Incidence of Congenital Anomalies among White and Black Live Births with Long-Term Follow-Up

ROBERTA E. CHRISTIANSON, MA, BEA J. VAN DEN BERG, MD, LUCILLE MILKOVICH, BA, AND FRANK W. OECHSLI, PHD

Abstract: The incidence of congenital anomalies at birth and accumulated to age five years is presented for live-born children in a large prospective study. Congenital anomalies are not all diagnosable at birth; our data demonstrate that the incidence rate increases approximately three-and-one-halffold for Blacks and fivefold for Whites between six days of age and five years of age.

The incidence of congenital anomalies at birth was higher among Black children than White children, but there were no notable differences between the groups in incidence accumulated to age five years. At five years, the incidence rate of severe and moderate (but not trivial) congenital anomalies amounted to 15 per cent; for severe congenital anomalies, 4 per cent. Severe congenital anomalies diagnosed through age five years were observed to have a much higher incidence among children who weighed 2500 gm or less at birth than among those who were heavier. (*Am J Public Health* 1981;71:1333–1341.)

Introduction

Although efforts are being made to standardize information on congenital anomalies, it is widely recognized that the reported incidence of congenital anomalies is subject to considerable variation. The factors primarily responsible for this variation include the definition of congenital anomalies applied, the method of their ascertainment, the length of time the population is under observation, and the ethnic and socio-economic characteristics of the population studied. Longitudinal cohort studies with special follow-up examinations provide high incidence figures for congenital anomalies as contrasted with studies based only on birth certificate information. Even longitudinal studies, however, have demonstrated striking variation in incidence rates because of different definitions of what is included as a congenital anomaly. The study of Myrianthopoulos and Chung[1] observed an overall incidence of congenital anomalies among infants followed for one year of 15.56 per cent, while McIntosh, *et al*,[2] observed an incidence of 7.5 per cent in a cohort of births also followed for one year; the latter study did not include many of the minor anomalies counted in the former study.

The present study illustrates the effect of definition and length of follow-up on the incidence rates observed for White and Black live-born children and documents incidence rates in conjunction with prematurity as defined by birth weight and gestational age.

From the Child Health and Development Studies, School of Public Health, University of California, Berkeley. Address reprint requests to Roberta E. Christianson, Child Health and Development Studies, University of California at Berkeley, 3873 Howe Street, Oakland, CA 94611. This paper, submitted to the Journal September 8, 1980, was revised and accepted for publication May 26, 1981.

Materials and Method

This analysis is based on data from the Child Health and Development Studies (CHDS), a longitudinal study of pregnancy and the subsequent development of the offspring. The women who enrolled in CHDS early in their pregnancies were members of the Kaiser Foundation Health Plan. This prepaid health plan provides comprehensive medical care to the family in any of the Kaiser Foundation clinics. Subscribers to this health plan constitute a predominantly employed urban population representing a wide range of economic, social, and educational characteristics deficient only in extremes. The gravidas enrolled in CHDS, including nearly 100 per cent of those eligible for the study, delivered their infants at Kaiser Hospitals in the San Francisco East Bay Area of California during the years 1959–1967. The children were examined frequently and followed intensively by pediatricians and other specialists in the Kaiser Foundation facilities until the youngest child in the study was five years old. Subgroups of the children were still being seen in 1979/80.

Information on the mothers and children is derived from several sources. The mothers were interviewed extensively early in pregnancy and the complete medical record of both mother and child was abstracted in detail. For each visit and hospitalization a child had at any Kaiser clinic or hospital, from birth until at least five years of age, the physician's diagnosis, prescribed treatment, laboratory test results, and anthropometric measurements were noted. Substantial efforts were made by CHDS to ascertain the health and whereabouts of the children who did not come to any of the Kaiser clinics for regular examinations. For each child, all information abstracted on definite or possible congenital anomalies was reviewed by at least two CHDS physicians and at least one biostatistician to be certain that the informa-

tion was uniformly and properly processed. A fuller description of CHDS is given in a previous publication.[3]

The congenital anomalies detected were ascertained through routine medical procedures; no special tests such as neurological or dental examinations were instituted on a routine basis. When in the course of routine care a child was suspected of having an anomaly, he/she was then referred for the appropriate specialized tests and examinations. Congenital heart disease was ascertained by these procedures and the resulting incidence rate observed is higher than that observed in other longitudinal studies.[4] The ascertainment level is high in CHDS because the children were seen frequently, and this provided many opportunities for detection.

