Incidence of congenital clubfoot in Sweden

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Background **Idiopathic clubfoot is one of the most common congenital orthopedic problems. Nationwide studies of the incidence are scarce. We performed a prospective multicenter study in order to assess the cumulative incidence in Sweden over 2 consecutive years.**

Patients and methods **44 clinics identified as treating clubfoot reported new cases prospectively. The medical records of 280 children with clubfoot born during 1995–1996 were collected and analyzed with special reference to gender, regional distribution and seasonal variation.**

Results **The average cumulative incidence of clubfoot during the study period was $1.4/10^3$ (95% CI 1.2–1.6). Three-quarters of the cases were boys. In half of the cases both feet were affected. There was significant regional heterogeneity, but no seasonal variation in occurrence of clubfoot.**

Interpretation **The cumulative incidence was higher than in earlier Scandinavian studies. Gender distribution and laterality were similar to those in previous reports. We found significant regional differences in incidence, but the cause of this observation must be investigated in greater depth.**

Idiopathic clubfoot is a common congenital orthopedic problem of unknown but complex etiology, and with a reported cumulative incidence of 0.64–6.8 per 1,000 live births (Ching et al. 1969, Cartlidge 1984). In several previous studies performed in different regions of Scandinavia (Nilsonne 1927, Thomasen 1941, Severin 1956, Reimann 1967, Somppi 1984, Danielsson 1992) the reported cumulative incidence of congenital clubfoot was stable (Table 1). During the observed period 1913–1990, the various authors revealed similar figures of between $0.8/10^3$ to $1.0/10^3$. There has, however, been no national study in Sweden to confirm these estimates, except for the one by Severin (1956). Furthermore, nationwide surveys and regional comparisons regarding the cumulative incidence of idiopathic clubfoot have been associated with complicating factors such as variations in diagnostic criteria and the registration methods used (Chesney et al. 1999). Such conceivable inaccuracies caused the Swedish Registry of Congenital Malformations to exclude clubfoot as a registry diagnosis during the period 1982–1999.

In an attempt to improve the accuracy of the registered diagnosis with coherent diagnostic criteria, we performed a prospective, nationwide multicenter study in collaboration with the Swedish Pediatric Orthopaedic Society with the main aim of assessing the cumulative incidence of congenital clubfoot in Sweden over 2 consecutive years.

Patients and methods

In 1994, all public Orthopedic Departments in Sweden (n 65) were approached and 44 clinics reported that they treated newly diagnosed cases of congenital club foot. At each of these hospitals, we recruited a contact person to be responsible for the local registration during the recruitment period. To our knowledge, no private hospitals in Sweden were treating clubfoot during the study period.

Children with the ICD-9 code of congenital clubfoot (754F) were included. The sampling period started on January 1, 1995 and ended on December

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Table 1. Cumulative incidence estimates of congenital clubfoot in previous Scandinavian studies

Author	Period	Country	Cases (n)	Number of living births (in thousands)	Estimated cumulative incidence per thousand (95% CI)
Nilsonne	1913–1926	Sweden	34	33´	1.03 (0.74–1.44)
Severin	1936–1945	Sweden	797	1,076´	0.74 (0.69–0.79)
Thomasen	1931–1938	Denmark	29	36´	0.81 (0.56–1.16)
Reimann	1956–1962	Denmark	226	235´	0.96 (0.85–1.10)
Sompii	1963–1978	Finland	66	67´	0.99 (0.78–1.25)
Danielsson	1946–1990	Sweden	128	138´	0.93 (0.78–1.10)

31, 1996. For this period, we requested the medical records of 292 children who had been identified and reported in a preliminary way as new cases of cub foot. All the records that were available were scrutinized and children with positional clubfeet (n 3) and cases with additional neurological findings (n 3) were excluded, together with 6 preliminarily reported cases that could not be identified later on or which had missing records. Thus, 280 children were finally included in the present study.

