Pediatric Deaths Attributable to Complex Chronic Conditions: A Population-Based Study of Washington State, 1980–1997

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Abstract. ***Objectives.*** **Advances in medical technology and public health are changing the causes and patterns of pediatric mortality. To better inform health care planning for dying children, we sought to determine if an increasing proportion of pediatric deaths were attributable to an underlying complex chronic condition (CCC), what the typical age of CCC-associated deaths was, and whether this age was increasing.**

Design. **Population-based retrospective cohort from 1980 to 1997, compiled from Washington State annual censuses and death certificates of children 0 to 18 years old.**

Main Outcome Measures. **For each of 9 categories of CCCs, the counts of death, mortality rates, and ages of death.**

Results. **Nearly one-quarter of the 21 617 child deaths during this period were attributable to a CCC. Death rates for the sudden infant death syndrome (SIDS), CCCs, and all other causes each declined, but less so for CCCs. Among infants who died because of causes other than injury or SIDS, 31% of the remaining deaths were attributable to a CCC in 1980 and 41% by 1997; for deaths in children 1 year of age and older, CCCs were cited in 53% in 1980, versus 58% in 1997. The median age of death for all CCCs was 4 months 9 days, with substantial differences among CCCs. No overall change in the age of death between 1980 to 1997 was found (nonparametric trend test).**

Conclusions. **CCCs account for an increasing proportion of child deaths. The majority of these deaths occur during infancy, but the typical age varies by cause. These findings should help shape the design of support care services offered to children dying with chronic conditions and their families.** ***Pediatrics*** **2000; 106:205–209;** ***complex chronic conditions, population-based study, pediatric deaths, mortality trends.***

ABBREVIATIONS. CCC, complex chronic condition; *ICD-9, International Classification of Diseases, Ninth Revision;* SIDS, sudden infant death syndrome.

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What kinds of children are likely to require end-of-life care in the future? Certainly, the experience of dying has altered drastically over the past century, shifting from infectious causes to chronic conditions, from younger to older, from swiftly to slowly.[1–3] Although these and other trends have motivated a burgeoning concern for adult end-of-life care, far less attention has been focused on changing patterns of childhood mortality and the implications of these changes for pediatric palliative care services. Although several longitudinal population-based studies have informed us regarding trends in specific causes such as injury,[4–6] cancer,[7] respiratory distress,[8] unexpected sudden natural death,[9] and birth defects or genetic diseases,[10] none have examined the contribution to pediatric mortality of all the complicated and chronic conditions that, at some point in their progression, might warrant palliative care.

Classifying a variety of pediatric health states as complex chronic conditions (CCCs) has 2 major advantages when examining childhood mortality. First, a wide array of discrete conditions account for many pediatric deaths, so that without aggregation into larger groups, patterns of an evolving forest of pediatric morbidity and mortality are lost for diagnosis-specific trees. Second, children with CCCs constitute an evolving population with shared but changing needs. As medical technology shifts the epidemiology of serious pediatric disease from acute to chronic illness, problems once rapidly lethal now linger, causing long-term disability and sometimes a protracted process of dying.[11]

For these reasons, we conducted a population-based retrospective cohort study of all deaths among children aged 0 days to 18 years in Washington State from 1980 to 1997, specifically to determine if an increasing proportion of childhood death is attributable to an underlying CCC, estimate the typical age of death, and test whether the age of death attributable to these conditions is rising.

METHODS

Data Sources

All deaths occurring in Washington State to individuals <1 years old from 1980 to 1997 were identified from vital statistics computer files. Throughout this period, death certificates noted the Underlying Cause of Death with a 4-digit *International Classification of Diseases, Ninth Revision* (*ICD-9*) code, as well as sex, race, date of birth, date of death, year of death, and city of residence. For rates of deaths, the year-specific sizes of the

population aged 0 to 18 were obtained from 1980 and 1990 census data along with annual intercensal and postcensal estimates calculated by the Washington State Office of Financial Management.