The definition of congenital anomalies used by CHDS is quite broad. It includes conditions thought to be of prenatal origin whether or not they were manifest at birth. The definition encompasses structural defects, functional abnormalities, inborn errors of metabolism, and chromosomal aberrations. Conditions which were considered to be the sequelae of postnatal events, such as hearing loss subsequent to otitis media, were not counted as congenital anomalies. The data on birth trauma and postnatal infections were carefully reviewed to evaluate their relationship to a given abnormality. Among the conditions thought to be of prenatal origin, the continuum of defects ranges from those of little or no consequence to those producing severe handicap; these have been divided arbitrarily into three groups which have been labeled "severe," "moderate," and "trivial." The "trivial" category, which includes such conditions as skin tags, umbilical hernias, supernumerary nipples, and minor leg-foot postural defects, has not been included in the count of congenital anomalies (inclusion of this category would label the majority of CHDS children as having congenital anomalies). Thus the two categories—"severe" and "moderate"—when combined represent total congenital anomalies as defined by CHDS.

To be included in this analysis, a congenital anomaly had to be definitely diagnosed; probable cases are excluded. Severe anomalies are defined as those that are potentially life-threatening or, if not corrected, would impair the child's development or well-being. While this definition requires subjective judgment, the resulting classification is arbitrary only because there has never been complete agreement or uniformity in the medical literature as to how certain anomalies should be classified. Conditions representing early embryological maldevelopment fall in the "severe" category. The anomalies remaining after removal of those designated "severe" are labeled "moderate." Again, as in the "severe" group, subjective judgment applies. A summary listing of conditions which gives incidence rates by system may be found in the Appendix; this listing identifies which conditions were designated "severe" and which were designated "moderate." It should be noted that this summary listing excludes much of the detail given in the original code and lists only conditions diagnosed repeatedly.

The life table method is used to compute cumulative incidence rates for congenital anomalies at various ages from birth to five years. This methodology allows each child to be considered at risk of having a congenital anomaly diagnosed until he or she either has such a diagnosis made or is lost from direct observation. The child may be lost from direct observation by being adopted, being institutionalized, dying, or by leaving the health plan. Those children who leave the health plan can be divided into two groups: those whose health status is known (follow-up cases), and those whose health status is unknown (lost cases). Follow-up cases were not considered to be under direct observation because information derived from follow-up contacts was not as detailed as that derived by direct contact. Once a diagnosis of a congenital anomaly is made, whether the child dies, is institutionalized, or survives, that diagnosis is picked up at the age level at which it was made and contributes to the cumulative incidence from that age onward (all institutionalized children were diagnosed to have an anomaly, and 27 per cent of those dying before age one year and 59 per cent of those dying between age one and five years were diagnosed to have an anomaly).

The cumulative incidence rate at a certain age thus represents the proportion of children in the cohort with a congenital anomaly diagnosed at that age or at an earlier age. In effect, it assumes that cases lost to observation would have had defects diagnosed at the same rate as those under observation.

The diagnosis has been entered on the life table at the age at which it was first noted whether it was definite at that time or was determined to be definite at a subsequent time. When a child had multiple *qualifying* anomalies, the date suspected for the first one detected was used in the life table. For example, a child noted to have a syndactyly of the toe (moderate) at one month of age, mental retardation (severe) at two years of age, and an inguinal hernia (moderate) at three years of age would be picked up at age two years as having a "severe" anomaly and at age one month when total or "moderate" anomalies are considered.

This analysis is based on all single White live births and single Black live births enrolled in CHDS. Respectively, these numbered 12,204 and 4,299. At age five years, 68.3 per cent of the original birth cohort were still Kaiser Health Plan members and another 21.1 per cent were follow-up cases. Approximately 3 per cent of the children had died, been institutionalized or adopted by age five. The number of children totally "lost" from observation was very small: 0.2 per cent at one year of age and 7.4 per cent by five years of age.

