Visiting Consultant Clinics to Study Prevalence Rates of Juvenile Rheumatoid Arthritis and Childhood Systemic Lupus Erythematosus Across Dispersed Geographic Areas

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ABSTRACT. ***Objective.*** Visiting consultant clinics (VCC) may provide pediatric rheumatologic care to children in rural populations, but the clinical demands have not been studied. We studied whether these clinics could be effective in determining prevalence rates of rheumatic illness like juvenile rheumatoid arthritis (JRA) and childhood systemic lupus erythematosus (SLE) across large dispersed geographic areas.
Methods. The study population included children diagnosed with JRA or SLE at the only civilian pediatric rheumatology center in the State of Hawaii. Prevalence rates of these illnesses were then calculated for the urban and more rural neighbor island areas. VCC and prevalence data were calculated over a 10-year period.
Results. We found a lower prevalence of JRA in the urban area (38.3 per 100,000) when compared to the rural neighbor islands (63.2 per 100,000). However, an equivalent prevalence of SLE was found in the urban (24.0 per 100,000) and neighboring islands (21.8 per 100,000). Clinical demands increased significantly with the success of the VCC, and with an increase in pediatric rheumatologic staffing.
Conclusion. We found an increased prevalence of JRA in rural areas when compared to urban areas. Similar prevalence rates of SLE suggested the finding was not due to referral bias alone. VCC are useful to estimate disease prevalence over large areas, and therefore make it possible to identify areas at greater risk. Further investigations are needed to elucidate the possible environmental and genetic factors that may explain the regional differences in JRA prevalence. (First Release Jan 15 2007; J Rheumatol 2007;34:425–9)

Key Indexing Terms:
PEDIATRIC RHEUMATOLOGY EPIDEMIOLOGY PREVALENCE
JUVENILE RHEUMATOID ARTHRITIS SYSTEMIC LUPUS ERYTHEMATOSUS

Rural or remote areas of low population density have limited access to pediatric specialty services. Such areas do not have a sufficient population of children with the condition of interest to support a pediatric specialist, but may still have children who suffer from the specific illnesses that the specialist is trained to care for. This problem is evident in pediatric rheumatology where the disease can cause disability or death if not treated adequately. Visiting consultant clinics (VCC) have been used as a method of providing rural areas access to other needed specialty services. Such programs have been successful in cardiology, orthopedics, urology, and oncology[1,2]. Little has been written about providing pediatric rheumatologic services utilizing VCC.

Few comparative studies of the prevalence of juvenile rheumatoid arthritis (JRA) and childhood systemic lupus erythematosus (SLE) in different geographic areas are available[3]. Mauldin, *et al*[4] recently reported a higher prevalence of JRA in rural Oklahoma when compared to urban areas. Their study was based on a disease registration database of Native Americans, so cases could not always be confirmed[4]. Descriptive studies of prevalence could help to identify possible risk factors important to the development of these conditions[3].

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Supported by grant number RR003061 from the National Centers for Research Resources, National Institutes of Health. Contents are solely the responsibility of the authors and do not necessarily represent the official views of the NCRR or NIH. Also supported by the Alexander and Baldwin Foundation, A&E Vidinha Charitable Trust, First Hawaiian Foundation, Garmar Foundation, Kapi'olani Children's Miracle Network, Shriner's Hospital for Children, and Weinberg Foundation.

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Accepted for publication November 9, 2006.

Utilizing VCC, we were able to assess disease point prevalence on our neighbor islands and compare them to the urban island of Oahu. In Hawaii, there is a single pediatric rheumatology center (PRC) providing specialty care for the state's entire civilian population. The situation in Hawaii is unique due to the geographic separation of the Hawaiian Islands by large expanses of ocean, isolating heavily populated Oahu from the less populated neighbor islands.

Prevalence rates of JRA and SLE historically have been determined either in defined geographic areas or in multicenter collaborative studies; however, with a single PRC servicing the 4 major islands (Oahu, Hawaii, Maui, and Kauai) through the utilization of outreach clinics, disease stratification can be easily ascertained.

