2001). In South Australia, which has a population of approximately 1.5 million people, all violent or unusual deaths are reported to the State Coroner. Children's autopsies were performed at the Forensic Science Centre or the Adelaide Children's Hospital. Of the 369 cases, there were 189 motor vehicle accidents, 63 drownings, 40 sleeping accidents, 24 fire/flame/scalds, 14 foreign body aspirations, 10 poisonings, six electrocutions, five miscellaneous asphyxias, four sports-related deaths, two falls, one industrial death, and one train death. Fifteen of the deaths occurred on farms. The details are summarized in Table 2.2.