Results

The probability of diagnosing a severe anomaly in specific age intervals by specified ages is shown in Table 1 for single White live-born children. By using the life table method, cumulative probabilities of diagnosing severe anomalies are derived where the cumulative probability equals one minus the probability that a severe anomaly was *not* diagnosed in the given age interval or in any previous age interval. These probabilities can also be expressed as incidence rates per 1,000 live-born children (e.g., cumulative

TABLE 1—Probability of Diagnosing a Severe Anomaly by Specified Ages among Single White Live-born Children

Age Interval	Number of Children Entering Interval-under Observation but Undiagnosed	Average Number of Children at Risk in the Interval	Number of Children Diagnosed in the Interval	Probability of Diagnosis in the Interval	Cumulative* Probability of Diagnosis up to and including this Interval
Birth	12,204	12,204.0	147	0.0120	0.0120
1–6 days	12,003	11,826.1	19	0.0016	0.0136
7–31 days	11,603	11,546.2	29	0.0025	0.0161
32–91 days	11,414	11,229.5	48	0.0043	0.0203
92–182 days	11,037	10,895.8	35	0.0032	0.0235
6–12 months	10,727	10,479.5	26	0.0025	0.0259
1–2 years	10,233	9,857.0	40	0.0041	0.0298
2–3 years	9,483	9,174.4	42	0.0046	0.0343
3–4 years	8,850	8,580.6	27	0.0031	0.0373
4–5 years	8,313	8,095.8	35	0.0043	0.0415

*The cumulative probability of diagnosis = 1 − probability of no diagnosis in this or any previous interval.

probability of severe congenital anomalies for single White live-born children diagnosed by age five years, .0415, cumulative incidence 41.5 per 1,000).

Incidence rates for severe congenital anomalies and for severe and moderate congenital anomalies combined as derived by this method are given in Table 2 for CHDS White and Black children at selected ages. The detailed Table 1 for Whites and the summarizing Table 2 for Whites and Blacks indicate that only a fraction of the severe and moderate anomalies are diagnosed in the neonatal period; the incidence rate for severe congenital anomalies over the first week of life is about one-third of that over the first five years. As for the total congenital anomalies, the incidence rate at the first week of life is about one-fifth for Whites and one-third for Blacks of total accumulated over the first five years.

The data given in Table 2 reflect the ethnic differences observed to exist for such anomalies as cleft lip and cleft palate, and polydactyly. Other studies have demonstrated considerable ethnic variation for many specific congenital anomalies.[1,5–7] The Table shows that the incidence of "severe" anomalies noted at birth is higher in White children than in Black children, but that the cumulative incidence of severe anomalies in both ethnic groups is equal at one year of age. When "severe" and "moderate" congenital anomalies are jointly considered, the rate at birth is higher for Blacks than for Whites, but by five years of age the cumulative incidence is higher for Whites than for Blacks. By age five years the cumulative incidence of congenital anomalies, as defined in this study, approaches 15 per cent for the total cohort.

TABLE 2—Cumulative Incidence Rates for Severe Congenital Anomalies and Total* Congenital Anomalies Diagnosed by Specified Ages for 12,204 White and 4,299 Black Singletons

Age	Severe Anomalies per 1000 Live-born Children		Total* Anomalies per 1000 Live-born Children	
	White	Black	White	Black
Day of birth	12.0	10.2	25.7	39.8
6 Days	13.6	11.2	29.6	41.4
6 Months	23.5	21.5	74.5	78.9
1 Year	25.9	25.5	85.1	89.6
3 Years	34.3	34.0	116.8	113.0
5 Years	41.5	38.9	154.9	142.2

*Total = severe + moderate

Table 3 subdivides the population into five birthweight-gestational age groups and contrasts those who were mature by both birthweight and gestation (Group V) with the other four groups. Group I children, those weighing 1500 grams or less at birth, are observed to have the highest severe anomaly rates diagnosed before they were five years old. Group III children who were small for gestational age at birth (birth weight of 1501–2500 grams; gestation ≥37 weeks) are also observed to have markedly increased rates. White children who were small at birth but who had a more appropriate gestational age for their size (Group II: birth weight 1501–2500 grams; gestation <37 weeks) had moderately increased severe anomaly rates while Black children in this group did not. These observations apply to each age level considered. White children who were of normal birthweight but who were born early (Group IV: >2500 grams; <37 weeks) also were observed to have a higher incidence of severe anomalies noted at birth as compared to Group V, but by age five years the difference between the two groups had almost disappeared. Black children in Group IV had lower rates than those in Group V.

