

Deaths Attributed to Pediatric Complex Chronic Conditions: National Trends and Implications for Supportive Care Services

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ABSTRACT. *Background.* Children with complex chronic conditions (CCCs) might benefit from pediatric supportive care services, such as home nursing, palliative care, or hospice, especially those children whose conditions are severe enough to cause death. We do not know, however, the extent of this population or how it is changing over time.

Objectives. To identify trends over the past 2 decades in the pattern of deaths attributable to pediatric CCCs, examining counts and rates of CCC-attributed deaths by cause and age (infancy: <1 year old, childhood: 1–9 years old, adolescence or young adulthood: 10–24 years old) at the time of death, and to determine the average number of children living within the last 6 months of their lives.

Design/Methods. We conducted a retrospective cohort study using national death certificate data and census estimates from the National Center for Health Statistics. Participants included all people 0 to 24 years old in the United States from 1979 to 1997. CCCs comprised a broad array of *International Classification of Diseases, Ninth Revision* codes for cardiac, malignancy, neuromuscular, respiratory, renal, gastrointestinal, immunodeficiency, metabolic, genetic, and other congenital anomalies. Trends of counts and rates were tested using negative binomial regression.

Results. Of the 1.75 million deaths that occurred in 0- to 24-year-olds from 1979 to 1997, 5% were attributed to cancer CCCs, 16% to noncancer CCCs, 43% to injuries, and 37% to all other causes of death. Overall, both counts and rates of CCC-attributed deaths have trended downward, with declines more pronounced and statistically significant for noncancer CCCs among infants and children, and for cancer CCCs among children, adolescents, and young adults. In 1997, deaths attributed to all CCCs accounted for 7242 infant deaths, 2835 childhood deaths, and 5109 adolescent deaths. Again, in 1997, the average numbers of children alive who would die because of a CCC within the ensuing 6-month period were 1097 infants, 1414 children, and 2548 adolescents or young adults.

Conclusions. Population-based planning of pediatric supportive care services should use measures that best inform our need to provide care for time-limited events (perideath or bereavement care) versus care for ongoing

needs (home nursing or hospice). Pediatric supportive care services will need to serve patients with a broad range of CCCs from infancy into adulthood. *Pediatrics* 2001;107(6). URL: <http://www.pediatrics.org/cgi/content/full/107/6/e99>; *chronic disease, causes of death, mortality, cohort studies, terminal care, hospice.*

ABBREVIATIONS. CCC, complex chronic condition; ICD-9, *International Classification of Diseases, Ninth Revision*; CI, confidence interval.

Children who are dying because of complex chronic conditions (CCCs)—such as cancer, cardiac malformations, cystic fibrosis, or neurodegenerative diseases—require special health care services. These supportive care services range broadly, from aggressive control of pain and effective symptom relief, to sensitive spiritual care and grief counseling, to respite and bereavement services for the family. The importance of providing such comprehensive care has recently been underscored by the American Academy of Pediatrics.¹ Accessing such care can be difficult, however, not only because of reluctance to confront dying openly with children and their families or because of a mismatch between the standard aggressive tertiary pediatric care practices and the philosophy of hospice or palliative care, but also because of a welter of logistic obstacles, such as a shortage of health care personnel trained to provide supportive care, inadequate funding, and constraining insurance benefit packages.^{1–5}

As a society, we would be better prepared to address these obstacles effectively if we knew how many children at a given moment need pediatric supportive care services, and how the pattern of need is shifting over time. Ideally, we would acquire this knowledge from population-based prospective surveys of children and their families. Lacking such information, several authors have offered estimates. The statement of the American Academy of Pediatrics states that “53 000 children in the United States die every year from trauma, lethal congenital conditions, extreme prematurity, heritable disorders, or acquired illness.”¹ Such a figure, if taken at face value, assumes that all children who die—even instantaneously or nearly so from trauma or sudden infant death syndrome—have similar needs and would benefit equally from supportive care services. Martinson⁶ put forward a more limited estimate that 4115 children between 1 and 14 years old will need

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hospice services annually, but this was based on data from 1992 only, used a limited number of cause of death conditions as indicating the need for possible hospice care, and did not consider infants.

