# An Evaluation of the Maryland Rheumatic Fever Registry

LEON GORDIS, M.D., Dr.P.H., ABRAHAM LILIENFELD, M.D., M.P.H., and ROMEO RODRIGUEZ, M.D.

THE SPECIFIC mechanisms in the pathogenesis of rheumatic fever are still unknown, but the demonstration that all attacks of rheumatic fever occur as sequelae to antecedent streptococcal infections has been the basis for a rational approach to both primary and secondary prevention. When first sulfa and then penicillin were added to the therapeutic armamentarium, rheumatic fever was not expected to recur. Today, however, in Baltimore, recurrent attacks still comprise 13 percent of the total hospital admissions for rheumatic fever (*1*).

Registries for rheumatic fever have been operating for some time in many States to facilitate efforts at secondary prevention (*2*), and reexamining a number of important issues relating to the objectives and effectiveness of one such registry, the Maryland Rheumatic Fever Registry, is appropriate. Using data from a review of cases of patients discharged from Baltimore hospitals with a diagnosis of rheumatic fever, the registry was evaluated as a source of statistical and epidemiologic data, as a mechanism for followup of rheumatic fever patients, and as a stimulus for physician and community education.

*Dr. Gordis is chief, department of community medicine, Sinai Hospital, Baltimore, Md., and assistant professor of pediatrics, Johns Hopkins University School of Medicine. Dr. Lilienfeld is professor and chairman of the department of chronic diseases, Johns Hopkins University School of Hygiene and Public Health. Dr. Rodriguez was a fellow in pediatrics at Johns Hopkins University School of Medicine when this study was conducted.* *The work was supported by National Institutes of Health grants HE 05555 and 5 T01 HE 5297, and general research support grant FR05445-06.*

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## Registry Operation

More than 10 years ago, the Maryland Heart Association established a registry to facilitate the distribution of free or low-cost penicillin to rheumatic fever patients for continuous prophylaxis against recurrent attacks. Currently, under an agreement between the Maryland Heart Association and the Maryland Pharmaceutical Association, pharmacists in the State sell penicillin at low cost to persons who are certified by the heart association to have rheumatic fever. There are no age or income limitations on eligibility.

The Maryland registry has been converted to electronic data processing, and each month the computer punchcards are sorted to determine which patients need prescription refills for penicillin. The pharmacists report to the registry when a patient has a prescription filled.

The cards containing this information are matched with those of the patients who were due back for refills. Thus a printout on "delinquent" patients is obtained monthly.

Since 1965, all new cases referred to the heart association's registry have been verified by a registry physician, who reviews the clinical features reported on the referral forms. Reporting physicians are encouraged to register as "suspects" all patients with a doubtful diagnosis of rheumatic fever. One year later the physicians are reminded that they referred a suspect patient to the registry and perhaps re-evaluation and final disposition are in order.

### Distribution of Registered Cases

The Maryland registry now includes 3,800 patients. In 1967, the first complete year of automated operation, 483 new patients were referred to the registry. Of these, 224 had acute (190 initial and 34 recurrent) attacks, with carditis present in 92 patients. The remaining 259 patients, who did not have an acute attack of rheumatic fever in 1967, were referred either with a history of rheumatic fever or with rheumatic valvular disease. Definite rheumatic heart disease was found in 214 patients, and the possibility of rheumatic heart disease could not be excluded for another 39 patients.

### Evaluation of the Registry

The value of rheumatic fever registries in providing statistical and epidemiologic data about the disease is limited. Although rheumatic fever is a reportable disease in some States, underreporting in these States is probably quite extensive. The degree of underreporting is difficult to determine since the true number of cases in a community at a given time is usually unknown. This contrasts, for example, with cancer registries, in which the lethal nature of the disease permits a determination of the completeness of reporting by comparing the number of reported cases with death certificates. Acute rheumatic fever generally is not lethal, however, and both the quality and severity of its symptoms are quite variable. Although the Jones Criteria have produced a degree of standardized reporting, diagnoses still vary considerably. Registries therefore suffer not only from underreporting but also from incorrect diagnoses for reported patients.

