Retrospective Survey of Congenital Rubella Syndrome in the State of Washington, 1963-68

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EFFICIENT techniques for surveillance of the congenital rubella syndrome (CRS) must be developed in order to assess the efficacy of rubella vaccination programs in preventing CRS. We, therefore, made a retrospective survey to ascertain how many children were born with this syndrome in Washington State in the period 1963–68 and to find an efficient method for the continuing surveillance of this syndrome. The 6-year period 1963 through 1968 was chosen because it encompassed the rubella epidemic of 1965, as well as nonepidemic years, and ended just before the licensing of rubella virus vaccines in 1969. The survey was begun in April 1969 and completed in December 1969.

Methods

Sources. Information was obtained from:

1. Birth certificates of babies born during the study period listing any of the following defects: congenital rubella syndrome, cataracts, purpura, micrognathia, and microcephaly.

2. Death certificates listing CRS as a cause of death of babies born in the study period.

3. Reports from hospitals. All general hospitals with more than 50 beds were asked by mail to report the number of children born during the study period who had a syndrome diagnosed as CRS. Detailed information was then requested on each such child.

4. Records of the hearing, speech, and congenital defects clinics of three large referral centers; these records were abstracted by Chinn. The three referral centers were the Children's Orthopedic Hospital and Medical Center, the Seattle Hearing and Speech Center, Inc., and the University of Washington Hospital.

5. Reports from physicians and teachers. With the cooperation of the crippled children's services section of the Washington State Division of Health, otolaryngologists, ophthalmologists, and teachers of special education (that is, teachers of the deaf or retarded and speech and hearing therapists) were sent a survey letter with a return postcard requesting a report of the number of children born in the period 1963–68 who were suspected of having CRS. Individual case report forms were sent to all who indicated knowledge of any such children.

6. Reports from parents. A letter and case report form were sent to parents of children born during the study period whose names had been entered on the waiting lists of institutions for retarded children.

Diagnostic criteria. The criteria for the retrospective diagnosis of congenital rubella syndrome were based on a review of the literature (*1–4*).

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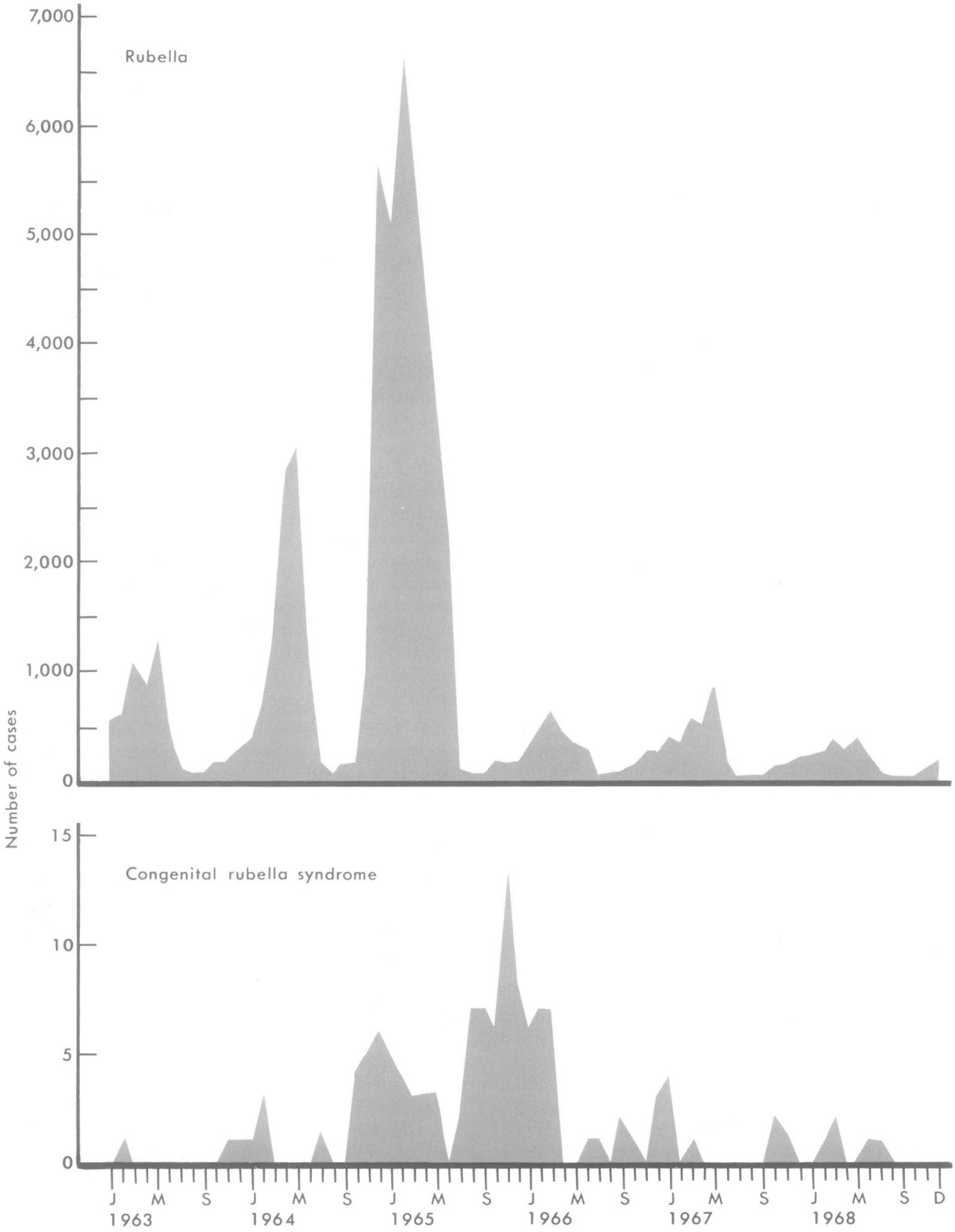
Figure 1. Comparison of month and year of report of 50,753 rubella cases with month and year of birth of 125 infants with congenital rubella syndrome