We sought to help redress these gaps in our knowledge by providing more refined information regarding several attributes of the population potentially in need of pediatric supportive care services. Specifically, avoiding the tendency to exclude infant deaths and to focus exclusively on cancer-related deaths, we estimated the number of children who died annually of cancer and noncancer CCCs in infancy (birth to 1 year of age), childhood (1–9 years old), and the teen and young adulthood stages (10–24 years old). We believed that this data would help us to gauge the need for 1-time services, such as perideath care for the child or bereavement care for the family. We also calculated the age-specific mortality rates over time to account for changes in the age structure of the US population when examining trends. Finally, we estimated the average number of children alive in the 6-month hospice-eligible interval¹ before death attributable to a CCC, who would often need ongoing services, such as home nursing or hospice. These various forms of data—counts, rates, and estimated prevalence—provide related yet different information useful to the planning of health care resources, workforce, and programs to meet the needs of these children and their families.

METHODS

Case Definition

We first constructed a list of CCCs, comprising a broad array of *International Classification of Diseases, Ninth Revision (ICD-9)* codes

for cardiac, malignancy, neuromuscular, respiratory, renal, gastrointestinal, immunodeficiency, and metabolic, genetic, and other congenital anomalies (Table 1). This list represented a minor modification of one that we used previously in a retrospective longitudinal study of mortality patterns in Washington State,⁷ which, in turn, had been based, in part, on studies of hospitalization patterns of children with costly illnesses⁸ and with congenital defects.⁹ We modified the list by including epilepsy and by expanding the range of *ICD-9* codes for the subcategories of mental retardation, central nervous system degeneration and disease, and malignancy.

Data Source

We then compiled data on CCC-attributed deaths to all US residents ages 0 to 24 years from 1979 to 1997. Using mortality data from the National Center for Health Statistics, we estimated the number of deaths occurring in the United States each year to US residents from national death certificate data, and estimated the population from census estimates for people >1 year old and from live-birth data for people <1 year old. Data were obtained from the Centers for Disease Control and Prevention WONDER web site (<http://wonder.cdc.gov>) in the spring of 2000, requesting from the compressed mortality/population file counts of deaths attributed to each of the 9 CCC categories of *ICD-9* codes for each year.

Statistical Analysis

We calculated annual age-specific mortality rates by dividing the number of deaths by the estimated total population for that age group. To determine changes in the annual age-specific number of deaths and mortality rates from 1979 to 1997, we first created a spline for the calendar year exposure variable¹⁰ (with intervals 1979–1982, 1982–1986, 1986–1990, 1990–1994, and 1994–1997), then we fit negative binomial regression models for the number of deaths and the mortality rates for each age category, and finally we took the linear combination of the estimated values of the spline intervals over the entire period. To calculate the average number of children who died because of a CCC within the ensuing 6 months (that is, living in the 6-month life expectancy definition of hospice eligibility as promulgated by governmental

TABLE 1. Categories of CCCs and Specific *ICD-9* Codes

CCC Categories	Subcategories	<i>ICD-9</i> Codes
Neuromuscular	Brain and spinal cord malformations	740.0–742.9
	Mental retardation	318.0–319.0
	Central nervous system degeneration and disease	330.0–337.9
	Infantile cerebral palsy	343.0–343.9
	Epilepsy	345.0–345.9
	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 and dysrhythmias	426.0–427.4, 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
Hematology and immunodeficiency	Sickle cell disease	282.5–282.6
	Hereditary anemias	282.0–282.4
	Hereditary immunodeficiency	279.0–279.9, 288.1–288.2, 446.1
	Human immunodeficiency virus disease	042
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		140.0–239.9

and private health insurance coverage), we assumed that the duration of this hospice-eligible phase comprised half of the lifespan of children who died under 1 year of age (that is, for deaths occurring under 1 day of age, 0.5 of a day was designated as hospice-eligible; deaths between 1 and 6 days of age, 3.5 days; 7–27 days of age, 17 days; 28–364 days of age, 146 days); and for deaths occurring at 1 year of age and older, designating the last 6 months of life (182 days) as hospice-eligible. For each cause of death, we summed these days and divided the total by 365. We performed all analyses with *Stata, Version 6.0*.¹¹

RESULTS

Of the 1.75 million deaths that occurred in 0- to 24-year-olds from 1979 to 1997, 5% were attributed to cancer CCCs, 16% to noncancer CCCs, 43% to injuries, and 37% to all other causes of death (which includes sudden infant death syndrome, infant respiratory distress syndrome, extreme prematurity, pneumonia, and other typically acute causes of death). The proportion of all deaths attributed to each cause varied by age (Table 2). Noncancer CCCs were cited as responsible for one quarter of all infant deaths, 20% of childhood deaths, and 7% of adolescent deaths. Cancer CCCs were cited as the cause of <1% of infant deaths, 11% of childhood deaths, and 6% of adolescent deaths. Injuries accounted for 3% of infant deaths, 47% of childhood deaths, and 76% of adolescent deaths. Among just the CCC-related deaths, the predominant causes of death likewise varied with age (Table 3), with cardiovascular, malignancy, neuromuscular, genetic, and respiratory being the leading causes overall.

