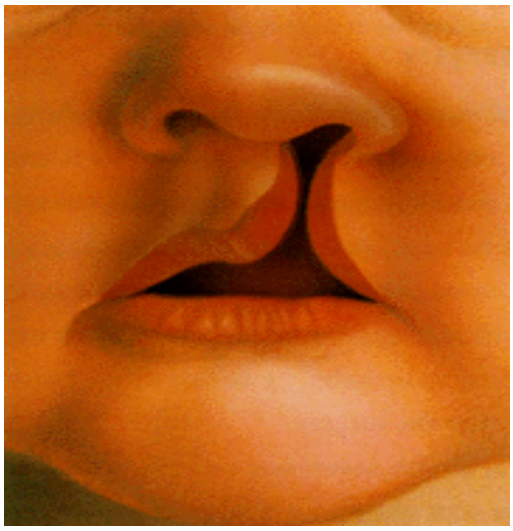

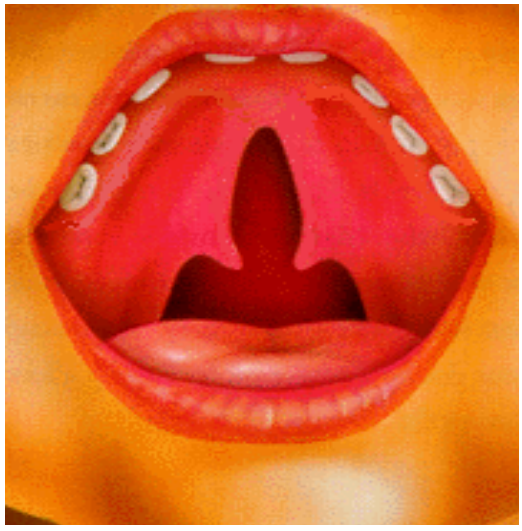

SNAP REPORT ON CLEFT LIP AND PALATE

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November 1998

FOREWORD

This SNAP report was commissioned by the SNAP Oral Health Group in 1997 and has been produced by a small working group. Most health professionals in Scotland who provide cleft lip and palate services meet on a regular basis in a forum known as the Scottish Cleft Lip and Palate (SCALP) Group. The SCALP group was asked to suggest members for the SNAP working group since it was felt to be important that a broad range of professionals who worked in the Cleft Lip and Palate field were represented. The SCALP group as a whole were given the opportunity to comment on the SNAP report at a draft stage.

Cleft lip and/or palate is a congenital anomaly which affects some 100 babies born each year in Scotland. Whilst the numbers are relatively small the active treatment lasts throughout childhood and beyond and it is important that there should be a proper assessment of need and appropriate commissioning of health services. The preparation of this report has coincided with the publication of a report on cleft lip and palate by the Clinical Standards Advisory Group (CSAG) which has identified a number of areas of concern in current service provision in the UK.

It has been a significant advantage to have the CSAG report available and it is recommended that it be read in conjunction with this document. In view of the likely outcomes of the CSAG report, the SNAP working group has made a number of recommendations for commissioning cleft lip and palate services in Scotland and proposes a revised configuration for those services.

CONTENTS PAGE

1. EXECUTIVE SUMMARY (INCLUDING RECOMMENDATIONS)

BACKGROUND

1.1 Cleft lip and/or palate is the most common abnormality in the cranio facial region with about 100 new cases occurring each year in Scotland.

1.2 Genes, environment and diet are thought to be implicated in the aetiology of orofacial clefting and further research is ongoing.

1.3 At present, treatment takes place in six Scottish centres with considerable variation in surgical workload between centres. There is also evidence of variation in the outcome of cleft care in the UK as a whole and this may, in some cases, be poorer than in some European centres. There is a general lack of information on outcomes from the six Scottish Centres and on how they compare with other units.

1.4 Cleft care is complex and varied and there is a need to improve our knowledge of the quality of cleft treatment in Scotland through audit to ensure continuous improvement.

RECOMMENDATIONS

Aetiology and prevention

1.5 Further research is needed to determine the possible protective effect of folic acid and other micronutrients in orofacial clefting and the guidelines on the use of these in the periconceptional period should be further promoted and the uptake monitored.

1.6 Further research is needed to identify the possible effect of other environmental and nutritional factors and how they interact with certain genes to predispose to orofacial clefting.

Scottish Cleft Lip and Palate (SCALP) registry and database

1.7 The SCALP registry and database is strongly supported as a tool for monitoring activity, assessing outcomes and audit and should be properly resourced.

Health needs of adult cleft patients

1.8 Research is needed into the health needs of adults who have partially treated or untreated orofacial clefts.

Diagnosis

1.9 There is a need to ensure appropriate training from the cleft team for clinicians who may make the initial diagnosis (including radiologists and paediatricians) and for midwives, nurses, health visitors and other health professionals who are involved in care at an early stage.

1.10 Staff in maternity units should have rapid access to the cleft team so that a designated team member can visit without delay.

Treatment

1.11 Cleft patients should have access to a full range of specialist care during the progress of their treatment.

1.12 Cleft centres without a comprehensive team (as defined by the Royal College of Surgeons criteria) should be able to demonstrate that satisfactory alternatives are in place.

1.13 Mechanisms should be in place to ensure effective communication between the cleft team and community services including the community paediatricians.

1.14 Written reports on individual cases should be provided to parents when requested and, possibly, where treatment is particularly complex.

1.15 The sessions available (see Table 4, p29) for professionals working in cleft teams should be reviewed with a view to ensuring optimum input, including services delivered in the community setting.

1.16 Audit and research should be conducted on a prospective multicentre basis to enable proper assessment of process and treatment outcome. Local Cleft Lip and Palate Association (CLAPA) groups should be involved in the production of regular consumer audit.

1.17 Patient records should be collected systematically and according to the Scottish Cleft Lip and Palate Group (SCALP) criteria and minimum data set, with cleft teams co-ordinating the record keeping, assessment and treatment of adult cleft patients.

1.18 Effective psychosocial support for cleft patients and their families should be available and further research into this aspect of care is needed.

1.19 Cleft teams should ensure that patients should have the benefit of a co-ordinated dental disease prevention programme from birth - possibly delivered by a dental hygienist in the cleft team.

Configuration of services

1.20 There should be a staged move towards no more than two cleft centres in Scotland whilst maintaining a network of high skill levels locally. This should fit with the concept of a managed clinical network as envisaged in the Acute Services Review.

Economics of change

1.21 Many issues need to be addressed in implementing the recommendations of this report. In particular, the recommendations on service reconfiguration have operational implications which need to be supported by an economic appraisal.

2. THE SCOPE OF THE REPORT

2.1 Cleft lip and/or palate is the most common abnormality in the cranio-facial region (OPCS, 1994). About 15% of clefts occur in association with a known genetic syndrome (syndromic clefts), approximately 10% may be shown to have an environmental link but most occur with no single apparent genetic or environmental cause. About 25-30% of syndromic clefts involve isolated cleft palate.

2.2 This report will primarily consider non-syndromic clefting in children and the information presented on birth prevalence refers to non-syndromic clefts. However, it is impractical to disassociate the two entirely since cleft teams treat patients with both syndromic and non-syndromic clefts. There are currently some 300 known syndromes associated with clefting and it is conceivable that this number will rise as research and our understanding of the genetic influences progresses.

2.3 The report does not attempt to include other congenital cranio-facial abnormalities (though it is recognised that the treatment of such conditions may also impact on the work of a cleft team), nor does it address the needs of adults who may have partially treated or untreated clefts. This is an important research area.

2.4 A range of issues are considered - from aetiology to rehabilitation - and a number of recommendations are made for the future commissioning of services for patients with cleft lip and palate.

3. DEFINING THE PROBLEM

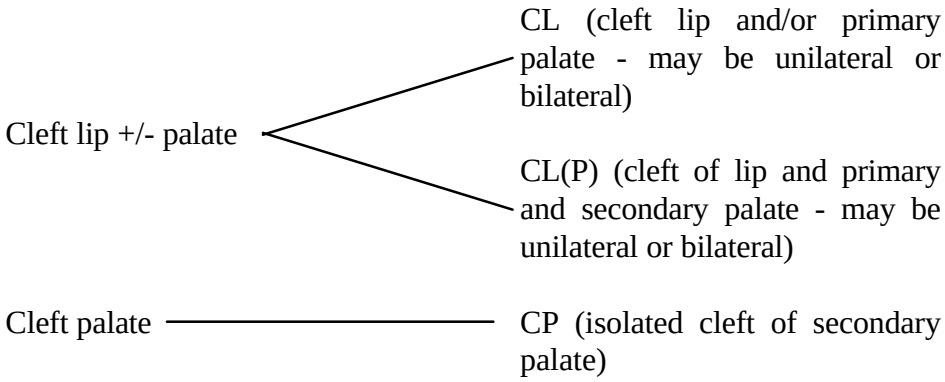
3.1 Introduction

3.1.1 Non-syndromic cleft lip +/- palate (CL(P)) and isolated cleft palate (CP) are distressing congenital malformations which occur in ~1.35:1,000 live births (Ross and Johnston, 1978; Piggott, 1992). In the UK population there are currently around 50,000 people with orofacial clefting with a birth prevalence rate of approximately 650 per year, presently accounting for 65% of all congenital craniofacial abnormalities (Williams et al, 1994).

3.1.2 The Scottish population is characterised by a particular predilection to congenital malformations including orofacial clefting and has the highest rate of isolated CP of all the EUROCAT (European Central Registry for Congenital Malformations at birth) centres (Stone and Dolk, 1994). There is also a unique CL(P) to CP ratio of almost 1:1 in the populations studied in Scotland and Northern Ireland (Fitzpatrick et al, 1994; Gregg et al, 1994). This is not typical of the rest of the UK or of other foreign studies where a ratio of 2:1 is more common.

3.2 Classification

3.2.1 Cleft lip (and/or palate) and cleft palate should be considered as separate entities (Kernahan and Stark, 1958). It is therefore helpful to divide orofacial clefts into three broad categories (see Figure 1):



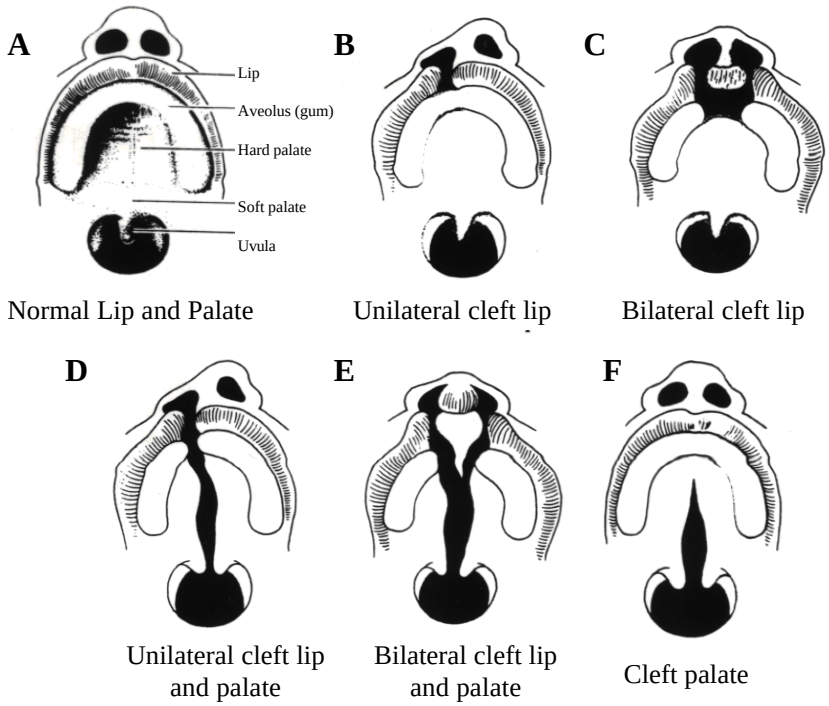


Figure 1: Diagrammatic examples of the variation in morphology of orofacial clefts. The structures of the roof of the mouth are viewed from below. Figure A identifies normal palatal structures in a non-cleft palate. Figures B, C, D and E are all classified as CL(P) - cleft lip with or without cleft palate. Figure F is classified as cleft palate (CP).

3.3 Aetiology of Orofacial Clefting

3.3.1 CL(P) and CP are considered as separate entities because of differences in respect of embryology, timing of fusion, epidemiological characteristics and mode of inheritance. They are both considered to have a polygenic multifactorial aetiology.

3.3.2 Unlike diseases determined by a single gene defect and inherited in a simple Mendelian fashion, the recurrence risk for orofacial clefting is based on evidence from familial studies. On the basis of such studies the recurrence risk in children of affected parents varies according to a number of characteristics such as the type of cleft, the number of affected family members and the severity of the malformation.

3.3.3 The contribution of environmental factors to adverse reproductive outcome is complicated by the fact that most studies in this field have been retrospective. There is, however, some evidence for an environmental contribution to both CL(P) and CP. The most powerful evidence for environmental contribution is from the study of twins, and it is noted that in monozygotic twins (i.e. identical twins who by definition have the same genotype) the frequency of both children being affected is 35% for cleft lip and palate and less than 30% for isolated cleft palate. The environmental factors which are thought to contribute to orofacial clefting are expanded upon in Appendix 1.

3.3.4 In recent years the influence of maternal nutrition, in particular folic acid, has aroused interest in the field of maternal reproductive health. Some aspects of maternal nutrition, such as folic acid, appear to have a protective effect and therefore deficiencies lead to adverse reproductive outcomes; while others such as Vitamin A are embryotoxic in excess. More detail on the role of maternal nutrition in orofacial clefting is given in Appendix 1.

3.3.5 Environmental factors may act as a trigger. Much of the contemporary research effort in orofacial clefting is aimed towards the analysis of gene/environment interaction. An example of such a study which is under way in Scotland at present is a Wellcome Trust funded study entitled 'Interaction Between Maternal Nutrition and Folate Metabolic Factors in the Aetiology of Orofacial Clefting'.

3.3.6 Further information on the aetiology and pathogenesis of orofacial clefting is provided in Appendix 1.

3.4 Complexities of cleft management

3.4.1 The treatment of clefts does not always follow a straight forward path. A number of well recognised complications can have implications both for the complexity and length of treatment. The following are among the more common:

Lip reconstruction

3.4.2 Many surgical techniques have been described which aim to produce a lip of normal length and thickness and with normal internal muscle positions. The aim is to enable the patient to have a lip which looks normal at rest and in function and which grows in proportion to the rest of the face.

3.4.3 In practice, these ideals are not totally achievable and some degree of lip scarring and nasal asymmetry are almost inevitable.

Palate Reconstruction

3.4.4 Primary surgical closure of the palate may fail to eliminate nasal speech and nasal escape of food or liquids. Poor valve function (velopharyngeal insufficiency) also demands further surgery in 20% of cases (Hall and Golding-Kushner, 1989).

3.4.5 Residual holes (fistulae) in the palate may require further surgery in about 20% of cases (Sell et al, 1994). Even small fistulae may affect both speech and resonance (Henningson and Isberg, 1987) although some children develop entirely normal speech (Harding, 1993).

Speech

3.4.6 Approximately 50% of children with repaired cleft palate will have speech and language difficulties which require specific interventions (Spreistersbach et al, 1973). Stengelhofen (1989) found that 40% have long standing speech problems.

3.4.7 Speech and/or language and its development may be affected by a number of factors associated with the cleft and as a result of some aspects of treatment such as:

- lip posture
- occlusion
- airway
- tongue posture
- scarring
- hearing
- velopharyngeal incompetence (VPI)
- psychosocial development

Cognitive skills and other aspects of development not specific to cleft palate may also influence speech and language development.

3.4.8 Historically, speech evaluation in judging the success of surgery has mainly concentrated on nasality but assessments varied and were therefore difficult to compare. Dalston (1988) suggested assessment should broaden to include hypernasality, hyponasality, intelligibility, nasal emission and articulation. Instrumental investigations of VPI and a distinction between cleft and non cleft errors are also suggested.

3.4.9 The speech and language therapy National Special Interest Group for Cleft Palate has recommended that for detailed assessment the Great Ormond Street Speech Assessment (GOSSPAS) should be used (Sell, Grunwell and Harding, 1994) and that for audit purposes the Cleft Audit Protocol for speech (CAPS) be used (Sell et al, 1996). This would enable meaningful comparisons to be made between centres.

