CRANE Database

www.crane-database.org.uk

Annual Report on cleft lip and/or palate 2013

On behalf of the Cleft Development Group

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Abbreviations

BCLP	Bilateral cleft lip and palate
BINOCAR	British Isles Network of Congenital Anomaly Registers
CAPS-A	Cleft Audit Protocol for Speech—Augmented
CARE	Craniofacial Anomalies Register
CDG	Cleft Development Group
CFSGBI	Craniofacial Society of Great Britain and Ireland
CI	Confidence interval
CL	Cleft lip only
CLEFTSiS	The National Management Clinical Network for Cleft Service in Scotland
CP	Cleft palate only
CSAG	Clinical Standards Advisory Group
CRG	Clinical Reference Group
dmft	Decayed, missing and filled teeth
DoH	Department of Health
ENT	Ear, nose and throat
GOSH	Great Ormond Street Hospital
HES	Hospital Episode Statistics
ICD-10	International Classification of Disease 10th Revision
MCN	Managed Clinical Network
OPCS-4	Classification of Surgical Operations and Procedures 4th Revision
PEDW	Patient Episode Data Wales
RCPCH	Royal College of Paediatrics and Child Health
SCG	Specialised Commissioning Group
SD	Standard deviation
SE	Standard error
SIG	Special Interest Group
UCLP	Unilateral cleft lip and plate
VTCT	Vocational Training Charitable Trust
WHO	World Health Organization

Glossary

Alveolus / alveolar	The part of the jaw that supports the teeth and contains the tooth sockets.
Administrative Unit	A hospital that provides cleft surgery and submits data to the CRANE Database, sometimes as part of a wider cleft centre or network.
Cleft	A failure of tissues to join during development.
Cleft Development Group (CDG)	NHS National group representing all stakeholders in cleft care that is responsible for the CRANE Database as well as oversight and guidance on all aspects of the delivery of reorganised cleft care.
Cleft surgeon	A surgeon undertaking cleft repair surgery in an Administrative Unit
Clinical Standards Advisory Group (CSAG)	A group established in 1991 to act as an independent source of expert advice on standards of clinical care for, and access to and availability of services to, NHS patients.
Craniofacial anomalies	A diverse group of deformities in the growth of the head and facial bones.
Craniofacial Society of Great Britain and Ireland (CFSGBI)	An inter-specialty group set up to study cleft lip and palate and other craniofacial anomalies. www.cfsgb.org.uk
Hospital Episode Statistics (HES)	A national database containing records on all admissions to NHS hospitals in England.
LAHSAL	A code used to classify clefts. Each letter (LAHSAL) relates to one of the six parts of the mouth that can be affected by a cleft.
Managed Clinical Network (MCN)	A formally organised network of clinicians.
National Information Governance Board (NIGB)	An independent statutory body established to promote, improve and monitor information governance in health and adult social care. http://www.nigb.nhs.uk
Patient Episode Data Wales (PEDW)	A national database containing records on all admissions to hospitals in Wales.
Submucous Cleft Palate	The term *submucous* refers to the fact that the cleft is covered over by the lining (mucous membrane) of the roof of the mouth. This covering of mucosa makes the cleft difficult to see when looking in the mouth.

Executive summary

Craniofacial abnormalities are among the most common of all birth defects [1]. Cleft lip and/or palate can affect a variety of functions, including speech and hearing. Appearance and psychosocial health may also be compromised in those with a cleft. Typically, children with a cleft need multidisciplinary care from birth to adulthood, and they have higher morbidity and mortality throughout life compared with unaffected individuals [2].

The CRANE Database is a national register that collects information on children born with a cleft lip and/or palate in England, Wales and Northern Ireland. The database was established in 2000 and transferred to the Clinical Effectiveness Unit of the Royal College of Surgeons in 2005. CRANE has two broad aims:

- to register birth and demographic data related to all children born in England, Wales and Northern Ireland with the congenital abnormality of cleft lip and/or palate;
- to record the treatment of children and adults with a cleft lip and/or palate and the outcome of such treatment.

Data are submitted to CRANE by the 15 hospitals (otherwise known as Administrative Units) providing surgical treatment to cleft patients in England, Wales and Northern Ireland. This Annual Report describes the results of on-going analyses of the CRANE Database, examining trends in registrations and the timing of cleft diagnosis, patient referral to Administrative Units and first contact between Administrative Units and the parents of children born with a cleft.

For the third year running, we present information on cleft-related outcomes for children at five years of age (born 2004-2007). The completeness of these data, which is essential for CRANE to perform meaningful analyses, is presented according to Administrative Unit.

This report also describes the analyses of data from Hospital Episode Statistics (HES), a database containing records on all NHS hospital admissions in England. These data are used to derive information on children diagnosed with cleft lip and/or palate. This year, we have examined neonatal (<28 days of age) and infant (<1 year of age) mortality among children with a cleft in England. We present the results according to the absence or presence of additional anomalies or syndromes and by cleft type classification.

Key findings

Children born with a cleft lip and/or palate in 2012

Overall, 12,863 children born between 1 January 2000 and 31 December 2012 with a cleft lip and/or palate were registered on the CRANE Database by 18 September 2013. Of these, 1,127 were born in 2012. This represents the highest number of registrations CRANE has received since 2000. This high number should reflect all children born with a cleft lip and/or palate, referred to one of the 15 Administrative Units in England, Wales and Northern Ireland, regardless of their consent status. The increase in CRANE registrations represents the improved function of the database as a national register of cleft births. CRANE case ascertainment is very high, being around 95%, according to comparisons with HES and Patient Episode Data Wales (PEDW) [3]. Out of the children born in 2012 whose consent status had been verified, the parental consent rate was 98.5% (ranging from 91.1% to 100% between Units). Out of all children registered, 20.1% (ranging from 0% to 49.0% between Units) had not had their consent status verified by the Administrative Unit at the time of writing this report.

Among children born in 2012, CRANE analyses revealed:

- 39% of all children with clefts and 69% of those with a cleft affecting the lip were diagnosed in the antenatal period through screening. The NHS Fetal Anomaly Screening Programme has a target of 75% for the antenatal diagnosis of clefts affecting the lip.
- Only 1% of children with cleft palate only (CP) were diagnosed during antenatal screening; 67% were diagnosed at birth, leaving 32% who were diagnosed late according to the national standard [4]. The proportion of children diagnosed late has increased by 4% compared with last year. Seven per cent of children with a cleft palate alone are diagnosed after one month of age. This proportion has increased by 2% since last year.
- 53% of children were referred by a maternity unit to an Administrative Unit within 24 hours of birth. This is similar to last year's rate of 54%.
- Referrals from maternity units within one day of birth varied from 31% to 77% according to the Administrative Unit receiving the referral. Some regions have seen substantial changes in this area over the last year, with improvements occurring in eight of the regions.
- Administrative Units established contact with 90% of parents within 24 hours of their child's referral. This has not changed over the last two years.

