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The Incidence of Cleft Lip and Palate Deformities in the South-east of Scotland (1971–1990)

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Abstract: *This retrospective study reports the incidence of infants born with the cleft lip and palate anomaly within the Edinburgh Cleft Units catchment area, between 1 January, 1971, and 31 December, 1990. The importance of accurate data collection for local, regional, and national data bases is discussed with reference to the recent CSAG report on cleft lip and palate services in the UK.*

Five-hundred-and-two cleft lip and palate patients were identified (291 males, 211 females). The incidence is reported as 1.4 per 1000 live births (1 in 711). Twenty-five per cent of clefts affected the primary palate, 45 per cent affected the secondary palate, and the remaining 30 per cent were clefts of both the primary and secondary palate. Overall, a higher percentage of males were affected (58 per cent males to 42 per cent females). Clefts of the secondary palate, however, were more common in females (56 per cent females to 44 per cent males).

Data presented in this study is similar to that previously reported from UK centres. It is suggested the accuracy of the UK cleft lip and palate data collection needs to be improved. Prospective data collection in a standardized format carried out on a national basis has to be a priority as recommended by the CSAG report.

Index words: Cleft Lip and Palate, Data Base, Epidemiology, Incidence.

Refereed Paper

Introduction

It is now widely recognized that accurate record keeping and data collection is of paramount importance in the running of Health Services. This is particularly true in the planning and development of service provision and resource allocation, as well as clinical audit and research. The recording and documentation of cleft lip and palate deformities in the UK has historically been voluntary, and carried out by the Office of National Statistics (ONS), formally known as the Office of Population Censuses & Surveys (OPCS), in England and Wales, and in Scotland by the Information and Statistics Division NHS in Scotland. The Clinical Standards Advisory Group (CSAG) report into cleft lip and palate services in the UK has stated its concerns relating to the accuracy of this data.

Regional data bases as described by Greg *et al.* (1994), Luther and Cook (1994), and the Scottish Cleft Lip and Palate Group (SCALP), have set the standard for data collection in the 1990s. CARE was initially set-up by the Craniofacial Society in 1989 as an additional voluntary register of cleft birth data nationally. This proved less reliable than the ONS data initially. However, in 1995, regional cleft co-ordinators were introduced to improve the system of collection (personal communication). Registrations have subsequently improved dramatically. However, a few units still prefer not to inform this group of their cleft birth data.

National registration systems for cleft anomalies are not new. In Denmark, the government introduced compulsory recording of children with facial clefts in 1937 (Bixler *et al.*, 1971). Norway established a medical register of births, including information on congenital malformations in 1967. A European initiative, EUROCAT, was established in 1979 (European Registration of Congenital Anomalies). This was supported by the European Commission with the aim of improving the methodology for population studies throughout the community. Initially, EUROCAT was set-up as a feasibility study to test the ability of pooling data across national boundaries. By 1991, as a result of its success, it is no longer supported as a research project, but as a fully funded service in its own right.

Studies in Denmark by Jensen *et al.* (1988) have demonstrated an increased incidence of cleft births between 1942 and 1981. Between 1938 and 1942 there were 1·5 per 1000 births, which increased to 1·89 per 1000 births between 1978 and 1981. Similar increases have been reported by Rintala (1986), and Srivastava and Bang (1990) in Finland and Kuwait, respectively. No such increases have been reported in UK studies. This demonstrates the importance of having base-line values easily available to determine any change in incidence.

Since 1989, the Edinburgh cleft unit has been participating in the Scottish Cleft Lip and Palate Project (SCALP). This is a voluntary system for collecting standardized data on all cleft lip and/or palate infants born in Scotland. No accurate cleft data was available prior to this date in an easily accessible form.

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Methods

A retrospective study was undertaken to identify all cleft lip and palate births within the regions of Lothian (East, West, and Central), Borders, Fife, and the Highlands. The aim was to gather information from 1970 onwards specifically noting the cleft type, date of birth, presence of an associated syndrome, medical history where applicable, and details of surgical care received since birth. The cleft deformities were recorded using an anatomically descriptive classification (as used in the CARE registration document). The classification therefore depicted the cleft types as follows:

(1) cleft lip and alveolus (right, left, or bilateral);
(2) cleft lip and palate (right, left, complete, and incomplete);
(3) bilateral cleft lip and palate (complete or incomplete);
(4) cleft palate (soft and hard);
(5) submucous cleft.