Feeding

3.4.10 At birth 25% of cleft children will have feeding difficulties (Witzel et al, 1984) and appropriate advice on bottles, teats and feeding techniques will be required. Dental plates may be of value. Children born with a combination of a cleft palate and a small lower jaw (Pierre Robin syndrome) may have significant breathing and feeding difficulties and may require intensive nursing care and other treatment (including surgery) within the first year of life.

Other Anomalies

3.4.11 Fifteen percent of clefts occur as part of a wider syndrome (Shprintzen et al, 1985) which may involve oro facial, cardiac, skeletal or other abnormalities. Of the remaining 85% (non syndromic clefts), 50% have other less well defined anomalies (Shprintzen et al, 1985). Appropriate specialist paediatric input and adequate genetic advice is needed to confirm such diagnoses and to give advice on the possibility of recurrence in the family.

Hearing

3.4.12 Children with cleft palate are very prone (97%) to fluid collection in the middle ear (glue ear) and supervening infection (otitis media) due to poor structure and function of the drainage (Eustachian) tube. This compares to about 20% in the general population (Grant, 1988). Otitis media leads to problems with hearing and consequently this may affect speech and language development. Therefore, hearing will require longitudinal monitoring and possible treatment by an audiologist and ENT specialist.

Occult Clefts

3.4.13 Sub-mucous cleft palate may not be evident or recognised at birth but may result in the same speech, hearing and feeding problems as in more overt clefts. Referral of submucous clefts is often late because they are usually diagnosed as a consequence of the speech problems.

3.4.14 VPI may account for up to 50% of the speech and language therapy workload (Sell and Ma, 1996). Full investigation prior to possible surgery is required in such cases.

Dento-facial Anomalies

3.4.15 Approximately 50% of UK children have abnormalities of their occlusion (the way that their teeth fit together) (Brook and Shaw, 1989; O'Brien and Weyant, 1993). In addition, cleft children are likely to have some missing, small, malformed or mis-aligned teeth, especially if the gum (alveolus) is cleft.

3.4.16 By skeletal maturity, up to 50% of cleft patients will have an upper jaw which is so small that they will require further surgery in the late teens to bring the jaw forward into a more normal position (Shaw et al, 1992). Stability of such a correction is not totally guaranteed.

3.4.17 The recent report on cleft lip and palate from the Clinical Standards Advisory Group (CSAG) also noted that in the UK, 16% of the complete unilateral cleft lip and palate patients had not received their bone graft by age 12, and that only 58% of bone grafts had taken successfully (CSAG 1998).

3.4.18 Primary repair of the structures derived from the primary and secondary palate may result in interference with growth with narrowing of the dental arch. This may require expansion with braces before bone grafting can take place.

3.4.19 Should orthognathic surgery (surgery of the jaws to correct their relative position) be required, this would normally be carried out after facial growth has been completed, usually not before age 17 years. Orthodontic treatment may be required prior to surgery and for a short time afterwards.

3.4.20 Variation in jaw growth and potential osteotomy rate is regarded as an indicator of the quality of the early surgery and the amount by which it has tethered the upper jaw and prevented its normal downward and forward growth. The 1996 audit of UK cleft services (Shaw et al, 1996) showed that 70% of twelve year olds with a unilateral cleft lip and palate had some measurable 'smallness' of the upper jaw with respect to the lower jaw and in 39% of cases this was severe. In Norwegian cleft patients (Semb and Shaw, 1996), regarded as a good outcome group, the rate was 6%.

Dental Care

3.4.21 Scotland's poor dental health is well documented (Scottish Health Boards' Dental Epidemiological Programme reports 1987/88-1996/97). Should caries occur and subsequent early extraction of teeth arise this can cause or exacerbate malocclusion in cleft patients. Furthermore, poor oral hygiene is a contraindication to the wearing of orthodontic appliances due to an increased risk of further caries or decalcification of teeth and gingivitis. There is some recent evidence from a study in the Netherlands (Bokhout et al, 1997) that cleft patients are presenting with much higher caries rates than their non-clefted peers.

Psychosocial Problems

3.4.22 Evidence exists to support the role of facial appearance in stereotyping of perceived personality traits (Bradbury, 1994). Attractive individuals are perceived as being more popular, socially desirable and of greater intelligence. They receive more help and gain better marks and fewer punishments for work or misdemeanours than less attractive individuals.

3.4.23 Basic research in developmental psychology has shown that the nature of early social interaction is critical in a child's later development, and conditions which negatively affect such interaction often prove detrimental for a child's developmental outcomes. The limited amount of research that has been done on this aspect in craniofacial abnormalities suggests that such psychosocial difficulties do arise (e.g. Field and Vega-Lahr, 1984: Pannbacker, 1988; Barden et al, 1989). Unattractiveness equates with being less likable, more anti-social, having lower educational expectations and poor self-esteem (Clifford and Walster, 1973; Dion and Berschied, 1974).

3.4.24 Cleft children have problems in relation to quantity and quality of social relationships and interaction which may relate not just to appearance, but also to difficulties with speech and hearing (Strauss et al, 1988; Lansdown 1990). Parental concerns can also affect a cleft child as can differences in parent child perceptions of treatment needs. Several researchers have reported breakdown

in parent/child interactions either due to lack of parental knowledge of how to read their children's cues or limited capacity of some children with disabilities to stimulate their carers (Bowlby, 1958; Ainsworth, 1979; Belsky, Rovine and Taylor, 1984).

3.4.25 Further research on the psychosocial aspects of orofacial clefting is needed. In particular this should seek to identify the role and impact of the social environment in relation to cleft patients and their carers.

4. THE EXTENT OF THE PROBLEM

4.1 Birth prevalence of cleft lip and palate

(i) Global studies

4.1.1 For over fifty years investigators have been carrying out birth prevalence studies on patients with CL(P) and (CP) and from these reports the average birth prevalence of facial clefting in caucasian populations is 1:1000 total births for CL(P) and 1:2000 total births for CP.

4.1.2 Generally CL(P) occurs more frequently in males whereas for CP the reverse is true.

4.1.3 Significant racial differences in the birth prevalence of orofacial clefts exist. The birth prevalence of CL(P) varies from 2.1:1000 in Japan and 2.7:1000 in Canadian Indians to 0.4:1000 in Nigeria and 0.42:1000 in African Americans (Leck, 1972), with the geographical variation being less important than ethnic differences. Cleft palate alone (CP) has a lower average birth prevalence and shows little variation in different racial groups. The racial differences in CL(P) birth prevalence are likely to have a genetic basis as shown by Ching and Chung (1974) and are thought to be independent of environment.

4.1.4 Two thirds of all cases of unilateral CL (P) have left sided defects regardless of gender, race and severity of defect (Fraser and Calnan, 1961).

(ii) UK studies

4.1.5 At least seven localised studies (Birmingham, Northumberland, Liverpool, Trent, Edinburgh, Glasgow and Northern Ireland) concerned with the birth prevalence of cleft lip and palate have been reported in the last 40 years and these are summarised in Table 1. Further UK and Ireland data are presented in Appendix 2.

Table 1. Birth prevalence of Cleft Lip and/or Cleft Palate in the UK Reported in the Literature

Reference	Source	No. of Clefts	Population	Time Span	Rate per 1000 Births	Ratio clefts live births	Cleft palate only (%)
McMahon and McKeown (1953)	Clinical records	285	Birmingham	1940-50	1.30	1:769	40
Knox et al (1962)	Hospital records	574	Northumber-land	1949-58	1.42	1:704	33
Owens et al (1985)	Registry	456	Liverpool	1960-82	1.40	1:701	34
Womersley and Stone (1987)	Registry	247	Glasgow	1974-85	1.56	1:641	52
Coupland et al (1988)	Hospital activity analysis	930	Trent	1973-82	1.82	1:505	39
Gregg et al (1994)	Regional database	398	N Ireland	1980-90	1.28	1:781	53
Bellis & Wohlgemuth (1998)	Regional database	503	SE Scotland & Highlands	1971-90	1.4	1:709	45

The number of cleft cases per year in the UK depends in part on the total number of live births which have fallen from 799,258 in 1990 to 731,645 in 1995.

4.1.6 There is an apparent reduction in the number of babies born with cleft lip and/or palate in England and Wales between 1990-94 and this may be due to changes in reporting which, it should be noted, is a voluntary system. It might well be that some (although not necessarily all) of the variation in the birth prevalence of both cleft lip and cleft palate is due to a change in the completeness of recording over the period. Termination of pregnancy following pre-natal diagnosis is a possible further factor.

4.1.7 In Scotland the situation is different. There is a more complete reporting system to the Scottish Congenital Anomalies Register at the Information and Statistics Division (ISD) of the Common Services Agency. Figures have recently been obtained from ISD which have enabled regional comparisons to be made with regard to the number of cases of CP and CL(P) between 1988 and 1995. (Tables 2 and 3 and Appendix 4).

Table 2. Number of cases of Orofacial Clefting (OFC) in Scotland (1988-1995)

	1988	**1989**	**1990**	**1991**	**1992**	**1993**	**1994**	**1995**
CL(P)	71	52	63	64	80	51	58	58
CP	36	44	43	40	48	44	52	43
Total	107	96	106	104	128	95	110	101

4.1.8 Two independent studies in the West of Scotland by Womersley and Stone (1987) and Fitzpatrick et al. (1994) reported a remarkably high birth prevalence of isolated CP - 0.81 and 0.79 per 1000 live births respectively.

Table 3. Regional distribution of cases of Cleft Lip and/or Palate - Scotland (1988-1995)

Area	**1988**	**1989**	**1990**	**1991**	**1992**	**1993**	**1994**	**1995**
Glasgow	35	38	33	42	57	35	38	35
Edinburgh	29	25	32	24	38	24	28	31
Aberdeen	17	11	19	14	15	15	18	6
Dundee	8	6	12	12	7	8	13	14
Ayr	9	7	8	8	4	7	9	8
Falkirk	9	9	2	4	7	6	4	7
Total	107	96	106	104	128	95	110	101

This table should be read in conjunction with the geographic distribution of clefts in Scotland.

4.1.9 Table 3 shows the number of cases requiring care in the various Scottish centres. There are currently six centres for cleft lip and palate treatment in Scotland (Aberdeen Royal Hospitals NHS Trust, Dundee Teaching Hospitals NHS Trust, Edinburgh Sick Children's NHS Trust, Falkirk and District Royal Infirmary NHS Trust, South Ayrshire Hospitals NHS Trust and Yorkhill NHS Trust in Glasgow). It should be noted that some cross boundary flow occurs.

4.1.10 Figure 2 presents a dot density map showing the regional distribution of patients undergoing cleft procedures for the period April 1992-March 1998.

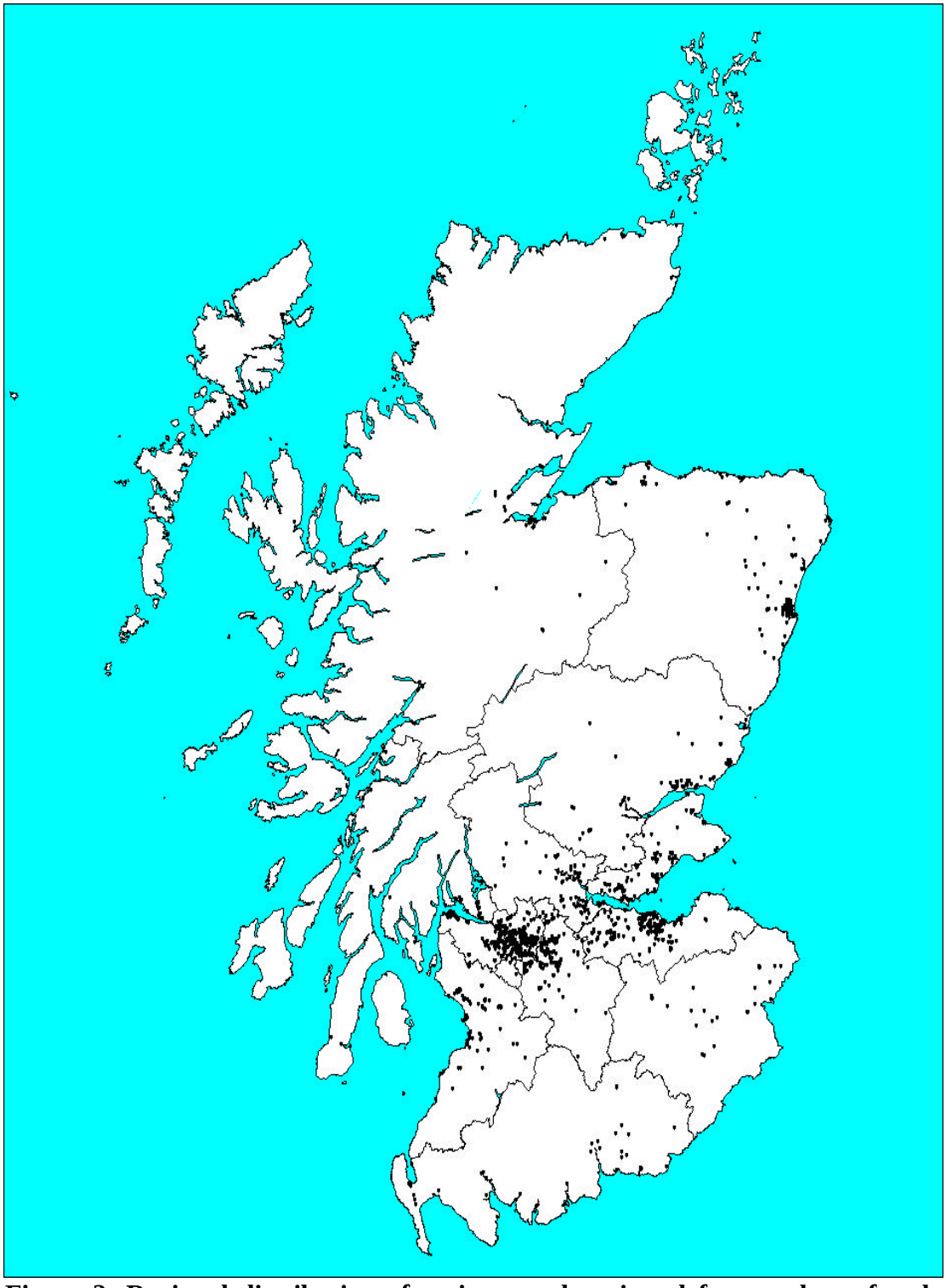


Figure 2: Regional distribution of patients undergoing cleft procedures for the period April 1992-March 1998

(iii) Trends in the birth prevalence of orofacial clefting

4.1.11 Appendix 2 shows cleft lip/palate surgery discharges for the period April 1992 to March 1998 by Carstairs deprivation category. This shows that patients in the highest deprivation category appear to suffer disproportionately in comparison to those in the lowest. It is not clear whether this is due to a higher birth prevalence of clefting or to poorer surgical outcomes or to a combination of the two. Rates of access to antenatal care and screening in high socio-economic population groups might also lead to a lower birth prevalence.

4.1.12 To have true figures on the total birth prevalence of a particular birth defect in the population, figures on the rate of spontaneous and induced abortions as well as stillbirths would be required. However the cause of early perinatal death is heterogenous and as well as a malformed embryo or foetus, complications such as infections, immunological incompatibility and disorders in the maternal reproductive organs can lead to prenatal death. The diagnosis of the disease or anomaly in question in the aborted foetus (in this case CL(P) or CP - syndromic or non-syndromic) would also be necessary. Complete and consistent collection of such data presents major problems.

4.1.13 Up to 10% of perinatal mortality and 15% of postnatal mortality is caused by congenital malformation. In Scotland, miscarriage accounts for 10-15% of all pregnancies (Regan et al 1989) and 50% of miscarried foetuses are found with genetic abnormalities (Creasy et al, 1976 and Boué et al, 1975). Recurrent early pregnancy loss (REPL) occurs in 1% of all women (Rae et al, 1996) and parental chromosomal abnormality presents in 3-5% of couples with REPL (Stray Pedersen B, Stray Pedersen S, 1984; De Braekeleer M, Dae T N, 1990; Hay P E et al, 1994)

4.1.14 There is also concern that trends in the incidence of OFC may reflect differences in the level of completeness of recording (or ascertainment). Considerable efforts are being made to ensure that these differences are minimised. Initiatives such as the European Registry for Congenital Anomalies (EUROCAT) produce data which is consistent and validated.