Cleft-related clinical outcomes at five years of age

CRANE collected clinical outcomes at five years of age among children born between 2004 and 2007. These outcomes include height and weight, the number of decayed, missing and filled teeth (a measure of oral health), Five Year Old Index scores (2004-2006 births only), which reflect dental arch relationships and the effects of primary cleft repair surgery on the facial growth of children with a complete unilateral cleft lip and palate (UCLP), and a speech assessment recorded using the Cleft Audit Protocol for Speech – Augmented (CAPS-A) scoring system (2006 births only). Although there is still a high proportion of missing data, some Units have reported outcomes for more than 75% of their eligible patients, suggesting that the reporting of these outcome data is feasible. For those children with reported outcomes:

- 43% had at least one decayed, missing or filled tooth (>0 dmft), which, although still significant, is only slightly higher than the background rate in the general population. The proportion of children with >0 dmft varied significantly according to cleft type. Bilateral cleft lip and palate (BCLP) was associated with the poorest oral health, with 50% of BCLP patients having >0 dmft at five years of age.
- Of the 239 children with a complete UCLP who had externally validated Five Year Old Index scores, 26% had scores of '4' or '5', reflecting poor dental arch relationships. This represents a significant improvement compared to the CSAG findings that 36% of (223) cleft children had poor dental arch relationships at five years of age in 1996.
- Of the 315 children born with a cleft affecting their palate, for whom CAPS-A scores were provided, 49% had speech scores that would suggest their speech was not significantly different from their non-cleft peer group. 23% of children received at least one score indicating a possible structural problem with the palate that may require further surgery.

Unfortunately, outcomes are still not collected consistently across Units:

- Height and weight measures are not collected routinely by five of the fifteen Administrative Units.
- The dmft score was not reported by Cambridge as they have not had a paediatric dentist who would examine children to determine the dmft. However, the reporting of this outcome should improve in future years as this post has now been appointed in Cambridge. A couple of Units reported very few data to CRANE, despite collecting dmft. Inadequate administrative support has been cited previously as the reason for the lack of data submitted.

- The Five Year Old Index score was not reported by three Administrative Units. Of these, Belfast reported that their Orthodontist does not routinely see patients at five years of age.
- Speech data were not provided by three Units, despite collecting the data.
- CRANE will explore methods for improving communication and links with cleft teams to facilitate the submission of data to the database.

Infant mortality

We analysed Hospital Episode Data (HES) linked to the Office for National Statistics (ONS) mortality dataset to examine national neonatal (<28 days of life) and infant (<1 year of life) mortality rates among children born between 2001 and 2010 in England. A total of 12,589 children with a cleft were identified. Of these, 388 died within the first year of life. The main findings are outlined below:

- Children with a cleft have an infant (<1 year of life) mortality rate (IMR) of 30.8 per 1,000 live births. This is 6.3 times higher than the background rate of 4.9 per 1,000 live births occurring across the same time period.
- Children with a cleft alone (and no additional anomalies) have an IMR of 7.0 per 1,000 live births, which is 1.4 times higher than the background rate. The increased risk of death was restricted to the early neonatal (<7 days of life) period only and may be partly explained by the presence of additional anomalies that were not diagnosed or reported.
- Children with a cleft and additional anomalies or syndromes have an IMR of 95.4 per 1,000 live births – a rate that is 19.5 times higher than the IMR for the general population. The increased risk of death was most pronounced in the post-neonatal (28-364 days of life) period.
- Among children without additional anomalies, those with a cleft lip only (CL) had the lowest mortality rates, while those with a CP had the highest rates. Conversely, among those with additional anomalies or syndromes, children with a CP had the lowest mortality rates.

Recommendations

Clinical care

- Late diagnosis of cleft palate (CP) remains an important issue that must be addressed. Among children born in 2012, 32% of those with CP were diagnosed late according to the national standard [4]. This represents a 4% increase in the late diagnoses rate compared with the previous year. Since the publication of our findings in last year's annual report, which highlighted the problem of late diagnosis, the Royal College of Paediatrics and Child Health (RCPCH) has set up a working group to develop a best practice guide and an e-learning module on the palate examination in the neonate. The impact of the RCPCH training tool should be assessed.

- Preventative dental support for children with a cleft seems essential to reduce dental decay, particularly among those with UCLP and BCLP who appear to be at the greatest risk of caries. However, further analyses on more complete data are recommended.

Outcome measures and reporting to CRANE

- Units must improve data submission to CRANE. This is related to the National Service Specification which now contractually obliges Units to submit data. The submission of data for all eligible patients is required so that CRANE can report data to the Quality Dashboard.

- CRANE's collection of CAPS-A data should be modified so that data can be analysed and reported more clearly in future.

- Further outcome measures need to be developed to reflect a wider age range of patients and a broader range of cleft-related outcomes, including hearing, psychology, and patient and/or parent satisfaction. The CFSGBI have tasked the Psychology Special Interest Group (SIG) with reviewing potential tools or measures that could be adapted or developed for the cleft population. Further outcome measures that are planned to be collected should reflect the CFSGBI minimum dataset and the Quality Dashboard requirements.

- Following the Quality Dashboard pilot, further outcome development work should be considered with commissioner involvement. This should aim to capture data that can be used to inform the commissioning process for cleft-related services.

1. Introduction

Craniofacial abnormalities are among the most common of all birth defects [1]. Cleft lip and/or palate can affect a variety of functions, including speech and hearing. Appearance and psychosocial health may also be compromised in those with a cleft. Typically, children with a cleft need multidisciplinary care from birth to adulthood, and they have higher morbidity and mortality throughout life compared with unaffected individuals [2].

The CRANE Database is a national register that was established in 2000 to collect information on children born with a cleft lip and/or palate in England, Wales and Northern Ireland. The Database collects birth, demographic and cleft diagnosis information. It also collects information about cleft-related treatment and outcomes. Hospital Episode Statistics (HES) is used to further examine treatment for cleft lip and/or palate in England. The HES database contains records on all NHS hospital admissions in England. It holds diagnostic and procedure information on each patient, allowing us to identify those with a cleft lip and/or palate and those undergoing cleft-related treatment.

This Annual Report includes trends in CRANE registrations over the last 10 years, comparing the 15 Administrative Units and the four different types of cleft. Using CRANE data, we also report the proportion of babies born in 2012 who were diagnosed at birth, referred within 24 hours of birth, and contacted within 24 hours of referral. For the third year running, cleft-related outcomes at five years of age are presented. These outcomes include height and weight, number of decayed, missing or filled teeth, and Five Year Old Index scores. For the first time, we report speech-related outcomes at five years of age in the form of Cleft Audit Protocol for Speech—Augmented (CAPS-A) scores.

This year, we have analysed Hospital Episode Statistics (HES) data linked to the Office for National Statistics (ONS) mortality dataset to assess national neonatal (<28 days of life) and infant (<1 year of life) mortality among children with a cleft in England. These data span a 10 year period and we present the results according to the absence or presence of additional anomalies or syndromes and by cleft type classification.

1.1. Background to the CRANE Database

The CRANE Database was established in 2000 in response to the report of the Clinical Standards Advisory Group (CSAG) on cleft care in the UK in 1998 [5]. The report suggested that the outcome of cleft care in the UK was inferior to other countries in Western Europe. The CRANE Database can be considered a continuation of the Craniofacial Anomalies Register (CARE) that since 1990 was maintained by the Craniofacial Society of Great Britain and Ireland (CFSGBI).

The CSAG report recommended that cleft care should be centralised into regional cleft teams that would treat larger numbers of patients. The rationale for this recommendation was that it would increase the experience of the cleft teams and facilitate genuine multi-disciplinary care. At the same time, it would also enable meaningful and statistically significant audit. The Health Services Circular 1998/238, which set out arrangements for commissioning cleft services according to the CSAG report, stated that 'a craniofacial anomalies register, with which all patients should be registered [should] form the basis of national audit' [6]. A high-quality national database could furthermore contribute to comparisons between countries.

Currently, the CRANE Database collects information about children born with a cleft lip and/or palate in England, Wales and Northern Ireland. Scotland maintains a separate database which is part of CLEFTSiS, the National Management Clinical Network for Cleft Service in Scotland.

The Cleft Development Group (CDG) is responsible for making arrangements for the running and commissioning of the CRANE Database (see Appendix 3 for CDG's membership). The funding for CRANE was provided by the Specialist Commissioners based on repeated two-year contracts. The CRANE team has responded to a number of requests for information from a commissioner led comprehensive review of all databases relating to specialised services; the outcome of this has been an agreement to continue to fund CRANE in 2013/14.

