Incidence of Vasculitis in Children in the Czech Republic: 2-Year Prospective Epidemiology Survey

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ABSTRACT. ***Objective.*** To determine the incidence and presenting features of primary and secondary vasculitides in children across the Czech Republic.

Methods. The population of 2.02 million children under 17 years of age was surveyed over 2 years. Cases were identified through monthly questionnaires posted to consultant pediatricians in all hospital pediatric departments in the country. Patients were included in the analysis if they met established inclusion criteria for each diagnosis and had disease onset between 1997 and 1999. Incidence rates were calculated from population rates derived from the 1991 Census.

Results. We identified 452 new cases of vasculitis and connective tissue disease. The estimated annual incidence of Henoch-Schönlein purpura (HSP) was 10.2/100,000 children, with a mean age at onset of 7 years. At disease onset palpable purpura was present in all cases; arthritis/arthralgia in 52%; abdominal pain and/or gastrointestinal bleeding in 40%; hematuria/proteinuria in 15%; and genital involvement in 2.8%. Forty-nine percent of all patients with HSP received short term corticosteroids. The estimated annual incidence of Kawasaki disease (KD) was 1.6/100,000 children under 5 years. Thirteen percent of patients with KD had transient dilatation of coronary arteries; 75% received high dose intravenous immunoglobulin. Other primary systemic vasculitides were extremely rare in this population. Secondary vasculitides of connective tissue diseases had an estimated annual incidence of 0.22/100,000 for systemic lupus erythematosus and 0.19/100,000 for dermatomyositis.

Conclusion. We determined the incidence of different childhood vasculitides within a hospital based population throughout the Czech Republic. HSP was the most common, with a relatively high proportion of the patients treated with a short course of corticosteroids. A lower incidence than expected of KD raised the suspicion that some cases were not identified. Other childhood vasculitides were rare. (J Rheumatol 2004;31:2295–9)

Key Indexing Terms:
VASCULITIS HENOCH-SCHÖNLEIN PURPURA KAWASAKI DISEASE INCIDENCE
SYSTEMIC LUPUS ERYTHEMATOSUS JUVENILE DERMATOMYOSITIS
PRIMARY SYSTEMIC VASCULITIS CHILDREN

Among vasculitic disorders in childhood, Henoch-Schönlein purpura (HSP) and Kawasaki disease (KD) are the most common. Other primary systemic vasculitides, rare in adults, are even less common in children. This is also true for conditions characterized by the presence of variable degrees of secondary vasculitis, such as juvenile dermatomyositis (JDM) and systemic lupus erythematosus (SLE).

Recognition of epidemiologic data is important in searching for etiologic factors such as infection, environmental influences, and genetically determined factors. It is also needed for appropriate planning of health service requirements. We present the first report on the incidence of these conditions from Central Europe. We prospectively determined the incidence and short term outcome of different types of primary and secondary vasculitic disorders in the whole country over a 2-year period.

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Submitted September 4, 2003; revision accepted May 12, 2004.

MATERIALS AND METHODS

Study setting and design. In the Czech Republic the pediatric health care system is based on a network of primary care physicians, most of whom are trained pediatricians. Every child aged 0–19 years has to be registered with a pediatric practitioner. Where hospital care is needed patients are referred to the pediatric department of a district (primary), regional (secondary), or university (tertiary) hospital, led by pediatric consultants. The pediatric rheumatology service is not evenly distributed around the country. Where available, the service is covered by pediatricians with an interest in rheumatology and in a few university hospitals, by fully trained pediatric rheumatologists who work at or in close connection with the hospital. The total number of hospital pediatric units in the Czech Republic at the beginning of the study was 126.

Pediatric population of the Czech Republic. Population data were derived from estimates of the Czech Health Statistics for 1998, based on the 1991 Census[1,2]. Within the total population of 10.3 million inhabitants, 2.02 million were under the age of 17 years with a boy:girl ratio of 1.3:1.2. National rather than ethnic origin is collected by the census, and is highly

homogeneous, with over 97% of the population of Czech, Moravian, or Slovak origin. The largest non-Caucasian ethnic group is Romani, with estimates by the International Romani Union at 2.4–2.9% of the total Czech population.