4.1.15 One of the most comprehensive data sets comes from Denmark, where there is a mandatory reporting system and a rigorous ascertainment procedure. The reported birth prevalence of OFC in Denmark has risen from 1: 667 live born infants in 1942 to 1:529 in 1981 (Jensen et al, 1988). This increase may be due to better reporting and recording, decreased neo-natal mortality, increasing environmental teratogens (e.g. drugs) and increased frequency of marriage in cleft patients because of better care.

4.1.16 Some trends are detectable from the available data (of which the EUROCAT data are regarded as being the most reliable) and these show that globally there is no reduction in the birth prevalence of orofacial clefting despite considerable improvements in nutrition. This might well be due to a concomitant increase in exposure to other environmental teratogens, or be a genetic phenomenon.

4.1.17 In the UK, the number of reported cases of orofacial clefting fell from 759 in 1990 (1:930 live births) to 630 in 1995 (1:1054 live births). The national reporting system, for reasons already explained, is likely to be an underestimate of the number of orofacial clefts born each year and makes it difficult to undertake meaningful audit of the outcome of cleft care in England and Wales (Shaw et al, 1996).

4.1.18 In Northern Ireland, considerable efforts have been made to create a reliable database. Overall the birth prevalence of CLP was 1:719 (Gregg et al, 1994), although the figures varied considerably from year to year, partly because of the small number of cases.

4.1.19 In Scotland the trend has remained fairly static and, for the reasons stated above, the national reporting system is likely to be more accurate.

4.2 SCALP database

4.2.1 In 1989 a project was initiated by the cleft teams in the Scottish centres to produce a national registry and database. This has become known as the Scottish Association for Cleft Lip and Palate (SCALP) registry and database.

4.2.2 The aim of this registry is to provide a validated database of all children born with syndromic and non-syndromic clefts in Scotland. In addition, data collection protocols have been devised for each speciality - cleft surgery, orthodontics, speech and language therapy and audiology so that consistent and co-ordinated records will be available for audit and research.

4.2.3 The Craniofacial Society of Great Britain has recently awarded a grant to enable a validation against the two existing registries in Scotland, the Scottish Congenital Anomalies Register (SCAR), Information Services Division and the European Concerted Action on Congenital Anomalies and Twins (EUROCAT) registry which is a well ascertained database in a defined geographic region in Greater Glasgow.

4.2.4 This is likely to become a valuable research resource in the future and there is scope for the database to be a major tool in prospectively measuring and comparing surgical outcome using the following key indicators:

- post-operative naso-labial soft tissue profile and nasal symmetry;
- study casts and lateral cephalograms at 5 year intervals as a measure of dental arch and skeletal discrepancy;
- quality of speech and velopharyngeal competence during function;
- post-operative outcome of bone grafting;
- hearing at various key stages of development;
- patient and parent satisfaction with the service and quality of outcome.

4.2.5 The Scottish cleft units are currently monitoring compliance (through the SCALP group) with a suggested minimum dataset (Appendix 7). Minor modifications may still be necessary but it is recommended that cleft teams all aim to comply with this dataset as soon as possible.

4.2.6 Finally, it should be noted that the figures given do not include submucous clefts and other problems (eg. velopharyngeal dysfunction) which are referred to a cleft team - often by speech and language therapists.

4.2.7 Weatherley-White (1972) suggested a birth prevalence of submucous clefting of 1:2000 live births but the birth prevalence of other anomalies such as non-cleft velopharyngeal dysfunction is unknown. It is the experience of speech and language therapists who are involved in managing such cases that non-cleft velopharyngeal problems may account for up to half their workload (Sell and Ma, 1996).

4.3 Prevalence of cleft lip and palate

4.3.1 In many ways the critical factor in orofacial clefting is the birth prevalence and this should not be confused with the incidence (which is essentially the number of affected embryos). With an overall birth prevalence of 1:700 there are probably well in excess of 7000 people in Scotland who have had some form of orofacial clefting.

4.3.2 A small proportion of these will be adults who have had no treatment, or rudimentary treatment, and who may require ongoing intervention from any of the cleft specialities. There is little information currently available on the treatment needs of such patients and this should be addressed.

5. CURRENT SERVICE PROVISION

5.1 Prevention

5.1.1 Waitzman et al. (1994) estimated the lifetime costs of cleft lip/palate defects for children born in California in 1988. It was found that approximately 85% of costs were the result of losses in productivity due to premature mortality and morbidity, the remaining 15% were due to medical costs and cost of developmental services and special education. The results were based partially on an estimate of 11.7% of sufferers being unable to work and 17.2% limited in work. Clearly, prevention has the potential for considerable economic, social and personal benefits.

5.1.2 Most current global research efforts are focused on the association between orofacial clefting and vitamins, including folic acid. It is still not clear whether vitamins are protective or whether the apparent protective effect is due to other characteristics of vitamin users compared to vitamin non-users.

5.1.3 If some vitamins are protective, it is unclear whether this is due to the action of folic acid or some other factor. In Scotland a multicentre research team has begun a population based case control study in three UK locations in order to try to determine whether there is an association between orofacial clefting and maternal dietary intake of vitamins during the periconceptional period and to identify genetic factors that mediate such susceptibility.

5.1.4 The nutritional epidemiology of orofacial clefts has not been investigated in the UK. The proportion of women taking folic acid supplementation is low - despite recommendations from the Scottish Office Department of Health. Some Health Boards in Scotland have sought to encourage folate supplementation with local policies, training and materials.

5.1.5 Currently, there is global interest in the idea of periconceptional multivitamin supplementation and in some countries food fortification programmes have been approved. This is mainly due to the evidence for the protective effect of folic acid against the occurrence and recurrence of neural tube defects (Czeizel and Dudas, 1992). In addition there is evidence for a preventive effect against vascular diseases, conotruncal heart defects, urinary tract malformations, limb reduction defects (Czeizel, 1998) as well as orofacial clefting (Tolarova, 1982; Shaw et al, 1995).

5.1.6 Periconceptional multivitamin supplementation is undoubtedly the most efficacious way of delivering the appropriate levels of nutrients including folic acid for the prevention of congenital anomalies. Consumption of foods which are rich in folate may not provide the same level of protection. However, as a large proportion of pregnancies are unplanned, the widespread use of bread fortified with folic acid, vitamin B12 and vitamin B6 may have a considerable protective effect against neural tube defects and other congenital abnormalities. Some countries, such as Hungary, have agreed that these vitamins will be added to bread from the second half of 1998 (Czeizel, 1998).

5.1.7 In the absence of conclusive data on orofacial clefting at present it is nevertheless recommended that the guidelines on folic acid supplementation in the periconceptional period continue to be promoted in Scotland.

5.2 Detection and Diagnosis

5.2.1 Problems for a family with a cleft child may begin even before birth. Some clefts of the lip are diagnosed by prenatal ultrasound, yet the radiologist or obstetrician may have little knowledge of the condition or its treatment possibilities. In the case of syndromic clefts, the diagnosis may be made following serum alfa fetoprotein (AFP) estimation. In cases where a prenatal diagnosis is made it is important that immediate advice is available from the cleft team.

5.2.2 Most clefts however are detected in the labour ward where there is a need for timely and accurate information to be available for all parents. If this is not given, parents may be left confused and distressed. Lack of accurate information can also lead to false expectations regarding treatment time, complexity and success.

5.2.3 CLAPA (The Cleft Lip And Palate Association) is currently piloting a project in the UK in association with the Kings Fund with the aim of increasing knowledge about cleft lip and palate among midwives, nurses, health visitors and obstetricians, who are normally the first point of contact for parents of a cleft baby.

5.2.4 We recommend that cleft teams in Scotland should ensure that all maternity units have staff who are appropriately trained and updated in giving first line advice and support to parents of cleft babies - wherever the diagnosis is made. This should include the use of protocols to ensure appropriate pathways of care and include provision of counselling and social support.

5.2.5 It is important that such staff have rapid access to the cleft team itself so that a designated team member can visit without delay.

5.2.6 Most cleft babies are born without any known family history. The input of a clinical geneticist or counsellor at an early stage is important. The geneticist may also offer prenatal testing in the case of a future pregnancy.

5.3 Treatment and oral disease prevention overview

5.3.1 The treatment of children born with a cleft lip and/or palate normally follows a well defined sequence - ongoing until adulthood. In addition to the original treatment, speech and language therapists, orthodontists, otologists and audiologists, dental hygienists, restorative and paediatric dentists, paediatricians, geneticists and specialist nurses are among the wide range of specialists who may be involved in the care of cleft children - with the emphasis firmly on the multi-disciplinary team approach.

Cleft Lip

5.3.2 In Western society the requirements of aesthetics and function dictate that surgical reconstruction of lip defects is mandatory - usually within the first six months of life. This is a difficult procedure given the complex three dimensional distortion of many of the internal and external structures of the lip and nose.

Alveolar (gum) Cleft

5.3.3 If the alveolus (gum) is cleft there is often a lateral displacement of the two segments of the upper jaw - greatly increasing the width of both the alveolar and the lip cleft in a new born child.

5.3.4 Some surgeons feel that lip repair is facilitated by the use of neonatal dental plates (pre-surgical orthopaedics) - sometimes combined with facial strapping to narrow the cleft. This is to obtain improved arch and lip positioning prior to surgical repair.

5.3.5 In bilateral clefts, the central section of alveolus and lip may protrude noticeably from the rest of the face and in these cases pre-surgical alignment by means of suitable oral appliances and extra oral strapping is often felt to be beneficial.

Palate Repair

5.3.6 If the palate is cleft, the defect is normally closed after the lip repair and usually before 18 months of age. Sometimes soft and hard palates are repaired in separate stages.

5.3.7 The aim is to produce an intact palate, with proper orientation of the internal muscles, so that the posterior or movable part of the palate can function successfully as a component of a valve mechanism to prevent air, food, or liquids escaping into the nose during swallowing and speech.

Speech

5.3.8 The specialist speech and language therapist will access, record and monitor patients until adulthood as well as carrying out specialist evaluation, audit and treatments. Whenever possible therapy/intervention is carried out locally under the direction of the specialist therapist. There is evidence to suggest that babble patterns can predict late speech difficulties - early intervention is therefore desirable.

5.3.9 Specialist speech instrumentation evaluation is initiated, co-ordinated and interpreted by the specialist speech and language therapist. This includes:

- videofluoroscopy (in conjunction with radiologists);
- nasopharyngoscopy (usually in conjunction with surgeons);
- computer linked airflow/nasalence/nasal and oral air flow instruments such as an anemometer, a nasometer, SNORS or aerophonscope - there is a wide

range of instruments available. Each measures a different parameter but few have comparable standardised scores;

- electropalatography (EPG) to analyse and treat tongue/palate placement during speech.

Referral is recommended to the newly published 'Clinical Guidelines by Consensus for Speech and Language Therapists' (RCSLT, 1998).

Orthodontics

5.3.10 After preliminary orthodontic expansion, bone grafting and eruption of the majority of the second teeth, further comprehensive orthodontic treatment with full fixed braces is normally required to align the teeth adequately. Often, if only one tooth in the line of the cleft is missing, other teeth can be moved to fill the space. In other cases, artificial replacements in the form of bridgework, dentures or implants may be required. The Index of Orthodontic Treatment Need (IOTN) is now nationally recognised as a means of prioritising orthodontic patients and classifies cleft patients in a maximal orthodontic need category.

5.3.11 It is essential if treatment is to be undertaken successfully that basic routine dental care is adequately provided. It is important that this dental care be underpinned by a preventive approach and that patients have full and ready access to routine caries and gingival disease prevention and, if necessary, specialist paediatric dental care. Older patients will also require adequate preventive advice, routine maintenance and access to advanced restorative care where required.

Bone Grafting

5.3.12 Where there is a cleft involving the alveolus, surgical repair of the bony defect in the alveolus (bone grafting) is usually recommended and is normally carried out around the age of 9 or 10 years - before second teeth have erupted in this area. This will improve the chances of the teeth appearing in a favourable position and their later alignment with orthodontic braces. It may well also improve lip and nose appearance as well as closing any residual oronasal fistula.

Surgery

5.3.13 The primary surgery, including the closure of any palatal defect, is often performed by a plastic surgeon (Piggot R W, 1992) and occasionally by paediatric or oral and maxillofacial surgeons. However, Piggot found in his UK survey that the average annual surgical workload was only 10.2 cases.

5.3.14 ENT Surgeons are involved with drainage procedures on the middle ear. Oral and maxillofacial surgeons or plastic surgeons are usually involved at the stage of alveolar bone grafting and again in late teenage years for orthognathic surgery when it is needed.

Current treatment services

5.3.15 In 1996 a Steering Group from the Surgical Audit and Epidemiology Unit at the Royal College of Surgeons of England recommended a set of minimum standards for the management of cleft lip and palate. These are presented in Appendix 5.

5.3.16 The background to the work of the group was an intercentre audit in the late 1980s which revealed poorer outcomes for facial growth for British patients with cleft lip and palate than for equivalent patients in Northern Europe.

5.3.17 A subsequent survey of surgical practice in England and Wales revealed the widespread involvement of low volume operators in cleft care, a tendency for these low volume operators to have an incomplete network of associated professionals and non-standardised record keeping protocols (Williams et al, 1994).

5.3.18 For the purposes of the SNAP report the working group decided to audit current practice in Scotland against the RCS criteria as the gold standard and at the same time to conduct an inter-centre comparison of protocols for surgical interventions, timing and record keeping. The comparison also included an overview of the sessional commitments of key clinicians in the six Scottish centres.

5.3.19 The audit took the form of a questionnaire which was circulated to the six Scottish centres.

5.4 Current Service Provision

Comprehensive service

5.4.1 Most centres had a fully comprehensive team. Exceptions were:

- one centre did not have specialised counselling or specialist nursing;
- one centre had no plastic surgeon or specialist dental advice in the team;
- two centres had no clinical geneticist within the team (access by referral);
- two centres had no developmental paediatrician available to the cleft team (except by referral).

Comment

5.4.2 The current provision of cleft services in Scotland is divided amongst six centres. Some smaller units are not able to offer the same access to specialist services as the larger units can. This is not ideal though the significance is unclear since the effect on patient outcome is not able to be quantified.

5.4.3 It is therefore recommended that centres without a comprehensive team (as defined by the RCS criteria) should be able to demonstrate that there are satisfactory alternatives. Where this involves a referral, there should be mechanisms to ensure that patients are not disadvantaged.

Co-ordinated treatment

5.4.4 Each centre had centralised registrations, record keeping and treatment planning functions; all but one engaged in multi-disciplinary audit. Two centres did not hold regular instructional courses for non-team members such as health visitors and midwives.

Comment

5.4.5 The SCALP database (see section 4.2) has been a significant development in Scotland and should ensure that a mechanism is available for assessing and comparing outcomes. The issue of instructional courses for non-team members is considered to be important given the comments in section 5.2.4 and all centres should therefore hold regular courses for non-team members.

Antenatal and neonatal counselling

5.4.6 All teams provided rapid assessment of new cases on the day of birth, advice to parents and information on future management by an experienced member of the team - apart from one team in one peripheral location.

Comment

Cleft teams should be taking the lead in ensuring that trained staff are available in all locations in their geographical catchment areas.

5.4.7 In peripheral units the initial contact will not necessarily be with a member of the core cleft team and teams should take steps to ensure that families always have accurate and timely information available from a local health professional who works in conjunction with the cleft team.

Neonatal nursing

5.4.8 One centre did not have a nurse specialist but a speech and language therapist provided early instruction on feeding, nursing and continuing community care.

Surgery

5.4.9 In all centres, key surgical procedures were performed by experienced surgeons with extended cleft lip and palate training but only one surgeon (working in two centres) carried out a minimum of 30 new cases per year.

5.4.10 Numbers of new cases per year for the other surgeons varied considerably - from 2 to 26 cases. The issue of surgical workload is discussed in section 6.2.

Orthodontic treatment

5.4.11 The variability of patient numbers per orthodontist was greater than for surgeons; only one orthodontist treated more than 30 new cases per year. The issue of orthodontic workload is discussed in section 6.2.

Dental care

5.4.12 Reported mechanisms for delivering patient education were uniform across all centres but referral to a paedodontist was variable. Three centres had no access to a paedodontist, and it was not clear how caries and gingival disease prevention was managed in each centre.

