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An Audit of the Yorkshire Regional Cleft Database

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Abstract *This study assessed the validity of the Yorkshire regional orofacial cleft database by comparing the computer-based records with locally collated records of primary surgical events for babies born over a 2-year period (1994–1995). One-hundred-and-thirty-two infants with clefts (excluding submucous cleft palate) were identified from the latter source with an equal proportion of unilateral cleft lip/palate and isolated cleft palate births. However, only 62 per cent of cases were recorded on the database and the reporting rate of individual cleft units was highly variable (43–85 per cent). In addition, there was a significant under-reporting of both cleft lip and isolated cleft palate cases (42 and 50 per cent ascertainment, respectively). Consequently, the database figures understated the prevalence of all cleft births, but especially of these two cleft subtypes. Conversely, the relative frequency of combined cleft lip and palate cases was exaggerated. The reasons for such discrepancies and possible improvements to data collection are discussed.*

Index Words: Audit, Cleft lip and palate, Database, Prevalence.

Introduction

Orofacial clefts are comprised of a range of anomalies: clefts of the lip/alveolus (CL), clefts of the lip and palate (CLP), and isolated cleft palate (CP). The former two subtypes are often grouped together as cleft lip with or without cleft palate [CL(P)]. Statistics on the prevalence of these clefts and their clinical outcome are essential for the progress of research, clinical audit, and for the planning of clinical services and specialist training (Sandy *et al*., 1998; Hammond and Stassen, 1999). Therefore, it is important that such statistics are based on valid and accurate data. These data are collated at a variety of levels ranging from individual units' records to national registers. Regional databases of orofacial cleft cases have existed in several parts of the United Kingdom (UK) since the 1980s (Gregg *et al*., 1994; Luther and Cook, 1994; Bellis and Wohlgemuth, 1999). Furthermore, a national data registry, covering England and Wales, was established in 1982 under the auspices of the Craniofacial Society of Great Britain (Hammond and Stassen, 1999). This database subsequently became known as the Craniofacial Anomalies Register (CARE). However, since the registration of patients has been voluntary, either incomplete or no data has been provided by many of the units involved in cleft care.

In the UK, data on congenital anomalies is also collected by a government agency, the Office for National Statistics (ONS), and by centres participating in the European Registration of Congenital Anomalies (EUROCAT) initiative. The ONS collects data from health authorities and local clinicians involved in the neonatal period. These figures are pooled with other data sources, e.g. CARE, via the British Isles Network of Congenital Anomaly Registers. Again, these registers receive orofacial cleft reports on a voluntary basis. As such, there is no robust estimate of the UK birth prevalence of orofacial cleft anomalies. Recently, the process of data collection has received fresh impetus with the compilation and publication of the Clinical Standards Advisory Group (CSAG) Report (1998) on the management of cleft anomalies in the UK. In particular, the implementation of this report's recommendations requires accurate data on the regional and subtype prevalences of orofacial clefts in order to forecast the service provision, workloads, and specialist staff and facility requirements of prospective cleft centres. For example, children born with isolated CL anomalies will not require secondary palate surgery nor speech therapy, whereas primary lip repair and alveolar bone graft surgeries are not necessary in isolated CP cases.

The Yorkshire cleft database was established in 1990 following a successful pilot study and audit (Luther and Cook, 1994). This database is compiled in the Orthodontic Department of the Leeds Dental Institute, and covers the North and West Yorkshire areas (except the Northallerton Health District). Reporting of cleft births occurs on a voluntary basis, though compliance is actively encouraged by the regional cleft (CARE) co-ordinator. Ideally, a CARE registration form is completed in the immediate post-natal period by a consultant or specialist registrar in Orthodontics, and forwarded to the co-ordinator to be entered on the database. Currently, there are four cleft units within this large but geographically defined area. Since this Yorkshire cleft database is used as a source of regional and national data it was decided to test its validity by comparing its figures with those of a locally-collated ascertainment source. Arguably, population ascertainment from maternity hospital records may be inclusive of all live births and anomalies, but this source would not be without its practical difficulties and deficits, e.g. insufficient staff involvement, home births, and undiagnosed CP and small defects. The use of regional health authority records, collated using hospital operation codes, was also considered. However, a preliminary assess-

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ment of this source revealed an unsatisfactory level of accuracy due to coding errors, e.g. the miscoding of lip trauma as primary cleft lip surgery. Therefore, theatre records of primary cleft lip and palate surgical repairs were selected as this source. A 95 per cent reporting level was arbitrarily set as the gold standard for the database's accuracy. This figure was selected to reflect the high cleft ascertainment rate in Denmark, where figures are collated under compulsory reporting conditions (Christensen, 1999). In addition, it is likely that a discrepancy of at least 5 per cent would be required to significantly affect extrapolations and service-commissioning arrangements derived from orofacial cleft statistics.