The following list indicates the sources of information for this particular study:

1. Plastic surgery records.
2. Hospital operating lists, Edinburgh Sick Childrens Hospital.
3. Edinburgh Dental Hospital patient records.
4. Fife hospital patient records.
5. Information and Statistics Division NHS in Scotland.
6. Patients records at the Edinburgh Sick Childrens Hospital.
7. Western General Hospital patient records.
8. Leith Hospital patient records.

An initial register of 721 possible patients were identified. All case notes were searched and many cases were excluded on the following grounds:

1. No cleft present.
2. Birth location outside catchment area.
3. Birth date not within the period 1 January, 1971, to 31 December, 1990.
4. Submucous clefts (23 cases).

The resulting cohort comprised of 502 cleft lip and palate patients (291 males and 211 females).

A computer data base was formulated and a paper copy generated to record the data in a standardized format. A single operator was used to sift and record the data, and then enter the data into the computer data base. Live birth data was obtained from the Scottish Home and Health Birth Statistics, published annually. Statistical analysis was carried out using the Chi-squared analysis with 1 degree of freedom. Probability values of 0·05 or less were taken to be statistically significant.

Results

The overall incidence for cleft births over this 20-year period was 1·4 per 1000 or 1 in 711 live births. The incidence of cleft births varied from year to year, ranging from 1 in 1013 in 1973, to a high of 1 in 540 in 1987 (Table 1).

Forty-three infants presented with Pierre Robin sequence and 92 with an associated syndrome. During the period of record collection, 12 post-natal deaths had been recorded from within the 502 cohort group (Table 2).

Comparison of the birth data for the first 10 years (1971–1980) with that of the second 10 years (1981–1990) show a remarkable overall consistency. While the cleft births have increased from 249 to 253 over this period, the overall birth rate dropped from 181,004 to 175,729. Thus, the incidence changed only slightly, increasing from 1·37 per 1000 to 1·4 per 1000 live births (1 in 727 to 1 in 695). The increase in cleft births between the first and the second 10-year period was not statistically significant (Table 3).

Twenty-five per cent of clefts affected the primary palate, while 45 per cent affected the secondary palate. The remaining 30 per cent affected both the primary and secondary palate. Left-sided cleft lip and palate defects (13 per cent) were more common than right-sided clefts (6 per cent; Table 4). The distribution of cleft types is shown in Table 4 and pictorially presented in Figure 1.

TABLE 1 *Cleft births 1971–1990*

Year	Male	Female	Total	Total births	Per 1000 births	Ratio per live births
1971	20	12	32	21,832	1·5	1:682
1972	12	11	23	20,010	1·1	1:870
1973	12	7	19	19,254	1	1:1013
1974	12	9	21	18,104	1·1	1:862
1975	15	10	25	16,090	1·5	1:643
1976	16	7	23	16,906	1·4	1:735
1977	14	4	18	16,212	1·1	1:901
1978	14	9	23	16,726	1·3	1:727
1979	18	14	32	17,894	1·8	1:559
1980	20	13	33	17,976	1·8	1:545
1981	20	11	31	18,011	1·7	1:581
1982	12	12	24	17,208	1·4	1:717
1983	14	13	27	16,831	1·6	1:623
1984	14	10	24	17,149	1·4	1:714
1985	16	11	27	17,710	1·5	1:656
1986	14	6	20	17,650	1·1	1:882
1987	15	18	33	17,829	1·8	1:540
1988	13	10	23	17,852	1·3	1:776
1989	11	7	18	17,407	1	1:967
1990	9	17	26	18,082	1·4	1:695
Total	291	211	502	356,733	1·4	1:711
	57·96%	42·03%				

TABLE 2 *Cleft births identified with an associated syndrome, Pierre Robin Sequence and recorded post-natal deaths.*

	Male	Female	Total
Syndrome	46	46	92
Pierre Robin	29	24	53
Post-natal deaths	8	4	12

TABLE 3 *Cleft births between 1971 and 1980, and 1981 and 1990*

10-year period	Cleft births	Births	Ratio for 1000	Ratio for live births
1971–1980	249	181,004	1·37	1:727
1981–1990	253	175,729	1·4	1:695

$x = 0{\cdot}0319$ ($P > 0{\cdot}50$), NS.