In Table 3 the rates for Group I have been bracketed because although the trend shown is clear the rates cannot be considered as unbiased. In the life table analysis of diagnosis of congenital anomalies we have computed the probability of being diagnosed in a given interval given that one is under observation at risk at the beginning of the interval. Excluded from being at risk are those dying, those

TABLE 3—Cumulative Incidence Rates for Severe Congenital Anomalies Diagnosed by Specified Ages within Birthweight-Gestational Age Groups for Black and White Singletons per 1000 Live Births

Age and Ethnic Group	Birthweight-Gestational Age Groups				
	I* ≤1500 gm	II 1501–2500 gm <37 wk	III 1501–2500 gm ≥37 wk	IV >2500 gm <37 wk	V >2500 gm ≥37 wk
	(Children with severe anomalies per 1000 live births)				
Day of birth					
White	95.9	21.6	66.9	19.7	9.9
Black	40.0	6.1	27.0	5.5	9.2
1 year					
White	(130.0)	82.0	98.1	27.6	22.7
Black	(142.2)	12.9	78.3	20.1	22.8
5 years					
White	(337.1)	101.3	132.3	40.4	37.6
Black	(142.2)	35.7	111.3	23.2	35.8
Number of Children in the Birth Cohort					
White	73	232	239	406	11,086
Black	75	163	148	364	3,460

*Figures in parentheses are based on very small numbers, as only 20 White and 22 Black children in this group survived the first month of life (see "Discussion" in text).

diagnosed with an anomaly, and those lost to observation before the interval begins. It is tacitly assumed, particularly when computing a cumulative probability of diagnosis, that those not at risk are essentially similar to those that are at risk. It is clear that where this assumption is not justified, the damage that can occur is partly a function of the relative numbers of those excluded from risk as compared with those remaining at risk. In particular, where, for example, we have the majority excluded from risk because they are already dead, we have a maximum opportunity for error due to faulty assumptions. Thus, for example, where we do a life table analysis of congenital anomalies among those whose birth weight is less than 1500 grams we cannot assume that those who die in the neonatal period (first 28 days of life) are as likely to have been anomalous as those who survive. It is hard to see which way the biases go. Some could have been diagnosed with an anomaly because they died (and hence were subject to autopsy). Some could have had an anomaly which was not diagnosed because they died before symptoms, behavior, etc., led to a suspicion of anomaly. Probably the latter is more likely than the former. In any event, where large numbers are removed from risk because of death, which we observe only in Group I, the life table will not ensure that there is no bias.

Since a high proportion of neonatal deaths are due to congenital anomalies, and since the neonatal death rate varies strongly with both birth-weight and gestation, the long-term burden of severe congenital anomalies in the various birthweight gestation groups is more clearly depicted by considering the incidence among children who survived the first 28 days of life (68.4 per cent of the deaths that occurred between birth and age five were neonatal deaths). Table 4 gives the incidence of severe congenital anomalies among these survivors. Groups I and II have been combined because there were relatively few survivors in Group I. Although this eliminates the equal birthweight classification set up for Groups II and III, it serves to point out strong ethnic differences. Among the Black neonatal survivors, the only immature group subject to increased severe congenital anomaly rates was Group III representing infants who were small for gestational age. Among the White neonatal survivors severe congenital anomaly rates were observed to be highest in Group III but they were also quite high among small babies with short gestations (Groups I and II combined).

Table 5 gives the incidence rates for each birthweight-gestational age group when the moderate anomalies are combined with the severe congenital anomalies; the "contribution" of moderate anomalies, the combined rate less the rates for severe anomalies, is also shown for age five. The combined cumulative incidence by age five for children who were of normal birthweight and gestation is 148.6/1000 for Whites and 139.4/1000 for Blacks. The rates for children weighing 2500 grams or less are considerably higher; 302.4/1000 and 281.2/1000 of the White children in the two groups weighing 1501–2500 grams with respective gestations of <37 weeks and ≥37 weeks had been diagnosed to have congenital anomalies by this age level. The Black children in the comparable groups had smaller increases yielding rates of 173.6/1000 and 225.1/1000, respectively. With the addition of the moderate anomalies, the children who were small for gestational age no longer stand out as being particularly subject to anomalies when compared with children of a similar birthweight but shorter gestation.

In Tables 3–5, ethnic differences were noted for infants who were immature by birthweight and/or gestation. If gestational age alone is used to subdivide the population as in Table 6, a very striking ethnic pattern emerges. Whites

TABLE 4—Cumulative Incidence Rates for Severe Congenital Anomalies Diagnosed by Specified Ages within Birthweight-Gestational Age Groups for Black and White Singletons per 1000 Survivors of the Neonatal Period*

Age and Ethnic Group	Birthweight-Gestational Age Groups			
	I & II* ≤2500 gm <37 wk	III 1501–2500 gm ≥37 wk	IV >2500 gm <37 wk	V >2500 gm ≥37 wk
	(Children with severe anomalies per 1000 alive at 28 days of age)			
Day of birth				
White	18.2	48.5	15.0	8.8
Black	5.7	28.0	2.8	7.8
1 year				
White	61.8	67.6	22.9	21.2
Black	17.6	79.2	17.4	21.2
5 years				
White	97.8	103.0	35.8	36.1
Black	37.6	112.2	20.5	34.1
Number of Children in the Cohort of Neonatal Survivors				
White, (Gp I-20) (Gp II-200)	220	227	400	11,047
Black, (Gp I-22) (Gp II-154)	176	143	361	3,441

*First 28 days of life.