NOTE: The month of birth was reported for 125 of the 126 CRS patients identified by the survey.

A case was considered as laboratory-confirmed if rubella virus was isolated from an infant who had defects consistent with CRS or if there was serologic evidence of congenital rubella infection (elevated hemagglutination-inhibiting antibody titer when the child was 6–12 months of age or, in neonates, increased IgM rubella antibodies).

A case was accepted as congenital rubella syndrome without laboratory confirmation if the child had (*a*) two or more of the following defects: purpura (or thrombocytopenia), congenital heart disease, deafness, cataracts or glaucoma, and long-bone radiolucencies; (*b*) one of these and either hepatosplenomegaly or microcephaly (excluding the combination of hepatosplenomegaly and congenital heart disease); or (*c*) the child had only one defect from the first-mentioned group, but the mother had a history of a rubella-like illness during pregnancy.

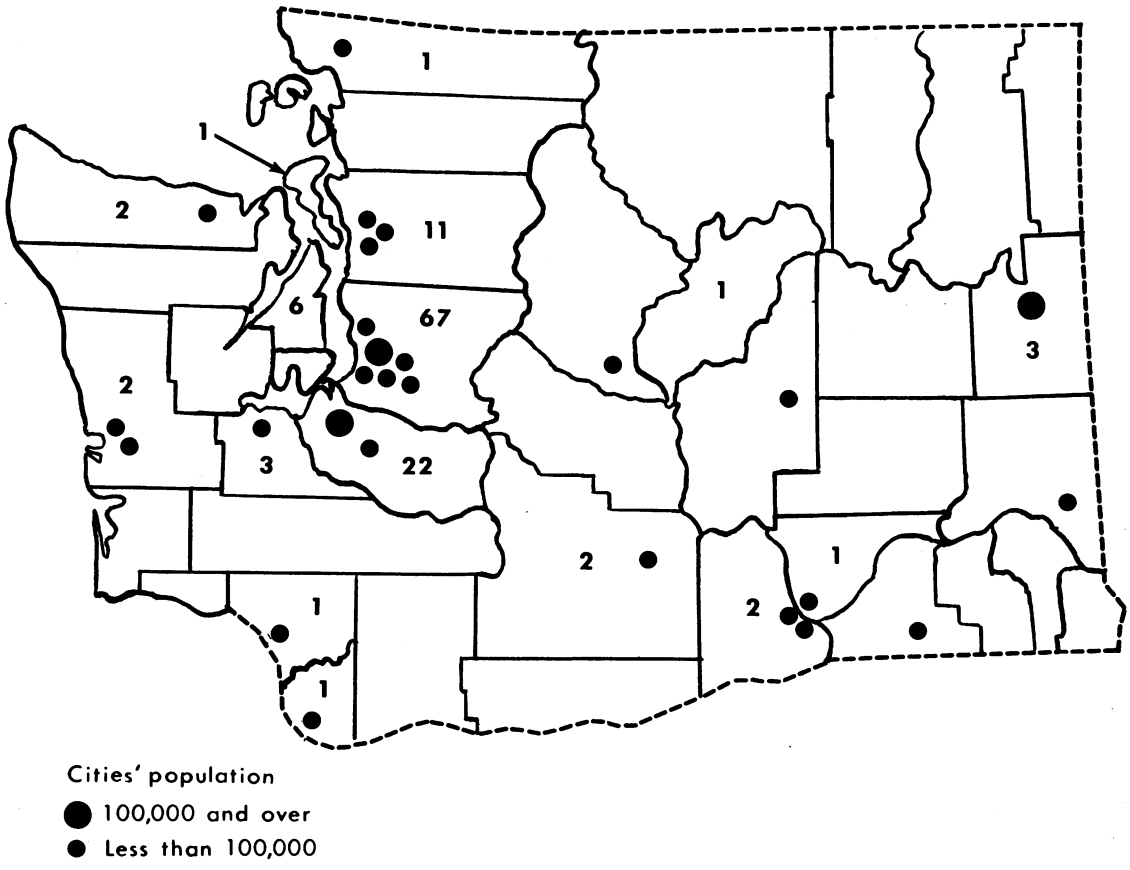
Results

One hundred and seventy cases met these criteria. There were 28 duplicate reports, and 16 reports pertained to children born outside Washington State. Thus, 126 children met the study's criteria for congenital rubella syndrome. Four of the 126 patients died during the study period. Because the survey was retrospective, complete information could not be obtained on all 126 patients.

In figure 1, the month of birth of 125 patients is compared with the reported incidence of rubella during the study period. The rubella epidemiologic year begins in October and runs through September; the number is of the calendar year in which the epidemiologic year ends. An epidemic occurred in the State in the epidemiologic year 1965. Sixty-nine CRS children identified by the survey were born in the 14-month period beginning 7 months after the start of that epidemiologic year and ending 9 months after its end.

The distribution of the 126 patients by county of birth in relation to the State's major population centers is shown in figure 2. Sixty-seven patients were born in King County. This county includes

Figure 2. County of birth of 126 patients with congenital rubella syndrome



NOTE: Population data are from Washington Vital Statistics Summary, 1967. Public health statistics section, Washington State Department of Health, Washington State Board of Health, Olympia, December 1968.

Seattle and contains approximately one-third of the State's population. As the following table shows, 112 of the 170 case reports came from the three large referral centers in King County.

Source of case reports	*Cases*
3 large referral centers in King County:	
Hospital records	57
Speech, hearing, and congenital defects clinics	55
Teachers of special education	34
Physicians	10
Birth certificates	8
General hospitals	5
Institutional waiting lists	1
Death certificates	0
Total reports	170
Less duplicate reports	28
Less reports of patients born out of State	16
Patients born in State during study period	126

Information on almost all (119 of 126) of the cases came from the hospital or clinic records of the three large referral centers or from the teachers of special education. The survey of birth and death certificates, general hospitals, institutional waiting lists, otolaryngologists, and ophthalmologists yielded only seven cases that would not otherwise have been identified.

Cases are categorized by reporting criteria in table 1. Twelve cases were laboratory-confirmed, and 47 were diagnosed on the basis of a combination of two or more defects. In 32 of these 59 cases (52 percent), there was a history of maternal rubella-like lness during pregnancy. Deafness, congenital heart disease, and cataracts were the defects most commonly reported (table 2).

In figure 3, the distribution of birth weights among 74 infants with CRS is compared with the distribution among all infants born alive in the State during 1967. The population identified by the study had significantly lower birth weights ($X_2^2 = (\text{collapsed table}) = 116.0$, $P \ll 10^{-5}$). Of the 54 infants for whom gestational age at birth was reported, four were premature.

The mother's age at the time of birth of the infants with CRS was reported for 65 cases. The distribution of these 65 mothers' ages did not differ from that of all women giving birth in the State during the study period.

Figure 4 shows the age at diagnosis by the year of birth for 83 of the 126 patients. Children born in 1963 were 5 to 6 years old at the time of the survey, whereas those born in 1968 were only 5 to 16 months old. Thus, the length of time during which the diagnosis could have been made depended upon the child's year of birth.