5.4.13 The opportunity for a consultant in restorative dentistry to contribute to treatment planning at a stage early enough to influence outcome was also variable. Two centres reported no formal mechanisms to ensure that this took place.

Comment

5.4.14 The availability of a specialised paedodontic service is considered to offer advantages although there is also evidence that dental caries in cleft patients can be effectively prevented via the input of other dental health professionals - e.g. dental hygienists (MacFadyen and McLennan, 1984). It may therefore be more appropriate to recommend that all cleft teams at least have access to a dedicated dental hygienist in each centre. The use of agreed protocols on preventive dental care is important. The arrangements for involvement of a consultant in restorative dentistry, without being a core member of the cleft team, presents a difficulty. There are advantages for teams based in cities with local access to a wide range of dental specialties.

5.4.15 There is also anecdotal evidence that some cleft children who attend the orthodontist regularly may regard this as sufficient dental input and not be under ongoing care and review in the primary care dental setting.

5.4.16 It is important that the total dental care package for cleft patients includes the primary care dimension and specific mechanisms should be developed by cleft teams to ensure lines of communication with primary care are effective.

Speech and Language therapy

5.4.17 All the centres reported meeting the criteria for speech and language therapy but only one centre had a team therapist involved with over 30 new cases per year. The issue of speech and language therapy workload is discussed in section 6.2.

Hearing assessment and treatment

5.4.18 The survey did not look at assessment methods. All centres tried to co-ordinate any middle ear drainage operation with others to reduce the number of anaesthetics given. Protocols need to be established and audit carried out.

Clinical genetics

5.4.19 Only three centres had a geneticist in the cleft team - the other centres relying on secondary referral. Whilst a clinical geneticist may not need to be a core member of the cleft team it is considered essential that appropriate and timely access is available.

Psychological counselling

5.4.20 No teams included a psychologist or counselling professional although most were able to refer to a specialist service. Arrangements for assessment at key stages were variable.

Comment

5.4.21 It is difficult to suggest a precise model for providing psychology or counselling services. On one hand, the working group considered that too strong an emphasis might bring stigmatisation whilst on the other hand the team needed to be able to identify those patients who would clearly benefit from counselling or psychology services. This may vary at different stages in the patient's life.

5.4.22 Cleft teams should review their protocols to ensure that specialist psychologist or counselling professionals work effectively as part of the wider cleft team and that patients who would benefit from such input are identified without delay.

5.4.23 Very little research has been conducted to address the question of whether orofacial clefting alters the interactions between a parent and infant, despite the fact that the findings may hold implications for treatment - including the age at which lips should be surgically repaired, the multi-disciplinary care protocols that should be recommended, the kinds of support that should be offered to families and its effect on outcomes.

5.4.24 Basic research in developmental psychology has shown that the nature of early social interaction is critical for the child's later development and conditions which negatively affect such interaction often prove detrimental for developmental outcomes. For example, research has shown that interactions between mother and child where there is infant visual impairment, hearing impairment, or Down's syndrome, exhibit a decreased amount of face-to-face contact, eye gaze, positive facial expression and maternal vocalisations (e.g. Jones, 1980; Jamieson and Pederson, 1993; Moore and McConachie, 1994; see Zeedyck, 1996 for review).

5.4.25 Such deficits give rise to problems in a child's longer term development, in domains such as cognitive and social skills, self-esteem and academic performance (e.g. Meins et al, in press; Panaccione and Whaler, 1996). The very limited amount of research that has been done on early interaction and

cranio-facial abnormalities suggests similar difficulties may exist as in these other groups (Barden et al, 1989; Field and Vega-Lahr, 1984; Pannbacker, 1988).

5.4.26 Further research is needed to investigate the effect of pyschosocial factors on the development of the cleft child and to determine any link to treatment protocols.

5.4.27 It should be emphasised that the involvement of professional services will always run the danger of stigmatisation and that the importance of local and national support groups as well as mechanisms of social support are very important and should be recognised as such by health professionals. Teachers may well have an important role too.

Paediatric developmental medicine

5.4.28 Arrangements for liaising with the community paediatricians varied from centre to centre. All centres provided information from the cleft team to community services and had mechanisms in place to investigate suspected developmental or growth delays.

Comment

5.4.29 All children with clefts should be known to the community paediatric consultants and although this is already likely to be the case in Scotland it is suggested that cleft teams send copies of letters to the paediatricians to ensure that no children are missed. The role of the general medical practitioner is also important in the links between the cleft team and other community child health professionals. Cleft teams should routinely copy correspondence to the community paediatric consultants to ensure good information channels. The community paediatricians provide an important link to a wider community team including health visitors, social workers and other professionals.

Communication with parents

5.4.30 Four centres undertook regular consumer audit although in one this had not happened for some time. No centres automatically provided families with written reports of multi-disciplinary assessments. There is a UK CLAPA network with three CLAPA groups currently in Scotland and a Scottish regional CLAPA representative. These groups work closely with the Cleft Teams.

Comment

5.4.31 It is recommended that families should be actively encouraged to see the patient support groups as part of the overall care process. Local groups

should be involved in the production of regular consumer audit. Further research into the information needs of patients and carers is needed.

5.4.32 The issue of providing families with written multi-disciplinary assessment reports is contentious. It is recommended that these be available if patients request them and that the practicality, benefit and medico-legal implication of making these available to all patients should be assessed. It may be of value to designate a member of the cleft team as the single point of contact for a patient. The use of patient 'advocates' may also have merit and information on these should be routinely available to patients and carers.

Clinical audit, research and records

5.4.33 All six centres participated in clinical audit and are working through the SCALP database to collect comprehensive records for all patients. The initiative to develop the SCALP database is of particular importance.

Review of surgical protocols and timing

5.4.34 There is a vast amount of literature on surgical techniques and timing for repair of the primary cleft defect, both of the lip and the palate. This is an indication of the complexity and heterogeneity of the problem. It is important to note that this is not merely a cosmetic operation like the surgical closure of a wound - the aim is to restore the anatomy which has been disrupted by the clefting defect.

5.4.35 Different techniques may have different short-term and long-term outcomes and one of the major challenges in audit and research in orofacial clefting is to establish which are best. It is however clear that a long-term multi-disciplinary approach is essential for optimum anatomical, physiological and functional results.

5.4.36 It is known that there appear to be differences in various UK centres in the proportion of patients with a repaired cleft lip and palate who develop maxillary retrusion and there is evidence to suggest that facial growth is related to the effects of primary surgery in infancy (Ross, 1987; Mars and Houston, 1990; Semb, 1991).

5.4.37 It has also been demonstrated (Witzel et al, 1994; Sell, 1995) that delayed repair probably results in less facial growth deficiency, but such benefits must be balanced against the disadvantages of prolonged feeding problems and speech abnormalities which may result from a delay in closing the secondary palate.

5.4.38 For the purposes of the SNAP report, protocols and timing of interventions were reviewed for pre-surgical orthopaedics, primary repair, palatal repair, secondary surgery, alveolar bone graft and orthognathic and other later surgery.

5.4.39 There was substantial uniformity in reported surgical practice for lip/nose reconstruction amongst 5 centres. One unit favoured a functional (Delaire) approach and one unit carried out neonatal lip reconstruction on selected patients. There was variation in the palatal reconstruction between and within centres. There was also some variation in the use of pre-surgical orthopaedics with three centres using it, two centres rarely using it and a third "possibly".

Comment

5.4.40 The degree of consistency in surgical practice was considered to be substantial for a lip/nose reconstruction. However, it should be emphasised that evidence for any technique will only become available through proper inter-centre audit of outcomes and the development of strict and detailed protocols.

5.4.41 There is no nationally or internationally agreed treatment regime for early orthopaedic management of clefts. A recent review of pre-surgical orthopaedics concluded that "not one research paper has ever been published investigating the possible benefits to cleft surgery resulting from pre-surgical orthopaedic treatment". A prospective study into the use of pre-surgical orthopaedics is planned as part of the BIOMED funded 'Eurocleft' project.

Clinical sessions

5.4.42 Units were asked to estimate the sessional commitment for a number of members of the core cleft team - surgeons undertaking primary repairs, orthodontists, speech and language therapists, surgeons undertaking bone grafting, ENT surgeons, paediatricians, team co-ordinators and specialist nurses.

5.4.43 The results showed a considerable degree of variation. This could simply be due to the difficulty of estimating the time spent on treating clefts when this is only part of a larger workload. It may also reflect the tendency for members of smaller teams to develop dual roles.

5.4.44 However, some variations could be due to real differences in staffing of cleft teams and the following variations are highlighted as being of concern.

Surgeon

5.4.45 The sessional time for surgeons undertaking primary surgery appeared to vary from one unit where there were two weekly sessions available to treat an annual workload of twelve new patients, to a unit where there were five sessions available to treat an annual workload of fifty new patients.

Orthodontist

5.4.46 There was also a variation in reported orthodontic sessional time which ranged from four sessions to treat an annual workload of twenty six patients, to three sessions to treat an annual workload of fifty patients.

Speech and Language Therapist

5.4.47 Similar variations were observed with reported sessions ranging from five sessions to treat an average annual workload of twelve new cases to seven sessions to treat an annual workload of fifty new cases.

5.4.48 The reported sessional data should be treated with caution as there is no standardised way in which sessions are recorded in each centre. However, in view of the fact that there may be real differences present it is recommended that work be undertaken to establish a minimum level of sessional input for each core team member related to the expected annual new patient workload.

Table 4: Sessional commitments: ratio of sessions per week to annual number of patients

	High ratio	Low ratio
Surgeons	2:12 (1:6)	5:50 (1:10)
Orthodontists	4:26 (1:6.5)	3:50 (1:16.6)
Speech and language therapists	5:12 (1:2.4)	7:50 (1:7.1)

Other cranio-facial abnormalities

5.4.49 This report addresses the issue of cleft lip and palate but it is emphasised that the work of the cleft team encompasses other abnormalities which have direct implications for workload and team structure.

Syndromic Clefting

5.4.50 Clefts of the lip and/or palate may occur as part of a syndrome. Although these occur less commonly than non-syndromic clefts the treatment may be more complex. There are now over 300 recognised syndromes of which isolated CP or CLP are part, and even in 'non-syndromic orofacial clefting' there are often other associated abnormalities which may or may not be diagnosed at birth.

5.4.51 These more rare conditions in aggregation make up a significant proportion (20-25%) of the workload in a cleft palate clinic. Clinically it is very important to make these diagnoses as early as possible. This is because it can have a major influence in prognosis, may impact on management or have inheritance implications. In addition, the clinical geneticist can record and interpret the family pedigree, examine patients for other abnormalities, organise appropriate screening tests and interpret the results of these, as well as carry out genetic counselling where appropriate (Reid, 1998).

5.4.52 Patients with hypernasality problems, submucous clefts and other cranio-facial abnormalities which do not exhibit clefting may all impact upon the workload of the cleft team. In one cleft unit the speech and language therapist estimated her sessional commitment as:

- 60% on routine non-syndromic clefting;
- 40% on other craniofacial syndromes and non-cleft velopharyngeal incompetence (VPI) eg. velocardiofacial syndrome.

Non-cleft cases presented on average every two weeks and required about two hours of clinical time.

Untreated/partially treated adult clefts

5.4.53 Section 4.3.2 introduced the issue of untreated or partially treated adult clefts. Little data exists on the problem but it is clear that some cases will present to the cleft team for assessment or treatment. It is recommended that research is done to quantify the health needs of adults with untreated and partially treated clefts and that cleft teams have a lead role in co-ordinating their treatment.

6. EFFECTIVENESS OF CURRENT PROVISION

6.1 Outcome measures

6.1.1 Various methods have been devised to define whether the outcome of the treatment sequence for a cleft patient has been effective or not. A number of outcome measures are in common usage and the SCALP database project (see section 4.2) is designed to provide a series of objective and comparable outcome measures.

Skeletal relationship

6.1.2 The relationship of the upper and lower jaws relative to the skull base is assessed using lateral cephalometric radiographs. The extent to which the upper jaw is underdeveloped relative to the lower jaw is considered to provide one indicator of the quality of the early surgery.

6.1.3 The debate on outcomes has centred on the apparent differences in surgically induced growth disturbance between cleft units in the United Kingdom and those in other parts of Europe. A simple measure of growth disturbance has been the proportion of cleft patients who require an osteotomy in their late teens to correct the under development of the upper jaw and mid-face. At present it is not known what proportion of orofacial clefting cases in Scotland require osteotomies.

6.1.4 However, a study of patients who had had their operations in the UK in the late 1970s compared clinical outcomes for one group of children with complete unilateral clefts with those for a similar group of patients in Norway, Denmark and the Netherlands. 48% of patients from one UK centre were judged to have surgically induced growth disturbance of such severity that an osteotomy would be required in the late teens to correct maxillary retrusion (Shaw et al, 1992). The equivalent figure for the Norwegian centre was 6%.

Dental arch relationship

6.1.5 The relationship of the upper and lower teeth is assessed by taking impressions and making plaster models of the teeth. Dental arch relationship in orofacial clefting is often a reflection of maxillary growth deficiency. The relationship is usually measured using the GOSLON yardstick (Mars et al, 1987), a dental measure with outcomes ranked on a scale of 1-5. The SCALP database contains sufficient information to allow a baseline analysis of current outcome with respect to dental arch relationship in the Scottish cleft treatment centres.

Alveolar bone grafting

6.1.6 The outcome of the alveolar bone grafting is assessed using oblique anterior occlusal radiographs.

6.1.7 In the CSAG study 36% of 5 year olds and 39% of 12 year olds had a poor dental arch relationship reflecting an underlying skeletal discrepancy. 16% of 12 year olds had not received an alveolar bone graft and, in those who had, only 58% were judged to have been successful. (CSAG, 1998)

Speech

6.1.8 The Eurocleft Cross Centre Speech Study of the same cohort found similarly unfavourable results in terms of speech outcome for the UK group compared to the Norwegian centres. Whilst this is suggestive of an inferior standard of surgical and perhaps non-surgical care in the UK centres, there are a number of confounding factors which complicate this analysis.

6.1.9 The CSAG report also carried out an outcome audit of cleft care in the UK by measuring dental arch relationship, success of alveolar bone grafting, patient profile, naso-labial appearance, speech, hearing and patient satisfaction including psychosocial status. This showed that, although parents were generally satisfied with the outcome of care received, there were significant residual treatment needs in surgery, speech therapy, hearing and dental care (CSAG, 1998).

Improving the assessment of outcome

6.1.10 The main difficulties in auditing outcomes of treatment of orofacial clefting are the relative rarity of the condition, the heterogeneity of the deformity and the long time span over which the audit must be done.

6.1.11 In both audit and research there is a need for more multi-centre collaborative studies. Cleft centres with small case loads do not generate sufficient data quickly enough for meaningful inter-centre comparisons to be made.

6.1.12 The data collection protocols set up by SCALP will allow comparisons to be made of various outcome measures of the quality of surgery in terms of dental arch relationship, skeletal relationship, success of alveolar bone grafting and speech outcome. However, there are no international comparisons for speech, hearing and facial appearance that allow meaningful comparisons to be made at the moment. There are no internationally agreed comparisons.

6.1.13 The Eurocleft study highlighted the importance of good clinical records, so that clinical audit can be carried out in a systematic manner. Without good records it is impossible to quantify the appropriateness of the treatment provided. It should be recognised that the satisfactory collection of records has inherent manpower costs which must be considered.

6.1.14 Records must be taken in a systematic way at appropriate ages to enable inter-centre audit to be carried out. Clinicians should ensure that this information is obtained for every child. (see recommended minimum dataset in Appendix 7)

6.1.15 Consideration should be given to the inclusion of the views of patients and carers as an outcome measure. This could, for example, encompass measures of appearance, self esteem, social and physical functions.

6.2 Workload

6.2.1 There is a body of opinion that the poor results observed in the UK are at least partly related to workload. Thus, the minimum standards for the management of cleft lip and palate (Shaw et al, 1996) from the Surgical Audit and Epidemiology Unit, The Royal College of Surgeons of England, suggested a minimum case load of 30 new patients annually for primary surgery, 20 cases for alveolar bone grafting and VPI surgery, 15 cases for maxillary osteotomy, 30 cases for orthodontics and 30 cases for speech and language therapy.