Methods

The cleft database records were compared with the primary surgical records of CL(P)/CP babies born during the 2-year period from 1 January 1994 to 31 December 1995. Submucous cleft cases were excluded from the study because these anomalies are often identified at a later stage and may represent a distinct cleft subgroup. The time lapse between the selected study period and the data analysis provided an opportunity for the delayed database registration of cleft cases, yet it preceded the next major registration triage which would occur at the stage of 5-year old audit records collection. The audit involved a systematic search of the operating theatre logbooks at each hospital site in the North and West Yorkshire region where primary cleft lip and palate surgery was undertaken routinely during the study period. The five sites were the Hull Royal Infirmary, Leeds General Infirmary, Pinderfields Hospital (Wakefield), St James's Hospital (Leeds), and St Luke's Hospital (Bradford). However, only four units were involved since the two Leeds sites function as a single cleft team. These theatre records were cross-checked with individual hospital clinical files.

Figures were compiled according to the cleft units and cleft subgroups involved. A chi-squared test was utilized to assess the statistical significance of the results where appropriate. In addition, the Yates correction was applied if a subgroup being tested consisted of less than 25 cases.

Results

One-hundred-and-thirty-two patients (64 male, 68 female) were identified in the surgical records as having been born within the study period and having undergone one or more primary cleft repair procedures. Eighty-two of these cases were positively recorded in the cleft database (Table 1). The database's overall reporting score was 62·1 per cent and there was a wide variation in the ascertainment of individual units, ranging from 43 to 85 per cent. In relation to the years audited there were 67 and 65 live cleft births during 1994 and 1995, respectively, of which 61·2 and 61·5 per cent were reported to the database. Therefore, there was no significant difference between these 2 years in terms of either birth prevalence or ascertainment rates.

Table 2 illustrates the ascertainment rate of the cleft database in relation to each cleft subtype. These figures show that the database registration levels for CL and CP cases, at 42·1 and 50 per cent, respectively, were much lower than for the CLP anomalies (83 per cent). Both of these differences were statistically significant ($P < 0{\cdot}0001$). Bilateral CL(P) patients (i.e. CL + CLP) were reported at a slightly higher rate than unilateral cases (80 and 70·2 per cent, respectively), but this difference was not statistically significant ($P = 0{\cdot}77$), especially after incorporation of the Yates correction ($P = 0{\cdot}94$). Similarly, the ascertainment of bilateral CL (100 per cent) cases was higher than that of unilateral CL (35·3 per cent), but the number of cases involved was small and not suitable for meaningful statistical analysis. Conversely, for CLP anomalies the unilateral and bilateral cases were recorded on the database at 85 and 76·9 per cent, respectively, but this difference was not statistically significant ($P = 0{\cdot}207$). Consequently, there was an important and statistically significant difference between the database and surgical records sources in terms of their prevalence figures (Table 2). In effect, the database figures give the impression of a higher proportion of CLP cases, and conversely a lower proportion of CL and CP, than is evident from the surgical records. For example, the proportion of CLP cases was 54 per cent in the database compared with an actual relative frequency of 40 per cent in the surgical records.

TABLE 1 *Recorded cleft cases according to their surgical unit and database registration*

Centre	Surgical records	Database	Percentage in database
A	30	20	66·7
B	33	28	84·8
C	20	13	65
D	49	21	42·9
Region	132	82	62·1

Discussion

The Yorkshire regional cleft database's overall low ascertainment score of 62·1 per cent was significantly below the arbitrary gold standard of 95 per cent and was certainly disappointing. Although one cleft unit achieved a score of 85 per cent, this still fell short of the desired reporting accuracy. Furthermore, the problem of collecting data from multiple sites is exemplified by both the lowest score (43