Trends in Annual Number of Deaths

From 1979 to 1997 (Fig 1), the annual number of deaths attributable to cancer CCCs has declined 30% for children (95% confidence interval [CI]: 24–36) and 38% for adolescents and young adults (95% CI: 34–42), whereas the annual number of deaths because of noncancer CCCs has declined 33% for infants (95% CI: 31–35), 34% for children (95% CI: 29–38), and 14% for adolescents and young adults (95% CI: 3–23). In 1997, a total of 15 186 children died with CCCs. Examining this figure more closely, cancer CCC-related deaths numbered 154 infants, 1085 children, and 2284 adolescents; noncancer CCC-related deaths numbered 7088 infants, 1750 children, and 2825 adolescents and young adults.

Trends in Mortality Rates

Trends closely paralleling those seen in the annual number of deaths are observed in the changing population-based mortality rates from 1979 to 1997 (Fig 2), namely significantly diminished cancer CCC mortality rates among children (43%; 95% CI: 36–46) and

adolescents and young adults (32; 95% CI: 28–36), and noncancer CCC mortality rates among infants (39%; 95% CI: 37–41) and children (44%; 95% CI: 41–48). Nonsignificant declines are found for the annual cancer mortality rate among infants (8%; 95% CI: 27% decrease to 15% increase) and for the noncancer CCCs mortality rate among adolescents and young adults (6%; 95% CI: 17% decrease to 6% increase). An additional exploratory analysis, using each of the 6 major age groups identified in the compressed mortality file data, revealed as shown in Fig 3 that the rate of decline in the age-specific mortality rates for both cancer and noncancer CCCs displays a U-shaped pattern, with the most rapid fall occurring in the mid-childhood years, and the least rapid decrease or even increase of mortality rate in infancy and the late adolescent years.

Average Number of Hospice-Eligible Children With CCCs

Because most infant deaths occur within hours to days of birth, infants who die because of CCCs spend few days alive in the 6-month period of time during which they would have been eligible for hospice insurance coverage benefits. For this reason, trends in the average number of children alive during this hospice-eligible phase of a terminal illness do not neatly correspond to the patterns seen in annual number of deaths or mortality rates (Fig 4). For both cancer and noncancer CCCs, there are more adolescents and young adults alive who might benefit from supportive care services than infants or children. Indeed, among noncancer CCC-related deaths, the average number of living hospice-eligible adolescents and young adults has—in distinction to the other age groups—remained fairly steady, at 1500 per year. Focusing on the most recent year, in 1997, there averaged 1050 infants living within 6 months of their death because of a noncancer CCC, and another 47 infants living within 6 months of their death because of cancer. For children, the averages were 873 (noncancer CCCs) and 541 (cancer CCCs). For adolescents and young adults, the averages were 1409 (noncancer CCCs) and 1139 (cancer CCCs).

DISCUSSION

We estimate that each year 15 000 infants, children, adolescents, and young adults die from conditions that suggest that they and their families might benefit from pediatric supportive care services. On any given day, 5000 of these patients are living within the last 6 months of their lives. Although the last 2 decades have shown marked medical progress, with

TABLE 2. Number of Deaths by Cause and Age

Cause of Death	Number of Deaths			
	<1 Year Old	1 to 9 Years Old	10 to 24 Years Old	Total
Noncancer CCCs	175 319	43 389	57 286	275 994
Cancer CCCs	3058	24 114	52 108	79 280
Injury	24 006	100 881	620 790	745 677
Other causes	511 651	48 358	91 360	651 369
Total	714 034	216 742	821 544	1 752 320

TABLE 3. Proportion of CCC-Attributed Deaths Attributable to Specific Subtypes of CCCs