Statistics

To compare the annual number of newborn children with congenital clubfoot with the total number of live births during 1995 and 1996, we used official reports concerning native data from the Swedish Board of Statistics (SCB). The cumulative incidence was calculated as the ratio between the number of children with clubfoot born during 1995–96 and the number of native births during the same period. For the estimated incidence, the 95% confidence intervals (CIs) were thus calculated, both for the total number of cases and for the subgroups year and sex. Abortive cases and stillbirths were not included in the population at risk and therefore, theoretically, the term prevalence proportion would be the most appropriate disease measure (Rothman 2002). However, we prefer to use the term cumulative incidence to avoid confusion when comparing the results of this study with those from earlier reports, which have generally used incidence (meaning cumulative incidence and not rate) as a description of clubfoot occurrence.

To assess the geographical distribution of clubfoot deformities, the country was divided into 6 regions commonly used by Health Service Authorities for administrative purposes (Figure 1). The chi-square test was used for grouped data and a p-value of < 0.05 was considered significant.

We also assessed seasonal variation by describing the monthly distribution of clubfoot deformities around the year (monthly cumulative incidence). We used a circular scale starting with January at 0º and the following months at 30º increments (Fischer 1993, Robertson and Corbett 1997) (Figure 2). The angle 0º (range 345–15) represents January and the scale ends up with December at a mean angle of 330º (range 315–345). The average vector represents mean birth month for the children. We estimated one vector for children born with

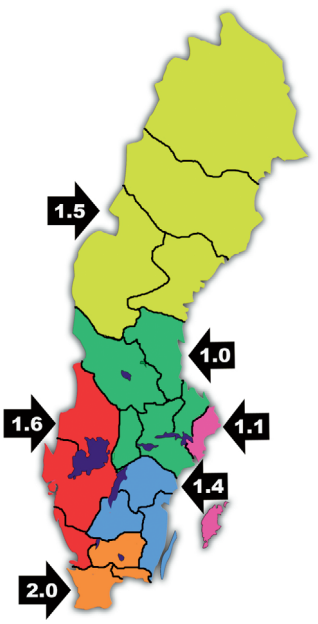

Figure 1. Cumulative incidence of idiopathic clubfoot during 1995–96 in 6 regions of Sweden. See Table 5 and text.

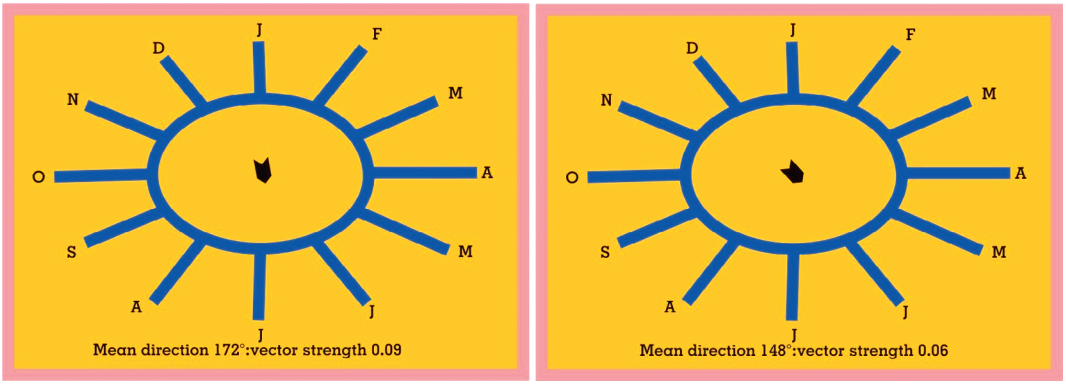

Figure 2. Monthly distribution of births in Sweden during 1995–96 for A. Congenital clubfoot (n 280) mean vector corresponding to July. B. Native births (n 198,719) mean vector corresponding to June.