1.2. Geographical representation of the cleft Administrative Units

The CRANE Database covers England, Wales and Northern Ireland. Cleft care is currently delivered by eight Regional Cleft Centres and two Managed Clinical Networks. Each of these 10 geographical hubs, with the exception of Northern Ireland, treats at least 80 new children born with a cleft lip and /or palate each year. Several of the Regional Cleft Centres are split

between two hospitals, where the primary surgery is usually undertaken. There are 15 Administrative Units (hospitals) who submit data to the CRANE Database (*Table 1*).

Table 1. Regional Cleft Centres and Managed Clinical Network and their associated Administrative Units

Regional centre / MCN	Administrative Unit
Northern & Yorkshire	Royal Victoria Hospital, Newcastle Leeds General Infirmary, Leeds
North West & North Wales & Isle of Man	Alder Hey Children's Hospital, Liverpool Royal Manchester Children's Hospital, Manchester
Trent	Nottingham City Hospital, Nottingham
West Midlands	Birmingham Children's Hospital, Birmingham
East	Addenbrooke's Hospital, Cambridge
North Thames	Great Ormond Street Hospital, London Broomfield Hospital, Chelmsford
The Spires	John Radcliffe Hospital, Oxford Salisbury District Hospital, Salisbury
South Wales & South West	Morriston Hospital, Swansea Frenchay Hospital, Bristol
South Thames	Guy's and St Thomas' Hospital, London
Northern Ireland	Royal Belfast Hospital for Sick Children, Belfast

MCN, managed clinical network

1.3. Aims and objectives of the CRANE Database

The aims of the CRANE Database are:

- to register birth, demographic and epidemiological data related to all children born in England, Wales and Northern Ireland with the congenital abnormality of cleft lip and/or palate;
- to record the treatment of children and adults with a cleft lip and/or palate and the outcome of such treatment.

These data will provide the basis for national audit of cleft care.

In line with these broad aims, the CRANE Database has the following specific objectives:

- to ensure there is an up-to-date register of all children with cleft lip and/or palate;

- to monitor the frequency and incidence of clefting in the population;
- to audit and report on the quality of care for patients with clefts, thus promoting high standards in clinical management;
- to work with and receive advice from the CFSGBI to improve the delivery of cleft care in the UK;
- to work in partnership with Specialised Commissioning Groups (SCGs) to inform commissioning of cleft services;
- to support research and focused studies.

2. Methods

This report contains information on patterns of care and outcomes derived from two sources of data: data from the CRANE Database and Hospital Episode Statistics (HES) linked to the Office for National Statistics (ONS) mortality dataset.

2.1. CRANE

2.1.1. Data source

CRANE is an online custom-built secure database that holds information on children born with a cleft lip and/or palate in England, Wales and Northern Ireland. CRANE collects data pertaining to a patient's birth, demographics, type of cleft, time of diagnosis, time of referral to a cleft team, and time of first contact between a patient and cleft team. CRANE also collects information about cleft-related treatment and outcomes. These data are reported to CRANE by the 15 Administrative Units, listed in *Table 1*. Each child born with a cleft in England, Wales and Northern Ireland should be referred to one of these Units shortly after having their cleft diagnosed.

Since January 2012, CRANE has been able to act as a national register of cleft-affected births by collecting some basic information on all children born with a cleft and being treated by the specialist cleft Administrative Units. Additional information, including patient referral and contact time, as well as cleft-related outcomes, is collected for children whose parents have consented to their child's data being submitted to the national database. Parental consent is obtained by the Administrative Unit, usually at some point between referral and the first primary repair. A coordinator within each Unit submits data to CRANE on the children referred to them. Once a record has been created on CRANE for a particular child, it can later be updated with further information.

2.1.2. Patients

All data entered into the CRANE Database by 18 September 2013 pertaining to children born between 1 January 2003 and 31 December 2012 have been included in the analyses described in this Annual report. Patients whose parents did not consent to their data being used by CRANE have been excluded from *Tables 9-14*.

2.1.3. Data validation and cleaning

Logical and systematic data cleaning was undertaken to identify any potential data errors. Continuous data variables (birth weight, five-year weight and five-year height) were assessed in relation to valid ranges. Valid ranges for five-year body weight and five-year height were defined according to growth charts published by the World Health Organisation (WHO) [7].

2.1.4. Analyses

Data have been analysed according to year of birth, unless otherwise stated. Five-year outcome data were restricted to children born between 2004 and 2007, depending on the outcome of interest. Children dying before five years of age were excluded from these analyses.

Cleft type

Cleft type was defined according to reported *LAHSAL* codes. The *LAHSAL* code is used to classify clefts, with each letter relating to one of the six parts of the mouth that can be affected by a cleft:

L	**A**	**H**	**S**	**A**	**L**
Right Lip	Right Alveolus	Hard palate	Soft palate	Left Alveolus	Left Lip

The code also indicates whether there is a complete cleft (upper case letter, e.g. H), an incomplete cleft (lower case letter, e.g. h), or no cleft (left blank). Where *LAHSAL* has not been reported (10.1% of children born in 2012), cleft type is based on the type reported by the Administrative Unit registering the child. Children with a unilateral cleft lip and palate (UCLP) were categorised according to whether the UCLP was complete or incomplete. A complete UCLP was defined as *LAHS* or *HSAL* codes, indicating a complete cleft affecting all three components of the mouth on either the right or left side.

Decayed, missing and filled teeth (dmft)

The dmft score describes the amount of dental caries in an individual and is a measure of oral health. A dmft score reflects the total number of teeth that are decayed, missing or filled. Analyses on dmft data were restricted to consented children born between 2004 and 2007 (excluding children with a submucous cleft palate).

Five Year Old Index

Dental models of five-year old children with UCLP can be assessed using the Five Year Old Index to examine dental arch relationships. The index evaluates the effects of primary

surgery on the facial growth of children with UCLP before any other interventions are performed, such as orthodontics or alveolar bone grafting, which may influence this growth further [15]. CRANE collected both internal and external Five Year Old Index scores for consented children born between 2004 and 2006 with a complete UCLP (*LAHSAL* codes *LAHS* or *HSAL*). Some cleft teams score the models of children treated in their Unit (internal scores) before they are sent off to be scored externally (external scores) by a blinded process undertaken by calibrated examiners. For the purpose of this report, we have analysed external scores only.

Cleft Audit Protocol for Speech – Augmented (CAPS-A)

CAPS-A scores collected at five years of age among children born in 2006 were reported to CRANE for consented children only. The parameters of speech assessed include hypernasality, hyponasality, nasal airflow, nasal turbulence and four cleft speech summary categories (anterior oral Cleft Speech Characteristics (CSCs), posterior oral CSCs, non-oral CSCs and passive CSCs), summarising 12 CSCs in total.

Missing data

Missing data have been excluded from the denominators presented in *Tables 5* to *8* and *10* to *14*. All Units have some degree of missing data. The number of patients with missing data for five-year outcomes is high. A variety of reasons were reported by units. Reasons out of a Unit's control include children not attending an appointment or moving away from the area.

2.2. Hospital Episode Statistics (HES)

2.2.1. Data source

HES is a national database containing records on all admissions to NHS hospitals in England. It includes data on private patients treated in NHS hospitals, patients who were resident outside of England and care delivered by treatment centres (including those in the independent sector) funded by the NHS. Data on admissions are available for every financial year from 1989/90 onwards. Since the 1997/98 financial year, a unique patient identifier has been available that enables records belonging to the same patient to be identified across years.