6.2.2 These threshold numbers for competence were based partly on minimum numbers for other forms of surgery and partly on recognition that meaningful audit required a minimum case load.

6.2.3 The CSAG report recommended a reconfiguration of cleft centres in order to achieve a sufficient throughput of cases in each.

6.2.4 Surgeons need a sufficient number of cases on which to operate so that effects of technique and changes in technique can be monitored objectively.

6.2.5 The GOSLON index is widely regarded as the best available objective measure in cleft treatment. In order to detect a difference of 0.5 on the GOSLON scale (1 - 5) at 5% probability and with 80% power, an annual caseload of some 60 patients is needed if reliable results are to be obtained in less than a decade.

6.2.6 A high volume operator is no guarantee of high quality outcome. However, because of the lengthy period of time which is required to follow up cleft patients and the variety of initial presentation, the quality of outcome for a cleft surgeon can only be timeously assessed in a unit with a high caseload.

6.2.7 This kind of centralisation of surgery would enable a surgeon in training to pursue further training in order to become a full time cleft surgeon. They may be selected from those specialities with most involvement in clefts (usually plastic or oral and maxillofacial surgery). This would also enable sub-specialisation in subjects such as surgery for speech problems, bone grafting, osteotomy and bone distraction work and rhinoplasty as these are areas of work needing concentrated practice.

6.3 Audit

6.3.1 The audit carried out by the SNAP working group showed that only one surgeon in Scotland currently meets the "threshold numbers for competence" suggested by Shaw et al. However, the SNAP working group did not consider that the relationship between increased case load and improved outcomes is fully proven - nor that the outcomes in any of the Scottish centres are necessarily poor. Many features of cleft services in Scotland are considered to be excellent and in some cases well in advance of other UK centres.

6.3.2 Centres with small numbers are not viable when it comes to evidence based assessment of quality of care - not because the quality of care is necessarily poor, but because the minimum volume required to demonstrate quality cannot be achieved within a reasonable time period. The SCALP database is an ideal vehicle for data collection for prospective audit of process and outcomes.

7. CHANGES TO CURRENT PROVISION

7.1 There is inevitably a conflict between centralisation of services to achieve better assessment of clinical outcome and dispersal of services to make them more accessible to patients.

7.2 On the basis of the evidence of considerable variation in outcome from other cleft units and the need to promote comparable audit of process and outcome of the treatment of orofacial clefts within Scotland, it is proposed that the number of centres carrying out primary cleft surgery be reduced. The CSAG report recommended a similar reduction within the UK.

7.3 However, it should be recognised that cleft services in Scotland have developed in a much more homogeneous way than in many other parts of the UK and that the services currently provided are already well integrated. There is therefore a need to build on the high standards of service provision of the cleft teams already in existence.

7.4 It is proposed that the critical areas of treatment planning and case overview, record keeping and primary surgery should eventually be carried out in no more than two centres. These central cleft teams should direct and co-ordinate care and might be regarded as the centre for training (including the training of staff working in the peripheral units).

7.5 The recently published Acute Services Review in Scotland (Acute Services: Review Report 1998) recommends the development of managed clinical networks. The recommended changes in cleft lip and palate services should be a suitable case for piloting this principle.

7.6 It is considered important that the current expertise in the peripheral units should be retained and developed such that much of the continuing work could still be provided locally. Further, it is not envisaged that the relationship between the centre and the other units would be a hierarchical one but rather an extended network of cleft clinicians. Consideration should also be given to the effect of the proposed changes in the peripheral units so that appropriate clinical leadership is retained.

7.7 Whilst there is little objective data on the user perceptions of the services currently provided in Scotland, there is a view that service users might be prepared to trade some diminution of local service for better outcomes. This should be monitored independently.

7.8 There are not thought to have been any service specifications or specific contracting arrangements for cleft services in Scotland in the past and it is recommended that commissioners give priority to developing these in conjunction with this report. This process would be facilitated by a move towards fewer centres.

8. ECONOMICS OF CHANGE

8.1 There are many economic issues to disentangle when considering the impact of a move to two surgical centres in Scotland. The main issues relate to the effects on patient access, cost per case, outcomes, the feasibility of such a move and the effect on service agreements.

Access costs

8.2 As section 7.1 indicates, any consideration of the implications of a two centre model of surgical services must account for the increase in the cost of access to patients.

8.3 CSAG (1998) found that in the UK 30% of parents lived more than one hours travelling time from home to clinic. Thirty six per cent of parents expressed difficulties in travelling to clinics. There are five principal patient costs incurred in the treatment of cleft lip and/or palate: transport, subsistence, childcare, loss of earnings and the cost of lost education to the child.

8.4 Appendix 6 undertakes an exploratory analysis of access costs for patients, under the status quo and under a situation where cleft lip/palate surgery is delivered in two centres. For the purposes of the analysis, Glasgow and Edinburgh (the units with the highest current throughput) have been shown as the two centres although it should be emphasised that this is for illustrative purposes only, and should not be interpreted as a recommendation of this report.

8.5 The analysis finds that the access costs incurred by patients' families currently amounts to approximately £84,000. With a move to these two centres, family costs would rise to approximately £144,000 for Scotland. The brunt of these cost increases would be incurred by families in Grampian, Tayside and the three island Boards. Given that the greatest level of cleft surgery activity is in the lowest socio-economic group (see section 4.1.11), the increase in access costs might disproportionately affect the most deprived in society.

Cost per case

8.6 There is no strong evidence that the proposed move to two centres would lead to a reduction in cost per case. It is sometimes argued that, centralisation in general may allow human and physical capital to be used to its full capacity. In the case of human capital, this would mean that expensive training would be fully utilised in large centres, rather than under-utilised in small centres. This should reduce training requirements, forcing down costs. However, as central clinics increase in size, lines of communication may become distorted or discontinued, leading to a reduction in clinicians' efficiency and an increase in cost per case. Therefore, although intuitively one might think that increasing surgical volume would reduce cost per case, there is little evidence to suggest that this is the case.

Outcomes

8.7 A further factor concerns the economic benefits of improved outcomes. If a volume-outcome relationship exists in the treatment of cleft lip/palate, then improvements in outcomes in the early years of life should avert remedial treatment in later years. As well as having an intrinsic value, improved surgical outcomes should lead to a reduction in treatment costs and costs to patients and their families. It should be emphasised that other treatment outcomes would also be influenced by an improved surgical result.

8.8 This is difficult to estimate in Scotland since there is currently insufficient available data on outcomes. If CSAG's figures of 48% of UK cleft patients requiring osteotomy in their late teens are robust and if it is assumed that this could be reduced to just 18% (in Norway just 6% of patients required osteotomy - CSAG, 1998) then the approximate discounted value of costs averted would be £13,200 to the NHS and £2,430 to patients and their families (see Appendix 6)

8.9 Therefore the net effect of a move to two centres could be to increase costs by approximately £45,500. It is strongly emphasised that these figures are based on a number of assumptions.

Feasibility of Centralisation and Impact on Other Hospital Services

8.10 The impact of a move towards no more than two centres would have some effect on sites where services would cease as it would in those sites where services would expand. In hospitals where services would cease, there may be an impact on staffing and training, as well as the provision of surgical and diagnostic tools and support facilities. Nevertheless, smaller centres in Scotland do as few as 12 new cases annually and these centres should be able to deal adequately with the reconfiguration of the service.

8.11 In those hospitals where services expand, an examination of their capacity to expand, and the effect on related services, is also complex. It is impossible to estimate the eventual cost per case in the proposed larger centres, as it will depend on their levels of spare capacity and potential for expansion. A rigorous costed service specification for prospective centres would be required to determine eventual cost per case.

Effect on Service Agreements

8.12 The final issue that needs to be considered is more a financial than a strictly economic one. While centralisation may lead to improved outcomes for a similar cost per case, leading to efficiency gains, it will be difficult to realise financial savings in those centres where surgical workload is discontinued. As section 7.8 outlines, purchasers usually have block agreements for plastic surgery and oral surgery services with hospital sites, so there would be a need to reconfigure agreements to account for the discontinuation of cleft care in certain sites.

9. CONCLUSIONS

- Orofacial clefting is a relatively common condition with an birth prevalence of approximately 1 in 650 live births in Scotland;
- The present provision of services for people with cleft lip and palate is not optimal and shows considerable variation;
- Treatment outcomes are not as good in the UK as is achieved elsewhere. It is unclear how treatment outcomes in Scotland compare with other areas;
- Multidisciplinary long term planning and provision of treatment is essential for good clinical outcomes;
- The volume of activity in some existing centres is too low to demonstrate quality of outcome within a reasonable time period and the future configuration should operate on the basis of a managed clinical network.

10. RECOMMENDATIONS

Aetiology and prevention

10.1 Further research is needed to determine the possible protective effect of folic acid and other micronutrients in orofacial clefting and the guidelines on the use of these in the periconceptional period should be further promoted and the uptake monitored.

10.2 Further research is needed to identify the possible effect of other environmental and nutritional factors and how they interact with certain genes to predispose to orofacial clefting .

Scottish Cleft Lip and Palate (SCALP) registry and database

10.3 The SCALP registry and database is strongly supported as a tool for monitoring activity, assessing outcomes and audit and should be properly resourced.

Health needs of adult cleft patients

10.4 Research is needed into the health needs of adults who have partially treated or untreated orofacial clefts.

Diagnosis

10.5 There is a need to ensure appropriate training from the cleft team for clinicians who may make the initial diagnosis (including radiologists and paediatricians) and for midwives, nurses, health visitors and other health professionals who are involved in care at an early stage.

10.6 Staff in maternity units should have rapid access to the cleft team so that a designated team member can visit without delay.

Treatment

10.7 Cleft patients should have access to a full range of specialist care during the progress of their treatment.

10.8 Cleft centres without a comprehensive team (as defined by the RCS criteria) should be able to demonstrate that satisfactory alternatives are in place.

10.9 Mechanisms should be in place to ensure effective communication between the cleft team and community services including the community paediatricians.

10.10 Written reports on individual cases should be provided to parents when requested and, possibly, where treatment is particularly complex.

10.11 The sessions available (see Table 4, p29) for professionals working in cleft teams should be reviewed with a view to ensuring optimum input, including services delivered in the community setting.

10.12 Audit and research should be conducted on a prospective multicentre basis to enable proper assessment of process and treatment outcome. Local CLAPA groups should be involved in the production of regular consumer audit.

10.13 Patient records should be collected systematically and according to SCALP criteria and minimum data set, with cleft teams co-ordinating the record keeping, assessment and treatment of adult cleft patients.

10.14 Effective psychosocial support for cleft patients and their families should be available and further research into this aspect of care is needed.

10.15 Cleft teams should ensure that patients should have the benefit of a co-ordinated dental disease prevention programme from birth - possibly delivered by a dental hygienist in the cleft team.

Configuration of services

10.16 There should be a staged move towards no more than two cleft centres in Scotland whilst maintaining a network of high skill levels locally. This should fit with the concept of a managed clinical network as envisaged in the Acute Services Review.

Economics of change

10.17 Many issues need to be addressed in implementing the recommendations of this report. In particular, the recommendations on service reconfiguration have operational implications which need to be supported by an economic appraisal.

APPENDIX 1
Page 1 of 3

AETIOLOGY OF OROFACIAL CLEFTING

Orofacial clefting such as CL(P) and CP clusters in families but its inheritance does not conform to Mendelian laws. It is thought that the inheritance of orofacial clefting is explained by the concept of genetic liability or susceptibility to a given characteristic, governed by many different genes, and a threshold, determined by both genetic and environmental factors (Fraser, 1970).

This concept of inheritance makes several predictions - namely that:

- the defect in question will cluster in families;
- the risk for first-degree relatives of affected individuals (parents, siblings, and offspring) will approximate the square root of the population risk;
- the risk for second-degree relatives (uncles, aunts, half-siblings) will be sharply lower than the risk for first-degree relatives;
- the more severe the malformation, the greater the risk for recurrence;
- the greater the number of affected family members, the greater the risk for recurrence;
- the risk for recurrence will be increased for relatives of the least affected sex, if gender differences are noted;
- consanguinity will increase the risk.

There is firm evidence for an aetiological distinction between cleft lip with or without cleft palate (CL(P)) and isolated cleft palate (CP) alone (Fogh-Andersen, 1942; Curtis et al, 1961; Ferguson, 1987). In both, the genetic basis is complex but, unlike population data for CL(P), data regarding CP rarely conformed to any model predictions, suggesting the probability of a greater environmental component (Shields et al, 1981; Carter et al, 1982; Czeizel, and Tusnady 1984).

The epidemiological evidence and recurrence rates for CP and CL(P) suggest genetic predisposition, and an increasing number of candidate genes for both types of orofacial clefting are currently under investigation. Evidence suggests that major genes predispose to non-syndromic OFC in certain individuals and families (e.g. Murray et al, 1986). Segregation analysis on a West of Scotland CP population (Fitzpatrick and Farrall, 1993) suggests that the number of genetic susceptibility loci may be as few as three or four. To date a number of studies, including one carried out in Scotland, have reported an association of CL(P) with the transforming growth factor alpha (TGFα) locus (Ardinger et al., 1989; Chenevix-Trench et al., 1991; Holder et al., 1992; Stoll et al., 1992; Shiang et al., 1993; Sassani et al.1993; Mossey, 1994).

Genetics

Evidence is also emerging for a general distinction between CL(P) and CP with different candidate genes being implicated in each and there is evidence or a difference even at the same genetic locus. Allele frequency distribution at the transforming growth factor alpha (TGFα) locus differs in parents of children with CL(P) and CP (Mossey, 1994).

Unlike monogenetically determined diseases, the recurrence risk for orofacial clefting has only been estimated in the past, on the basis of family studies. Such studies have shown that the recurrence risk in children of affected parents varies between 4% and 15% (Fogh-Andersen, 1942; Tolarova 1971, 1984). However the predictive value of this process is lower than expected and it is now thought that this might be because of a major gene effect.

Adverse environmental factors

It is not easy to demonstrate the contribution of environmental factors to human orofacial clefts. The vast majority of studies have been retrospective and may therefore have introduced a memory recall bias. Review of the literature on orofacial clefting however does reveal several significant environmental teratogenic factors.

Alcohol produces a characteristic craniofacial abnormality known as foetal alcohol syndrome. This occurs in approximately 1:1000 live births and is characterised by microcephaly and a typical appearance of short palpebral fissures, short nose, long upper lip with deficient philtrum, small midface and small mandible (Jones, 1983). Clefts of either lip or palate are only an occasional feature of the foetal alcohol syndrome. Apart from foetal alcohol syndrome, maternal alcohol use during pregnancy has been shown to increase the risk of non-syndromic cleft lip with or without cleft palate, but not isolated cleft palate (Munger et al, 1996).

Other teratogens have been implicated in the cause of cleft palate in man through epidemiological studies and have also been found to induce cleft palate in laboratory animals, viz. corticosteroids, aspirin, diazepam and retinoids (Wilson, 1977). Teratogenesis in animals however does not imply teratogenesis in humans (Jelinek, 1984). Some studies suggest that maternal smoking may increase the relative risk for clefts between two and sixfold (e.g. Erikson et al, 1979; Werler et al, 1990 and Keels, 1991) whereas other studies (e.g. Hill, 1988) have yielded negative results. This confounding evidence might be explained by genetic predisposition and therefore future studies will concentrate on gene/environment interaction. Hwang (1995) and Shaw (1996) have both demonstrated an increased teratogenic effect of smoking in mothers with certain TGFα gene polymorphisms. These studies have illustrated the influence of genetic predisposition on the potency of environmental factors in the production of orofacial clefting.

The fathers of new-borns with clefts tend to be older on average than the fathers of neonates without an orofacial cleft. However, teratogens may only have an effect where there is a parental genetic susceptibility. Further, clefts will only result if there is exposure to certain noxious environmental factors in specific doses, in specific combinations and at a specific time during pregnancy (Saxen, 1974).

APPENDIX 1
Page 3 of 3

Nutrition

Folic acid supplements may have a protective effect for orofacial clefting as this has proven effective in the prevention of neural tube defects (MRC vitamin study, 1991). The essential role of folic acid in DNA methylation, protein synthesis and cell replication may be one of the reasons why its insufficiency during early embryonic development could contribute to the aetiology of birth defects, such as neural tube defects and OFC.