We wondered if JRA and SLE point prevalence might differ in different geographic settings. VCC provided a mechanism to study the prevalence in the State of Hawaii, and allowed the comparison of these 2 conditions. We calculated disease point prevalence for JRA and SLE over a 10-year period in the civilian population for all of the main islands noted above.

MATERIALS AND METHODS

The patient population included all children seen in the PRC, either at the center or through the VCC. The PRC is located in the only full service children's hospital in the state, Kapi'olani Medical Center for Women and Children (KMCWC), and is located on Oahu, in downtown Honolulu, the state capital. Patients are referred to the PRC only through their primary care physicians, and children from the entire state are referred to it. There are no rheumatologists in practice on these neighboring islands, with the exception of Oahu. This then allows for studies to be done on the prevalence of childhood rheumatic illnesses.

Diagnostic criteria. All patients were categorized into disease-specific groups. We focused on the 2 more common illnesses: JRA and SLE. The diagnostic criteria used were the consensus criteria for SLE by Tan, *et al*[5] and for JRA, the American Rheumatism Association JRA criteria[6]. As in other studies of rheumatic populations, we separated the spondyloarthropathies from the JRA population[7-10]. This may have had the effect of lowering the prevalence rates when compared to other historical studies[11].

Location of study. There are 4 major islands in the state of Hawaii: Oahu, Maui County (which includes the smaller islands of Lanai and Molokai), Hawaii (Big Island), and Kauai. These islands possess approximately 99% of the total population of the state (2000 census)[12]. Regular VCC were held on Maui, Hawaii, and Kauai. When hospitalization was necessary, the patients were usually flown to Oahu for hospitalization at KMCWC. All patient encounters, both PRC and VCC visits, had the diagnosis, age, and basic demographics recorded. A tally of unduplicated patients was generated on each illness seen, and then de-identified. The Institutional Review Board granted an exemption for this study.

Time period. Cases were accrued over 10 years, from January 1, 1993, to December 31, 2002.

Statistical analysis. Data were analyzed by comparing cases of JRA and SLE on the neighbor islands (more rural) to Oahu (urban). The point prevalence of these disorders was then calculated over the 10-year period using 2000 census data[12]. Confidence intervals (CI) of 95% from these data were estimated using the normal approximation for a binomial distribution for descriptive comparisons. Statistical significance of differences between prevalence rates in rural versus urban areas was performed by Poisson regression using PROC GENMOD procedures with SAS version 8.2 (SAS Institute, Gary, IN, USA).

With increased physician staffing size in 1996, we studied this effect on the number of neighbor island clinics and patient visits. To account for these changes the t test was used to calculate significance using the calculated means pre- and post- 1996.

RESULTS

A total of 207 clinics were held on the neighbor islands during the 10-year period (Table 1). Children with chronic rheumatic disease usually need monthly followups, and this was nearly accomplished on Hawaii and Maui. The number of outreach clinics, and patient visits seen in these clinics increased over time (Figure 1). The greatest numbers of patients evaluated in these outreach clinics were on the islands of Hawaii and Maui, which also have the greatest number of children outside of Oahu (see Table 1). A total of 1723 patient visits occurred during the 10-year period.

During 1996, the PRC added an additional pediatric rheumatologist (KSY). Up to 1995, there had been only one (DKK). From 1993 through 1995, the PRC averaged 16 outreach clinics per year. This increased to 23 clinics per year during the period 1996 to 2002 ($p < 0.001$). Similarly, the number of patient visits increased from a mean of 106/year to 201/year during the same period ($p < 0.005$). This increase of patient visits was found to be 90%, a finding similar to that of pediatric cardiology outreach clinics, which found an increase of 61% in a 5-year time frame from 1991 to 1996[13]. The increase in PRC visits was most likely due to an increase of staffing, but may also have been accountable by an increased demand for these services[13].