Case identification. All hospital pediatric departments in the country were surveyed over a 2-year period from November 1997 to October 1999. Cases were identified by monthly postal questionnaires directed to the clinical heads of the pediatric units to be filled by them or by other delegated consultant pediatricians, one from each of the units. Members of the Czech pediatric rheumatology group were also included in the mailing list.

The questionnaires were based on those used in our previous epidemiological study in the West Midlands, UK[3]. The primary questionnaire, distributed regularly at the beginning of each month, briefly outlined the diagnostic/classification criteria for HSP[4], KD[5], SLE[6], and JDM[7], and requested cases of other vasculitides where there was strong clinical or histological support for the diagnoses. Recipients were asked to indicate "nothing to report" or to provide the number of patients with each of the diagnoses identified over the past calendar month. On receipt of a positive identification, a further detailed disease-specific questionnaire appropriate for the notified diagnosis was sent, requesting demographic, diagnostic, laboratory, and management information. Personal data apart from the sex and date of birth were not recorded. Organ involvement at disease onset was further analyzed in patients with HSP and related to the use of corticosteroids. For KD, information on presence of coronary artery abnormalities assessed by echocardiography and use of high dose intravenous IgG (IVIG) were analyzed.

Case confirmation. All returned questionnaires were reviewed by one individual (PD). Where there were insufficient data, or where doubt about the diagnosis existed, the hospital discharge summary was requested and/or relevant physicians were contacted by the investigator via telephone. Cases were included only when sufficient information to fulfil the inclusion criteria was available. To exclude case duplication the database was carefully examined for duplicates according to unique birth codes, dates of disease manifestations, and referring physician's name.

Demographic criteria. Cases fulfilled demographic criteria for inclusion if disease onset occurred between November 1, 1997, and October 31, 1999. Disease onset [or 4 of 10 American College of Rheumatology (ACR) criteria for the diagnosis of SLE] must have occurred before the patients reached their 17th birthday, and when they were resident in the Czech Republic.

Inclusion criteria. Children with the following diagnoses were included in our study: (1) HSP that fulfilled a modification of the ACR classification criteria[4]: at least 2 of palpable purpura without thrombocytopenia; age less than 20 years at onset; biopsy showing granulocytes around arterioles or venules; and gastrointestinal (GI) bleeding. (A modification was required because all children automatically satisfy the ACR age criterion, and diagnostic biopsies are seldom performed in children.) This gave heavy weighting to the presence of palpable purpura, which has a sensitivity of 88.2% and specificity of 79.9% for HSP[4]. The distribution of the rash was recognized by Mills, *et al* to further improve the specificity of the criteria[4]. In our study, therefore, children with isolated palpable purpura and no other features of HSP were excluded if the rash was not in the classical distribution. (2) KD that fulfilled the criteria defined by the American Heart Association[5]. (3) Primary systemic vasculitis (PSV) that fulfilled established classification criteria sets including Wegener's granulomatosis[8], polyarteritis nodosa (PAN)[9], and Takayasu arteritis[10]. (4) Either SLE[6] with disease onset between November 1997 and October 1999 or juvenile dermatomyositis/polymyositis (DM/PM) diagnosed using classification criteria of Bohan and Peter[7]. For cases of polymyositis magnetic resonance imaging (MRI) and/or histopathology evidence of muscle inflammation was necessary. (5) Children with other or undefined vasculitic disorders were also included.

Children with either infection or drug related vasculitis were excluded from the study.

Statistical analysis. Incidence rates were calculated using the number of incident cases as the numerator and the population of each subgroup derived from the Census of 1991 as the denominator. Ninety-five percent confidence intervals (95% CI) were calculated assuming a Poisson distribution using exact methods. Specific age-related incidence rates were calculated to allow comparison to previously published studies for KD (under 5 years) and HSP (under 14 years).