Whilst folic acid deficiency is thought to have adverse affects on the developing embryo, some other vitamins in excess such as vitamins A D and E may be embryotoxic. Hypervitaminosis A has been extensively studied in rodents and is a potent teratogen causing cleft palate in strains that are genetically predisposed (Seller et al, 1979); and there are anecdotal reports of congenital abnormalities in human embryos exposed to excess vitamin A (Bernhardt and Dorsey, 1974). Further human studies on the effects of vitamin A and retinoic acid and in particular the influence of gene environment interaction are urgently required.

Drugs

Various drugs have been implicated in the cause of cleft palate in man through epidemiological studies and some have also been found to induce cleft palate in laboratory animals, *viz* anticonvulsant drugs, corticosteriods, aspirin, diazapam and retinoids (Wilson, 1977). Of these the anti epileptic drug Phenytoin has a proven teratogenic effect and the suspected mechanism is its folate antagonistic property. A host of other drugs which are also folate antagonists, such as Methotrexate, Carbamazapine, Sulphonomides and antimalarial drugs may also be implicated. There are still many unanswered questions regarding the mechanisms involved and possible interaction between vitamins, drugs and illnesses, (Dostal and Blahova, 1986). The relevance of animal and tissue culture experiments to clinical practice remains unknown.

APPENDIX 2
Page 1 of 1

Orofacial clefting birth prevalence data: UK and Ireland

Country	Date of Study	Live/ Still	Syndromic/ Non-syndromic	CL ± P	CP	Ref
West Scotland	1974-1985	Live	Non-Syndromic	0.75	0.81	1
West Scotland	1980-1984	Live	All Inclusive	0.677	0.763	2
Glasgow	1980-1992	Live	Non-Syndromic	0.72	0.77	3
Glasgow	1990-1994	Live	Non-Syndromic	0.79	0.79	4
England-Wales	1974-1988	------	------	0.92	0.92	5
Belfast	1980-1992	Live	Non-Syndromic	0.62	0.57	3
Belfast	1990-1994	Live	Non-Syndromic	0.27	0.21	4
Dublin	1980-1992	Live	Non-Syndromic	0.84	0.66	3
Dublin	1990-1994	Live	Non-Syndromic	0.83	0.73	4

APPENDIX 3
Page 1 of 2

Cleft Lip/Palate Surgery discharges: April 1992-March 1998

By Carstairs Deprivation Category

Category	Number	% distribution	Population[1]	Discharges per 100000 pop
Total	**1 741**	**100.0**	**4 998 256**	**34.8**
1 Least Deprived	61	3.5	300 690	20.3
2	219	12.6	706 249	31.0
3	364	20.9	1 104 648	33.0
4	458	26.3	1 247 924	36.7
5	272	15.6	751 278	36.2
6	177	10.2	548 934	32.2
7 Deprived	169	9.7	338 533	49.9
Unknown	21	1.2		

1. From the 1991 census

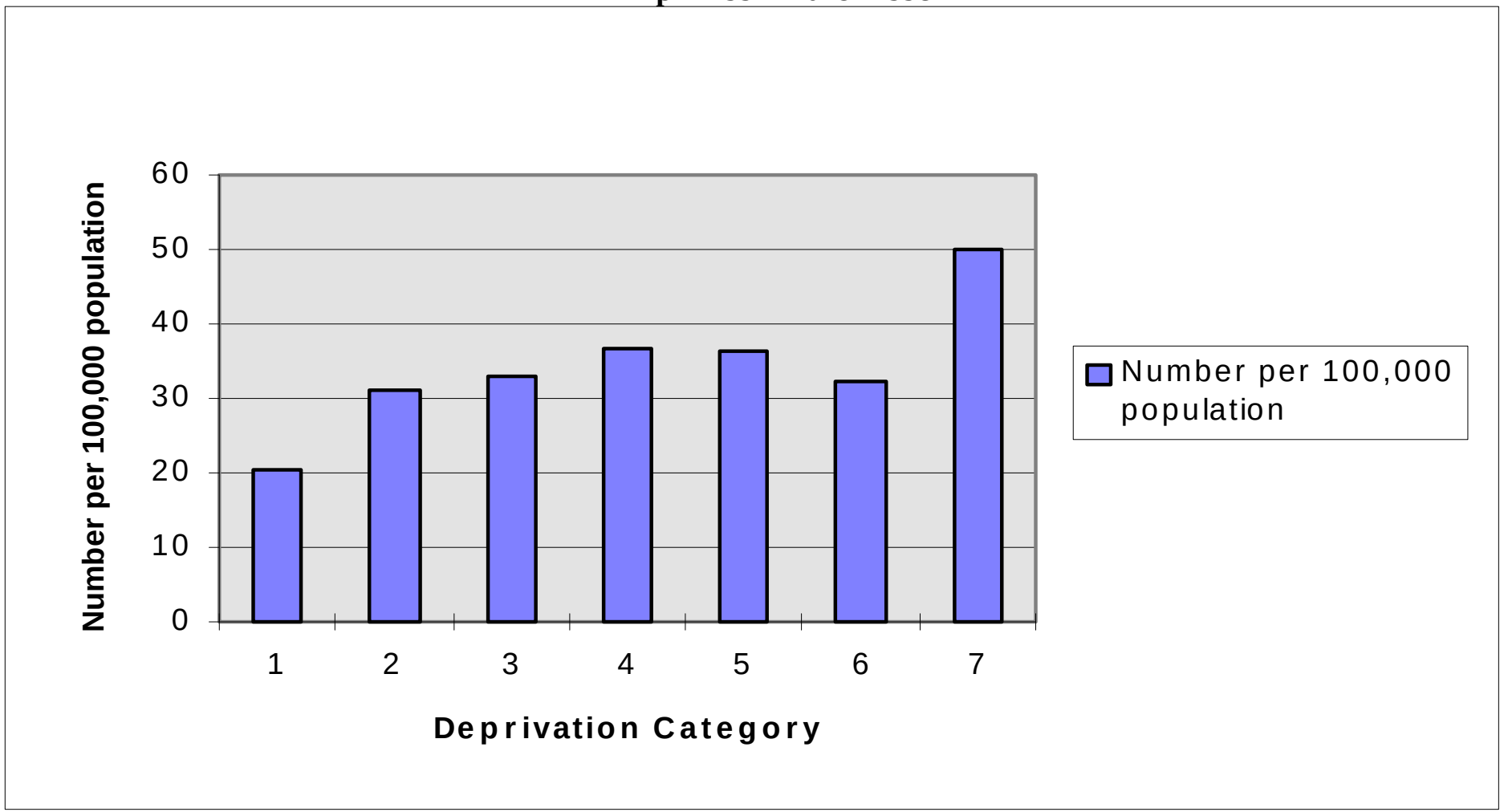
Source: SMR1

Produced by
WACIU,
ISD Scotland

Ref: MK8/080

APPENDIX 3
Page 2 of 2

Cleft Lip/Palate Discharges per 100,000 population by Carstairs Deprivation Category: April 1992-March 1998



APPENDIX 4
Page 1 of 4

Cases of cleft palate and cleft lip - Scotland*
By area of residence, year and diagnosis: 1988-92

Area of Residence		**1988**				**1989**				**1990**				**1991**				**1992**			
Health Board	**Local Government District**	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip
Argyll & Clyde	Argyll & Bute	1	-	-	1	1	-	-	1	2	-	1	1	5	2	2	1	3	-	-	3
	Dunbarton	3	-	2	1	-	-	-	-	1	-	-	1	3	1	2	-	4	1	2	1
	Inverclyde	1	-	1	-	-	-	-	-	3	1	1	1	2	-	-	2	2	-	2	-
	Renfrew	1	-	-	1	2	1	-	1	2	-	2	-	4	1	1	2	6	2	-	4
Area Total		**6**	**-**	**3**	**3**	**3**	**1**	**-**	**2**	**8**	**1**	**4**	**3**	**14**	**4**	**5**	**5**	**15**	**3**	**4**	**8**
Ayrshire & Arran	Cumnoch and Doon Valley	1	1	-	-	-	-	-	-	1	-	-	1	-	-	-	-	-	-	-	-
	Cunninghame	1	1	-	-	3	3	-	-	4	-	1	3	3	1	1	1	1	1	-	-
	Kilmarnock and Loudon	3	2	-	1	2	1	-	1	2	-	-	2	4	3	-	1	-	-	-	-
	Kyle and Carrick	4	1	1	2	2	-	-	2	1	1	-	-	1	-	-	1	2	-	-	2
Area Total		**9**	**5**	**1**	**3**	**7**	**4**	**-**	**3**	**8**	**1**	**1**	**6**	**8**	**4**	**1**	**3**	**3**	**1**	**-**	**2**
Borders	Berwickshire	1	1	-	-	1	1	-	-	1	1	-	-	-	-	-	-	2	2	-	-
	Ettrick and Lauderdale	1	1	-	-	1	-	-	1	-	-	-	-	-	-	-	-	2	-	-	2
Area Total		**2**	**2**	**-**	**-**	**2**	**1**	**-**	**1**	**1**	**1**	**-**	**-**	**-**	**-**	**-**	**-**	**4**	**2**	**-**	**2**
Dumfries&Galloway	Annandale and Eskdale	-	-	-	-	3	2	1	-	-	-	-	-	-	-	-	-	1	-	-	1
	Nithsdale	1	1	-	-	-	-	-	-	-	-	-	-	1	-	1	-	1	1	-	-
	Wigtown	1	-	-	1	1	-	-	1	-	-	-	-	1	-	-	1	1	-	-	1
Area Total		**2**	**1**	**-**	**1**	**4**	**2**	**1**	**1**	**-**	**-**	**-**	**-**	**2**	**-**	**1**	**1**	**3**	**1**	**-**	**2**
Fife	Dunfermliine	5	3	-	2	2	2	-	-	3	1	1	1	5	1	-	4	5	3	-	2
	Kirkcaldy	4	-	3	1	4	3	-	1	2	-	1	1	4	-	1	3	7	3	1	3
	North East Fife	3	1	-	2	-	-	-	-	1	1	-	-	2	1	-	1	1	1	-	-
Area Total		**12**	**4**	**3**	**5**	**6**	**5**	**-**	**1**	**6**	**2**	**2**	**2**	**11**	**2**	**1**	**8**	**13**	**7**	**1**	**5**
Forth Valley	Clackmannan	4	1	1	2	3	1	1	1	-	-	-	-	-	-	-	-	1	1	-	-
	Falkirk	2	-	2	-	3	-	3	-	1	-	1	-	2	-	2	-	3	1	1	1
	Stirling	3	2	1	-	3	1	-	2	1	1	-	-	1	1	-	-	2	-	-	2
Area Total		**9**	**3**	**4**	**2**	**9**	**2**	**4**	**3**	**2**	**1**	**1**	**-**	**3**	**1**	**2**	**-**	**6**	**2**	**1**	**3**
Grampian	City of Aberdeen	7	2	-	5	4	2	1	1	5	1	1	3	5	1	1	3	9	4	-	5
	Banff and Buchan	1	-	-	1	1	-	1	-	4	2	-	2	2	-	-	2	2	-	1	1
	Gordon	1	-	1	-	2	-	-	2	4	2	-	2	-	-	-	-	1	-	1	-
	Kincardine and Deeside	1	-	-	1	2	1	-	1	-	-	-	-	1	-	-	1	-	-	-	-
	Moray	2	2	-	-	1	-	-	1	3	2	1	-	3	1	-	2	2	2	-	-
Area Total		**12**	**4**	**1**	**7**	**10**	**3**	**2**	**5**	**16**	**7**	**2**	**7**	**11**	**2**	**1**	**8**	**14**	**6**	**2**	**6**
Glasgow	Bearsden and Milngavie	1	-	-	1	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-
	Clydebank	1	-	1	-	-	-	-	-	1	-	-	1	1	-	-	1	2	1	1	-
	Eastwood	-	-	-	-	1	1	-	-	1	1	-	-	4	1	3	-	-	-	-	-
	City of Glasgow	12	3	3	6	15	8	2	5	15	6	6	3	14	8	4	2	25	8	8	9
	Strathkelvin	-	-	3	1	1	1	-	-	1	-	1	-	1	-	-	1	2	2	-	-
Area Total		**18**	**3**	**7**	**8**	**17**	**10**	**2**	**5**	**18**	**7**	**7**	**4**	**20**	**9**	**7**	**4**	**29**	**11**	**9**	**9**

APPENDIX 4
Page 2 of 4

Area of Residence		**1988**				**1989**				**1990**				**1991**				**1992**			
Health Board	**Local Government District**	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip	Tot al	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip	Tot al	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip
Highland	Bademoch and Strathspey	-	-	-	-	-	-	-	-	1	-	-	1	-	-	-	-	-	-	-	-
	Caithness	2	1	-	1	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-
	Inverness	-	-	-	-	1	1	-	-	1	-	-	1	2	1	1	-	2	1	-	1
	Lochaber	1	-	1	-	-	-	-	-	-	-	-	-	-	-	-	-	1	1	-	-
	Nairn	-	-	-	-	1	1	-	-	1	-	-	1	1	-	1	-	-	-	-	-
	Ross and Cromarty	2	2	-	-	2	-	-	2	1	-	-	1	-	-	-	-	-	-	-	-
	Skye and Lochalsh	-	-	-	-	-	-	-	-	1	1	-	-	-	-	-	-	-	-	-	-
	Sutherland	-	-	-	-	-	-	-	-	1	1	-	-	-	-	-	-	-	-	-	-
Area Total		**5**	**3**	**1**	**1**	**4**	**2**	**-**	**2**	**6**	**2**	**-**	**4**	**3**	**1**	**2**	**-**	**3**	**2**	**-**	**1**
Lanarkshire	Cumbernauld and Kilsyth	1	-	-	1	4	1	1	2	-	-	-	-	-	-	-	-	-	-	-	-
	East Kilbride	-	-	-	-	2	-	2	-	-	-	-	-	4	1	1	2	2	1	1	-
	Hamilton	-	-	-	-	2	1	-	1	1	-	-	1	-	-	-	-	-	-	-	-
	Clyesdale	1	-	1	-	-	-	-	-	1	-	1	-	-	-	-	-	1	1	-	-
	Monklands	7	3	1	3	6	3	1	2	3	2	1	-	1	1	-	-	2	-	-	2
	Motherwell	1	-	1	-	3	2	-	1	2	1	1	-	3	1	1	1	8	1	5	2
Area Total		**10**	**3**	**3**	**4**	**17**	**7**	**4**	**6**	**7**	**3**	**3**	**1**	**8**	**3**	**2**	**3**	**13**	**3**	**6**	**4**
Lothian	East Lothian	2	1	-	1	2	2	-	-	1	-	1	-	2	1	-	1	3	1	-	2
	City of Edinburgh	6	1	-	5	7	4	1	2	9	5	3	1	6	4	-	2	10	2	4	4
	Midlothian	1	1	-	-	1	-	1	-	3	1	1	1	1	1	-	-	1	1	-	-
	West Lothian	4	1	2	1	2	1	-	1	7	3	1	3	2	1	-	1	5	3	1	1
Area Total		**13**	**4**	**2**	**7**	**12**	**7**	**2**	**3**	**20**	**9**	**6**	**5**	**11**	**7**	**-**	**4**	**19**	**7**	**5**	**7**
Tayside	Angus	-	-	-	-	-	-	-	-	5	3	-	2	2	1	-	1	2	1	-	1
	City of Dundee	4	2	1	1	2	-	-	2	5	3	2	-	6	4	1	1	3	1	1	1
	Perth & Kinross	-	-	-	-	3	-	1	2	1	1	-	-	2	1	-	1	-	-	-	-
Area Total		**4**	**2**	**1**	**1**	**5**	**-**	**1**	**4**	**11**	**7**	**2**	**2**	**10**	**6**	**1**	**3**	**5**	**2**	**1**	**2**
Islands	Orkney	2	1	-	1	-	-	-	-	1	1	-	-	-	-	-	-	-	-	-	-
	Shetland	2	-	2	-	-	-	-	-	1	-	1	-	1	-	-	1	-	-	-	-
	Western Isles	1	1	-	-	-	-	-	-	1	1	-	-	2	1	-	1	-	-	-	-
Area Total		**5**	**2**	**2**	**1**	**-**	**-**	**-**	**-**	**3**	**2**	**1**	**-**	**3**	**1**	**-**	**2**	**-**	**-**	**-**	**-**
Other/Not Known	Not Known	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	1	1	-	-
Area Total		-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	1	1	-	-
TOTAL		**107**	**36**	**28**	**43**	**96**	**44**	**16**	**36**	**106**	**43**	**29**	**34**	**104**	**40**	**23**	**41**	**128**	**48**	**29**	**51**