To determine the extent to which both factors can introduce bias into epidemiologic and statistical data based on registries, the medical records of all patients discharged with a diagnosis of rheumatic fever from hospitals in the City of Baltimore from 1960 through 1964 were reviewed. For each case, the presenting manifestations were ascertained, and a judgment was made as to whether the case met the revised Jones Criteria. Cases meeting the criteria were classified as valid; those that did not meet the criteria were classified as invalid—although it was recognized that some of these patients might subsequently develop rheumatic heart disease. This review of medical records has been reported (*1*). Each case ascertained in this review was checked against the Maryland registry to determine whether the patient had been reported initially to the registry in 1960–64 and whether the case was still active with the registry in 1966.

During the study period, 413 patients with a diagnosis of acute rheumatic fever were discharged from Baltimore hospitals; 315 cases were valid, 85 were invalid, and the validity of 13 cases could not be determined. Of the 400 cases with a definite determination of validity, 244 (61 percent) were reported to the registry.

**Table 1. Reporting of hospitalized rheumatic fever patients to Maryland registry, by validity of diagnosis, 1960–64**

| Reported to registry | Valid diagnosis | | Invalid diagnosis | | Total | |
|---|---|---|---|---|---|---|
| | Number | Percent | Number | Percent | Number | Percent |
| Yes | 211 | 67.0 | 33 | 38.8 | 244 | 61.0 |
| No | 99 | 31.4 | 51 | 60.0 | 150 | 37.5 |
| Questionable | 5 | 1.6 | 1 | 1.2 | 6 | 1.5 |
| Total | 315 | 100.0 | 85 | 100.0 | 400 | 100.0 |

**Table 2. Registry followup of patients hospitalized with rheumatic fever, by validity of diagnosis, 1960–64**

| Registry status in 1966 | Valid diagnosis | | Invalid diagnosis | | Total | |
|---|---|---|---|---|---|---|
| | Number | Percent | Number | Percent | Number | Percent |
| Active | 145 | 68.7 | 21 | 63.6 | 166 | 68.0 |
| Inactive | 63 | 30.0 | 11 | 33.3 | 74 | 30.3 |
| Questionable | 3 | 1.3 | 1 | 3.0 | 4 | 1.7 |
| Total | 211 | 100.0 | 33 | 100.0 | 244 | 100.0 |

**Table 3. Reporting of rheumatic fever patients to Maryland registry, 1960–64, by presence or absence of rheumatic heart disease at time of hospital discharge**

| Reported to registry | Rheumatic heart disease present | | Rheumatic heart disease absent | | Questionable | | Total | |
|---|---|---|---|---|---|---|---|---|
| | Number | Percent | Number | Percent | Number | Percent | Number | Percent |
| Yes | 82 | 76.6 | 158 | 53.8 | 8 | 66.8 | 248 | 60.0 |
| No | 24 | 22.5 | 133 | 45.2 | 2 | 16.6 | 159 | 38.5 |
| Questionable | 1 | .9 | 3 | 1.0 | 2 | 16.6 | 6 | 1.5 |
| Total | 107 | 100.0 | 294 | 100.0 | 12 | 100.0 | 413 | 100.0 |

When analyzed separately according to the validity of diagnosis (table 1), there was a tendency for invalid cases not to be reported. Nevertheless, 33 (13.5 percent) of 244 cases referred to the registry were invalid. Of the 315 valid cases, 99 (31 percent) were not referred to the registry.

Table 2 presents data on the proportion of the 244 patients registered in 1960–64 that were still active on the registry in 1966: 74 patients (30.3 percent) were lost to followup by 1966; 63 of 211 with valid cases and 11 of 33 with invalid cases. Physicians tended not to report patients with invalid cases to the registry; nevertheless, once reported, regardless of the validity of diagnosis, approximately one-third of the patients were lost to registry followup by 1966.

These data suggest the extent of error that can be introduced by using registry data as the basis for estimating the incidence of rheumatic fever. Since 99 of 315 valid cases were never reported to the registry, the incidence based on registry data would have been underestimated by 31 percent. Part of the error would be masked, however, by the 33 invalid cases that were reported.

The degree of error is even more pronounced if one uses the registry data for 1966 as the basis for estimating the number of cases that occurred in 1960–64. Of the valid cases, 31 percent were never reported, and of those reported to the registry, 30 percent were lost to followup. Thus by 1966 only 148 (39 percent) of the valid cases that had been ascertained during 1960–64 were still active in the registry.