TABLE 2 *Distribution of cleft subtypes according to the data source (surgical records/cleft database). For each source, the relative frequency of each subtype is denoted in parenthesis (CL = cleft lip/alveolus, CLP = cleft lip and palate, CL(P) = cleft lip ± palate, CP = isolated cleft palate)*

Cleft subtype		Surgical records	Database	Percentage in database
CL:	Unilateral	17 (12·9%)	6 (7·3%)	35·3
	Bilateral	2 (1·5%)	2 (2·4%)	100
	Total	19 (14·4%)	8 (9·8%)	42·1
CLP:	Unilateral	40 (30·3%)	34 (41·5%)	85
	Bilateral	13 (9·8%)	10 (12·2%)	76·9
	Total	53 (40·2%)	44 (53·7%)	83
CL(P):	Unilateral	57 (43·2%)	40 (48·8%)	70·2
	Bilateral	15 (11·4%)	12 (14·6%)	80
	Total	72 (54·5%)	52 (63·4%)	72·2
CP		60 (45·4%)	30 (36·6%)	50
Patient total		132 (100%)	82 (100%)	62·1

per cent) and the ascertainment rate variations (Table 1). This regional reporting rate of approximately 41 cases per annum also compares unfavourably with the number of cleft cases recorded during the database's initial three years (1990–1992) when 48–59 new cases were reported per annum (Luther and Cook, 1994). This indicates that the database's ascertainment rate peaked at 89 per cent (range 73–89 per cent) within a few years of its inception. Notably, variable ascertainment is not restricted to the region and units examined here since a recent study of the Scottish Association for Cleft Lip and Palate (SCALP) database revealed an average rate of 74·6 per cent (range 62–85 per cent) over 8 years (Mossey and Clark, 1999).

The significant under-reporting of cleft births, especially of isolated cleft lip/alveolus (CL) and cleft palate (CP) anomalies, highlights the risk of skewed data arising from incomplete database ascertainment. This problem is not new since the database's current subtype frequency distribution is very similar to that reported by Luther and Cook (1994). The 45 per cent CP prevalence apparent from the surgical records in this study is both concordant (Womersley and Stone, 1987; Fitzpatrick *et al*., 1994; Gregg *et al*., 1994; Bellis and Wohlgemuth, 1999) and discordant (Owens *et al*., 1985; Coupland and Coupland, 1988; Srivastava and Bang, 1990) with other contemporary UK studies. Ascertainment bias partly accounts for this wide variation in the relative frequencies of CL, CLP and CP cases in the UK. Whilst this does not exclude other influential factors (e.g. geographic and racial variations) it does indicate that care should be taken when evaluating data on the relative frequency of cleft subtypes.

The UK ascertainment figures contrast with the Danish Facial Cleft Register's 99 per cent reporting rate (for non-syndromic cases, excluding submucous clefts). The latter's accuracy appears to arise from compulsory cleft case reporting in Denmark, centralized cleft surgical services, and the use of multiple ascertainment sources (Christensen, 1999). By inference, contemporary Danish cleft prevalence figures may be viewed as being highly reliable: 1·4–1·5 per 1000 (approximately 1:690) and 0·7–0·9 per 1000 (approximately 1:1250) live births for CLP and CP, respectively, yielding a combined prevalence of 2·2 per 1000 (1:450). In contrast, it has been possible only to estimate the current UK average cleft prevalence from contemporary local studies at approximately 1·5 per 1000 or 1:650 live births (range: 1·28–1·82 per 1000 or 1:550–800; Owens *et al*., 1985; Womersley and Stone, 1987; Coupland and Coupland, 1988; Srivastava and Bang, 1990; Fitzpatrick *et al*., 1994; Gregg *et al*., 1994; Bellis and Wohlgemuth, 1999; EUROCAT, 1999). Arguably, the true UK frequency statistics would approximate those of Denmark if the former's databases had higher ascertainment levels, though other variables may be partly responsible for these high prevalence figures, e.g. geographic and racial variations.