* Each diagnosis has been defined using the Interruption Classification of Diseases as follows: Cleft palate (749.0), Cleft lip (749.1), and Cleft palate and cleft lip (749.2) Source: Congenital Anomalies Register

Ref: Information and Statistics Division (Scotland)
Common Services Agency
Edinburgh

Adhoc Number: u297 1059

Cases of cleft palate and cleft lip - Scotland*
By area of residence, year and diagnosis: 1993-95

Area of Residence		1993				1994				1995			
Health Board	Local Government District	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip
Argyll & Clyde	Dunbarton	1	-	-	1	-	-	-	-	-	-	-	-
	Inverclyde	2	1	-	1	1	-	-	1	2	1	-	1
	Renfrew	5	2	1	2	4	1	-	3	4	1	1	2
Area Total		8	3	1	4	5	1	-	4	6	2	1	3
Ayrshire & Arran	Cumnoch and Doon Valley	1	1	-	-	-	-	-	-	2	-	1	1
	Cunninghame	5	1	-	4	3	2	1	-	2	1	-	1
	Kilmarnock and Loudon	-	-	-	-	3	1	1	1	2	1	1	-
	Kyle and Carrick	1	-	-	1	2	1	-	1	2	1	-	1
Area Total		7	2	-	5	8	4	2	2	8	3	2	3
Borders	Berwickshire	1	-	-	1	-	-	-	-	-	-	-	-
	Tweedale	-	-	-	-	-	-	-	-	1	1	-	-
Area Total		1	-	-	1	-	-	-	-	1	1	-	-
Dumfries&Galloway	Annandale and Eskdale	-	-	-	-	1	-	1	-	2	-	-	2
	Stewarty	-	-	-	-	-	-	-	-	1	-	-	1
	Wigtown	2	1	-	1	2	-	1	1	-	-	-	-
Area Total		2	1	-	1	3	-	2	1	3	-	-	3
Fife	Dunfermliine	2	1	-	1	-	-	-	-	2	1	-	1
	Kirkcaldy	6	4	1	1	2	2	-	-	1	1	-	-
	North East Fife	2	-	1	1	3	1	-	2	4	2	1	1
Area Total		10	5	2	3	5	3	-	2	7	4	1	2
Forth Valley	Clackmannan	-	-	-	-	1	1	-	-	1	-	-	1
	Falkirk	4	3	1	-	1	-	-	1	5	2	1	2
	Stirling	2	1	1	-	2	1	-	1	1	-	-	1
Area Total		6	4	2	-	4	2	-	2	7	2	1	4
Grampian	City of Aberdeen	8	4	3	1	3	2	-	1	-	-	-	-
	Banff and Buchan	-	-	-	-	4	1	1	2	1	-	-	1
	Gordon	-	-	-	-	3	1	1	1	1	1	-	-
	Kincardine and Deeside	1	-	-	1	1	1	-	-	1	-	-	1
	Moray	4	2	-	2	3	2	-	1	1	-	-	1
Area Total		13	6	3	4	14	7	2	5	4	1	-	3
Glasgow	Bearsden and Milngavie	1	-	1	-	-	-	-	-	1	-	-	1
	Clydebank	1	1	-	-	-	-	-	-	2	1	-	1
	Eastwood	-	-	-	-	1	1	-	-	1	-	1	-
	City of Glasgow	17	10	1	6	21	8	6	7	11	4	3	4
	Strathkelvin	1	-	-	1	2	-	2	-	1	-	-	1
Area Total		**20**	**11**	**2**	**7**	**24**	**9**	**8**	**7**	**16**	**5**	**4**	**7**

APPENDIX 4
Page 4 of 4

Area of Residence		**1993**				**1994**				**1995**			
Health Board	Local Government District	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip	Total	Cleft Palate	Cleft Lip	Cleft Palate with Cleft Lip
Highland	Badenoch and Strathspey	-	-	-	-	1	1	-	-	1	1	-	-
	Caithness	1	1	-	-	-	-	-	-	2	1	-	1
	Inverness	-	-	-	-	4	3	-	1	-	-	-	-
	Lochaber	1	1	-	-	-	-	-	-	1	-	-	1
	Ross and Cromarty	1	1	-	-	1	-	-	1	1	1	-	-
Area Total		3	3	-	-	6	4	-	2	5	3	-	2
Lanarkshire	Cumbernauld and Kilsyth	1	-	1	-	1	1	-	-	1	-	1	-
	East Kilbride	-	-	-	-	2	1	-	1	-	-	-	-
	Hamilton	2	1	-	1	1	-	-	1	3	1	1	1
	Clydesdale	-	-	-	-	2	-	1	1	1	1	-	-
	Monklands	1	-	1	-	-	-	-	-	1	-	-	1
	Motherwell	1	1	-	-	2	1	-	1	6	-	1	5
Area Total		5	2	2	1	8	3	1	4	12	2	3	7
Lothian	East Lothian	2	1	1	-	6	2	-	4	2	2	-	-
	City of Edinburgh	6	1	2	3	7	5	1	1	10	8	1	1
	Midlothian	1	-	1	-	2	2	-	-	3	1	1	1
	West Lothian	3	2	1	-	4	2	-	2	6	2	1	3
Area Total		12	4	5	3	19	11	1	7	21	13	3	5
Tayside	Angus	2	2	-	-	3	1	1	1	4	3	-	1
	City of Dundee	2	-	1	1	3	2	-	1	3	1	1	1
	Perth & Kinross	2	1	1	-	4	3	1	-	3	2	1	-
Area Total		6	3	2	1	10	6	2	2	10	6	2	2
Islands	Shetland	2	-	2	-	1	1	-	-	1	1	-	-
	Western Isles	-	-	-	-	2	1	-	1	-	-	-	-
Area Total		2	-	2	-	3	2	-	1	1	1	-	-
Other/Not known		-	-	-	-	1	-	-	1	-	-	-	-
Area Total		-	-	-	-	1	-	-	1	-	-	-	-
TOTAL		95	44	21	30	110	52	18	40	101	43	17	41

Source: Scottish Morbidity Record (SMR)1 and SMR11.

* Each diagnosis has been defined using the International Classification of Disease as follows: Cleft palate (749.0), Cleft Lip (749.1), and Cleft palate and cleft lip (749.2).

Ref: Information and Statistics Division (Scotland)
Common Services Agency
Edinburgh

Adhoc Number: u297 1059

APPENDIX 5
Page 1 of 2

Recommendations for Minimum Standards for the Care of Children with Clefts of the Lip and/or Palate

ROYAL COLLEGE OF SURGEONS OF ENGLAND SURGICAL AUDIT UNIT

CLEFT LIP AND PALATE STEERING GROUP

MINIMUM STANDARDS FOR THE CARE OF CHILDREN WITH CLEFTS OR THE LIP AND/OR PALATE

Introduction

1. About 750 new patients with cleft of lip and/or palate are born annually in England and Wales.

2. Cleft of lip and/or palate can result in a wide variety of disabilities, including difficulties in breathing and feeding after birth, poor speech, dental anomalies, impaired facial growth and problems with psychological well-being and social relations.

3. A cleft may sometimes be one feature of a syndrome or genetically determined disorders affecting more than one system in the body.

4. In many patients, treatment and clinical surveillance needs to continue throughout childhood and adolescence.

5. The cleft itself, and its functional and developmental consequences require treatment by members of several medical specialities and allied professions.

The Royal College of Surgeons Steering Group on Cleft Lip and Palate therefore recommends the following as minimum standards:

1. **Comprehensive Service** - Children with clefts require care from a Team - including specialised counselling, specialised nursing, plastic surgery, maxillofacial surgery, orthodontics, dental care, speech and language therapy, otology/audiology, clinical genetics, developmental paediatrics. The Steering Group stresses the important of team work between these specialities.

2. **Co-ordination** of all treatment by the team from a single regional centre where registration of cases, record keeping, treatment planning and multi-disciplinary audit are undertaken. The regional centre should have a paediatric setting. The Team should hold regular instructional courses for non-team members e.g. health visitors, midwives, paediatricians, speech and language therapists.

3. **Neonatal Counselling** - An experienced professional under the direction of the Team should assess the child on the first day of life and provide patients with reassurance and information on the future programme of management. Similar antenatal counselling should be provided in the event of foetal diagnosis. An identified member of the Team should be available to give further information support and counselling as necessary.

4. **Neonatal Nursing** - A Nurse Specialist in the Team should give early instruction to the mother on feeding and nursing. Continuing care in the community by the Specialised Nurse should include the availability of advice by telephone and home visit. Separation of mother and child should be kept to an absolute minimum.

5. **Surgery** - Primary lip and palate repair and revision, pharyngoplasty, alveolar bone grafting, surgical exposure of teeth, and jaw osteotomies should be performed only by experienced surgeons with extended cleft lip and palate training and a frequent involvement in this work. The Steering Group recommends 30 new cases per operator per year as a minimum involvement for primary surgery. Further rationalisation will be desirable to ensure appropriate concentration of skills for secondary procedures such as alveolar bone grafting, surgical correction of VPI and maxillary osteotomy, Annual experience should not fall below 20 cases for alveolar bone grafting and for VPI surgery and maxillary osteotomy 15 cases, possibly including some non-cleft subjects, is the recommended minimum.

APPENDIX 5
Page 2 of 2

Surgical technique, timing and sequencing should conform to well-tried protocols unless new alternative procedures are introduced as part of an ethically approved surgical trial. Decisions to perform secondary operations should be reached in a multi-disciplinary forum. Bone grafting and osteotomies should not be performed before joint surgical/orthodontic discussion, nor pharyngoplasty before joint surgical/speech and language evaluation including nasendoscopy/videofluoroscopy. Surgical records should include photographs and detailed anatomical description of the pre-operated cleft, preferably augmented by dento-facial casts obtained at the time of primary surgery.

6. **Orthodontics** - Orthodontic treatment should be performed only by experienced Orthodontists with extended cleft lip and palate training and frequent involvement in this work. The Steering Group recommends 30 new cases per orthodontist per year as a minimum involvement. Treatment should be concentrated into a pre-bone graft phase in the mixed dentition (only if necessary), and permanent dentition arch alignment. Additional early treatment should be carried out only when essential for facilitating surgery. It is a primary duty of the orthodontist to feed back to the surgeon information on patients with surgically related growth disturbance so that inappropriate surgical practices may be remedied. Orthodontic audit records should not duplicate those obtained for clinical care.

7. **Dental Care -** Dental Health education should be commenced by a named member of the Team. The Cleft Team should ensure that dental health education, fluoride supplementation and dental attendance is maintained throughout childhood. Children with repaired clefts should have priority access to a Consultant in Paediatric Dentistry where necessary. Adolescents requiring advanced restorative care including crown and bridgework, dental implants and obturators should have priority access to a Consultant in Restorative Dentistry. If this Consultant is not a Team member, access should be available for treatment planning at an early enough stage to be able to influence outcome.

8. **Speech and Language Therapy** - A Specialist Speech and Language Therapist should carry out early counselling and diagnostic assessments, and provide necessary therapy directly or through liaison with a local therapist. The Standing Committee recommends 30 new cases per year as a minimal involvement for the team therapist. It is a primary duty of this individual to feed back to the surgeon information on patients with surgically related speech impairment so that inappropriate surgical practices may be remedies, and to report outcome following speech related surgical or prosthetic procedures.

9. **Otology/Audioloy** - The Cleft Team should include an Otologist responsible for the co-ordination of regular audiometric evaluation. Wherever possible placement of grommets and other procedures should be co-ordinated with other surgical episodes to minimise the need for multiple anaesthetics.

10. **Clinical Genetics -** The Team should include a Geneticist/Dysmorphologist with access to a Genetics Laboratory. Patients with any possibility of syndromic association must have timely appraisal. All parents and patients should be offered the opportunity of access to genetic counselling.

11. **Psychological Counselling -** The Team should include a Psychologist or a professional in the Team suitably treated in counselling skills. All patients and parents should be assessed at times of transition and have access to counselling where indicated.

12. **Paediatric Developmental Medicine -** For each patient, the Team should provide information to the Consultant Community Paediatrician of the child's district of residence. Any suspicion of developmental or growth delay should be notified early to allow appropriate investigations to be undertaken.

13. **Audit and Research** - The Cleft Team should subject itself to regular consumer audit, wear name badges, provide families with written report of multi-disciplinary assessment, and take other measures to adhere to the "Patients' Charter". The Team should participate in multi-centre audits and in national and international research.

14. **Patent Support Group -** The Team should maintain good relations with the local Parent Support Group and put parents in touch with this organisation at an early opportunity.

APPENDIX 6
Page 1 of 8

ECONOMICS OF CHANGE: AN ILLUSTRATIVE EXAMPLE

This appendix expands the principles stated in section 8.4 and examines the cost of access borne by patients and their families, and the value of averted costs due to a move towards two surgical centres. It then combines both these costs to produce a net cost of such a move.

1. ASSESSMENT OF ACCESS COSTS

1.1 First of all, a note of caution: the analysis below requires some quite restrictive assumptions, especially with respect to patterns of referral, so the analysis should be treated merely as an exploration of access costs.

1.2 Essentially this appendix examines the cost of rail or air fares, subsistence and child care of patients and their families in attending for surgery. Visiting families are assumed to consist of two adult visitors and 0.8 children. As outlined in the main report, there are five principal costs to patients and their families of cleft lip/palate surgery: transport, subsistence, childcare, loss of earnings and the cost of lost education to the child. Due to difficulties in measuring the last two costs, we shall examine the first three only.

1.3 Two scenarios are examined - the current provision of services and reconfiguration centred on Glasgow and Edinburgh. It is emphasised that Glasgow and Edinburgh are chosen for illustrative purposes only - as described in the main report, there are many options for the reconfiguration of cleft lip/palate services in Scotland.

Regional Distribution of Referrals

1.4 The first step in the analysis is to determine the regional referral patterns of cleft lip/palate cases. Referral patterns are assumed by assigning patients from each Health Board area to each site of treatment, based on geographical proximity. The restrictiveness of the assumption is acknowledged. The assumed referral patterns are as follows:

Table 1: Assumed Referral Patterns from Each Health Board to Each Treatment Centre at Present

Site	Referring Health Board (assumed)	Average Annual Number of New Cases
Glasgow	Argyll and Clyde Greater Glasgow Health Board Dumfries and Galloway Lanarkshire	8.125 20.25 2.5 10
Total		*40.875*
Edinburgh	Borders Fife Lothian Highland	1.25 8.125 15.875 4.375
Total		*29.625*
Aberdeen	Grampian Islands	11.75 2.125
Total		*13.875*
Dundee	Tayside	*7.625*
Ayr	Ayrshire & Arran	*7.25*
Falkirk	Forth Valley	*5.875*

Source: ISD, Edinburgh.

Note: Highland and Fife patients are operated on in Edinburgh while out patient consultations are offered on an outreach basis.

1.5 By moving the service to Glasgow and Edinburgh, we assume that Glasgow would get referrals from Ayrshire and Arran and the three island Boards. Meanwhile Tayside, Forth Valley and Grampian patients would go to Edinburgh.

1.6 We assume that each patient receives 5 operations over their lifetime, and undergo 1 routine visit to the cleft team per annum up to the age of 18 years. Therefore, given an annual average number of new cases of 105, in any one year there should be 525 operations conducted and 1,890 outpatient visits. The average length of stay for an operation is assumed to be 4 days.

Travel Costs

1.7 We assume that the cost of rail travel from each local authority area is a reasonable proxy for travel costs from that area. In the case of the three island Boards, we use air travel. We assume a zero cost of intra-area travel, for the sake of simplicity. Thus, the cost of travel from all points within Kyle and Carrick to Glasgow is the price of a rail ticket from Ayr to Glasgow.

1.8 The number of journeys will vary between parents who have to stay over at the treatment centre and those parents who can commute. Therefore we assume that for parents who have to stay over, there is a total of 3.5 return journeys (one adult staying over, and therefore making 1 return journey, one adult making 2 return journeys and one child return). For parents who commute, we assume a total of 6.5 rail journeys. For annual visits, we assume that only one adult travels with the child, producing 1.5 return journeys per annum.