Since risk of recurrence and further heart damage is greatest in rheumatic fever patients who have rheumatic heart disease, the extent to which such patients are reported to the registry is important. Table 3 shows that the presence of rheumatic heart disease at the time of the acute attack increases the likelihood that a patient will be reported to the registry. Of 107 patients with rheumatic heart disease, 77 percent were reported as compared with only 54 percent of 294 patients without rheumatic heart disease; 24 patients with rheumatic heart disease were never reported. By 1966 only 58 (71 percent) of the 82 reported patients with rheumatic heart disease were still active on the registry as compared with 106 (67 percent) of 158 patients without rheumatic heart disease (table 4).

Thus, although the presence of rheumatic heart disease was correlated with initial reporting to the registry, once a patient was reported,

the presence or absence of rheumatic heart disease did not seem to be related to his continued followup in the registry. If registry data alone were used to calculate the incidence of rheumatic heart disease in Baltimore in 1960–64, it would be underestimated by 23 percent. Furthermore, if only data on patients active with the registry in 1966 were used for determining the incidence of rheumatic heart disease in 1960–64, it would be underestimated by 46 percent.

On the other hand, registry data are often used for estimating the proportion of rheumatic fever cases with rheumatic heart disease. If such an estimate were based on the 248 rheumatic fever cases reported to the Maryland registry for 1960–64, the frequency of rheumatic heart disease in attacks of acute rheumatic fever would be estimated at 33 percent, an overestimate of 7 percent. If only the 1966 registry data were used for estimating the frequency of rheumatic heart disease in rheumatic fever patients reported to the registry in 1960–64, 34 percent (58 of 170 patients still active with the registry in 1966) would be observed to have rheumatic heart disease, an overestimate of 8 percent.

Thus use of registry data would result in a slight overestimate of the frequency of rheumatic heart disease in new attacks of acute rheumatic fever, but would result in a significant underestimate of the incidence of rheumatic heart disease in the community.

The data presented demonstrate that, insofar as the Maryland registry is concerned, the followup program is incomplete. Approximately 30 percent of the cases reported to the registry in 1960–64 were lost to followup by 1966. Moreover, activity in the registry indicates only a patient-registry contact in transmitting a prescription, not that the patient has been maintained on regular prophylaxis. Indeed, among patients who attended rheumatic fever clinics, more than one-third did not comply with penicillin prophylaxis (*3*). The specific structure of a registry followup program is of critical importance and will be discussed further.

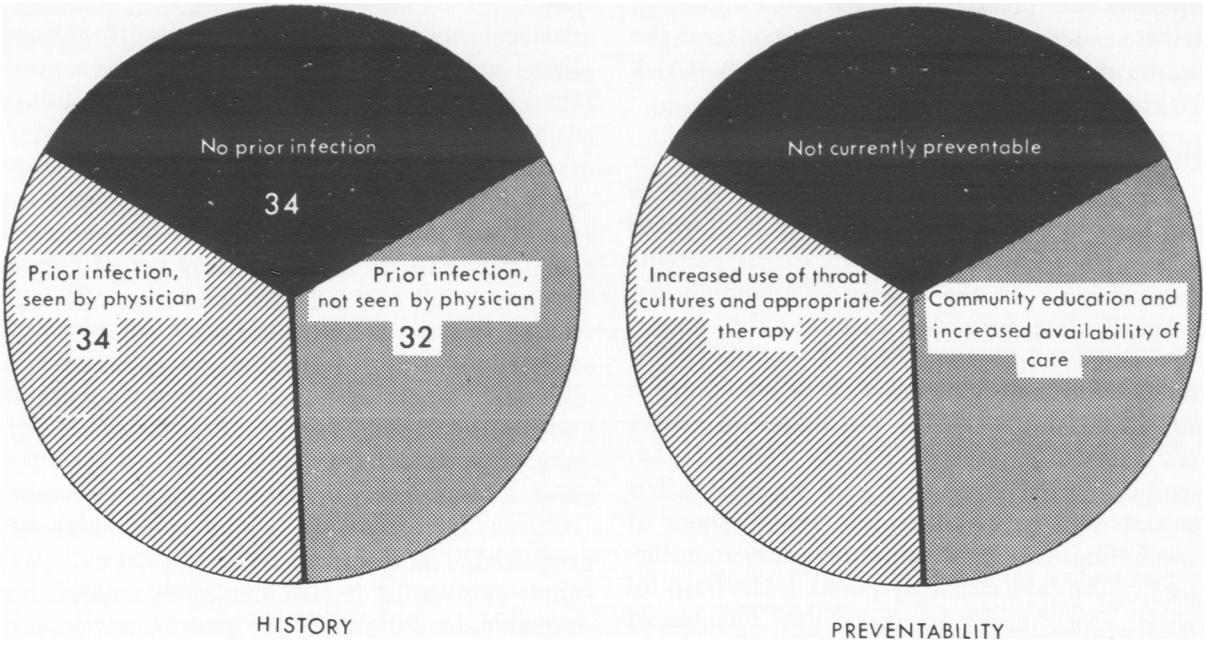
### Management of Streptococcal Infections