Table 1. Criteria on which the retrospective diagnosis of congenital rubella syndrome was based on 126 cases

Criteria	Cases
Cases with laboratory confirmation:	
Isolation of rubella virus in the neonatal period from an infant having defects consistent with CRS	12
Serologic evidence of congenital rubella infection	0
Cases without laboratory confirmation:	
History of maternal rubella-like illness during pregnancy and 1 of following:	
a. 2 or more defects from groups I and II, 1 of which must be from group I [1]	31
b. A single defect from group I [1,2]	67
No history of maternal rubella-like illness, but with 1 of the following:	
a. 2 or more defects from group I [1]	14
b. 1 defect from group I plus 1 or more defects from group II [1]	2
Total	126

[1] Group I: Neonatal purpura, congenital heart disease, deafness, cataracts or glaucoma, long-bone radiolucencies. Group II: Microcephaly, hepatosplenomegaly. The combination of congenital heart disease and hepatosplenomegaly was not considered sufficient for diagnosis.

[2] Of these 67 cases, the number of diagnoses based on each defect was as follows: deafness 61, purpura 2, congenital heart disease 2, cataracts or glaucoma 2.

Table 2. Defects reported in 126 patients with congenital rubella syndrome

Defect	Present	Absent	Unknown	Percent with defect when known
Deafness	99	3	24	97.0
Congenital heart disease	39	32	55	54.9
Cataracts	31	39	56	44.2
Developmental retardation	23	35	68	39.6
Neonatal purpura	23	28	75	45.0
Hepatosplenomegaly	23	27	76	46.0
Long-bone radiolucencies	15	27	84	35.7
Microcephaly	10	21	95	32.2
Neonatal jaundice	5	17	104	22.7
Glaucoma	3	24	99	11.1