**The contribution of anomalies observed in Group I to the combined Group I-II rates was particularly high for Whites. The Group II rates alone at age 5 were 68.0 for Whites and 36.0 for Blacks, as contrasted with the respective Group I-II combined rates of 97.8 and 37.6 shown above.

demonstrate increased risks for congenital anomalies at each age level considered when gestation was <37 weeks; Blacks do not. This holds true for severe and moderate anomalies combined as well as for severe anomalies alone.

Discussion

It is widely recognized that an increased risk of congenital malformation is associated with certain population characteristics. It is also widely acknowledged that overall incidence rates are dependent on the definition applied and the period of ascertainment. However, the degree to which these factors influence incidence levels is generally not recognized. The present study has used life table methodology and ethnic comparisons to illustrate the very strong influence these two factors have on the congenital anomaly rates that are observed in a birth cohort.

Many specific congenital anomalies have been shown to have substantial ethnic variation.[1,5–7] Some of these conditions are severe and some are moderate. For example, anencephaly, spina bifida, hypospadias, polydactyly, syndactyly, congenital dislocation of the hip, hemangioma, and branchial cleft cyst are among the conditions recognized as having substantial ethnic variation. Polydactyly stands out in this group because it is usually noted at birth, has a high incidence level, and occurs at a rate among Blacks that is approximately nine to ten times greater than the rate observed among Whites.[1,5] In CHDS, 37 per cent of the Black children observed to have a congenital anomaly at birth had polydactyly while the comparable figure for White children was 3 per cent. Classification of this anomaly as "moderate" is thus seen to have a strong effect on the ethnic patterns shown in Table 2. If polydactyly had been classified as "severe," the incidence of severe congenital anomalies at birth would have been higher among Blacks than Whites, a pattern opposite to that actually observed. Myrianthopoulos and Chung[1] included supernumerary nipples in their study and observed rates for this condition of .9/1000 among Whites and 11.4/1000 among Blacks. Addition of just this one anomaly (which we exclude as trivial) to CHDS data would thus have a strong effect on the rates shown.

A long period of ascertainment picks up congenital anomalies not generally detected in studies covering earlier time periods. Whereas many conditions are readily detected in infancy, others such as color blindness, myopia, dyslexia, slight mental retardation, minimal hearing loss, and anomalies of the teeth will not be detected until the child is older. Much of the change in rates over time is due to the inclusion of different conditions.

The period of ascertainment affects both the magnitude of the congenital anomaly rate and the relationship of other factors associated with it. The cumulative incidence rate for severe congenital anomalies increased by more than 50 per cent between one and four years of age; the rates for moderate anomalies increased by a larger amount in the same time interval. The relative ethnic patterns also demonstrated considerable change over time. The White to Black total anomaly rate ratio changed from 0.65 at birth to 1.09 at age five years.

TABLE 5—Cumulative Incidence Rates for Total* Congenital Anomalies Diagnosed by Specified Ages within Birthweight-Gestational Age Groups for Black and White Singletons

Age and Ethnic Group	Birthweight and Gestational Age Group				
	I** ≤1500 gm	II 1501-2500 gm <37 wk	III 1501-2500 gm ≥37 wk	IV 2501+ gm <37 wk	V 2501+ gm >37 wk
	(Rate per 1000 live-born children)				
Day of birth					
White	109.6	64.7	79.5	34.5	22.7
Black	53.3	36.8	74.3	30.2	38.4
1 year					
White	(257.2)	199.5	212.9	107.0	78.9
Black	(285.9)	126.0	147.6	68.0	85.1
5 years					
White	(399.8)	302.4	281.2	160.8	148.6
Black	(330.5)	173.6	225.1	103.5	139.4
	"Contribution" of moderate congenital anomalies by age 5				
White	(62.7)	201.1	148.9	120.4	111.0
Black	(188.3)	137.9	113.8	80.3	103.6

*Total = severe + moderate

**Figures in parentheses are based on very small numbers, as only 20 White and 22 Black children in this group survived the first month of life.