RESULTS

The mean primary questionnaire return rate was 72.5%. A total of 562 children were referred to the study. Sixty-one did not fulfil demographic or inclusion criteria for any of the disorders and were excluded from further analysis. Forty-nine children were reported as having some other sort of vasculitis or vasculitis-resembling disorders (Table 1). The remaining 452 children were included in the analysis. The estimated annual incidence rates for all diagnoses in comparison to other published series are summarized in Table 2.

HSP. A total of 410 children with HSP were referred to the study. The estimated annual incidence of HSP was 10.2 per 100,000 (95% CI 8.8–11.6 per 100,000) for all children under 17 years, and 10.1 per 100,000 (95% CI 8.8–11.9 per 100,000) in children under 14 years. The highest disease incidence was between the age of 5 and 9 years, with a mean age of 7.2 years (range 2–16.8). There appeared to be 2 seasonal peaks, in November and March, with the lowest incidence during the summer months. Individual organ involvement rates at initial evaluation are outlined in Table 3. All patients had palpable purpura, about half of them had joint involvement and/or GI manifestations, and 14% had microscopic hematuria either alone or in combination with mild proteinuria. Short term (less than 1 month) corticosteroid therapy was given to 49% of the patients with no clear correlation with the severity of clinical symptoms. Organ involvement in relation to the use of corticosteroids is shown in Table 4.

Kawasaki disease. A total of 23 cases of KD were reported with a mean age at onset of 2.8 years (range 0.2–13) and a ratio of boys to girls of 2.3:1. Nineteen patients were under 5 years of age. Of the 19 patients for whom an ethnic origin was reported, 18 were Caucasians and one was of Romani origin. The estimated annual incidence rate was 0.6 per

Table 1. Other vasculitic or vasculitis-resembling disorders.

Diagnosis	No. of Cases
Cutaneous purpura (other than HSP)	36
Erythema nodosum	8
Cutaneous polyarteritis nodosa	1
Isolated CNS vasculitis	2
Mucha-Habermann disease	1
Urticarial vasculitis	1
Total	49

CNS: central nervous system.

Table 2. Annual incidence of pediatric vasculitides per 100,000 children in comparison with different series.

Diagnosis (age)	Czech Republic	Austria[28]	Denmark[12]	UK[3,11,22,26,32]	Finland[29,30]	Sweden[24]	Australia[25]	USA[19,27,33]	Canada[20]	Japan[21,31]
HSP (< 14 yrs)	10.1		14–18	13.5–22.1						
KD (< 5 yrs)	1.6			3.4–5.5	3.1–7.2	6.2	3.7	8.1–18.5	7–46	108–111
JDM	0.19			0.19–0.4	0.30			0.4–0.8	0.15	0.16
SLE	0.22	0.48		0.8	0.37–0.9			0.4	0.28	0.47

Table 3. Organ involvement in HSP.

	No. of Patients (%)				
Total Followup Data	This Study	Stewart 1988[11]	Kaku 1998[13]	Balmelli 1996[14]	Reinehr 2000[15]
Palpable purpura	410 (100)	270	194 (100)	139 (100)	171 (100)
Arthritis/arthralgia	209 (51)			110 (79)	110 (64)
Gastrointestinal (pain and/or bleeding)	165 (40)			92 (66)	99 (58)
Renal*	58 (14)	55 (20)	65 (33.5)	60 (43)	49 (29)
Scrotal/penile	11 (2.8)			11 (7.9)	7 (4)

* Isolated hematuria or hematuria with proteinuria.

Table 4. Corticosteroid (CS) use in relation to the clinical presentation of HSP.

	No. of Patients (%)		
Organ/System Involved	Total	CS+	CS–
GI only*	129	66	63
Renal only*	27	19	8
GI + renal*	31	31	0
Genitals (± renal, GI)*	11	10	1
Skin ± joints only	212	74	138
Total	410	200 (49)	210 (51)

CS+: corticosteroids used; CS–: corticosteroids not used. * Internal organ involvement with palpable purpura with or without joint involvement.

