Birth Defects Surveillance: Assessing the "Gold Standard"

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ABSTRACT

Objectives. This study assessed the sensitivity of the Metropolitan Atlanta Congenital Defects Program (MACDP) by capitalizing on the delayed receipt of a data source.

Methods. In 1997, we reviewed the medical records of potential cases from the 1995 birth certificates that had not previously been identified by the MACDP. Capture–recapture methods produced an estimate of total cases.

Results. We identified 1149 infants with defects, including 20 exclusively from birth certificates. The estimated sensitivity of the MACDP when data from birth certificates were included was 86.9% (95% confidence interval [CI] = 80.6%, 91.9%) at 1 year after birth, increasing to 94.8% (95% CI = 90.3%, 97.8%) at 2 years after birth.

Conclusions. The MACDP underestimates defects by 13% at 1 year after birth and by 5% at 2 years after birth. (*Am J Public Health.* 1999;89: 1238–1240)

The Metropolitan Atlanta Congenital Defects Program (MACDP) was established in 1968 as the nation's first multiple-source, population-based birth defects surveillance system with active case-finding.[1] It has been considered the most comprehensive system in the United States[2,3] and has been used as a "gold standard" to evaluate other surveillance systems.[4] While unpublished data from the Centers for Disease Control and Prevention that indicate an overall sensitivity above 99% have been previously cited,[5] the sensitivity of the system has not been objectively assessed. Delayed receipt of the 1995 birth certificates provided a natural experiment to assess the sensitivity of the MACDP and to identify weaknesses in the usual methods of data collection.

Methods

The MACDP conducts active surveillance of all major birth defects among live births and fetal deaths (at 20 or more weeks of gestation) in the 5 counties of metropolitan Atlanta. A birth defect is defined as a structural or chromosomal anomaly that is present at birth and diagnosed before 6 years of age.

Case sources include obstetric and pediatric hospitals, a genetics laboratory, and vital records. At the obstetric hospitals, MACDP abstractors review medical records of all newborns with a defect noted in the logs or disease indices and of infants meeting at least one of the following criteria: birthweight of less than 2500 g, 5-minute Apgar score of less than 7, gestational age of less than 36 weeks, admission to neonatal intensive care unit, any surgery except circumcision, or diagnosis of certain other conditions specified in the MACDP manual.[6]

MACDP abstractors also review the medical records of all infants not currently in the MACDP database whose birth or death certificate indicates a major defect. Although birth certificates are normally reviewed in conjunction with all other sources, an administrative oversight resulted in a late request for the 1995 birth certificates. The abstractors began follow-up of the 1995 certificates in January 1997.

Capture–recapture methods can be used to estimate total incidence and to evaluate the relative contribution of various case sources.[7–10] We used capture–recapture techniques to estimate the true number of birth defect cases in metropolitan Atlanta, using 2 independent sources that each had incomplete case ascertainment: birth certificates and all other MACDP sources. We calculated the 95% confidence intervals for this estimate by fitting a log-linear model to the data and observing the change in the goodness-of-fit statistic that occurred when estimated trial values of the total population were varied.[11] The estimated total number and its 95% confidence interval were then used to assess the sensitivity of the MACDP. We characterized missed cases to identify potentially correctable gaps in MACDP surveillance methods.

Results

Among the 40266 live births in 1995 to Atlanta residents, we identified 1149 cases of birth defects (3%) by using both birth certificates and all other MACDP sources. The birth certificates identified 137 cases, including 20 that were not previously identified by the MACDP (Table 1). Using capture–recapture methods, we estimated that 1322 cases occurred in 1995 (95% confidence interval [CI] = 1250, 1426). At a minimum of 1 year after birth, the estimated sensitivity of the MACDP was 1129/1322, or 85.4% (95% CI = 79.2%, 90.3%), without birth certificates and 1149/1322, or 86.9% (95 % CI = 80.6%, 91.9%), including birth certificates.

A review of disease indices indicated that 12 of the 20 cases identified only by birth certificates (birth certificate–only cases) would probably have been picked up from the abstractors' review of backlogged disease indices. Therefore, we recalculated the capture–recapture estimates by assuming that there were 8 missed cases and estimated that the total number of cases was 1212. The

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sensitivity of the MACDP, including both the 12 "late" cases and the birth certificates, was then 94.8% (95% CI = 90.3%, 97.8%).

The characteristics of the 20 birth certificate–only cases indicate that only 1 met any of the criteria for record review in the MACDP guidelines (Table 2). Non-Hispanic Whites were underrepresented ($P = .02$) in the birth certificate–only cases. In addition, a disproportionately high number of musculoskeletal defects ($P = .16$) and a disproportionately low number of digestive system defects ($P = .08$) were missed.

TABLE 1—Distribution of Found and Missed Cases of Birth Defects, by Source of Report: Metropolitan Atlanta Congenital Defects Program, 1995

Baby's Defect Identified by All Other Sources	Baby's Defect Identified by Birth Certificates		
	Yes	No	Total
Yes	117	1012	1129
No	20	173	193
Total	137	1185	1322

Note. The cases missed by both sources (173) and the total number of infants with defects (1322) were calculated with capture–recapture methodology. Missed cases = (20 × 1012)/117 = 173.