Table 2. Number of children with idiopathic clubfoot in relation to number of native births

Year	No. of children		Cumulative incidence per thousand (95% CI)
	with clubfoot	born alive	
1995	157	103,422	1.5 (1.3–1.8)
1996	123	95,297	1.3 (1.1–1.5)
Total	280	198,719	1.4 (1.2–1.6)
Boys	198	101,874	1.9 (1.7–2.2)
Girls	82	96,845	0.8 (0.7–1.1)

Table 3. Distribution according to gender, side and bilateral occurrence of clubfoot

	Girls (n 82)	Boys (n 198)
Right side	27	56
Left side	21	48
Bilateral	34	94

Sex difference of bilateral vs. unilateral clubfoot, p = 0.3.
Left vs. right side, p = 0.7.
Sex difference of side p = 0.5.

congenital clubfoot and another vector for all other children born in Sweden during the same period. The advantages and methodology of analyzing circular data have been thoroughly described by Zar (1999). We used the Rayleigh z-test (Fischer 1993) to test whether there was a uniform distribution of seasonal cumulative incidence for the children with clubfoot and for all other live births, respectively. To test whether the two distributions of month of birth (mean vector) were equal between the clubfoot group and other newborn children, the non-parametric Watson's test was used (Zar 1999).

Results

The cumulative incidence of idiopathic clubfoot in each of the 2 years studied was similar, at 1.5‰ and 1.3‰, (p for difference = 0.2) (Table 2). The average cumulative incidence was 1.4‰ (95% CI 1.2–1.6). As commonly found, the majority (72%) of cases were boys. We found bilateral clubfeet in 46% of the cases. We also analyzed whether the distribution of patients with left or right side affected or bilateral clubfoot was similar for boys and girls. No gender-related side difference could be detected (p = 0.7) and the proportion of bilateral clubfeet was similar in boys (47%) and girls (41%) (p = 0.3; Table 3).

Most clinics (82%) had up to 5 cases per year, while 8 larger clinics (18%) treated 43% of the children with a range of 8–13 children per year.

When the cumulative incidence in different regions was determined, a statistically significant heterogeneity was found (p = 0.007) (Table 4). In comparison, the lowest cumulative incidence was $1.0/10^3$ (95% CI 0.7–1.4) in one region as compared to $2.0/10^3$ (95% CI 1.5–2.6) in the region with highest cumulative incidence (Figure 1, Table 4).

The seasonal variation in children with clubfoot peaked at 172º, corresponding to birth in July (Figure 2A). The strength of this mean vector was, however, weak (r = 0.09). The distribution by

Table 4. Cumulative incidence of idiopathic clubfoot during 1995 and 1996 in 6 regions of Sweden

Region	No. of native births over 2 years	No. of children with clubfoot	Cumulative incidence per thousand
1	30,775	62	2.0
2	45,248	73	1.6
3	21,660	30	1.4
4	45,704	52	1.1
5	35,022	35	1.0
6	18,848	28	1.5

month might in this case be uniform and the hypothesis of uniform distribution could not be rejected ($p > 0.05$). This means that for the children with clubfoot, there was no predominance of any month regarding birth. The angle of the mean vector for all live births was 148º ($r = 0.06$), corresponding to June. This is probably not a uniform distribution, i.e. the null hypothesis can be rejected ($p < 0.001$). Even so, there was no significant difference in distribution of birth month between clubfoot children and all other live births ($p > 0.5$).

Discussion

We found a higher cumulative incidence of congenital clubfoot during the 2 years of the study than has been found in previous studies from Scandinavia (Table 1). Does this mean that our sampling method is more accurate than in previous studies, or that the cumulative incidence is increasing—or both? It is difficult to provide a definite answer. We do, however, agree with Severin (1956) that the derived cumulative incidence is an underestimation of the true estimate. There are always a number of cases missing. Thus, our study is most likely not an exception, but a confident estimation of cases overlooked is difficult to make. The clubfoot diagnosis is easily set at birth, and since practically all children in Sweden are born in hospital, we can conclude that there are merely no nondiagnosed cases.