A total of 204 patients with JRA and SLE were seen during the 10-year period for all islands. Of this group, 135 (66%) had the diagnosis of JRA, and 69 (34%) SLE. On Oahu we cared for 80 patients with JRA, while there were 55 in the neighbor islands combined. For SLE we followed 50 patients on Oahu and 19 combined on the neighbor islands. Since the majority of the population of the state lives on Oahu, calculating the prevalence data allows for a more accurate comparison.

The point prevalence of JRA and SLE was calculated for each island and compared, as depicted in Table 1. JRA was found to be more prevalent than SLE in all of the islands studied. This finding is consistent with previous studies of pediatric rheumatology clinics[7,8,10]. The larger islands of Hawaii and Maui had prevalence rates similar to those of previous studies on prevalence of JRA (Table 1).

Due to the small numbers on each individual neighbor island, the data were grouped from these sites and then compared to the larger urban site of Oahu. The comparison of this prevalence is displayed in Table 1. Although JRA prevalence CI show a small overlap based on the approximate CI, when urban is compared to rural populations, using Poisson regression, the difference between these populations was statistically significant ($p < 0.01$). In contrast, the SLE prevalence rate CI showed dramatic overlap, and were not statistically significant in Poisson regression ($p = 0.78$).

Table 1. Point prevalence over a 10 year period for JRA, childhood SLE, and JAS over the State of Hawaii. The point prevalence for JRA, SLE, and JAS was calculated for the time period of 1993 to 2002. Census data from 2000 was used to calculate these prevalence rates presented in # of children per 100,000 children at risk. 95% confidence intervals are included. The point prevalence of JRA was higher on the more rural neighbor islands when compared to the urban island of Oahu. Point prevalence of JAS revealed a similar trend to JRA, but did not reach a significant difference, possibly due to lower numbers. The point prevalence of SLE was similar throughout the state. The neighbor islands were considered rural, as they do not have access to pediatric rheumatologic specialty care, except for outreach clinics.

Island	2000 Census (Ages < 1 to 19)	No. of JRA Patients	JRA Prevalence	No. of SLE Patients	SLE Prevalence	No. of JAS Patients	JAS Prevalence
Oahu	208,758	80	38.3 (29.9–46.7)	50	24.0 (17.3–30.6)	20	9.6 (5.4–13.8)
Hawaii	38,852	25	64.3 (39.1–89.6)	10	25.7 (9.8–41.7)	5	12.8 (1.6–24.0)
Maui	32,711	23	70.3 (41.6–99.0)	6	18.3 (3.7–33.0)	3	9.2 (–1.2–19.6)
Kauai	15,443	7	45.3 (11.8–78.9)	3	19.4 (3.9–34.9)	3	19.4 (–2.6–41.4)
Rural (ave)	87,006	55	63.2 (46.5–79.9)	19	21.8 (12.0–31.7)	11	13.8 (5.6–22.0)

JRA: juvenile rheumatoid arthritis; SLE: systemic lupus erythematosus; JAS: juvenile ankylosing spondylitis.