TABLE 2—Comparison of Infants With Birth Defects Identified Only by Birth Certificates (BC Only) and All Infants With Birth Defects Identified by the Metropolitan Atlanta Congenital Defects Program (MACDP) in 1995

	BC Only (%)	MACDP (%)	P[a]
Review criteria			
Birthweight <2500 g	0	297 (28)	.001
Gestational age <36 wks	0	239 (25)	.004
5-min Apgar score <7	1 (5)	115 (11)	.34
Race/ethnicity			
Black	10 (50)	386 (37)	.16
Asian/Pacific Islander	2 (10)	32 (3)	.13
Hispanic	2 (10)	55 (5)	.29
Non-Hispanic White	6 (30)	576 (55)	.02
Affected organ system[b]			
Central nervous system	1 (5)	95 (9)	.45
Facial (ear, eye)	4 (20)	245 (23)	.49
Cardiovascular system	6 (30)	303 (29)	.54
Digestive system	1 (5)	206 (20)	.08
Genitourinary system	5 (25)	316 (30)	.42
Respiratory system	1 (5)	79 (8)	.55
Musculoskeletal system	9 (45)	338 (32)	.16
Integument	3 (15)	134 (13)	.48

[a]One-tailed P values based on Fisher exact test.
[b]Some infants had defects in more than one organ system.

Discussion

The delay in receiving the 1995 birth certificates created a natural experiment to assess the sensitivity of the MACDP. We estimated that when the use of birth certificates was included, the sensitivity of the MACDP was 86.9% at a minimum of 1 year after birth, with the sensitivity increasing over time. The gradual increase in sensitivity was due primarily to late abstracting, but diagnoses and hospitalizations after the initial newborn period also contributed.

Characterization of the birth certificate–only cases revealed differences by race/ethnicity and organ system affected. Digestive system defects were disproportionately low among birth certificate–only cases, probably because of the birth certificate's low sensitivity to many types of digestive system defects that may not be readily apparent at birth.[12] On the other hand, musculoskeletal defects were disproportionately high among birth certificate–only cases, owing primarily to a few cases with "Polydactyly/Syndactyly/Adactyly" noted on the birth certificate. Abstractors may not have initially pursued these cases because postaxial polydactyly type B among Black infants is not monitored by the MACDP because of its high frequency.

The MACDP may be compared with the New York State Congenital Malformations Registry (NYCMR), which uses passive case-finding consisting of case reports from physicians and other health care providers.[13] On the basis of a capture–recapture estimate using data from 1983 to 1986, the sensitivity of the NYCMR without using birth certificates was 86.4%. This estimate is similar to that of the MACDP at a minimum of 1 year after birth; however, the sensitivity of the MACDP increased markedly during the following year because of the active case-finding of the system and some backlogged abstracting.

The capture–recapture methodology used here assumes independent data sources, yet the birth certificates and other MACDP sources probably have positive ascertainment dependence. For example, the most visible and severe defects are likely to be noted both on the birth certificate and in the medical record. This positive dependence means that the capture–recapture estimate tends to be an underestimate of the true prevalence[14] and that we have probably overestimated the sensitivity of the MACDP to some degree.

Conclusion

We estimated the MACDP's sensitivity to be 86.9% when data from birth certificates were included; this figure increased to 94.8% when "late" cases from backlogged disease indices were included. To ensure that the MACDP remains a model program, we must continually improve case-finding. The sensitivity estimates and characterization of birth certificate–only cases may be useful for future decisions about MACDP procedures. Other surveillance systems should also consider employing capture–recapture methodology to assess their sensitivity. □

Contributors

M. A. Honein had primary responsibility for the analysis and interpretation of the data. L. J. Paulozzi had primary responsibility for the conception and design of the project. Both authors made substantial contributions to the writing of the paper.

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Twenty-Year Trends in the Reported Incidence of Mycosis Fungoides and Associated Mortality

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ABSTRACT

Objectives. Patterns of mycosis fungoides incidence and associated mortality in the United States were evaluated.

Methods. Data were taken from the Surveillance, Epidemiology, and End Results cancer registry program and the National Center for Health Statistics.

Results. The incidence rate from 1973 through 1992 was $0.36/10^5$ person-years. The age-adjusted incidence rate ratio of Blacks to Whites was 1.7; that of Asians to Whites was 0.6. There was no evidence of increasing incidence rates during the period 1983 through 1992. Mortality rates declined steadily from 1979 to 1991 and were less heterogeneous geographically than incidence rates. Mortality rate patterns with age, sex, and race were similar to the corresponding incidence patterns.

Conclusions. The incidence rate of mycosis fungoides has stabilized and the mortality rate has declined. For unknown reasons, the disorder varies greatly among demographic and geographic subgroups. (*Am J Public Health.* 1999; 89:1240–1244)

Mycosis fungoides is a form of cutaneous lymphoma. Previous studies of mycosis fungoides suggested that the incidence rate had increased, but details were limited.[1–4] We examined incidence patterns in a substantially larger database than had been used before, and we also assessed mortality rates.

Methods

Our incidence data were derived from the Surveillance, Epidemiology, and End Results (SEER) program of the National Cancer Institute for 1973 through 1992. Histologic confirmation of mycosis fungoides was noted for 97% of the cases registered. For this study, the "Asian" racial group was defined to include persons who were neither White nor Black. Thus, in addition to persons of East Asian background, the group included a modest proportion of persons of Polynesian ancestry and a much smaller number of others, including members of American Indian groups. We combined these diverse groups because of our lack of more specific population data. The mortality data (*International Classification of Diseases, Ninth Revision* [*ICD-9*] codes 202.1 and 202.2) came from routine death certifications reported to the National Center for Health Statistics for the United States for the periods 1979 through 1981, 1984 through 1985, and 1989 through 1991 and from the Current Population Survey of the Bureau of the Census. Incidence rate ratios (IRRs), exact confidence intervals, and *P* values were calculated conventionally.[5,6] Incidence and mortality rates and rate ratios were age-adjusted to the 1970 US standard million population unless otherwise indicated.

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