Selection of CCC *ICD-9* Codes

Before the analysis, we selected the *ICD-9* codes that would represent a CCC as an underlying cause of death. We began with a working generic definition[12,13] of a CCC as:

> *Any medical condition that can be reasonably expected to last at least 12 months (unless death intervenes) and to involve either several different organ systems or 1 organ system severely enough to require specialty pediatric care and probably some period of hospitalization in a tertiary care center.*

To operationalize this definition, we identified a list of possible CCCs, based on clinical knowledge and experience, for several physiologic systems and for malignancies and other congenital and genetic anomalies. We then reviewed studies that have used similar methodology to determine which codes they had employed to identify high-cost conditions suitable for a carve-out financing mechanism[14] and to assess the role of congenital defects in pediatric hospitalizations.[15] Having assembled a list of possible conditions, we examined the corresponding *ICD-9* codes, leading to the selection of diagnostic codes that, in the setting of identifying the underlying cause of death, would be highly specific for identifying individuals with a CCC. We then queried the death certificate data with this preliminary classification to assess whether any underlying causes of death appeared among those classified as not having a CCC were in fact CCCs; this led only to the inclusion of dwarfism (not elsewhere classifiable), 259.4; diaphragmatic hernia, 553.3; and kyphoscoliosis and scoliosis, 737.3 in the itemized list of CCC *ICD-9* codes. The final categories and codes, the basis for all subsequent analyses, are presented in Table 1. Of note, the nature of the conditions drove the classification, so that a childhood death occurring even on the first day of life could still be classified as attributable to a chronic complicated condition.

To determine the sensitivity of the single underlying cause of death *ICD-9* code to detect a case with a CCC, and to assess whether any secular trend in coding practice had occurred,[16] we used the 20 multiple causes of death coding fields that were added to each record from 1988 onward. Each record was classified as either having a CCC *ICD-9* code in any of the multiple cause of death fields or not. With this multiple-code assessment as the criterion gold standard, the single underlying cause of death field code was 87% sensitive for detecting a CCC code in the record, and no secular trend was detected (χ^2 trend of sensitivity; $P = .7$). Given the sufficiently high and consistent level of sensitivity, we felt justified in using the single underlying cause of death *ICD-9* code for classification of cases, which enabled the study to extend back to 1980.

Statistical Analyses

Comparisons of proportions used the χ^2 test. Trends in proportions were examined using the χ^2 linear test for trend, while change in the age of death were tested using Cuzick's nonparametric test for trend.[17] At the outset, significance was set at P values $<.05$ and 95% confidence intervals were calculated. Analyses were performed using Intercooled STATA 6.0 for Windows (Stata Corporation, College Station, TX).

TABLE 1. Categories of CCCs and Corresponding Four-Digit *ICD-9* Codes

Categories	Subcategories	*ICD-9* Codes
Neuromuscular	Brain and spinal cord malformations	740.0–742.9
	Mental retardation	318.0–318.2
	Central nervous system degeneration and disease	330.0–330.9, 334.0–334.2, 335.0–335.9
	Infantile cerebral palsy	343.0–343.9
	Muscular dystrophies and myopathies	359.0–359.3
Cardiovascular	Heart and great vessel malformations	745.0–747.4
	Cardiomyopathies	425.0–425.4, 429.1
	Conduction disorders	426.0–427.4
	Dysrhythmias	427.6–427.9
Respiratory	Respiratory malformations	748.0–748.9
	Chronic respiratory disease	770.7
	Cystic fibrosis	277.0
Renal	Congenital anomalies	753.0–753.9
	Chronic renal failure	585
Gastrointestinal	Congenital anomalies	750.3, 751.1–751.3, 751.6–751.9
	Chronic liver disease and cirrhosis	571.4–571.9
	Inflammatory bowel disease	555.0–556.9
Hematologic or immunologic	Sickle cell disease	282.5–282.6
	Hereditary anemias	282.0–282.4
	Hereditary immunodeficiency	279.00–279.9, 288.1–288.2, 446.1
	Acquired immunodeficiency	0420–0421
Metabolic	Amino acid metabolism	270.0–270.9
	Carbohydrate metabolism	271.0–271.9
	Lipid metabolism	272.0–272.9
	Storage disorders	277.3, 277.5
	Other metabolic disorders	275.0–275.3, 277.2, 277.4, 277.6, 277.8–277.9
Other congenital or genetic defect	Chromosomal anomalies	758.0–758.9
	Bone and joint anomalies	259.4, 737.3, 756.0–756.5
	Diaphragm and abdominal wall	553.3, 756.6–756.7
	Other congenital anomalies	759.7–759.9
Malignancy	Malignant neoplasms	140.0–208.9, 235.0–239.9