100,000 (95% CI 0.36–0.85 per 100,000) in the whole childhood population under age 17 years, rising to 1.6 per 100,000 (95% CI 0.99–2.57 per 100,000) in children under the age of 5 years. Transient dilatation of coronary arteries was found on echocardiography in 3 patients (13%) at initial evaluation: no aneurysms were detected. Three patients (13%) had pericardial effusions. Sixteen patients (70%) received high dose IVIG on day 3–32 (mean 9.5) after fever onset. Twelve had infusions of 2 g IVIG/kg over 1–2 days, one had 1 g/kg, and 3 had 0.4 g/kg for 4 consecutive days. Cardiac involvement was not reported in any of the 5 patients who did not receive IVIG. In 20 cases acetylsalicylic acid in doses ranging from 40 to 100 mg/kg/day was given during the acute stage of the illness. In one patient corticosteroids were added to the standard therapy; 2 patients who did not receive IVIG were treated with corticosteroids only.

Connective tissue diseases. A total of 7 cases of JDM were identified with an estimated annual incidence of 0.19 per 100,000 children (95% CI 0.08–0.4 per 100,000). Four patients had definite and one probable JDM; 2 patients did not have skin involvement and fulfilled criteria for the diagnosis of polymyositis. Nine patients with SLE were reported with annual incidence of 0.22 per 100,000 children (95% CI 0.04–0.4 per 100,000). The incidence was higher in girls aged 11–16 years [1.03 per 100,000 (95% CI 0.9-4.04 per 100,000)]. The mean age within the JDM group was 8 years (range 3–15) with a ratio of girls to boys of 1.3:1. The mean age of SLE patients was 13.7 years (range 9–16) and all were girls. All patients in this group were Caucasian.

Primary systemic vasculitis. Only 3 cases of PSV fulfilling classification criteria were identified, comprising one case each of PAN, Takayasu arteritis, and Wegener's granulomatosis (WG). The calculated annual incidence of PSV was 0.06 per 100,000 children (95% CI 0.012–0.7). In PAN and Takayasu arteritis the clinical diagnosis was confirmed by direct angiography; in WG it was confirmed by the presence of characteristic histopathological findings on renal biopsy.

DISCUSSION

This is the first report estimating the incidence of primary childhood vasculitis in a prospective population based survey using strict inclusion criteria in a single European country. We found the estimated annual incidence of HSP in children under 14 years of age is close to that of 13.5 and 14/100,000 children at risk reported by Stewart, *et al*[11] (England) and by Nielsen[12] (Denmark), respectively, suggesting that similar genetic and environmental factors might play a role in disease etiopathogenesis around Europe. Nevertheless it is substantially lower than those found in the

most recent surveys from the UK[3] and Copenhagen[12]: 22.1 and 18/100,000 children, respectively. It had been a common practice in the Czech Republic that patients diagnosed with HSP were routinely admitted to pediatric hospital wards for observation. Since changes to the healthcare system have been proceeding, this might have changed over time. Although we believe that the majority of HSP cases have been referred to hospitals it is not clear what proportion of HSP patients might have been followed by first-line practitioners, and therefore were not detected by our study.

The distribution of organ involvement (other than nephritis) does not differ significantly from published series[11,13-15]. The lower incidence of renal manifestations in our study reflects the short period of followup, where only presenting manifestations were reported. It has been shown that nephritis can manifest within a variably long interval ranging from weeks to years after acute illness[13,14]. Management of HSP is a matter of ongoing debate. Corticosteroids are commonly used for subjective relief of more severe GI or genital manifestations; however, their role in preventing development of renal disease has been advocated[16,17] but not confirmed[18]. Our study found that about half of the patients received corticosteroids during acute illness. Interestingly, there appeared to be no correlation with severity of internal organ involvement and corticosteroid usage. The extent of skin involvement was not specified in the questionnaire, so we can only speculate that patients without organ involvement who received corticosteroids had more severe skin manifestations.