In the Yorkshire context, regional health authority statistics (provided by the Regional Health Authority Headquarters, Durham) indicate that 82,265 live births occurred within the study area during 1994 and 1995. Based on an average live birth rate of 41,100 per year, a CL(P) and CP prevalence of either 1:650 (estimated UK figure) or 1:450 (Denmark figure) would have given rise to 63 or 91 births per annum, respectively. Interestingly, the number of cleft births recorded from surgical records in this study (66 per annum) gives rise to a birth frequency of 1·6 per 1000 (1:620) which approximates the estimated UK average prevalence. However, whilst the Yorkshire CP prevalence of 0·73 per 1000 (1:1370) is consistent with the Danish statistics (0·7–0·9 per 1000), the Yorkshire CL(P) prevalence of 0·88 per 1000 (1: 1140) is much lower than in Denmark (1·4–1·5 per 1000). Arguably, the precision of this extrapolation may be undermined by undetermined confounding factors, such as geographic or racial variations, and losses due to pregnancy terminations. However, one explanation for this specific CL(P) birth incidence disparity may be the more complete capture of both syndromic and mild cases in Denmark, where over a 50-year period there has been an apparent increase in the CL(P) prevalence through ascertainment rate improvements (Christensen, 1999). Perhaps in the present study the significant under-reporting of CL cases was due partly to a misconception that such anomalies are isolated cosmetic problems. In addition, some babies will not have featured in the Yorkshire surgical records if they had minor defects that did not require primary surgery. Despite this, it is evident that the use of surgical records as a source of ascertainment has a high degree of validity for live birth statistics.

Given that in the present context the surgical ascertainment data represents a gold standard, the reasons for the low cleft database accuracy in this audit are unclear. It may be postulated that various factors were responsible for this discrepancy:

1. Mild cleft defects, especially palatal ones, were not diagnosed in the initial post-natal period, although this is more relevant to submucous cleft palates which were excluded from this study.
2. Babies with clefts, especially those with isolated clefts of the lip or palate, were not reported to local orthodontic staff. Conversely, babies with cleft lip and palate may be reported more frequently because of more apparent neonatal feeding difficulties.
3. Non-viable babies with clefts died before being registered, although there is evidence that this number would be very small (Bellis and Wohlgemuth, 1999), especially if infants survive to the stage of primary cleft surgery.
4. CARE forms were not forwarded from each unit to the regional co-ordinator.
5. CARE forms were received centrally, but not input into the database.
6. Incorrect details were filed on the CARE form, e.g. name/date of birth.
7. The baby's name was changed during early infancy (after their initial registration). Anecdotally, this is not uncommon especially when the infant is transferred to an adopted family.
8. There was a mis-match between the births and surgical procedures performed within the region, e.g. babies entered the region prior to their primary cleft surgery but were not registered.

Strictly speaking, the third and eighth factors cannot be evaluated by the comparison of surgical records with the database. The analysis of birth (maternity hospital) records would be required in these instances, but the number involved in any such discrepancy is likely to be insignificant and maternity source reliability is questionable.

In the light of the finding of the Yorkshire cleft database's

unsatisfactory ascertainment performance several changes, both direct and coincidental, are being implemented. First, the cleft database has been updated with the patient details obtained from this study's additional sources. In clinical and organizational terms, a major change has occurred with a regional reduction in the number of surgeons and units performing primary cleft surgery. This has involved key staff changes, as well as the rationalization of services. Further centralization of cleft services will occur following implementation of the CSAG report recommendations. This should lead to the ascertainment of those cases referred for centralized care who have been missed during the initial perinatal triage. Prior to this change, some of those patients with clefts not recorded on the database may also not have benefited from multi-disciplinary team management. Finally, the CARE database and reporting system has gained a higher profile, especially since the CSAG review process was undertaken. It is hoped that compulsory registration of cleft cases would improve data collection further (Hammond and Stassen, 1999), although this does not necessarily guarantee accuracy (Abyholm, 1978). As such, statistics utilizing regional and CARE database information should be treated with caution until further audit demonstrates a reliable level of ascertainment. It is intended that the Yorkshire database will be re-audited in several years time following the re-organization and consolidation of cleft services.

Conclusions

This audit indicates that the Yorkshire regional cleft database has a low ascertainment rate (62 per cent) and, therefore, it should not be viewed as a reliable source for orofacial cleft statistics. In particular, both cleft lip/alveolus and cleft palate anomalies have been reported at a significantly lower level than cases with combined cleft lip and palate giving rise to skewed impression of subtype relative frequencies. By extrapolation, the accuracy of national (CARE and ONS) cleft statistics is also questionable. As such, pending future confirmation of UK database validity, surgical records appear to provide the most valid and feasible source of information on cleft lip and palate prevalence.

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