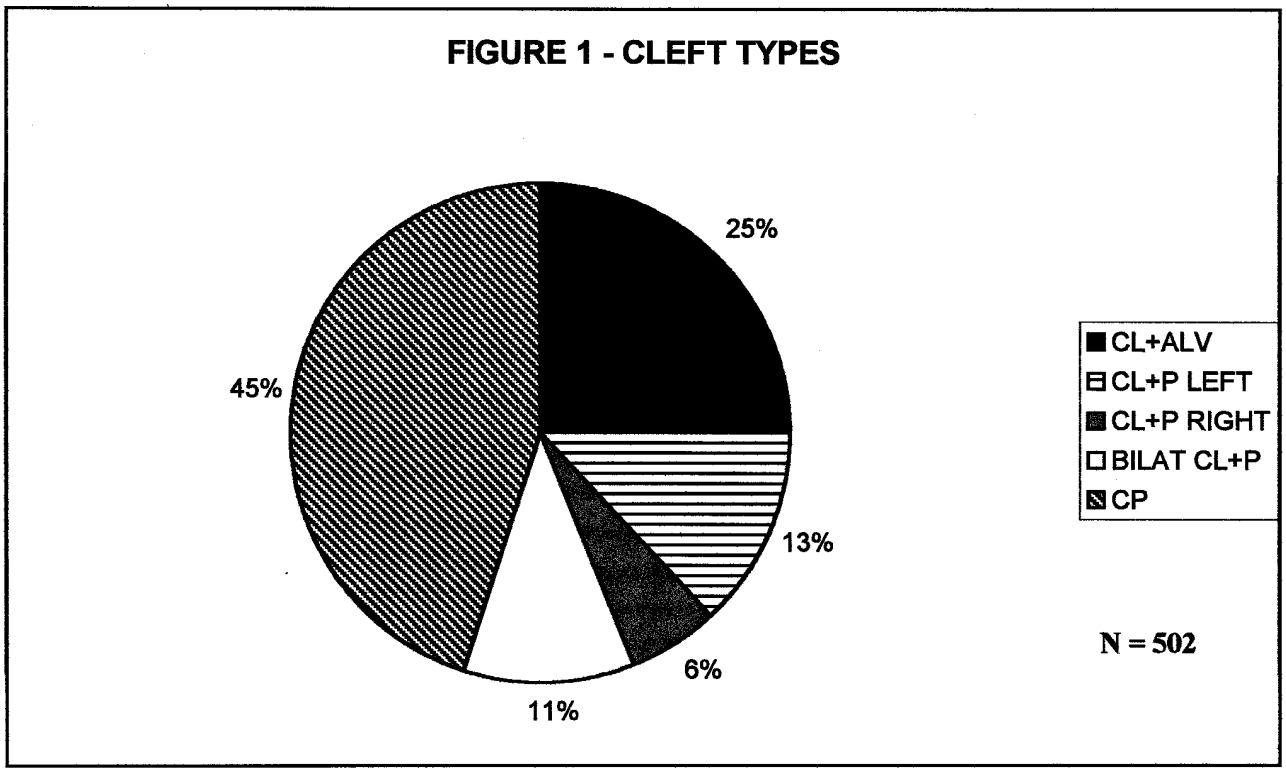

FIG. 1 *Distribution of cleft birth types in South East Scotland 1971–1990*

TABLE 4 *Distribution of cleft types*

	Male	%	Female	%	Total	Total %
CLIP	93	74	33	26	126	25
CPALATE	99	44	128	56	227	45
CL + P LEFT	40	62	25	38	65	13
CL + P RIGHT	20	65	11	35	31	6
BIL CL + P	39	75	14	25	53	11
Total	291	58	211	42	502	