The cause of missing data is therefore probably more often failure to properly record and report the diagnosis. In this respect, there may be several additional reasons for different cumulative incidence figures in our study and in earlier Scandinavian studies. Firstly, there is always the critical issue of how to define the population when assessing the cumulative incidence. All the previous studies in Scandinavia, except the one by Severin (1956), took place in only part of the respective country. Secondly, especially in retrospective studies, there is uncertainty as to how cases were collected in relation to the defined population. Since the collection of cases in our study was prospective, we believe that we have achieved a good level of accuracy in the figures presented. Thirdly, by collecting medical records rather than using plain registry data, it was possible to exclude non-idiopathic cases, for instance those associated with syndromes or positional clubfeet. Since there was no central registration in Sweden of children with this diagnosis during the study period, the only way to obtain accurate data was to maintain continuous contact with all orthopedics departments in Sweden treating clubfeet. Nonetheless, we did observe a tendency of a decrease in the number of cases reported during the second year, which might reflect the general difficulty in continuous registration work.

One critical issue is, of course, whether the true cumulative incidence of congenital clubfoot in Sweden is increasing or not. A study from southern Sweden for the period 1946–90 (Danielsson 1992) concluded that there was an increase in the cumulative incidence during the late part of the period, from approximately $0.6/10^3$ up to $1/10^3$ at the end of the study. The authors suggested that the increase could be explained by immigration from non-Nordic countries. The estimated cumulative incidence for the Nordic part of Europe has generally been demonstrated to be lower than for other European regions (Cartlidge 1984, Somppi 1984, Danielsson 1992). A further increase in cumulative incidence might therefore be due to a continuous ethnic dilution of the Swedish population. Several extrinsic factors have also been analyzed (Barker and Macnicol 2002), such as a higher prevalence of maternal smoking during pregnancy over the past decades (Honein et al. 2000, Skelly et al. 2002), with a possible association with the regional difference in clubfoot cumulative incidence seen in our study (http://www.sos.se/fulltext/125/2004-125-4/2004-125-4.pdf, Table 4). The highest incidence

of maternal smoking is found in southern Sweden, the region that also has the highest proportion of clubfoot deformity. The higher proportion of immigrants in southern Sweden might also explain the regional differences in cumulative incidence of clubfoot (Franzén 2005).

Theories related to gestation giving increased intrauterine compression have been both recognized and rejected (Somppi 1984), and at present there seems to be convincing evidence that intrauterine compression is not a major risk factor (Barker and Macnicol 2002).

Several authors have suggested a seasonal variation in cumulative incidence of clubfoot, with a peak birth rate during December to March (Pryor et al. 1991), July (Lochmiller et al. 1998), or March–April (Barker and Macnicol 2002), and peak month of conception in October (Robertson and Corbett 1997). We could not confirm the observations of these authors. Others (Wynne-Davies 1964, Carney and Coburn 2005) have found no seasonality, which is in line with our observations. The study group (n 330) in the work of Robertson and Corbett (1997) was collected during the years 1956–1994, and the control group was collected from 1989 through 1993, which raises the question of whether the environmental exposure was the same during these overlapping time periods (Barker and Macnicol 2002).

We could not find any gender difference regarding uni- or bilateral distribution, an issue that has attracted little attention in the literature. Our results are in accordance with the findings of Severin (1956).

To summarize, we found a somewhat higher cumulative incidence of congenital clubfoot in Sweden compared to earlier reports, although the sex ratio and laterality distribution is in accordance with previous reports. Regional differences were significant, but we could not detect any seasonal variation.

Contributions of authors

HW: collection of data, statistical analysis and manuscript writing. LH: Contributed in general discussion concerning incidence research and giving views on the manuscript. KM: contributed in the analysis and interpretation of the data and in writing the manuscript.

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Each author certifies that his or her institution approved the human protocol for this investigation, that all investigations were conducted in conformity with ethical principles of research, and that informed consent was obtained.

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