One important byproduct of a rheumatic fever registry is the interest it generates in a community concerning the disease. The registry is often the focal point of rheumatic fever programing. In many communities, registry-based activities give the physicians who are actively interested in this disease and its control their only opportunity for exchanging information and ideas on the subject. A program in a community also tends to stimulate general interest in rheumatic fever among physicians engaged in private practice. It would be expected that such an interest would be reflected not only in a greater awareness of the importance of prophylaxis among these physicians, but also a more rational approach to primary prevention of rheumatic fever through the appropriate management of respiratory infections.

To determine the level of physician and community knowledge in Maryland concerning streptococcal infections and sore throats, the medical histories of the 261 patients with first attacks of rheumatic fever identified in this study were reviewed to learn why the disease developed in these patients. More than 80 percent of these patients were less than 20 years of age. The results of the review are shown schematically (see chart). Thirty-four percent of the patients had no history of prior respiratory infection; therefore, they had no reason to con-

**Table 4. Registry status in 1966 of rheumatic fever patients reported to registry, 1960–64, by presence or absence of rheumatic heart disease at time of hospital discharge**

| Registry status, 1966 | Rheumatic heart disease present | | Rheumatic heart disease absent | | Questionable | | Total | |
|---|---|---|---|---|---|---|---|---|
| | Number | Percent | Number | Percent | Number | Percent | Number | Percent |
| Active | 58 | 70.7 | 106 | 67.1 | 6 | 75.0 | 170 | 68.5 |
| Inactive | 24 | 29.3 | 48 | 30.4 | 2 | 25.0 | 74 | 29.8 |
| Questionable | | | 4 | 2.5 | | | 4 | 1.7 |
| Total | 82 | 100.0 | 158 | 100.0 | 8 | 100.0 | 248 | 100.0 |




**History of preceding respiratory infection in 261 patients with first attacks of rheumatic fever, in percentages, Baltimore, 1960–64**

sult a physician. In other studies 6 to 33 percent of patients with rheumatic fever had no preceding symptomatic respiratory infection (*4–7*). Perhaps more important is the finding that 32 percent of patients in the present study had a respiratory infection but did not consult a physician. This finding suggests that additional efforts at community education, particularly in alerting parents to the importance of streptococcal infections, might reduce the incidence of initial attacks of rheumatic fever. Such efforts could be promoted through registry-associated programs, but it seems likely that two important factors in the lack of medical care among some patients were the limited quality and availability of medical services for the poor. Prevention of rheumatic fever must therefore go far beyond the efforts of a categorical disease-control program.

Thirty-four percent of the patients with prior respiratory infections and seen by physicians nevertheless developed rheumatic fever. The specific treatment of these patients could not be completely ascertained; throat cultures were performed for only 20 of the 89 patients, and 13 were negative for streptococci. The infrequent use of throat cultures in this group of patients raised the question of the extent to which physicians served by the Maryland registry utilized throat cultures in their daily practice.

Facilities for throat cultures are available in Maryland through the State health department as well as in hospital and private laboratories. In addition, some physicians use their own office incubators for throat cultures. To determine the extent to which cultures are used, all pediatricians, internists, and general practitioners in private practice in Maryland were surveyed and asked how frequently they used cultures. A questionnaire was sent to 1,307 physicians, and a second mailing went to nonrespondents. The response rate to the combined mailing was 76 percent. The results indicated that 69 percent of responding pediatricians and only 20 percent of responding general practitioners used throat cultures at least several times a week.