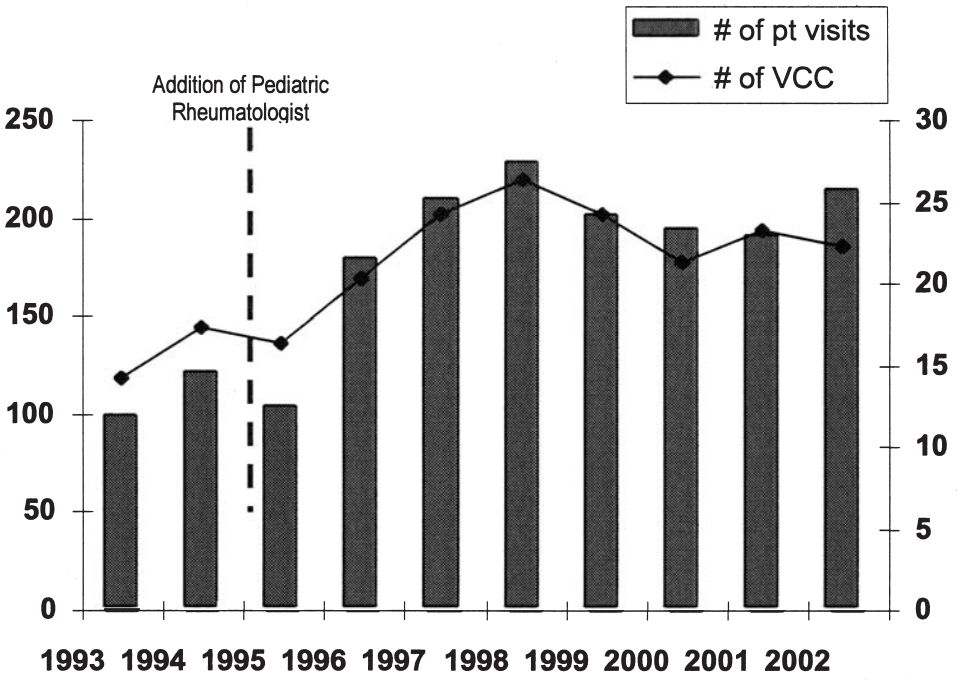

Figure 1. Number of VCC patient visits and rural/neighbor island VCC per year. Both the number of clinics and patient visits increased significantly after 1996 [clinics ($p < 0.001$) and patient visits ($p < 0.005$)]. An additional pediatric rheumatologist was added in that year, allowing for a consistent increase in patient visits and neighbor island outreach clinics. These increases were sustained because the 2 rheumatologists could now cover both the urban base and rural clinics more effectively.

The relative risk for development of JRA on the neighbor islands was calculated in comparison to Oahu and found to be 1.65 (95% CI 1.17–2.33) compared to SLE, which was found to be 1.10 (95% CI 0.65–1.86). To study this further, spondyloarthropathy point prevalence was also compared for Oahu versus the neighbor islands and not found to be statistically significant, although the trend was for a higher prevalence on the neighbor islands, but with limited numbers (Table 1).

DISCUSSION

The situation in Hawaii is unique and allowed us to study the prevalence of JRA and childhood SLE distributed over a large area separated geographically. The population tends to be fairly static due to the separation by large bodies of water. Commercial air flights are available between the islands, but travel by boat is difficult due to the long distance between islands (100 to 200 miles). There have been few published

reports concerning the prevalence of rheumatic disease over such widely dispersed geographic areas. One article surveyed adult RA in rural and urban areas of Taiwan[14]. A significantly higher prevalence of RA was found in the suburban/urban areas as compared to the rural areas. The opposite was seen here: the prevalence rates were higher on the more rural neighbor islands. A number of possibilities could explain these differences. Our study was based on JRA and childhood SLE and not adult RA, and this difference in age groups may account for a difference. Another explanation might be that disparities in living conditions and behavioral and cultural factors between rural and urban regions may differ in Taiwan compared to Hawaii. Another important difference between these 2 studies is that our data are based on physician specialist diagnosed cases, which are in general more accurate than survey data and self-reported diagnoses.

Hawaii may more closely mirror the report from Oklahoma, where an increased prevalence of JRA was found in rural areas versus the urban environments[4]. Our prevalence rates were lower; however, all of their cases were not confirmed by the research physicians and were reported from a large database of Native Americans. Our study investigates this finding more thoroughly by including only cases diagnosed by the pediatric rheumatology specialists. The cases in this study were all seen and followed by the PRC, and strict attention was paid to the diagnostic criteria. Additionally, clinical followup was provided through the urban and rural VCC to further strengthen the proper diagnosis. It is encouraging to see that the 2 studies, surveying very different geographic areas, have prevalence rates that fall in the range of previous studies, as reviewed by Schneider and Passo, who found prevalence rates ranging between 14.8 to 148.1/100,000[15]. A recent report examining the nationwide prevalence in Taiwan found very low prevalence rates for both juvenile chronic arthritis (JCA) and pediatric SLE of 3.8 and 6.3 per 100,000, respectively. These data were collected from Taiwan's national insurance program, not confirmed by rheumatologists, so it is conceivable that some cases were misdiagnosed or undiagnosed[16]. However, studying an entire nation's prevalence rate provides intriguing data.