For this report, data were extracted from the HES database linked to the Office for national Statistics (ONS) mortality dataset. Diagnostic information is coded using the International *Classification of Disease* 10th revision (ICD-10), and procedure information is classified

according to codes from the *Classification of Surgical Operations and Procedures* 4th Revision (OPCS-4).

Using the linked dataset, we examined neonatal (<28 days of life) and infant (<1 year of life) mortality among children with a cleft lip and/or palate.

2.2.2. Patients

Patients born between 1 January 2001 and 31 December 2010 were included and defined as cleft patients if they had at least one HES record with a diagnosis code for cleft lip and/or palate (ICD-10 codes Q35, Q36, Q37). Patients who were identified in HES as 'private' with an 'unavailable/not applicable' postcode were excluded from analyses as they are unlikely to represent the 'typical' cleft patient in England, and they are less able to be followed up.

2.2.3. Deaths

Deaths occurring within the first 364 days of life were assessed. Age at death was classified as early neonatal (0-6 days), late neonatal (7-27 days) or post neonatal (28-364 days).

2.2.4. Additional anomalies

Children were defined as having a syndrome or additional anomalies if any of their hospital episode or mortality records contained at least one of 33 ICD-10 diagnostic codes (listed in Appendix 5) representing congenital malformations and chromosomal abnormalities, in any one of the diagnosis or cause of death fields. Deaths were analysed separately for patients with additional anomalies, as the presence of these has a major impact on the risk of mortality.

2.2.5. Cleft type

Clefts were grouped as cleft lip only (CL), cleft palate only (CP) and cleft lip and palate (CLP) according to the diagnosis codes (ICD-10) in the available HES records. If a child's records provided more than one diagnosis code, the child was categorised according to the more severe diagnosis code. For example, if a child had both CL and CLP diagnoses in their records, they were categorised as having CLP. If they had separate records of CL and CP, they were also categorised as having CLP.

2.2.6. Analyses

The HES and ONS mortality linked data underwent validation checks and duplicate records were removed. Mortality rates are presented per 1,000 live births. Where appropriate, 95% confidence intervals (CI) are provided with mortality rates. These are calculated using the Poisson distribution. Data were analysed according to the absence or presence of additional anomalies and cleft type classification. All analyses were performed in Stata 11 (Statacorp, College Station, TX, USA).

3. CRANE

In this chapter, we present data on children with a cleft lip and/or palate, born between 1 January 2003 and 31 December 2012 in England, Wales and Northern Ireland. The consent status for all children born in 2012 who have been referred to a Cleft Administrative Unit for treatment and registered on CRANE is presented below. Data entered into the CRANE Database by 18 September 2013 have been analysed to assess registration patterns, the timing of diagnosis, referral and contact with Administrative Units around the time of birth, and cleft-related outcomes at five years of age.

3.1. Consent status

Out of 1,127 children born in 2012 and being treated by the 15 Administrative Units, the parents of 901 (79.9%) had been approached for consent. This figure ranged from 51.0% at Guy's and St Thomas' to 100% at Leeds and Belfast (*Table 2*). Of the children whose parents had been through the consenting process, 98.4% provided consent for their child's data to be submitted to CRANE, which is extremely positive. This proportion ranged from 91.1% at Swansea to 100% at ten Units (Newcastle, Liverpool, Nottingham, Cambridge, GOSH, Oxford, Salisbury, Bristol, Guy's and St Thomas' and Belfast). Administrative Units reported a total of 226 (20.1%) children born in 2012 whose parents had not yet been approached for consent. Of these, it was not possible to obtain consent for 13 (5.8%) children (1.2% of all children born in 2012). The overall proportion of children whose parents still need to be approached for consent ranged from 0% at Leeds and Belfast to 49.0% at Guy's and St Thomas'.

Overall, the consenting data are encouraging. The consent rate is very high for those children whose parents have been approached. However, there is still a relatively high proportion of children whose parents have not yet been through the consent process, and the varied rate between Units suggests different processes are being used across centres. We are aware that some Units do not obtain consent until the time of the primary repair, which may account for some of this lag. Units with a high proportion of unconsented children may want to review their consent-taking process, with the aim of obtaining consent in a more timely fashion.

Table 2. Number of children born in 2012 with a cleft lip and/or palate in England, Wales and Northern Ireland, according to Administrative Unit and consent status

		Consent status n (%)				
		Consent status verified		Consent status not verified		
Regional centre / MCN	**Administrative Unit**	Consented	Refused	Awaiting verification	Not possible to verify	All
Northern & Yorkshire	Newcastle	61 (93.8)	0 (0.0)	1 (1.5)	3 (4.6)	65
	Leeds	63 (96.9)	2 (3.1)	0 (0.0)	0 (0.0)	65
North West & North Wales	Liverpool	58 (90.6)	0 (0.0)	6 (9.4)	0 (0.0)	64
	Manchester	65 (94.2)	2 2.9)	2 (2.9)	0 (0.0)	69
Trent	Nottingham	92 (98.9)	0 (0.0)	1 (1.1)	0 (0.0)	93
West Midlands	Birmingham	102 (84.3)	4 (3.3)	12 (9.9)	3 (2.5)	121
East	Cambridge	49 (56.3)	0 (0.0)	35 (40.2)	3 (3.4)	87
North Thames	Gt Ormond St	101 (78.3)	0 (0.0)	28 (21.7)	0 (0.0)	129
	Chelmsford	40 (90.9)	2 (4.5)	2 (4.5)	0 (0.0)	44
The Spires	Oxford	30 (66.7)	0 (0.0)	12 (26.7)	3 (6.7)	45
	Salisbury	33 (62.3)	0 (0.0)	20 (37.7)	0 (0.0)	53
South Wales & South West	Swansea	41 (80.4)	4 (7.8)	6 (11.8)	0 (0.0)	51
	Bristol	47 (72.3)	0 (0.0)	17 (26.2)	1 (1.5)	65
South Thames	Guy's and St Thomas'	74 (51.0)	0 (0.0)	71 (49.0)	0 (0.0)	145
Northern Ireland	Belfast	31 (100.0)	0 (0.0)	0 (0.0)	0 (0.0)	31
All	All	887 (78.7)	14 (1.2)	213 (18.9)	13 (1.2)	1,127

MCN, managed clinical network

3.2. CRANE registrations

A total of 12,863 children born between 1 January 2000 and 31 December 2012 have been registered on the CRANE Database, of whom 1,127 have been added for 2012 births .

Table 3 shows the number of registrations for each Cleft Unit over the last 10 years (since 2003). Birmingham registered the most births over the last 10 years. The North West and North Wales region, consisting of two Administrative Units, is the region that has the most registrations overall.

CRANE received the highest number of registrations for births in 2012 compared to all other birth years. This increase in registrations can be attributed to the registration of children before the verification of consent. The greatest increase in registrations was observed at Guy's and St Thomas', which registered twice as many children born in 2012 than registered in 2010 or 2011.

The distribution of the four main cleft types is shown in *Table 4*. Cleft type was defined according to reported *LAHSAL* codes. Where *LAHSAL* has not been reported (10.1% of children registered in 2012), cleft type is based on the type reported by the Administrative Unit registering the child. Overall, 7.8% of the registered children born in 2012 did not have their type of cleft specified, which is twice as high as the 3.1% rate for 2011. The proportion of children without a specified cleft type ranged between Units from 0% (Leeds, Liverpool, Oxford) to 24.8% at GOSH. Last year, Chelmsford had the highest proportion of patients whose cleft type was not specified (12.0%). Their rate has improved substantially for 2012, with just one child (2.3%) missing a cleft type classification.