RESULTS

Demographics

From 1980 through 1997, a total of 21 617 children <19 years old died in the state of Washington (Table 2). Of these, 60% were males, 81% were white, and 55% were <1 year old. Death certificates cited a CCC code as the underlying cause of death in 24% of all cases. Females were more likely to have a CCC code, while children 1 to 12 months and 15 to 18 years, as well as Native American and black children, were all less likely to have a CCC code ($P < .001$ for each comparison).

During the study period, the total population aged 0 to 18 years rose from 1.2 million in 1980 to 1.58 million in 1997. Based on these figures, approximately 24.2 million pediatric person-years accumulated during this 18-year interval, yielding an overall rate of 89.4 pediatric deaths per 100 000 children per year.

Causes of Death

Table 3 summarizes the identified underlying causes of death as both proportions of all deaths and as death rates. Trauma accounted for 29.7% of the deaths and <.1% remained unassigned because of missing data. Nearly one-quarter (24.3%) of deaths were coded as having a CCC as the underlying cause. Among those deaths with a CCC coded as an underlying cause, cardiovascular and malignancy codes were most common; hematologic and immunologic were the least.

Trends in Cause of Death

Trends in the annual rates of death attributable to CCCs, the sudden infant death syndrome (SIDS), injuries, and all other causes differed markedly, as shown in Fig 1. In all 4 categories, the rates of death per 100 000 children 0 to 18 years old decreased between 1980 and 1997: from 25.8 to 15.8 with CCC codes; from 15.2 to 5.5 for SIDS; from 39.6 to 20.6 for injury; and from 45.7 to 18.0 without CCC codes.

Given the study's purpose of examining the potential future need for pediatric supportive care services, we eliminated cases attributable to SIDS and injury from our subsequent analyses, so as to focus on the changing contribution of CCCs to medical causes of death. In 1980, 36.1% of medical deaths were classified as having a CCC cause in 1980; by 1997, this had risen to 46.8% (χ^2 for trend; $P < .001$).

Quite different trends exist for deaths occurring before and after 1 year of age, as shown in Fig 2. The proportion of medical deaths during infancy attributable to a CCC increased, chiefly because the non-CCC deaths have declined substantially over this 18-year period. In 1980, 30.6% of deaths were coded as attributable to a CCC; by 1997 that proportion had risen to 41.2% (χ^2 for trend; $P < .001$). By contrast, during the same period the majority of deaths after the first birthday were cited as attributable to a CCC, fluctuating nonsignificantly from 52.8% in 1980 to 64.3% in 1985 to 58.1% in 1997 (χ^2 for trend across all 18 years; $P = .4$).

Trends in Age of Death

Considering all deaths attributable to CCCs together, the median age of death was 4 months 9 days, with the 25th percentile occurring on the second day of life, and the 75th percentile at 6 years 3 months of age. The typical age of death varied substantially among the CCC categories (Fig 3). We did not detect any statistically significant change in the age of death from 1980 to 1997 ($P = .7$ for nonparametric trend test).

TABLE 2. Demographics

Feature	Category	With CCC Code	All Deaths
Total number		5247	21 617
Sex	Female	43%	40%
	Male	57%	60%
	Unknown	.1%	.0%
	Total	100%	100%
Age	1 day or less	22%	19%
	2–30 d	18%	12%
	1–12 mo	19%	24%
	1–4 y	14%	11%
	5–9 y	9%	7%
	10–14 y	8%	8%
	15–18 y	10%	19%
	Missing data	.1%	.5%
	Total	100%	100%
Race or ethnicity	White	84%	81%
	Black	5%	7%
	Native American	2%	4%
	Asian or Pacific Islander	3%	3%
	Hispanic origin	5%	5%
	Other	1%	1%
	Total	100%	100%

DISCUSSION

This study of all childhood deaths that occurred in Washington State from 1980 to 1997 revealed that a substantial proportion of pediatric deaths is attributable to an underlying complex chronic condition. This proportion has increased, due mostly to the decline of other causes of death in children under 1 year old. Overall, both the annual number of deaths and the mortality rates have declined markedly, but deaths with an underlying CCC have declined less rapidly than deaths not associated with CCCs. Counter to one of our initial hypothesis, the typical age of death attributable to all CCCs has remained stable.