TABLE 5 *Cleft types by sex*

	C LIP	C PALATE	CL + P (L)	CL + P (R)	BIL CL + P
Male	93	99	40	20	39
Female	33	128	25	11	14
Total	126	227	65	31	53
	$\chi^2 = 28{\cdot}571$	$\chi^2 = 3{\cdot}704$	$\chi^2 = 3{\cdot}461$	$\chi^2 = 2{\cdot}612$	$\chi^2 = 11{\cdot}792$
	$P < 0{\cdot}001$	$P > 0{\cdot}05$	$P > 0{\cdot}05$	$P > 0{\cdot}10$	$P < 0{\cdot}001$
		NS	NS	NS	

TABLE 6 *Cleft lip only*

	Complete		Incomplete		Total	
	Male	Female	Male	Female	Male	Female
Left	6	3	44	12	50	15
Right	6	7	28	6	34	13
Bilateral	6	1	3	4	9	5
Sub-total	18	11	75	22	93	33
Total (M + F)		29		97		126

TABLE 7 *Cleft lip complete/incomplete + sex*

	Complete	Incomplete
Male	18	75
Female	11	22
Total	29	97
	$\chi^2 = 1{\cdot}689$	$\chi^2 = 28{\cdot}958$
	$P > 0{\cdot}10$	$P < 0{\cdot}001$
	NS	

TABLE 8 *Cleft lip by side + sex*

	R	L	Bilateral
Male	34	50	9
Female	13	15	5
Total	47	65	14
	$\chi^2 = 9{\cdot}383$	$\chi^2 = 18{\cdot}846$	$\chi^2 = 1{\cdot}142$
	$P > 0{\cdot}001$	$P < 0{\cdot}001$	$P < 0{\cdot}50$
			NS

Fifty-eight per cent of clefts were males, while 42 per cent were females. In all categories males dominated, apart from the isolated cleft palate group, where females dominated by 56–44 per cent . Statistically, however, male dominance was significant in the cleft lip group, as well as the bilateral cleft lip and palate group (Table 5). In the cleft lip group only (Table 6), male dominance was statistically significant for left-sided clefts (Table 8), and highly significant in the male incomplete left sided cleft lip group (Table 7).

Discussion

While retrospective studies such as this can easily be criticized for their inaccuracies, every attempt has been made to maintain consistency in this study. Several factors may affect the accuracy of regional data bases. These include:

1. Cross-boundary flow as previously described by Knox and Braithwaite (1962).
2. Fluidity of movement of personnel in and around her Majesty's armed forces bases (there are several large military bases within the Edinburgh catchment area).
3. Quality and availability of record keeping.

The data presented in this study would seem to be consistent with previous studies reported in the UK literature (McMahon and McKeown, 1953; Knox and Braithwaite, 1962; Owens *et al.*, 1985; Womersley and Stone, 1987; Coupland and Coupland, 1988; Srivastava and Bang, 1990; Greg *et al.*, 1994; Fitzpatrick *et al.*, 1994). In these studies, the incidence reported varies from a low of 1·12 per 1000 (Coupland *et al.*, 1988) to a high of 1·79 per 1000 live births (Srivastava *et al.*, 1990; Table 9).

The cleft incidence varies from year to year with quite dramatic swings. Table 1 shows the annual figures where the incidence varies from a low of 1 in 1013 births in 1973, to a high of 1 in 540 in 1987. These large swings, which are totally unpredictable, are consistent with previous findings (Knox *et al.*, 1962; Greg *et al.*, 1994).

Jensen *et al.* (1988), Rintala (1986), and Srivastava and Bang (1990) have previously reported an increase in cleft lip and palate incidence in recent years. No British authors have been able to show a similar trend. This study reports the incidence between 1971 and 1980, and 1981 and 1990, increased from 1·37 per 1000 to 1·4 per 1000 live births (Table 3). The increase is small and not statistically significant.

High levels of clefts affecting the secondary palate were reported by Womersley and Stone in 1987 (52·2 per cent) and again by Fitzpatrick *et al.* in 1994 (53 per cent) in the West of Scotland. Greg *et al.* (1994) also reported from Northern Ireland that clefts of the secondary palate were consistently high at 53 per cent . In this study, the clefts of the secondary palate occurred in 45 per cent of our sample.