One possible explanation for the overall increased proportion of children missing a reported cleft type could be that Units are now registering children earlier, before a cleft type has been confirmed. Units are encouraged to ensure that CRANE is updated as soon as new information about cleft type for a patient becomes available.

The distribution of cleft type is consistent over time. CP is the most common type of cleft, affecting over 40% of the cleft population. This proportion increases slightly, to around 45%, once late CP diagnoses are reported to CRANE. Bilateral cleft lip and palate (BCLP) is the least common type, affecting around 10% of people with clefts. A total of 185 children registered in 2012 had complete UCLP (defined by either '*LAHS*..' or '..*HSAL*' *LAHSAL* codes), representing 74.3% of the 249 children with UCLP. This is similar to the proportion in recent previous years.

Table 3. Number of CRANE-registered children born with a cleft lip and/or palate in England, Wales and Northern Ireland, according to Administrative Unit and year of birth, 2003-2012

		Year of birth										
Regional centre / MCN	**Administrative Unit**	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012	All
Northern & Yorkshire	Newcastle	68	59	77	55	87	65	61	64	64	65	665
	Leeds	78	71	71	75	70	74	67	71	70	65	712
North West & North Wales	Liverpool	61	65	85	54	62	85	79	86	63	64	704
	Manchester	84	84	85	105	91	80	67	90	81	69	836
Trent	Nottingham	74	92	105	94	85	98	81	91	90	93	903
West Midlands	Birmingham	119	110	104	116	92	107	94	94	105	121	1062
East	Cambridge	73	82	87	81	84	86	86	82	66	87	814
North Thames	Gt Ormond St	24	38	70	86	78	110	104	82	109	129	830
	Chelmsford	21	36	42	33	33	30	43	41	54	44	377
The Spires	Oxford	42	39	43	46	56	34	51	43	58	45	457
	Salisbury	40	58	46	69	72	55	38	54	54	53	539
South Wales & South West	Swansea	40	37	45	46	47	42	47	44	50	51	449
	Bristol	59	53	50	57	62	70	51	72	51	65	590
South Thames	Guy's and St Thomas'	57	86	98	101	111	104	83	60	76	145	921
Northern Ireland	Belfast	27	29	37	42	41	30	34	38	36	31	345
All	All	867	939	1045	1060	1071	1070	986	1012	1027	1127	10204

MCN, managed clinical network

Table 4. Number (%) of CRANE-registered children born with a cleft lip and/or palate in England, Wales and Northern Ireland, according to cleft type and year of birth, 2003-2012

	Year of birth n (%)										
Cleft type	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012	All
CL	169 (20.5)	209 (23.1)	196 (19.7)	234 (23.2)	254 (24.3)	256 (24.8)	203 (21.4)	243 (24.7)	243 (24.9)	239 (23.0)	2246 (23.0)
CP	403 (49.0)	409 (45.3)	491 (49.3)	465 (46.1)	448 (42.8)	465 (45.1)	444 (46.8)	431 (43.8)	430 (44.1)	455 (43.8)	4441 (45.5)
UCLP	187 (22.7)	198 (21.9)	232 (23.3)	214 (21.2)	234 (22.3)	235 (22.8)	200 (21.1)	204 (20.8)	215 (22.1)	249 (24.0)	2168 (22.2)
BCLP	64 (7.8)	87 (9.6)	76 (7.6)	96 (9.5)	111 (10.6)	76 (7.4)	102 (10.7)	105 (10.7)	86 (8.8)	96 (9.2)	899 (9.2)
Not specified	44 -	36 -	50 -	51 -	24 -	38 -	37 -	29 -	53 -	88 -	450 -
All	867 (100.0)	939 (100.0)	1045 (100.0)	1060 (100.0)	1071 (100.0)	1070 (100.0)	986 (100.0)	1012 (100.0)	1027 (100.0)	1127 (100.0)	10204 (100.0)

CL, cleft lip; CP, cleft palate; UCLP, unilateral cleft lip and palate; BCLP, bilateral cleft lip and palate

3.3. Characteristics of children born with a cleft lip and/or palate, 2012

Of the children born with a cleft in 2012, 45.9% were girls and 54.1% were boys. Thirty-nine (3.5%) children did not have their sex reported to CRANE. There are significant gender differences in the distribution of cleft type ($P<0.001$), as shown in the Annual Report published in 2010 [8]. CP is more prevalent among females (56.2% vs. 43.8% in males), while CL, UCLP and BCLP is more prevalent among males (59.3% vs. 40.7%; 62.5% vs. 37.5% and 67.4% vs. 32.6%, respectively).

Gestational age was reported for 457 (51.5%) consented babies born in 2012. This reporting has increased by 3% since last year; however, further improvements are required. The mean gestation for those born in 2012 was 38.6 weeks (95% CI 38.4 to 38.9 weeks) and ranged from 22 to 42 weeks. Sixty-four (14.0%) babies were premature (born before 37 weeks' gestation), which is higher than the six per cent background rate in England [9], although it should be noted that the gestation recorded in CRANE may not be representative of all babies born with a cleft lip and/or palate as 48.5% of registered children were missing this information.

A valid birth weight was reported for 478 (53.9%) babies born in 2012. The mean birth weight was 3.17kg (95% CI 3.11 to 3.23kg), which is consistent with the national average [9].

Among all the children born in 2012, there were 13 (1.2%) deaths reported to CRANE. The majority of deaths occurred between one month and one year of age. It is not known from CRANE whether these children had additional anomalies or syndromes.

3.4. Timing of diagnosis

3.4.1. Diagnosis times among children born in 2012

Of the 1,127 children born in 2012 with a cleft diagnosis, 89 (7.9%) did not have the timing of their diagnosis reported to CRANE. This is almost twice as high as last year's figures. Units with high levels of missing diagnosis time data include GOSH (30.2%), Bristol (12.3%) and Swansea (11.8%). All other Units had missing data rates below 10%.

Of the 1,038 children born in 2012 with a reported diagnosis time, 406 (39.1%) had their cleft diagnosed during the antenatal period. This is slightly lower (not statistically significant) than the 42.2% of children born in 2011 with an antenatal diagnosis. The proportion of children diagnosed antenatally varied between cleft types, as shown in *Table 5*. Fifty-six per cent of children with CL and almost 80% of children with UCLP and BCLP were diagnosed in the

antenatal period. Conversely, only 1.1% of children with a CP were diagnosed antenatally, which demonstrates the difficulty of identifying this type of cleft with current antenatal screening techniques.

Table 5. Number (%) of CRANE-registered children born in 2012 with a cleft lip and/or palate according to the timing of diagnosis and cleft type

	Time of diagnosis in relation to birth* n (%)						
Cleft type	Antenatal	At birth	≤1 week	≤1 month	≤6 months	>6 months	**All**
CL	131 (56.2)	84 (36.1)	11 (4.7)	3 (1.3)	3 (1.3)	1 (0.4)	233
CP	5 (1.1)	292 (66.8)	89 (20.4)	21 (4.8)	24 (5.5)	6 (1.4)	437
UCLP	189 (79.1)	44 (18.4)	3 (1.3)	1 (0.4)	1 (0.4)	1 (0.4)	239
BCLP	71 (78.0)	19 (20.9)	1 (1.1)	0 (0.0)	0 (0.0)	0 (0.0)	91
Not specified	10 (26.3)	18 (47.4)	4 (10.5)	1 (2.6)	2 (5.3)	3 (7.9)	38
All	406 (39.1)	457 (44.0)	108 (10.4)	26 (2.5)	30 (2.9)	11 (1.1)	1038

CRANE, 2012 births

* 89/1127 (7.9%) missing diagnosis time and excluded from the table; CL, cleft lip; CP, cleft palate; UCLP, unilateral cleft lip and palate; BCLP, bilateral cleft lip and palate

Of the 632 children not diagnosed during the antenatal period, 72.3% were diagnosed at birth. This is 4.5% lower than the rate in 2011. Of those children who did not have their cleft identified antenatally, the majority (82.5%) with a CL, UCLP and BCLP were diagnosed at the time of birth; however, 32.4% of children with a CP were not identified until later, with 6.9% of all children with a CP being diagnosed beyond one month after birth. It should be noted that some children born in 2012 with a CP may not yet have had their cleft identified. Each year, around ten children with CP are diagnosed after six months of age.