Our estimated annual incidence of KD differs from reports from other countries. Not surprisingly it is about 10 times lower than in North America[19,20] and 100 times lower than in Japan[21], but it is also 2–3 times lower than in other European countries and Australia[3,22-25]. Ethnic differences in incidence figures reflecting an importance of genetic makeup in disease susceptibility explain lower incidence rates in European countries in comparison with Japanese[21] and American populations[19,20], but they can hardly be responsible for differences seen in north-European studies[23,24]. In our cohort only one patient was reported as of non-Caucasian (Romani) origin. Nevertheless, the low KD incidence we found may reflect an insufficient awareness of the clinical manifestations leading to underdiagnosis of this entity. The occurrence of coronary heart disease, ranging between 14% and 25% of patients in reported series[22-25], is similar in the Czech patients (13%). Nevertheless, the true incidence of coronary abnormalities cannot be estimated by our study due to the short patient followup (usually less than 1 month). The diagnosis of KD was made by retrospective case analysis in 3 patients reported initially as having some other sort of vasculitis, who did not receive IVIG during an acute stage of the disease. For the majority of patients, a diagnosis of KD was immediately followed by IVIG administration. Still, there remained 5 additional patients who did not receive IVIG. The proportion of untreated patients (35%) was similar to some reports (21–39%)[22,25], but lower than others (9%)[24]. Overall, our study has raised an important issue of possible underdiagnosis and probably also undertreatment of KD in the Czech Republic.

It has been shown that the incidence of SLE and JDM is higher in black and Asian populations[3,20,26,27]. Relative ethnic homogeneity of the Czech population, with over 97% Caucasians, may have contributed to the lower incidence figures reported here. Nevertheless, for SLE it is also somewhat lower than the incidence of 0.48 or 0.9/100,000 children reported in other Caucasian populations in Austria and Finland, respectively[28-30]. For JDM our figures are similar to those reported from Canada, Japan, and the UK (0.15, 0.16, and 0.19, respectively)[20,31,32]. It is of interest that the diagnosis in 2 patients reported by their physicians initially as JDM without skin manifestations was questioned, and after reevaluation the final diagnosis turned out to be dystrophinopathy of Becker's type in one of them (boy) and carrier state of Duchenne's dystrophy in the other (girl). Neither of these patients was included in the analysis. The remaining 2 patients without typical JDM skin manifestations were diagnosed as having polymyositis. They both had, apart from proximal muscle weakness, muscle enzyme elevation, myopathic electromyographic inflammatory changes on T-2 weighted MRI images, and typical muscle biopsy findings. The proportion of polymyositis patients in this study (25%) is relatively high compared to larger series from the US and the UK (3, 0, and 6%, respectively)[26,32,33]. In contrast to its current rarity in the US, polymyositis had been reported in 17 out of 43 patients in Hanissian's series[34]. Differential diagnosis of chronic juvenile polymyositis can be far more difficult than that of dermatomyositis, as the diverse group of inherited muscle disorders can resemble most of its clinical signs.

In order to design our study as simply as possible we used only one system of data collection instead of more accurate case ascertainment methods like capture-mark-recapture analysis. We believed that in current pediatric practice in our country, the vast majority of seriously ill children were admitted to pediatric wards of general or faculty hospitals and if properly evaluated and reported, their diagnoses should not have been missed. However, the proportion of adolescents who might have been admitted to the adult departments or patients who were not referred to the hospital and were followed on an outpatient basis is not known.

We have estimated, for the first time, incidence rates of childhood vasculitides in a central European country with an ethnically homogeneous white population. We describe the distribution of initial manifestations of the more common primary vasculitides as well as current therapeutic practice. Low incidence figures for Kawasaki disease have raised the possibility of underdiagnosis of this important entity in the Czech Republic, with unfavorable prognostic implications.

ACKNOWLEDGMENT

We thank all the pediatricians who participated in the survey and enabled its realization. We also thank Dr. Janet Gardner-Medwin, Consultant Pediatric Rheumatologist, Glasgow, UK, for helpful comments; Dr. Ing Jaroslav Chládek, Faculty of Medicine, Hradec Králové, for statistical advice; and Blanka Aubrechtová for secretarial assistance.

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