We interpret these findings cautiously for several reasons. First, our classification of deaths was based solely on the underlying cause of death, a single attribute that may not accurately reflect the complexity or chronicity of illness before death. The underlying cause of death, moreover, was determined by a wide array of health care professionals with minimal standardization,[18] leading undoubtedly to some deaths being misclassified regarding their CCC status. We think, though, that the resulting classification errors would tend to miss cases of CCCs, thereby making the findings of this study conservative. Regarding our failure to detect any systematic change in the typical age of

TABLE 3. Cause of Death *ICD-9* Code Categories

Category	Subcategory	Percent of all 21 617 Deaths	Rate per 100 000 Child-years
Medical	No indication of chronic illness	45.9%	41.1
	SIDS	13.6%	12.1
	IRDS	4.3%	3.8
	Underlying CCC	24.3%	21.7
	Cardiovascular	6.6%	5.9
	Malignancy	5.4%	4.8
	Other congenital or genetic defect	3.8%	3.4
	Neuromuscular	3.8%	3.4
	Respiratory	2.8%	2.5
	Renal	.8%	.7
	Metabolic	.6%	.5
	Gastrointestinal	.3%	.2
	Hematologic or Immunologic	.2%	.2
Trauma	Any form of injury	29.7%	26.7
	Unintentional injury	22.3%	20.0
	Homicide	3.6%	3.2
	Suicide	3.1%	2.8
	Undetermined intent	.8%	.7
Missing data		.1%	.1
Totals		100.0%	89.4

IRDS indicates infant respiratory distress syndrome.

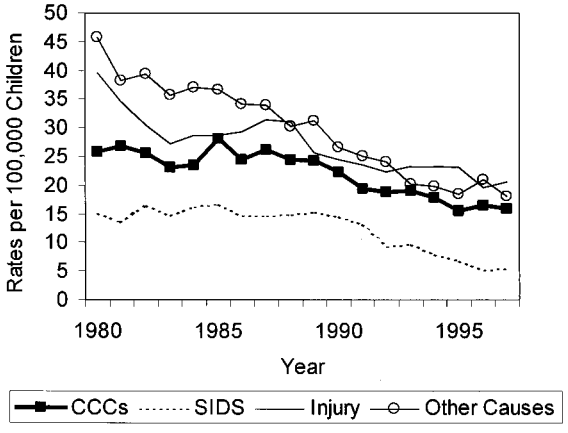

Fig 1. Annual rates of childhood death attributable to CCCS, SIDS, injuries, and other causes.

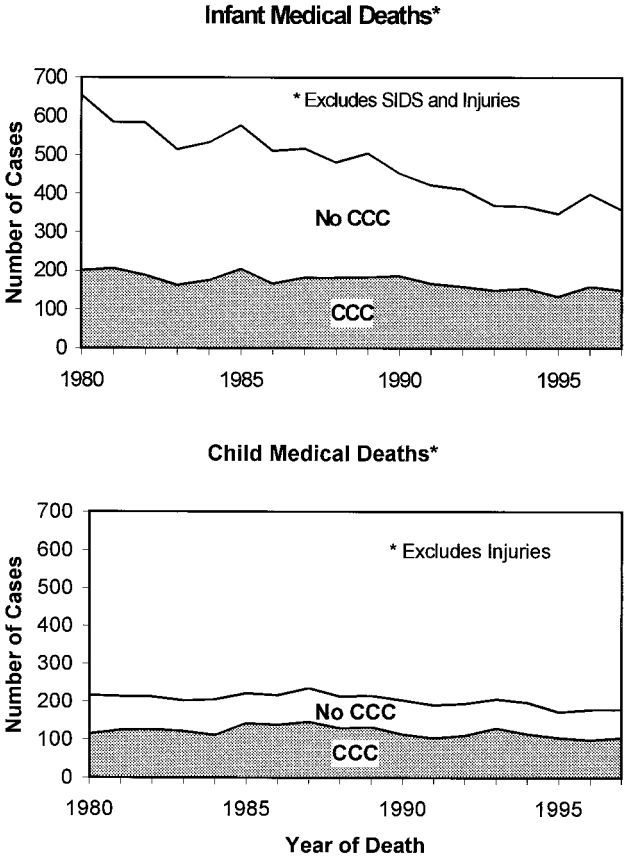

Fig 2. Deaths with or without an underlying CCC.