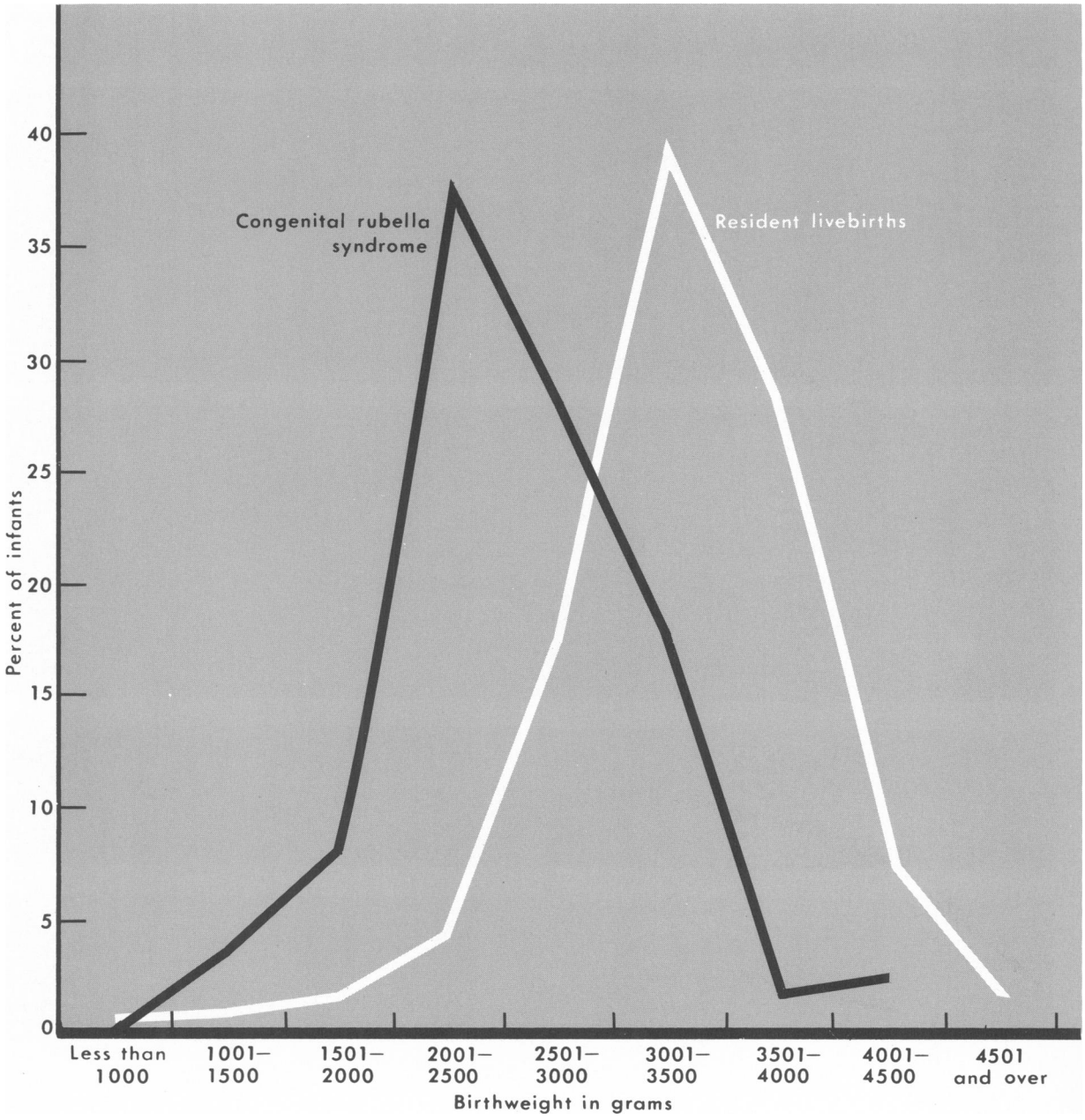
Discussion

The survey demonstrates that retrospective surveillance of congenital rubella syndrome is feasible and suggests methods for prospective surveillance. More rigorous criteria than we applied in this survey will be possible when laboratory facilities for rubella serologic testing are more widely available and there is more widespread understanding of this disease and the importance of reporting it.

The defects identified in this group of 126 chil-

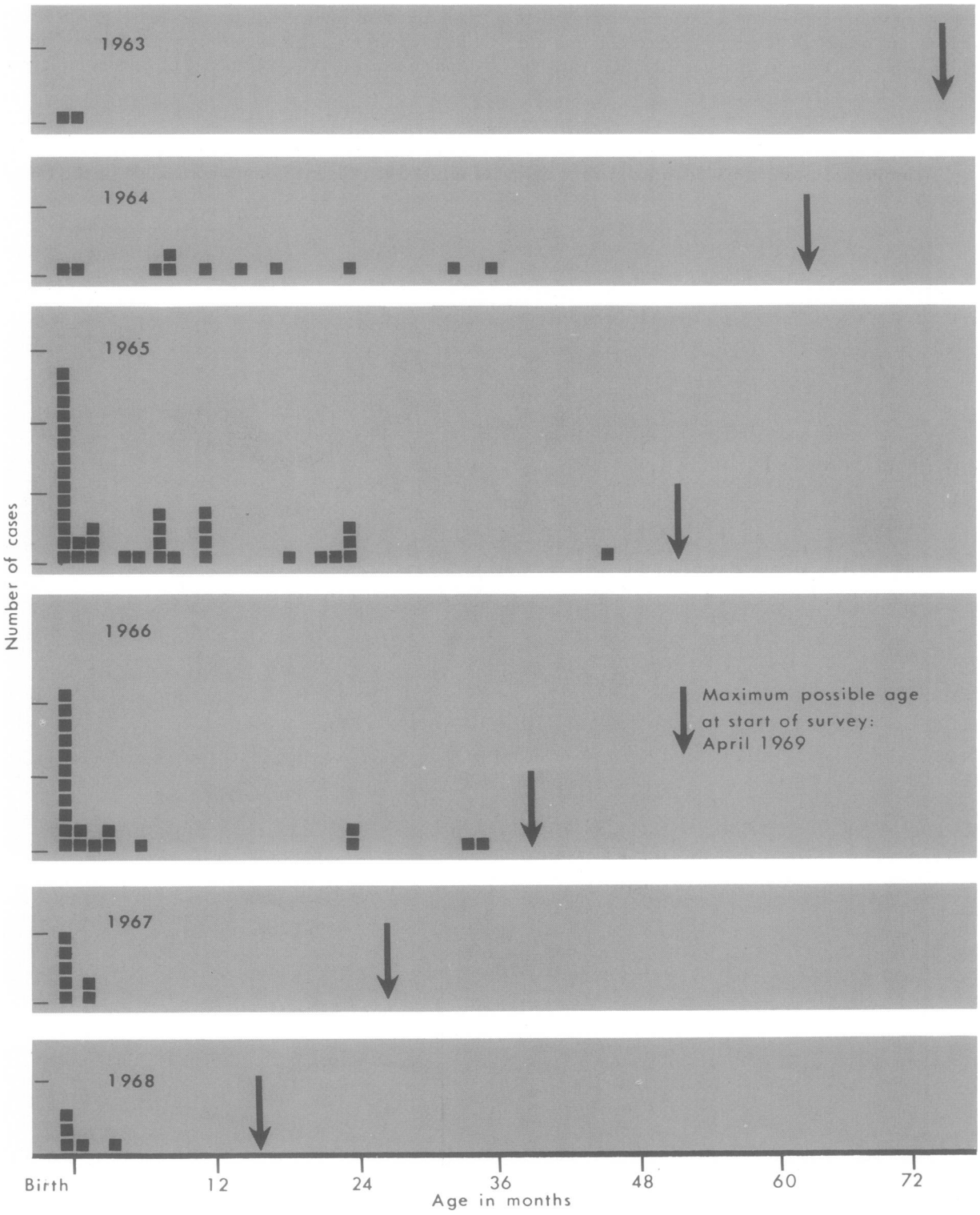
dren do not represent the true incidence of defects caused by fetal infection since the interests of the reporting source influence the frequency with which defects are reported. Severely affected children are more likely to come to medical attention early in life. Presumably, additional cases might be identified later among children born in the study period. An accurate estimate of the incidence of CRS defects would require mass serologic screening of pregnant women to obtain evidence of rubella infection