	Infants <1 Year of Age	Children 1 to 9 Years of Age	Adolescents and Young Adults 10 to 24 Years of Age	All Ages 0 to 24 Years of Age
Cardiovascular	37%	24%	18%	29%
Malignancies	2%	36%	48%	22%
Neuromuscular	15%	21%	17%	17%
Genetic	22%	5%	13%	13%
Respiratory	15%	4%	10%	10%
Renal	5%	0%	3%	3%
Hematologic and immunologic	1%	5%	3%	3%
Metabolic	1%	3%	1%	1%
Gastrointestinal	1%	2%	1%	1%
Total	178 377	67 503	109 394	355 274

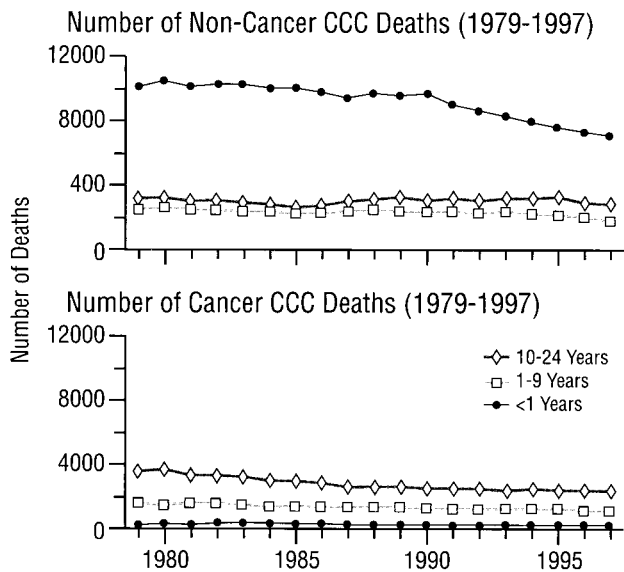


Fig 1. The number of deaths attributed to cancer and noncancer CCCs.

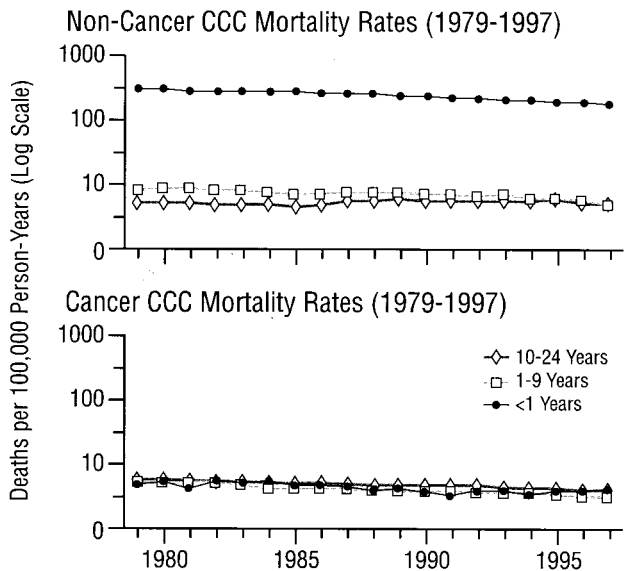


Fig 2. The rates of deaths attributed to cancer and noncancer CCCs.

mortality rates associated with CCCs generally declining across all age groups, the rate of decline is slowest for noncancer CCCs among older adolescents and young adults.

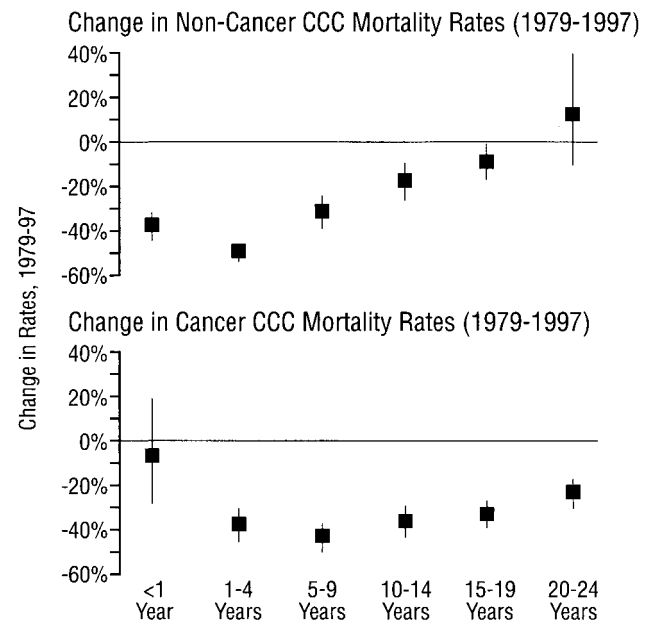


Fig 3. U-shaped pattern of the rate of decline in age-specific CCC mortality rates.