death, we may have erred by sampling only pediatric-age patients if in fact deaths are increasingly postponed beyond adolescence. Lastly, although tracking the nearly 20-year long experience of an entire state's population, we do not know whether these findings would be seen in other states, which would differ both by the composition of their pediatric populations as well as by the health care system serving children.

Despite these limitations, we believe that the patterns of childhood mortality observed in Washington State have 3 major implications for the planning of pediatric supportive care services for dying children and their families. First, because no single cause of death presents the full picture of how pediatric mortality is changing, the comprehensive planning of these supportive care services will need to use some unifying concept to assess this population and its needs. The CCCs classification that we used, although certainly amenable to improvement, provides one such scheme. The CCC rubric, while enumerating a disparate set of specific conditions, approximates a noncategorical approach to classifying childhood conditions, because these CCCs share common attributes including: management of voluminous information, skillful care planning and coordination, and often the need for extensive child and family support services.

Second, despite these commonalities, pediatric supportive care services must be flexible. Patients

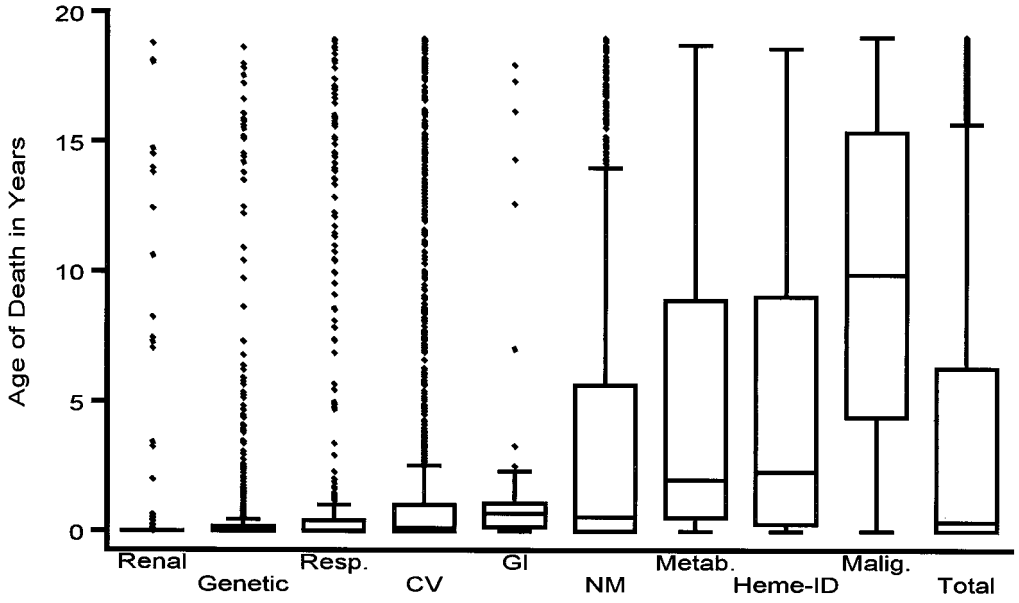

Fig 3. Typical age of death varies by underlying cause.

and families differ by important characteristics that influence needs, such as the underlying condition, the age of the child at the time of death, and the length of time that the family has cared for the child. Not even considering cultural and social features, we found that childhood deaths occurred across the range of these characteristics commonly enough to warrant a broad scope of practice for pediatric supportive care services.

Finally, and most fundamentally, we should no longer assume that most children who die from medical causes would not be suitable for supportive care services. For quite some time, a large proportion of childhood deaths have been attributable to CCCs that well might benefit from palliative, respite, or hospice care, and for infants this proportion is increasing. We should seek to understand the needs of this population far better than we currently do, and then set about meeting those needs.

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