3.4.2. Diagnosis times among children with a cleft palate alone

Last year's report highlighted the issue of late diagnosis among children with CP. We reported that 1% were diagnosed during antenatal screening and 71% were diagnosed at birth, leaving 28% who were diagnosed late according to the national standard [4]. This year, we have examined diagnosis time among CP patients born between 2008 and 2012. No significant differences were found between birth years, indicating diagnosis times have not improved in recent years.

Table 6. Number (%) of CRANE-registered children born between 2008 and 2012 with a cleft palate, according to the timing of diagnosis and Administrative Unit

		Time of diagnosis in relation to birth* n (%)						
Regional centre / MCN	**Administrative Unit**	Antenatal	At birth	≤1 week	≤1 month	≤6 months	>6 months	**All**
Northern & Yorkshire	Newcastle	2 (1.3)	97 (64.7)	14 (9.3)	18 (12.0)	9 (6.0)	10 (6.7)	150
	Leeds	1 (0.7)	98 (66.7)	28 (19.0)	15 (10.2)	3 (2.0)	2 (1.4)	147
North West & North	Liverpool	1 (0.7)	113 (73.9)	21 (13.7)	8 (5.2)	6 (3.9)	4 (2.6)	153
	Manchester	0 (0.0)	97 (63.4)	32 (20.9)	11 (7.2)	11 (7.2)	2 (1.3)	153
Trent	Nottingham	2 (1.8)	83 (75.5)	18 (16.4)	3 (2.7)	3 (2.7)	1 (0.9)	110
West Midlands	Birmingham	0 (0.0)	154 (81.1)	22 (11.6)	4 (2.1)	10 (5.3)	0 (0.0)	190
East	Cambridge	2 (1.2)	111 (66.5)	27 (16.2)	15 (9.0)	12 (7.2)	0 (0.0)	167
North Thames	Gt Ormond St	2 (1.2)	77 (46.1)	60 (35.9)	9 (5.4)	15 (9.0)	4 (2.4)	167
	Chelmsford	1 (1.1)	51 (58.6)	17 (19.5)	7 (8.0)	6 (6.9)	5 (5.7)	87
The Spires	Oxford	2 (1.9)	86 (80.4)	5 (4.7)	1 (0.9)	6 (5.6)	7 (6.5)	107
	Salisbury	2 (2.7)	57 (77.0)	9 (12.2)	4 (5.4)	2 (2.7)	0 (0.0)	74
South Wales & South	Swansea	2 (2.2)	68 (76.4)	9 (10.1)	6 (6.7)	3 (3.4)	1 (1.1)	89
	Bristol	1 (0.8)	82 (67.8)	25 (20.7)	6 (5.0)	6 (5.0)	1 (0.8)	121
South Thames	Guy's and St Thomas'	3 (1.6)	105 (56.1)	47 (25.1)	15 (8.0)	13 (7.0)	4 (2.1)	187
Northern Ireland	Belfast	1 (1.5)	50 (74.6)	7 (10.4)	2 (3.0)	1 (1.5)	6 (9.0)	67
All	All	22 (1.1)	1329 (67.5)	341 (17.3)	124 (6.3)	106 (5.4)	47 (2.4)	1969

CRANE, 2008-2012 births

* 256/2225 (11.5%) missing diagnosis time and excluded from 'All' values; MCN, managed clinical network

Table 6 shows the CP diagnosis times according to Administrative Unit. The proportion of CPs diagnosed at birth ranged from 46.1% among children registered by GOSH to 80.4% among those registered by Oxford. This wide variation suggests that practice varies considerably between maternity units, with some better than others at identifying clefting of the palate during the newborn examination. Overall, 14.1% of children with a CP were not diagnosed until they were at least one month old, which is concerning given that the National standard [4] states that clefts should be diagnosed within 24 hours of birth to enable immediate referral to a specialist hospital. This helps to ensure the baby, and their family, receive appropriate care and support as soon as possible. Administrative Units are advised to encourage their referring maternity units to identify all clefts as promptly as possible.

3.5. Referral to and first contact with a cleft team

3.5.1. Referral among children born in 2012

Out of the 887 consented children born in 2012, 104 (11.7%) were missing referral time. Almost one quarter of those without a reported referral time were registered by Salisbury: 69.7% of their consented children born in 2012 were missing this information. Other Units with high levels of missing referral time data were Belfast (45.2%), GOSH (20.8%), Nottingham (17.4%), Liverpool (13.8%) and Manchester (13.8%).

Of the 783 children with a reported referral time, 52.6% were referred to an Administrative Unit within 24 hours of birth. Seventy-three per cent of children whose clefts were diagnosed antenatally were referred to an Administrative Unit within 24 hours of birth. This compares to 39.6% of the 480 children without an antenatal diagnosis. This proportion is similar to that in 2011.

Table 7 shows that the proportion of referrals within 24 hours of birth varied according to cleft type ($p<0.001$), with CP patients having the lowest proportion (35.3%) out of those with a known cleft type, which corresponds with later diagnosis times for these children.

Referrals within one day of birth also varied significantly according to the Administrative Unit receiving the referral ($p<0.001$) (*Table 8*). Seventy-seven per cent of children registered by Belfast were referred from maternity units within 24 hours of birth, which is in contrast to only 31.0% of those referred to Oxford from maternity units. Progress in this area has been made in eight regions, with the greatest improvements observed at Manchester, Birmingham and Belfast.

Table 7. Number (%) of CRANE-registered consented children born in 2012 with a cleft lip and/or palate who were referred within 24 hours of birth to the Administrative Unit and contacted by the Administrative Unit within 24 hours of referral, according to cleft type

	Referral to Unit		Contact between Unit and parents of patient	
	Within 24h of birth	All*	Within 24h of referral to Unit	All[§]
Cleft type	n (%)	N	n (%)	N
CL	96 (57.1)	168	133 (89.3)	149
CP	120 (35.3)	340	268 (89.0)	301
UCLP	140 (74.1)	189	158 (93.5)	169
BCLP	47 (72.3)	65	56 (94.9)	59
Not specified	9 (42.9)	21	14 (77.8)	18
All	412 (52.6)	783	629 (90.4)	696

CRANE, 2012 births

*104/887 (11.72%) missing referral time, [§]191/887 (21.53%) missing contact time. Missing excluded in 'All' values; CL, cleft lip; CP, cleft palate; UCLP, unilateral cleft lip and palate; BCLP, bilateral cleft lip and palate

3.5.2. First contact between the Unit and parents of children born in 2012

Out of the 887 consented children born in 2012, 191 (21.5%) were missing the first contact time between Units and parents. One quarter of those without a reported contact time were registered by GOSH: 46.5% of their consented children born in 2012 were missing this information. The Unit with the highest proportion of children with a missing contact time was Oxford, with 83.3% of their registered children missing these data. Other Units with high levels of missing referral time data were Belfast (45.2%), GOSH (20.8%), Nottingham (17.4%), Liverpool (13.8%) and Manchester (13.8%).