Methods of ascertainment will also have a direct effect on the number and type of anomalies observed in a given population. The study of McIntosh, *et al*,[2] included a special dental examination and consequently they ascribe a much higher proportion of anomalies of the digestive system to the oral cavity than does the present study. Frequent physicial contact for routine care was the basis of ascertainment in the present study. The White children averaged 30.7 physician visits and the Black children 23.0 between birth and age five years. Although the Black children were seen less frequently, they still averaged three physician contacts per year between age three years and age five years. In both groups there was ample opportunity for detection of conditions readily diagnosed at clinic visits. Conditions diagnosed only through special screening procedures could still have gone undetected, however.

The impact of these different factors becomes extremely important because of present pressures to undertake studies on the influence of environmental factors such as occupational exposures or the use of drugs on the incidence of congenital anomalies. A recent CHDS study by Christianson[8] of the effect of maternal smoking on the incidence of congenital malformation illustrates their importance. In that study, heavy maternal smoking was associated with a statistically significant increase in the incidence of moderate congenital anomalies but not in the incidence of severe congenital anomalies. If the study had been limited to severe congenital anomalies, no significant associations would have been observed. In addition, the period of ascertainment was strongly reflected in the findings, as more than two-thirds of the cases of strabismus, one of the specific moderate anomalies associated with maternal smoking, were diagnosed after the age of one year.

The long-term ascertainment of congenital anomalies in this cohort has provided the opportunity to extend and refine the previously recognized association between the occurrence of congenital anomalies and prematurity. The increased frequency of congenital anomalies that occurs in conjunction with prematurity has been observed repeatedly.[7,9-12] The majority of studies have used birthweight alone

TABLE 6—Cumulative Incidence Rates for Severe Congenital Anomalies and Total Congenital Anomalies by Specified Ages within Two Gestational Age Groups for Black and White Singletons

Age	Severe Congenital Anomalies per 1000 Live-born Children				Total Congenital Anomalies per 1000 Live-born Children			
	White		Black		White		Black	
	<37 wk	≥37 wk	<37 wk	≥37 wk	<37 wk	≥37 wk	<37 wk	≥37 wk
Day of birth	28.4	11.1	10.0	10.0	52.5	23.9	35.1	39.9
1 year	55.0	24.3	27.3	25.0	146.9	81.7	97.2	87.6
5 year	75.9	39.6	35.9	38.8	219.3	151.4	136.7	142.7
Number of births*	705	11,326	599	3,609				

*Includes only cases with known gestational age; no requirement for birthweight.

to define prematurity, but Yerushalmy, *et al*,[10] demonstrated that the associations among the low-birthweight infants varied with duration of gestation. They noted that those weighing less than 1500 grams had a very high severe anomaly rate regardless of gestation and that those weighing 1501 to 2500 grams and having a gestation of 37 weeks or more had a rate that was twice as high as that observed for infants of the same weight but shorter gestation. The cumulative rates through age five in the present study also demonstrated very high severe congenital anomaly rates for those who weighed 1500 grams or less and those weighing 1501 to 2500 grams with long gestations. However, the relative difference between short and long gestation infants who weighed 1501–2500 grams at birth varied over time with the differences being more pronounced by Black children than for White children. Although ascertainment through age five demonstrates higher severe anomaly rates among children who were small at birth and particularly for those who were small for gestational age, it should be noted that less than one-fifth of all severe congenital anomalies were observed among children who weighed <2500 grams at birth.

Minor birth defects were observed to occur at a slightly higher rate among infants weighing less than 2500 grams at birth than among heavier infants in a study by Hook, *et al*,[13] of newborns who were without severe anomalies. The present data on moderate anomalies detected through age five also indicate somewhat increased rates among low-birthweight children; the increases are of a much lower magnitude than those observed for severe defects, however.

The present study has illustrated the transitions that occur in the recognition of congenital anomalies over a five-year time span in a closely studied cohort. The about fivefold increase between birth and five years of age in the cumulative incidence level of total congenital anomalies indicates the magnitude of the changes which take place. As more and more studies are undertaken on the relationship of various environmental factors to the incidence of congenital malformations, the composition of the group of anomalies under study must always be given due consideration.

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ACKNOWLEDGMENT

This research was supported by grant HD07256 from the National Institutes of Health.