Rheumatic fever has been found more frequently in Negroes (*1, 8*), primarily on a socioeconomic basis (*9*). Since children in low-income areas generally receive medical care from clinics, emergency rooms, or general practitioners, the infrequent use of cultures among general practitioners is an important concern. These findings suggest that any educational success of the rheumatic fever program in Mary-

land has been primarily among pediatricians in private practice, who generally do not treat the low-income population which is at highest risk for rheumatic fever.

## Discussion

The adequacy of a rheumatic fever registry has been evaluated by comparing its records with a complete ascertainment of all patients hospitalized with the disease in a community. From the data presented, it is clear that underreporting is a serious problem. Penicillin is now inexpensive, and its provision is no longer a sufficient inducement for physicians to report new cases to a registry. Efforts should be expended toward increasing such reporting, but greater need exists to explore new methods of casefinding. One approach is monthly monitoring of hospitals, either by phoning the hospital record rooms or by having a staff member of the registry program personally review the charts at each institution. An alternate approach is the use of computer listings by diagnostic category, such as the lists that can be obtained from the Professional Activities Service in Ann Arbor, Mich. In those communities where all hospitals subscribe to the system and are willing to cooperate with the registry, periodic lists of all hospital discharges of rheumatic fever patients would be relatively simple to obtain on an ongoing basis.

Such procedures, of course, exclude patients who are not hospitalized for acute attacks. Approximately one-third of patients with a diagnosis of acute rheumatic fever are treated without hospitalization (*1*), and ascertaining such patients is extremely difficult. The decision to support any program to ascertain this group, or to screen heart sounds in an attempt to find previously unrecognized cases of rheumatic heart disease, must be individualized for each community.

The extent of overdiagnosis of rheumatic fever described in this report demonstrates the need for a system of verifying the diagnosis for each new case referred to a rheumatic fever registry. Ideally, the best method is an examination of each new patient in a clinic staffed by expert clinicians experienced in the diagnosis of rheumatic fever, but in most U.S. communities this procedure is not feasible. The system described for the Maryland registry (namely, verification based on the clinical manifestations listed in the referral form) is therefore a practical if not an ideal alternative and is certainly a minimum requirement for an active registry.

The main objective of any rheumatic fever registry is the effective followup of patients for continued prophylaxis. Followup is concerned not only with patients who drop out of formal programs but also with patients who, although active in such programs, do not comply with physicians' instructions for oral prophylaxis. The structuring of an active followup program within the registry framework is needed. Additional personnel for direct followup activities and for the coordination of other available resources for followup in the community are required. The followup problem is so extensive that scarcities in health manpower make it impossible to follow all delinquent patients in registries of large communities. A priority scale could be developed for patients based on factors relating to the risk of recurrence (*1*, *10*, *11*): age, race, number of previous attacks, and presence of rheumatic heart disease; and factors relating to the risk of noncompliance (*12*): female, adolescent, large sibship, no hospitalization for acute attack, and no restriction on physical activity. Intensive followup services could then be directed to high-risk patients assigned priority ratings based on both sets of factors.

The study data show that the educational accomplishments of the Maryland registry are limited and warrant expansion and improvement, particularly in the appropriate and correct use of throat cultures by physicians. Furthermore, through mass media and educational programs geared to various groups, parent and other, the community should be informed of the importance of obtaining medical care for streptococcal infections.

## Summary

The increasing use of case registries for chronic diseases prompted a critical evaluation of the Maryland rheumatic fever registry. The adequacy of the registry as a source of statistical and epidemiologic data on rheumatic fever was evaluated by comparing registry records with the medical records of 413 patients dis-

charged from Baltimore hospitals with a diagnosis of acute rheumatic fever during 1960–64. Only 61 percent of these patients were reported to the registry. There was a tendency for invalid cases not to be reported, but 14 percent of the reported cases were invalid. Once patients were reported, regardless of diagnostic validity, approximately one-third were lost to registry followup by 1966. Presence of rheumatic heart disease correlated with initial reporting to the registry, but once reported, presence or absence of rheumatic heart disease was not related to continued registry followup. These data suggest the extent of the error introduced by using the registry as a basis for estimating the incidence of rheumatic fever and rheumatic heart disease.