Of the 629 consented children with a reported contact time, Units established contact with 90.4% within 24 hours of referral (*Table 7*). This is consistent with last year's rate of 89.7%. The proportion of patients contacted within 24 hours of being referred to an Administrative Unit did not vary significantly between cleft types. *Table 8 shows that* rates varied between Units (40.0% to 100.0%), but the majority contacted greater than 90% of their patients within 24 hours of being referred.

Table 8. Number (%) of CRANE-registered consented children born in 2012 with a cleft lip and/or palate who were referred within 24 hours of birth and contacted within 24 hours of referral, according to Administrative Unit

		Referral to Unit		Contact between Unit and parents of patient	
Regional centre / MCN	**Administrative Unit**	Within 24h of birth n (%)	All* N	Within 24h of referral to Unit n (%)	All[§] N
Northern & Yorkshire	Newcastle	32 (52.5)	61	54 (90.0)	60
	Leeds	41 (65.1)	63	61 (98.4)	62
North West & North Wales	Liverpool	23 (46.0)	50	45 (97.8)	46
	Manchester	35 (62.5)	56	55 (100.0)	55
Trent	Nottingham	40 (52.6)	76	75 (98.7)	76
West Midlands	Birmingham	61 (60.4)	101	81 (95.3)	85
East	Cambridge	30 (66.7)	45	37 (97.4)	38
North Thames	Gt Ormond St	32 (40.0)	80	33 (61.1)	54
	Chelmsford	16 (40.0)	40	17 (44.7)	38
The Spires	Oxford	9 (31.0)	29	2 (40.0)	5
	Salisbury	5 (50.0)	10	6 (75.0)	8
South Wales & South West	Swansea	16 (42.1)	38	37 (100.0)	37
	Bristol	18 (39.1)	46	43 (97.7)	44
South Thames	Guy's and St Thomas'	41 (57.7)	71	67 (94.4)	71
Northern Ireland	Belfast	13 (76.5)	17	16 (94.1)	17
All	All	412 (52.6)	783	629 (90.4)	696

CRANE, 2012 births

*104/887 (11.72%) children missing referral time; [§]191/887 (21.53%) children missing contact time. Missing excluded in 'All' values; MCN, managed clinical network

3.6. Five-year outcomes among children born with a cleft lip and/or palate

Five-year outcomes include height and weight, decayed, missing and filled teeth (dmft), the Five Year Old Index, and, for the first time, the Cleft Audit Protocol for Speech – Augmented (CAPS-A) scores.

3.6.1. Reporting of outcomes

Table 9 shows the number of consented children with reported outcomes at five years of age, according to Administrative Unit.

Table 9. Number (%) of CRANE-registered consented children born between 2004 and 2007 with reported outcomes at five years of age, according to Administrative Unit

Regional centre / MCN	Administrative Unit	Children alive at 5 years	Reported weight		Reported height		dmft*			5 year index[§]			Speech[¥]		
								Reported			Reported			Reported	
		N	n	(%)	n	(%)	N	n	(%)	N	n	(%)	N	n	(%)
Northern	Newcastle	260	187	(71.9)	183	(70.4)	226	196	(86.7)	22	7	(31.8)	43	33	(76.7)
&Yorkshire	Leeds	276	213	(77.2)	218	(79.0)	261	177	(67.8)	31	23	(74.2)	53	37	(69.8)
North West	Liverpool	220	26	(11.8)	25	(11.4)	211	150	(71.1)	28	24	(85.7)	33	24	(72.7)
	Manchester	264	47	(17.8)	47	(17.8)	260	182	(70.0)	20	11	(55.0)	60	32	(53.3)
Trent	Nottingham	362	24	(6.6)	24	(6.6)	352	23	(6.5)	45	9	(20.0)	74	34	(45.9)
West Midlands	Birmingham	397	188	(47.4)	185	(46.6)	393	353	(89.8)	52	47	(90.4)	77	0	(0.0)
East	Cambridge	241	22	(9.1)	18	(7.5)	236	0	(0.0)	32	18	(56.3)	45	18	(40.0)
North Thames	Gt Ormond St	233	5	(2.1)	5	(2.1)	214	39	(18.2)	0	0	-	47	0	(0.0)
	Chelmsford	143	24	(16.8)	24	(16.8)	139	64	(46.0)	29	0	(0.0)	21	0	(0.0)
The Spires	Oxford	161	1	(0.6)	1	(0.6)	158	118	(74.7)	24	18	(75.0)	34	22	(64.7)
	Salisbury	213	2	(0.9)	2	(0.9)	195	108	(55.4)	19	18	(94.7)	48	29	(60.4)
South Wales &	Swansea	174	56	(32.2)	52	(29.9)	172	95	(55.2)	22	8	(36.4)	33	26	(78.8)
South West	Bristol	209	59	(28.2)	60	(28.7)	205	86	(42.0)	21	13	(61.9)	40	29	(72.5)
South Thames	Guy's and St Thomas'	352	103	(29.3)	95	(27.0)	351	176	(50.1)	50	43	(86.0)	67	34	(50.7)
Northern Ireland	Belfast	141	0	(0.0)	0	(0.0)	138	51	(37.0)	21	0	(0.0)	29	23	(79.3)
All	All	3646	957	(26.2)	939	(25.8)	3511	1,818	(51.8)	416	239	(57.5)	704	341	(48.4)

CRANE, 2004-2007 births

62/3708 (1.7%) children died before 5 years and are excluded from table; *135/3646 (3.7%) children with submucous cleft palates excluded from dmft data; [§] Children born in 2004-2006 only, 177/593 (29.8%) incomplete UCLPs excluded from 5-year old index data; [¥] Children born in 2006 only, 218/956 (22.8%) children born with a CL and 34 (3.6%) children born with a non-specified cleft type excluded from Speech data.

There is a very high proportion of missing data for five-year height and weight. Belfast was the only Unit to not submit any height or weight data. However, very low levels of data were also reported by Oxford (0.6%), Salisbury (0.9%), GOSH (2.1%), Nottingham (6.6%) and Cambridge (7.5%), suggesting that these measures are not routinely collected. Conversely, Newcastle and Leeds managed to report height and weight for over 70% of their eligible patients. Although, overall, the reporting of these data is poor, there has been an improvement for the most recent birth cohort year for whom five-year old outcome data were collected. For patients born in 2007 who were alive at five years, weight was reported for 34.5% while height was reported for 33.1%. This compares to the reporting of both outcomes for just 25.2% born in 2006. It is hoped that the reporting of these outcomes improves substantially over the coming years.

Out of 3,511 consented children born between 2004 and 2007 (excluding 62 children dying before five years of age and 135 with submucous CPs), dmft scores were provided for 1,818 (51.8%). While dmft scores were submitted for 56.5% of children born in 2005 and 2006, they were reported for only 37.9% of those born in 2007.

The proportion of eligible children with reported dmft scores ranged from 0% (Cambridge) to 89.8% (Birmingham). Cambridge did not submit any dmft data as they do not have a paediatric dentist who would examine children to determine the dmft. Fortunately, this issue has been addressed by Cambridge this year, as a paediatric dentist has now been appointed. Nottingham and GOSH submitted data for only 6.5% and 18.2% of their eligible patients, respectively, despite collecting the data. Nottingham has previously informed CRANE that they have not had adequate administrative support to provide CRANE with dmft data. However, the provision of some data this year, albeit for only a few patients, represents a positive change towards providing CRANE with outcomes, and it is hoped that their outcome data submission continues to improve. Only Newcastle, Birmingham and Oxford provided data for at least 75% of their eligible patients.

Out of 416 consented children born between 2004 and 2006 with a complete UCLP and alive at five years, the Five Year Old Index was reported for 239, representing 57.5% of eligible children. The proportion of children with a reported Five Year Old Index score is 53.6% for those born in 2006, which compares to 62.4% for those born in 2004. It is not known why the proportion of children with reported data is lower for the most recent data collection year; however, one explanation may be the variation each year in attaining the record used to score the index due to the children's compliance.