APPENDIX—Cumulative Incidence Rates per 1000 of Severe and Moderate Congenital Anomalies at the Ages of 1 and 5 Years, by Organ System* (Includes only single live-born children)

Anomalies	Rate/1000 at Age 1		Rate/1000 at Age 5	
	White	Black	White	Black
(1) NEOPLASMS				
Severe Primarily malignant and benign brain neoplasms, leukemia, von Recklinghausen's disease, lymphangioma, and dermoid eye tumors	1.1	1.2	2.3	2.0
Moderate Primarily hemangioma of skin, lipoma, fibroma osteochondroma/bone cyst, pilonidal cyst, cysts of the oral cavity, and epidermoid cysts	10.6	4.8	13.6	5.9
(2) ENDOCRINE AND METABOLIC DISEASES				
Severe Primarily diabetes mellitus, hypoglycemia, celiac disease, fibrocystic disease of pancreas, and adrenal hypoplasia	1.3	0.2	2.7	0.2
Moderate Primarily precocious puberty and nevoxantho-endothelioma	0.6	0.5	1.5	1.7

APPENDIX (Continued)—Cumulative Incidence Rates per 1000 of Severe and Moderate Congenital Anomalies at the Ages of 1 and 5 Years, by Organ System* (Includes only single live-born children)

Anomalies	Rate/1000 at Age 1		Rate/1000 at Age 5	
	White	Black	White	Black
(3) DISEASES OF THE BLOOD				
Severe	0.4	1.8	1.6	2.6
Primarily sickle cell anemia, hemophilia, inherited thrombocytopenia				
Moderate	0.1	2.0	0.5	5.7
Primarily sickle cell trait, thalassemia trait, hypogammaglobulinemia				
(4) CIRCULATORY SYSTEM				
Severe	8.0	7.2	9.1	8.3
Primarily tetralogy of Fallot, transposition of great vessels, truncus arteriosus, atrial septal defect, ventricular septal defect, endocardial cushion defect, mitral insufficiency, patent ductus arteriosus, coarctation of the aorta, aortic stenosis, pulmonic stenosis				
Moderate	0.0	0.2	0.0	0.2
Congenital sinus type bradycardia				
(5) CENTRAL NERVOUS SYSTEM				
Severe	6.0	5.4	12.4	14.5
Primarily anencephaly, hydrocephalus, porencephaly, atrophy, agenesis, absence part of brain, agenesis of cranial nerves, spina bifida aperta, microcephaly, cerebral palsy, organic brain syndrome, a minimal brain damage syndrome, b mental deficiency, c aphasia, epilepsy, other convulsive disorders				
Moderate	1.8	1.9	2.1	2.5
Primarily facial palsy, spina bifida occulta, maturation delay, neuromotor				
(6) CONGENITAL ANOMALIES AFFECTING MULTIPLE SYSTEMS				
Severe	2.9	1.2	2.9	1.2
Primarily Down's syndrome, Marfan's syndrome, Turner's syndrome, Rubenstein-Taybia syndrome, Cornelia de Lange syndrome, other chromosomal aberrations				
Moderate	0.0	0.0	0.0	0.0
None				
(7) CONGENITAL ANOMALIES OF THE EYE				
Severe	0.5	1.7	0.5	2.0
Primarily blindness, total or partial; congenital cataract (operated)				
Moderate	10.8	7.9	45.9	32.3
Primarily congenital cataract (not operated), coloboma of iris, anisocoria of pupils, heterochromia of iris, corneal opacities, esotropia or exotropia, refractive errors (astigmatism, myopia, hyperopia), color blindness, ptosis palpebralis, nystagmus, oculomotor disturbance				
(8) CONGENITAL ANOMALIES OF EAR, FACE, AND NECK				
Severe	0.6	0.7	2.5	2.3
Primarily deafness-total or partial, agenesis external ear-canal, first/second arch syndrome				
Moderate	5.7	7.4	7.5	10.7
Primarily preauricular fistula, external ear malformations, asymmetry of face/skull, thyroglossal duct cyst, branchial cleft cyst				
(9) CONGENITAL ANOMALIES OF SKIN AND HAIR				
Severe	0.1	0.0	0.2	0.0
Ehlers-Danlos syndrome, ichthyosis				
Moderate	3.2	3.6	6.5	6.2
Primarily nevus (referred to Dermatology), telangiectasis, defect of scalp, vitiligo, hypertrichosis				
(10) CONGENITAL ANOMALIES OF THE MUSCULOSKELETAL SYSTEM				
Severe	4.0	4.5	4.9	5.3
Primarily craniostenosis, achondroplasia, limb reductions, partial adactylia (hand or foot), syndactyly (fingers), club foot (equinovarus)				
Moderate	17.2	22.9	26.9	29.1
Primarily hypognathia, anomalous ribs/vertebrae, torticollis, dislocation/dysplasia of hip, abnormal spinal curve (lordosis, kyphosis, scoliosis), polydactyly, syndactyly (toes), trigger thumb, hammer toe and other defects of digits, talipes, congenital hypotonia, hypoplasia-muscles, cranio-facial dysostosis, exostosis, Achilles M. shortening				
(11) CONGENITAL ANOMALIES OF THE RESPIRATORY SYSTEM				
Severe	0.1	0.2	0.1	0.2
Pulmonic aplasia				
Moderate	0.1	0.2	0.3	0.2
Primarily congenital tracheomalacia				
(12) CONGENITAL ANOMALIES OF THE DIGESTIVE SYSTEM				
Severe	5.4	4.3	5.7	4.8
Primarily cleft lip and palate, cleft palate, omphalocele, atresia of a portion of intestines, Meckel's diverticulum, imperforate anus, anal fistula, pyloric stenosis, diaphragmatic hernia, malrotation, volvulus, intussusception				