In our study, there may have been an element of referral bias. We were more likely to capture all cases on the rural islands because there was no other available rheumatologic care there. The lower number seen on Oahu may reflect referral bias as there are 10 adult rheumatologists and one military pediatric rheumatologist (DAP). Even if the total military population of children is subtracted from the population on Oahu, the prevalence rate of JRA would still be significantly lower than that of the rural areas (data not shown). However, it is interesting that the Oahu prevalence rate of SLE was similar to the other islands. An alternate explanation for this finding is that the rural populations are at increased risk of JRA versus the urban children. In SLE, there appear to be similar prevalence rates between the urban and rural communities. The finding that SLE is similar in prevalence rate and JRA is decreased in the urban area is an argument against a substantial effect from referral bias. However, it is possible that some of these other adult specialists would feel more comfortable caring for a child with JRA in comparison to one with SLE. This possibility is difficult to evaluate as no database exists for these other specialists, and would require a comprehensive prospective collaborative research study.

The other possibility for the finding of a higher prevalence of JRA on the neighbor islands is that these children may be at increased risk of developing JRA versus the population of the more urban Oahu. The increased risk of JRA on the more rural neighbor islands is not seen in childhood SLE, and provides evidence of different pathogenic mechanisms causing these 2 autoimmune diseases. The underlying reason for this difference may be due to genetic or environmental etiologies, and would require future studies to confirm. The impact of socioeconomic factors is difficult to study, as changes in the ethnic populations on the neighbor islands have occurred over time. Increases in the Caucasian ethnic group have elevated education levels and income levels in these areas, as seen in ongoing studies (AG, personal communication), so it is difficult to extrapolate from census data for the answer. To evaluate these factors within this population is very complex, as detailed stratification would reduce the groups to very small numbers, and make it difficult to answer these questions statistically.

The benefits of holding VCC have been well documented[1,2,17-22]. These clinics offer needed specialty services to underserved populations, helping to lessen health care disparities in those less accessible communities. In one review of 10 years' experience of pediatric outreach clinics, it was concluded that VCC increased parental and professional choice, access to pediatric consultant services, and service flexibility, reduced unnecessary hospital visits, and enabled more rational and relevant clinical decision-making[19]. A recent review of the Cochrane Database yielded 73 outreach intervention publications; 9 of these meet their inclusion criteria for specialist outreach clinics, with evidence that specialist outreach can improve access, outcomes, and service use, especially when delivered as part of a multifaceted intervention[20]. In addition to these benefits, this outreach provides a service to primary care providers to evaluate and diagnose their patients with rheumatic diseases earlier, resulting in improved health outcomes.

To maintain these pediatric specialty outreach clinics may require an increase in pediatric specialty staffing to maintain teaching, research, and service commitments at the tertiary center[13]. This point highlights some of the difficulty of comprehensively facilitating these clinics, especially when outreach staff are in solo practice. Time spent at the rural outreach clinics is time away from the central urban clinics and hospital consults, making it difficult for one person to cover both areas. However, by increasing staffing these clinics can

be held on a more consistent basis, leading to improved surveillance and more comprehensive care. Another successful alternative in the literature is utilizing telemedicine clinics, and our PRC with generous foundation support has started to develop this type of a care delivery system[21,22]. If the clinical and academic demands of the specialists are balanced, the VCC may provide an excellent means of studying the epidemiology of chronic illnesses across different regions. Our data suggest that, given the similarity of SLE prevalence rates reported in our study, referral bias is also greatly reduced, which may have the combined effect of enhancing patient care, as well as providing a means for estimation of the public health impact of pediatric rheumatologic disorders.

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