Clefts of the secondary palate have been shown to have female dominance (Knox and Braithwaite, 1962; Fitzpatrick *et al.*, 1994; Greg *et al.*, 1994). This report can confirm these previous findings with females dominating by 56–44 per cent.

Unilateral cleft lip and palate clefts are more prevalent on the left side (Table 4, 65 left to 31 right), as has been previously reported by Knox and Braithwaite (1962), Jensen *et al.* (1988), and Greg *et al.* (1994). Unlike Greg *et al.* (1994), the right-sided clefts of the lip and palate were still dominated by males (Table 4). There is one significant difference between this study and that reported by Greg *et al.* in 1994, this being that his unilateral cleft lip and palate group are separated into complete and incomplete clefts, which we have not carried out in this study.

A comparison of cleft data from recent publications has been tabulated in Table 9. Direct comparison between studies is often difficult due to differing classifications used to depict cleft types (Vanderas, 1987). Thus, enhancing the argument for standardizing record collection for cleft patients.

Currently, data collected by ONS, on cleft births, only describes a cleft in two forms:

(1) a cleft of the lip and palate;
(2) a cleft of the palate only.

It is essential that a full classification of cleft type is recorded for each infant. Vandaras (1987), in his comprehensive review of the literature on cleft incidence, suggests that the cleft data collected should be separated by:

1. Cleft type: cleft lip, cleft palate, and cleft lip and palate.
2. Racial type.
3. Sex.
4. Geographical area.
5. With and without associated malformations and syndromes.

In addition, live birth, still birth, and abortion statistics should also be reported.

The concern raised by the CSAG report regarding lack of completeness of recording the incidence of cleft lip and palate births, is a fundamental issue which needs to be addressed. The ONS data has been shown to be incomplete [Clinical Standards Advisory Group Report (CSAG

TABLE 9 *Previously reported Cleft Lip and Palate incidence studies*

Author	No. of births	Location	Time span of study	Incidence per 1000 live births	CL primary palate (%)	CP secondary palate (%)	CLP (L + R)-BILAT CLP (%)
McMahon *et al.* (1953)	285	Birmingham	1940–1950	1·3	23·2	40	36·8
Rank *et al.* (1960)	160	Tasmania	1945–1957	1·66	23·75	33·13	43·13
Knox *et al.* (1962)	574	Northumberland	1949–1958	1·42	31·60	32·80	35·70
Owens *et al.* (1985)	454	Liverpool	1960–1982	1·4	30	33·6	36·40
Womersley *et al.* (1987)	247	W. Scotland	1974–1985	1·56	13·40	52·20	34·40
Coupland *et al.* (1988)	930	Trent Region	1973–1982	1·12		39	61 CL + CLP combined
Jensen *et al.* (1988)	602	Denmark	1976–1981	1·89	34	27	39
Srivastava *et al.* (1990)	178	West Midlands	1985–1987	1·79		27.5	72.5 CL + CLP
Srivastava *et al.* (1990)	230	Kuwait	1985–1987	1·48		27·8	72·2 CL + CLP
Gregg *et al.* (1994)	398	N. Ireland	1980–1990	1·28	16	53	31
Fitzpatrick *et al.* (1994)	286	W. Scotland	1980–1984	1·44	19	53	28

Report) 1998]. The under reporting may well be due to the fact that this is a voluntary register, with several units choosing not to report the relevant information. Making a collection of cleft data mandatory may well improve its accuracy. However, Abyholm (1978) has reported that a compulsory national register alone is no guarantee of a comprehensive and accurate data collection system. In his article reviewing the registrations in Norway, he has shown an under-reporting of 14·46 per cent, when data from the national register was compared with the hospital operating statistics.

The CARE Group, using regional co-ordinators and standardized data collection forms, have recently shown that their collection of data has improved to such a degree that the number of clefts being reported is larger than that reported by the ONS data (*CARE News Letter*, March 1998). It would therefore seem sensible to try and merge these two separate data collection systems and make use of their respective strengths, as well as making registration notification compulsory.

Conclusions

Data presented here clearly demonstrate a similar distribution of cleft type, number and sex, to previous reports published from other UK centres. Prospective data collection in a standardized format carried out on a National basis has to be a priority as recommended by the CSAG report.

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