APPENDIX (Continued)—Cumulative Incidence Rates per 1000 of Severe and Moderate Congenital Anomalies at the Ages of 1 and 5 Years, by Organ System* (Includes only single live-born children)

Anomalies	Rate/1000 at Age 1		Rate/1000 at Age 5	
	White	Black	White	Black
Moderate Primarily anomalous dentition, dysplasia—teeth enamel, inguinal hernia (operated), anorectal stenosis or stricture	14.2	17.0	29.6	28.2
(13) CONGENITAL ANOMALIES OF THE GENITO-URINARY SYSTEM				
Severe Primarily renal agenesis, bifid kidney, hydronephrosis, urethral stenosis, double ureter, hypoplasia of the bladder, agenesis testis	1.5	2.4	4.9	3.7
Moderate Primarily horseshoe kidney, meatal stenosis, undescended testis (beyond age 2 years), hypospadias (1st degree)	9.0	7.5	12.9	12.4

* Children with multiple anomalies are counted in each system and severity category which applies; exceptions are the syndromes mentioned in subgroup (6).

a) The term "organic brain syndrome" in subgroup (5) refers to a neurologist's diagnosis of a generally unspecified CNS disorder with hard neurological signs and symptoms, e.g., hypsarrhythmia, ataxia, papiledema, etc.

b) "Minimal brain damage" in subgroup (5) is a neurological catch-all diagnosis with mixed neurobehavioral problems, including poor motor coordination, hyperactivity, and short attention span.

c) Mental retardation in subgroup (5) is a term with a wide range of deficiencies, and includes any definite diagnosis, whether indicating mild or severe retardation, by the child's pediatrician or neurologist.

ASTHO Inventory of Public Health Programs Available

The National Public Health Program Reporting System (NPHPRS) of the Association of State and Territorial Health Officials (ASTHO) has recently published its annual inventory of the programs of the nation's official state and territorial health agencies (SHAs) for fiscal year 1980. "Public Health Programs 1980: An Inventory of State Programs and Uses of Health Incentive Grants" provides complete listings of the programs and expenditures of each SHA. Program expenditures of Federal Health Incentive Grants are also listed.

NPHPRS is designated as the national public health program reporting system specified in Section 314(d) of the Public Health Service Act. As such, it is the only source of comprehensive, uniform, detailed data about these public health agencies.

Highlights from Public Health Programs 1980 include:

- The nation's state and territorial health agencies (SHAs) spent $4.5 billion for their public health programs in FY 1980;
- Seventy-one per cent of their expenditures were for personal health programs; 8 per cent for health resources programs; 7 per cent for environmental health programs; 6 per cent for general administration; and 4 per cent for the operation of public health laboratories. The remaining 4 per cent was granted by the SHAs to local health departments (LHDs) and not allocated to the program areas;
- The SHAs spent $64 million in Health Incentive Grants during FY 1980; $25 million of which was allocated to LHDs;
- SHAs and LHDs provided direct health services to an estimated 74 million people—one in every three Americans.

"Public Health Programs 1980" is available free of charge from:

National Public Health Program Reporting System
Association of State and Territorial Health Officials
962 Wayne Avenue, Suite 403
Silver Spring, MD 20910
(301) 589-2520