Effectiveness of the registry in stimulating physican and community education was evaluated by surveying the management of streptococcal infections in Maryland. Among 261 first attacks of rheumatic fever, 34 percent had no history of prior respiratory infection. Thirty-two percent had respiratory infections but did not consult a physician. The remaining 34 percent had respiratory infections and consulted physicians but, nevertheless, developed rheumatic fever. The data indicate that physicians make inadequate use of throat cultures, suggesting that these infections may have been inappropriately managed.

The reporting of new cases of rheumatic fever to the registry by physicians is important and efforts should be expended toward increasing such reporting, but a greater need is to explore new methods of casefinding. One approach is monthly monitoring of hospitals, either by phoning the hospital record rooms or by having a registry staff member review the charts at each institution. Ideally, the best method of verifying the diagnosis for each new case of rheumatic fever is examination of each new patient in a clinic staffed by expert clinicians experienced in the diagnosis of rheumatic fever.

The main objective of any rheumatic fever registry is effective followup of patients for continued prophylaxis. The structuring of an active followup program within the registry framework is needed. Additional personnel are required for direct followup and to coordinate other available resources for followup in the community. A priority scale could be developed for patients based on factors relating to the risk of recurrence and the risk of noncompliance. Intensive followup services could then be directed to the high-risk patients assigned priority ratings based on both sets of factors.

The approach described in this study can serve as a model for evaluating registries for chronic illnesses with low fatality rates.

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# A WAY TO ACQUAINT PEOPLE IN THE COMMUNITY WITH MENTAL HEALTH FACILITIES

With the opening of its three new mental health centers in Santa Clara County, Calif., the Santa Clara County Health Department was called upon to provide a formal public dedication and open house for each of its new centers early in 1968. Each open house was held on a separate date. Invited to these day-long events were mental health workers as well as other interested persons.

The mental health centers were built under the Federal Mental Health Services Act of 1963 and are programed to offer emergency, outpatient, day treatment, education and consultation, and training services. Inpatient services are available through financial contracts with local hospitals having psychiatric units. Each center serves a population of about 200,000.

The purpose in designing the open house events was to convert a routine function into an effective learning opportunity, and to minimize demands on staff time through a self-guided tour involving a guide sheet and easel display located at key points throughout each building.

The tour guide sheet included 10 stopping points at which black and white photographs (24 by 28 inches) were located to provide visual interpretation to reinforce the tour guide sheets. These photographs portray the mental health professional in his helping role by showing him in his relationship to a patient. The dynamic aspects of this relationship were stressed. For example, the photograph shown illustrates one of the roles of a psychiatrist, stressing the interpersonal dynamics of the helping process.

The format of the self-guided tour incorporates a number of elements of an effective communication process: (*a*) delineating the varied functions of a building which, for the most part, appears to be a series of similar offices, (*b*) freeing people to proceed at their own rate, allowing for lingering at one room longer than another, (*c*) succinctly describing


in the tour sheet the major functions involved in what the visitor is viewing, and (*d*) coordinating for maximum effect, the elements of the building structure and equipment, the professional in the photograph, and the described activities.

There were a few difficulties in the operation of these self-guided tours. Unless people were given the tour guide immediately upon entering the building, they usually did not use it. People appeared to need definite and firm directions. Throughout the buildings, directional arrows and numbers were placed to guide people to each of the tour points, but many persons did not look at the arrows; therefore a staff member stood at the starting point to provide personal guidance.

Movement to and from tour points was smooth. When large groups of people started through there was some confusion, but soon they thinned out due to differing paces. Although staff members were not needed as tour guides, some were casually available throughout the buildings to answer questions at various points during the tour.

Volunteer women from the Mental Health Association of Santa Clara County provided information, self-guided tour sheets, and coffee and cookies following ceremonies and during the tour hours.

After the self-guided tour is designed, it can be quickly set up and used again. In addition, the tour sheet alone can be used when a large group is taken through the facility, with a staff person pointing out the appropriate paragraphs in relation to what is being seen.—Dr. Alexander V. Monto, *psychiatrist and chief of the indirect services unit, community mental health program, Santa Clara County Health Department, Calif., and* William D. Miley, *associate director of public health education, Ventura County Health Department, Calif.*