This study, although based on national data over a 19-year period, nevertheless, is limited in several regards. First, the compressed mortality file, although providing nearly 2 decades of information, is based on death certificates, which may assign the cause of death inaccurately, and reports only the single underlying cause of death ICD-9 code from the original death certificate. Second, the changes observed in the annual number of deaths and mortality rates may, in part, be attributable to trends over time regarding what diagnoses were more likely to be cited as the underlying cause. Of note, our study of Washington state pediatric death certificates revealed that the single underlying cause of death ICD-9 code was 87% sensitive for detecting a record that had a CCC code in the other multiple diagnostic ICD-9 code variable fields, and we did not identify a trend over time in the underlying cause of death coding practice.⁷ Third, in this study, we can not determine which children would have benefited from pediatric supportive care services. Instead, we have estimated the average number of children who would have been eligible for such services based on the 'prevailing within 6 months of death' eligibility

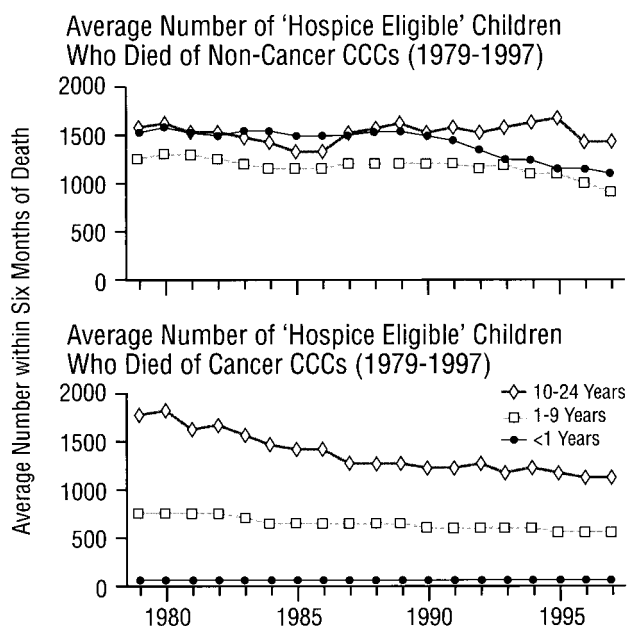


Fig 4. The average number of children with CCCs living within 6 months of their death.

criteria for hospice services, with the assumptions that infants who die from CCCs would benefit from such supportive care services for only half of their lifespan, whereas older children who die from CCCs would benefit for the entire last 6 months of their lives.

With these caveats in mind, several interrelated implications regarding pediatric supportive care services can be drawn from this study. First, the U-shaped relationship between age and the pace with which mortality rates attributed to CCCs are declining suggests that, over time, pediatric deaths attributable to these causes among children 1 to 9 years old are becoming increasingly rare compared with similarly caused deaths among infants, adolescents, and young adults. To the degree that enhanced life-extending technology postpones death into young adulthood, as was suggested by our subanalysis of the noncancer CCC-related deaths, the terminal phase of these illnesses of late adolescents and young adults may fall—and falter—during the precarious transition from pediatric to adult-oriented systems of health care.

Second, because these 2 age groups comprise an increasing proportion of all CCC-related deaths, the palliative and hospice care of pediatric patients might suitably assume 2 quite different forms in the future, each with a differing population of patients and, hence, differing methods of care, organizational structures, and financing mechanisms. One form, addressing the needs of infants born with severe congenital defects or nonviable prematurity, would be a fast-paced enterprise serving many patients, most likely predominantly provided in hospitals and even intensive care units during the neonatal period. The

other form of pediatric hospice care services, attending to the needs of children and increasingly adolescents and young adults with cancers and a host of other life-limiting complex chronic conditions, would be required by fewer patients, have a substantial home-based focus, and conceivably would have more time to explore the values and preferences of the patients and their families, to make decisions regarding the goals and limits of care and to enact these plans.

CONCLUSION

The 5000 infants, children, adolescents, and young adults living today within 6 months of their CCC-related deaths are spread across the nation, with undoubtedly some regions being very sparsely populated by such children. How to provide services at the local level of their home communities will prove a major challenge, similar to the provision of other services for rurally residing children with special health care needs.¹²⁻¹⁴

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