Figure 3. Comparison of birth weights of 74 infants with congenital rubella syndrome born alive in Washington State 1963–68 with birth weights of all 54,842 resident infants born alive in the State in 1967



NOTE: The birth weight was reported for 74 of the 126 CRS patients identified by the survey. Population data are from Washington Vital Statistics Summary, 1967. Public health statistics section, Washington State Department of Health, Washington State Board of Health, Olympia, December 1968.

Figure 4. Age at diagnosis of 83 patients with congenital rubella syndrome, by year of birth



NOTE: Age at diagnosis was reported for 83 of the 126 CRS patients identified by the survey.

and followup of their children until they reach school age, or even longer.

The low birth weights of patients with congenital rubella syndrome probably reflect retardation of intrauterine growth caused by rubella infection (*1*).

It is clear that not all CRS cases were detected. Only 35 percent of the State's live births occurred in King County; yet 53 percent of the patients identified were born there. Whether the true incidence of CRS is appreciably greater in King County than in the rest of the State would depend on several factors, such as the likelihood of maternal susceptibility and exposure to rubella. A baby with this syndrome born in a metropolitan area would be more likely to be identified than one born in a rural part of the State. If the incidence of CRS in infants born in King County during the study period is applied to infants born in the State as a whole, an additional 65 cases would have been expected in the rest of the State, and the total would have been 191 cases instead of the 126 detected.

Surveillance efforts should be directed toward those professionals most likely to encounter children with defects caused by rubella. In urban areas, where medical services are more readily available, surveillance could be confined to large medical referral centers and special education centers. In rural areas, where medical services are less accessible, physicians will need to be aware of, and to routinely report, this disease entity if we are to achieve a more accurate estimate of its incidence.

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MARCUSE, EDGAR K. (Center for Disease Control, Public Health Service), PETERSON, DONALD R., THOMPSON, ROBERT S., and CHINN, NINA: ***Retrospective survey of congenital rubella syndrome in the State of Washington, 1963–68. HSMHA Health Reports, Vol. 86, February 1971, pp. 161–167.***

Efficient techniques for the surveillance of congenital rubella syndrome (CRS) must be developed to assess the efficacy of rubella vaccination programs in preventing it. A retrospective survey was made to estimate the number of children born with CRS in Washington State from 1963 through 1968 and to seek an efficient method of continuing the surveillance of this syndrome. Sources surveyed included hospital and clinic records, birth and death certificates, otolaryngologists, ophthalmologists, teachers of special education, and parents of children whose names were entered on institutional waiting lists during the study period. One hundred and twenty-six reports met the study's criteria for the retrospective diagnosis of CRS. One hundred and nineteen of these cases were reported by teachers of special education or by three large referral centers. The results suggest that congenital rubella surveillance should be concentrated on these sources.