1.9 Thus, families who commute to the cleft team make an estimated 59.5 return journeys over the lifetime of the patient. Families where one adult stays at the treatment centre make an estimated 44.5 return journeys over the lifetime of the patient.

1.10 On the basis of the above assumptions and data on rail and air fares, the following results are produced:

Table 2: Estimated Cost of Rail Travel for Cleft Lip and Palate Treatment by Region

Area	Cost Now £	2 Centres £	Difference £
Argyll and Clyde	3,012	3,012	0
Ayrshire and Arran	7,105	8,989	1,885
Borders	924	924	0
Dumfries and Galloway	2,194	2,194	0
Fife	1,464	2,378	914
Forth Valley	904	2,803	1,900
Grampian	2,021	19,721	17,700
Greater Glasgow	0	0	0
Highland	3,353	3,353	0
Lanarkshire	2,172	2,172	0
Lothian	0	0	0
Tayside	1,926	6,103	4,177
Islands	9,401	14,418	5,017
Total	**34,473**	**66,066**	**31,593**

Source: National Rail Enquiries; ISD.

1.11 Therefore, under the status quo, the cost of travel to treatment sites for patients is £34,473. Those health boards that incur the majority of costs are the three island Boards, Highland and Ayrshire and Arran. By centralising to two treatment sites, the cost of travel almost doubles to £66,066. Those health boards that incur the majority of the cost increase are Grampian, the three island Boards and Tayside.

1.12 Patients sometimes travel for surgery using hospital transport. Therefore costs to patients may be overestimated to the extent of patients who use hospital transport. The overestimate is quite slight however, as the majority of transport costs are incurred by family visits, rather than by patients. Moreover, moving to two sites will increase transport costs - whether cost to patients or the health service.

Cost of Subsistence

1.13 As described above, we assume that parents of patients from certain areas stay at the treatment centres during their children's operations. We now make the further assumption that for annual visits, parents from the three island Boards are required to stay overnight.

1.14 The cost of accommodation and subsistence is proxied using the Whitley Guidelines (the guidelines for subsistence levels in the public sector). Thus, bed and breakfast is assumed to cost £55, evening meal £15 and lunch £5. Total cost of daily subsistence is £75.

1.15 For those who are required to stay at the treatment centre during the child's surgery, we assume 4 days subsistence per operation, by 5 operations by £75 per day is a total cost of £1,500 per patient. For parents of patients from the three island Boards the additional cost of subsistence during annual visits produces a total cost per patient of £3,750.

1.16 The regional distribution of estimated subsistence costs is illustrated in the table below:

Table 3: Estimated Cost of Subsistence for Cleft Lip and Palate Treatment by Region

Area	Cost Now £	2 Centres £	Difference £
Argyll and Clyde	2,250	2,250	0
Ayrshire and Arran	0	0	0
Borders	0	0	0
Dumfries and Galloway	3,750	3,750	0
Fife	0	0	0
Forth Valley	0	0	0
Grampian	0	17,625	17,625
Greater Glasgow	0	0	0
Highland	6,563	6,563	0
Lanarkshire	0	0	0
Lothian	0	0	0
Tayside	0	11,438	11,438
Islands	7,969	7,969	0
Total	**20,531**	**49,594**	**29,063**

Source: Whitley Guidelines; ISD.

1.17 Under the status quo, the cost of subsistence is £20,531. This increases to £49,594 under centralisation. The increase in costs is due to parents of patients from Grampian and Tayside having to stay over during the treatment of their children.

Child Care

1.18 The final element of patient costs that we wish to estimate is the costs of caring for the patient's siblings while parents are visiting the patient in hospital. The following assumptions are required:

- We assume that parents put the patient's siblings into childcare rather than bring them to hospital with them. As a result of the assumptions invoked, there is no difference in childcare costs for families where overnight stays are required and families who commute to hospital. Where parents arrange childcare informally, we assume that the market price for childcare represents the opportunity cost of caring to the informal carer;
- We assume that each family consists of 1.8 children, so 0.8 children require childcare;
- For the first 3 operations, we assume that 0.8 children are pre-school age and require a full day's care (since 2 operations occur in the first 2 years of life, it is reasonable to assume that the majority of the patient's siblings will be of pre-school age for these operations);
- For the fourth operation, we assume that the 0.8 siblings require 3 hours childcare each, while for the fifth operation no childcare is required;
- In the case of annual visits, we assume that, for the first 5 years, the 0.8 children require a full day's care. For the next 5 years, 3 hours per day for 0.8 children are required. We assume that no childcare is needed after age 10.

1.19 The cost of a full day's child care for pre-school children is £16.50, according to local estimates. The cost of 3 hours care for school age children is £6.60. Four days care is required for each operation leading to a total cost of approximately £180 during operations. Cost of annual visits is approximately £92 in all cases except for three island Boards where it is £184, as an overnight stay is required.

1.20 Thus, total lifetime childcare costs are approximately £272 per patient, except for the three island Boards' patients where they are approximately £364. Note that for single parent families, childcare costs are likely to be greater than estimated here. However, we are unable to estimate these additional costs without data on the number of cleft patients from single parent families, or their regional distribution. There is no difference in childcare costs between the status quo and a centralised service, since child care is required for both commuters and families who stay overnight. The regional distribution of childcare costs is outlined below:

Table 4: Estimated Cost of Childcare for Cleft Lip and Palate Treatment by Region

Area	Cost £
Argyll and Clyde	2,209
Ayrshire and Arran	1,971
Borders	340
Dumfries and Galloway	680
Fife	2,209
Forth Valley	1,598
Grampian	3,195
Greater Glasgow	5,506
Highland	1,190
Lanarkshire	2,719
Lothian	4,317
Tayside	2,073
Islands	774
Total	**28,782**

Source: Various Child Care Agencies; ISD.

Total Costs

1.20 Thus adding Tables 2, 3 and 4 produces the following:

Table 5: Total Costs of Access for Cleft Lip/Palate Patients by Region

Area	Cost Now £	2 centres £	Difference £
Argyll and Clyde	7,471	7,471	0
Ayrshire and Arran	9,076	10,961	1,885
Borders	1,264	1,264	0
Dumfries and Galloway	6,621	6,621	0
Fife	3,673	4,588	914
Forth Valley	2,501	4,401	1,900
Grampian	5,216	40,541	35,325
Greater Glasgow	5,506	5,506	0
Highland	11,105	11,105	0
Lanarkshire	4,892	4,892	0
Lothian	4,317	4,317	0
Tayside	3,999	19,614	15,615
Islands	18,144	23,161	5,017
Total	**83,786**	**144,441**	**60,655**

1.21 The cost of the status quo is £87,786. This rises to £144,441 under centralisation, a rise of £60,655. The majority of cost increases are incurred by patients in Grampian, Tayside and the three island Boards.

Sensitivity Analysis

1.22 Clearly the above results partly depend upon the assumptions on which they are based. Sensitivity analysis tests the robustness of the results by altering key variables. The effects of alterations are illustrated in summary in Table 6. Figures are rounded to the nearest thousand.

Table 6: Sensitivity Analysis of Access Costs Results (£000's)

Variable	Cost Now	2 centres	Difference
16 Routine Visits	81	140	59
18 Routine Visits	***84***	144	***61***
23 Routine Visits	92	157	66
Length of Stay : 3.5	79	163	57
Length of Stay : 4	***84***	***144***	***61***
Length of Stay : 4.5	89	153	64
4 Operations	77	129	52
5 Operations	***84***	***144***	***61***
6 Operations	93	162	69

1.23 The figures in bold and italics represent our best 'guesstimate' of access costs. The figures above and below represent the range inside which it is reasonable to assume that the actual costs of access exist.

1.24 Table 7 combines the results in Table 6 to produce the best and worst case scenarios.

Table 7: Best, expected and worst case scenarios (£000's)

Scenario	Cost Now	2 centres	Difference
Best Case	70	117	47
Expected Case	***84***	***144***	***61***
Worst Case	104	183	79

1.25 Access costs under the status quo are between £70,000 and £104,000, with an expected cost of £84,000. Under the centralised option, access costs are between £117,000 and £183,000, with an expected cost of £144,000. Therefore, it is estimated that a move to two centres will lead to an increase in access costs of between £47,000 and £79,000, with an expected increase of £61,000.

Conclusions

- The cost of access to treatment centres that is incurred by the patient's family currently amount to approximately £84,000. This may vary from £70,000 to £104,000, depending on the assumptions invoked;
- Under a move to two centres, family costs would rise to approximately £144,000 for Scotland. This may vary from £117,000 to £183,000;
- The brunt of these cost increases would be incurred by families in Grampian, Tayside and the three island Boards. The increase in costs in these 3 areas would be £35,325, £15,615 and £5017 respectively.

2. VALUE OF AVERTED COSTS THROUGH IMPROVED OUTCOMES

2.1 Suppose reconfiguration produces a reduction in the number of cleft lip/palate patients requiring osteotomy in their late teens from 48% to 18%. One operation and two routine visits are averted.

NHS Costs Averted

2.2 An osteotomy costs approximately £1,150, according to figures produced by Finance Departments of various hospitals where such procedures are undertaken. Since costs averted due to centralisation will occur approximately 17 years after centralisation, we need to discount averted costs to present value terms. Using a 6% discount rate, the value of averted costs is £427 per osteotomy. Given an annual birth prevalence of 105 cleft lip / palate patients per annum, a reduction in the number requiring osteotomy of 30% affects 31.5 patients. Therefore, the value of costs averted to the NHS is of the order of £13,450.

Patient Costs

2.3 The value of rail costs averted is £1,129, while the value of subsistence costs averted is £1,069. Since patients are in their late teens, it is assumed that no childcare costs are incurred.

3. NET COSTS OF MOVE TOWARDS 2 CENTRES

3.1 A move towards two centres would lead to an increase in access costs, coupled with potential to avert costs through fewer osteotomy procedures in late teenage years. The net cost of such a move is as follows:

Cost	**£**
Increase in Access Costs	60,655
Present Value of reduction in NHS costs	(13,450)
Present Value of reduction in Access Costs	(2,198)
Total	45,007

APPENDIX 7
Page 1 of 5

RECOMMENDED MINIMUM DATASET

CLEFT SURGERY

Basic Minimum (Summary)	Plastics
	Photographs
Birth	(√)
Pre-lip	√
Pre-palate	√
5 years	√
10 years	√
15 years	√
20 years	√

The vast majority of plastic surgeons and orthodontists use still photographs for documentation of clefts. Very few centres use video recording of clefts pre or post-operatively. There is no uniformity or standardisation of the way in which such photographs are taken.

For comparative studies to be done using photogrammetry, it is necessary to standardise the following:

1. The basic view of lip/nose to be taken in each patient
2. The dynamic view required e.g. smiling, whistling
3. The lighting and background colour
4. The framing of the view
5. The focal length of the lens and type of camera to be used

Basic views to be taken

AP, left and right, inferior (columellar) view.
3/4 facial (oblique) view.

Dynamic views

During smiling and whistling
In the co-operative older patient, these views will give an idea of function of the circumoral musculature. Video recording will be better for assessing circumoral movement but this will also need to be standardised and cannot be used routinely at the moment.

Lighting and Background

Lighting for the studio should be 2 fill-in lights and the main light synchronised with camera. The background should be pale green or pale blue.

Framing of the Picture

The international recommendations are as follows:

Full AP view, the camera should be set at a ratio of 1:8
Full lateral view, the camera should be set at a ratio of 1:8
Full inferior view, the camera should be set at a ratio of 1:4

Some of these frames may not be sufficient to show facial detail. Standardisation for computer analysis will be facilitated if the views are taken in portrait to show full height (forehead to chin with hair held off the forehead and behind the ears). In the absence of a cephlostat, horizontal may be judged by placing the top of the ear level with the eyebrow and the bottom of the ear level with the base of the nose in the A-P and with Frankfort plane horizontal in the side view. The base view should be taken at the same magnification as the A-P and with the skyline of the domes half way between the plane of the canthi and the eyebrows.

Camera and Lens

It is suggested standardisation of the camera body is not essential but 105mm lens should be used. Film type and speed need not be standardised.

Recommended Time-frame for Photographic Records

1. At birth/before lip repair
2. Before palate repair
3. Subsequently at 5 yrs, 10 yrs, 15 yrs and 20 yrs

The above schedule does not preclude photographs being taken at other times when the patient is admitted for surgery

Photographs taken during the standard times allow comparison between units of age matched patients.

ORTHODONTICS

Basic Minimum (Summary)

	Orthodontics		
	Models	**Lateral Skull Radiograph**	**Photos**
Birth	(√)		√
Pre-lip	√		√
Pre-palate	√		√
5 years	√	(√)	√
2^{o} Bone graft *			
10 years	√	√	√
15 years	√	√	√
Pre-osteotomy*			
20 years	√	√	√

This summary is self explanatory. The models at birth have been placed in brackets. Clearly if the unit uses pre-surgical orthopaedic appliances, than there is sense in collecting models at birth. If units do not use pre-surgical orthopaedic appliances then some consultants have expressed concerns over taking impressions of babies at birth. For this reason and because perhaps the amount of information derived from these models is limited, some units may choose not to produce these records.

Similarly the lateral skull radiograph taken at the age of 5 may have limited information for audit or research projects. Placing a 5 year old in a cephalostat is difficult with reference to reproducible head posture. The recent ALARA recommendations would also seem to suggest that at this age it may not be appropriate to expose a patient to radiation for a minimal amount of diagnostic information.

Recent guidelines for orthodontic radiography would suggest that at least 20% of radiographic examinations carried out in the United Kingdom are clinically unhelpful. If is important with the longitudinal care needed for clefts that they are not exposed to any more radiographs than absolutely necessary. For this reason the pan-oral view is not seen as a basic minimum necessary for cleft care outcome.

* The ages recommended do not of course prevent further records being taken such as when definitive orthodontic treatment is undertaken prior to surgery, secondary alveolar bone grafting or normal orthodontic treatment. These records should not be duplicated if they are within one year of the 5,10, 15 and 20 year old records. It is recommended that records for secondary alveolar bone grafting should comprise the following:

Pre grafting and one year later following full eruption and alignment of adjacent teeth;

a) Study models
b) Photographs
c) Intra-oral radiographs of graft site.

SPEECH AND LANGUAGE THERAPY

Basic Minimum (Summary)

	Speech & Language Therapists	
	Speech Recording	**Video Recording**
3 years	√	√
velpharyngeal surgery*		
5 years	√	√
10 years	√	√
15 years	√	√
osteotomy*		
20 years	√	√

It is considered that the timing of data collection for speech and language therapists should be at 3, 5, 10, 15 and 20 years.

At each age an audio speech recording using a high quality tape recorder, but ideally a video with an extended microphone played back through an amplifier and speakers.

The data sample should be standardised:

For 3 year olds

1. Single word data sampling PACS toys test
2. Nursery Rhyme

For 5, 10,15 and 20 year olds

1. Sentence repetition with the GOSSPAS list (Sell et al, 1994)

2. Spontaneous conversational speech

3. Counting from 1-20, 60-70
   The parameters to be assessed are:
   Nasal grimace
   Nasal emission
   Nasal turbulence
   Hypernasality
   Hyponasality
   Consonant sound system

   At velopharyngeal surgery:

   Speech records pre-operatively and at 3 months and 12 months post-operatively

   At surgical osteotomies:

   Speech recordings pre-operatively and at 12 months post-operatively

   * The ages recommended do not prevent further records being taken at any stage. These points of treatment (prior to velopharyngeal surgery or osteotomy) may also coincide with recommended ages. Records need not be taken if they are within one year of any of the recommended ages.

ENT SURGERY

Basic Minimum (Summary)

	Ear Nose & Throat Surgeons		
	Otoscopy	**Tympanometry**	**Audiometry**
Pre-lip	√	√	
Pre-palate	√	√	(√)
3 years	√	√	√
5 years	√	√	√
10 years	√	√	√
15 years	√	√	√
20 years	√	√	√

Audiometry testing from the age of 3 should comprise pure tone audiometry.

Pre-palate: distraction testing where available and if appropriate may be carried out.

Pure tone audiometry; air and bone conduction should be performed from 5 years of age.

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