Five Year Old Index data are not collected by Belfast because children are not routinely seen by Orthodontists at five years of age. Chelmsford did not provide any Five Year Old Index data, despite it being collected, and GOSH did not have any eligible children for whom to provide data. Nottingham has only recently started collecting these data, hence the low proportion of eligible children with scores. It is hoped that this proportion will increase dramatically over the next few years.

Only Liverpool, Birmingham, Oxford, Salisbury and Guy's and St Thomas' provided data for more than 75% of their eligible patients, as recommended by the Orthodontic Special Interest Group at the 2012 Craniofacial Society of Great Britain and Ireland Annual Conference, and, thus, scores for other Units should be interpreted with caution. The small number of patients with reported scores within each Unit (7-47) means that statistical comparison between Units is not currently appropriate. CRANE will continue to collect these outcomes over the next few years, and as numbers increase, meaningful comparison between Units will become possible.

Out of 704 eligible children born in 2006 with a cleft affecting the palate (CP, UCLP, BCLP), CAPS-A data were provided for 341, representing 48.4% of eligible children. Three units (Birmingham, GOSH and Chelmsford) did not provide any speech-outcome data. Birmingham reported support staff issues which prevented submission of the data. GOSH and Chelmsford felt the current lack of risk adjustment for speech outcomes prevented them from submitting data this year, despite agreement at a multidisciplinary meeting of the CFSGBI earlier in the year that CRANE would undertake risk adjustment research. Of those Units reporting data, the proportion of eligible children with CAPS-A scores ranged from 40.0% at Cambridge to 79.3% at Belfast. Given that this has been the first year that CAPS-A scores have been requested, CRANE is encouraged by the fact that the majority of Units have reported data and that some Units have reported data for the majority of their eligible children.

3.6.2. Height and weight (2004-2007 births)

Five-year height and weight were reported for 26% of the 3,646 children born in 2004-2007 who were alive at five years of age. The mean (SD) height was 111.4cm (5.8cm) while the mean weight was 19.7kg (3.1kg). Boys were marginally taller than girls (112.0 cm vs. 110.6cm) but no difference in weight existed between the sexes.

3.6.3. Decayed missing and filled teeth (dmft) (2004-2007 births)

The dmft describes the amount of dental caries in an individual and is a measure of oral health. A dmft score reflects the total number of teeth that are decayed, missing or filled. The risk of dental caries is thought to be higher among children with a cleft lip and/or palate compared to children without an oral cleft [10, 11]. We collect dmft data on CRANE-registered consented children at five years of age.

Among children with a reported dmft outcome, 42.6% of children with a cleft had at least one (>0) decayed, missing or filled tooth. The mean number of dmft at five years among children registered in CRANE was 2.1, with scores ranging from 0 to 24. Two hundred and fifty-nine children (14.3%) had a dmft score greater than 5. The dmft data, obtained in 2005, available for five-year old children in the general population in England and Wales show that 38.8% of five-year olds had >0 dmft, with a mean number of 1.5 [12]. The comparable figures for England and Wales among CRANE-registered children are significantly higher than the background rate.

Table 10. Number (%) of CRANE-registered consented children born between 2004 and 2007 with a cleft lip and/or palate according to the number of decayed, missing or filled teeth (dmft) at age five years and cleft type

	Number of decayed, missing or filled teeth (dmft)				
		0	>0		
Cleft type	Mean (95% CI)	n (%)	n (%)	(95% CI)	**All***
CL	1.4 (1.1 to 1.6)	236 (63.8)	134 (36.2)	(31.3 to 41.1)	370
CP	2.4 (2.1 to 2.7)	423 (57.9)	307 (42.1)	(38.5 to 45.6)	730
UCLP	2.0 (1.7 to 2.2)	272 (53.5)	236 (46.5)	(42.1 to 50.8)	508
BCLP	2.6 (2.0 to 3.2)	93 (50.5)	91 (49.5)	(42.2 to 56.8)	184
Not specified	1.6 (0.1 to3.1)	20 (76.9)	6 (23.1)	(5.7 to 40.4)	26
All	2.1 (1.9 to 2.2)	1044 (57.4)	774 (42.6)	(40.3 to 44.9)	1818

CRANE, 2004-2007 births

* 138/3708 (3.7%) children with submucous clefts excluded; 59/3570 (1.7%) children who died before the age of five excluded; 1693/3511 (48.2%) children with missing dmft data excluded; CL, cleft lip; CP, cleft palate; UCLP, unilateral cleft lip and palate; BCLP, bilateral cleft lip and palate

Table 10 shows the prevalence of dental caries according to cleft type. The mean dmft and the proportion of children with >0 dmft varied significantly according to cleft type (P=0.002). However, the number of dmft among children with a CL is not significantly different to the general population. Although mean dmft was significantly higher among CP patients compared to the background rate, the difference in the proportion of children with >0 dmft

is only borderline significant. Children with a UCLP and BCLP had significantly higher mean dmft scores than the general population and there were significant differences in the proportion of children with >0 dmft.

The fact that dmft were submitted for only 54.7% of children means that these data should be interpreted with caution. One Administrative Unit (Cambridge) did not provide dmft data for any of their patients. Thus, it is possible that the overall findings from the limited data made available to CRANE may not be representative of the cleft population. Analyses of data from a greater number of children are necessary to examine true differences that may exist between the cleft population and general population, and between cleft types.

Table 11 shows the prevalence of dmft according to Administrative Unit. There was a significant variation in dmft scores across Units. Children registered by Nottingham[1] had the highest number of mean dmft, which was significantly different to the overall mean. It should be noted that Nottingham submitted data for very few patients, and it is possible that dmft data were collected for only those who were referred to the dentist because of problems. This could explain their high caries rate. Data from a larger and more representative sample from Nottingham are required. Salisbury, Bristol and Chelmsford had mean dmft values that were significantly lower than the overall mean. In terms of the proportion of cleft children with >0 dmft, Salisbury had the lowest proportion (29.6%), which was significantly different to the overall proportion among cleft children. Whilst the proportion of cleft children with >0 dmft varies between regions (P=0.047), for the majority of regions their rate does not seem to differ substantially from their region's background rate [13]. The only substantial difference appears to be for Birmingham, whose cleft rate is a third higher than their background rate of 30.7%.

Regional differences in the levels of dental disease will not only be affected by the dental care received by children. Oral health will also be affected by deprivation, cultural differences in attitudes to dental health and water fluoridation levels. A systematic review found that water fluoridation is associated with an increased proportion of children without caries and a reduction in the number of teeth affected by caries [14]. Fluoridation levels vary within and between regions throughout the UK. For example, parts of the West Midlands and parts of the North East receive fluoridated water, whereas other areas do not. Interestingly, data from 2005 revealed the West Midlands had one of the lowest proportions of five year olds with >0 dmft in the general population; however the North East had the highest proportion (50%)

[1] Note that dmft data were reported for only 6.5% of eligible children registered by Nottingham

[13]. Accurate water fluoridation data will be useful for interpreting dmft regional differences and allowing for risk adjustment in the long term.

Table 11. Number (%) of CRANE-registered consented children born between 2004 and 2007 with a cleft lip and/or palate according to the number of decayed, missing or filled teeth (dmft) at age five years and Administrative Unit

		Number of decayed, missing or filled teeth (dmft)				
			0	>0		
Regional centre / MCN	**Administrative Unit**	Mean (95% CI)	n (%)	n (%)	95% CI	**All***
Northern & Yorkshire	Newcastle[§¥]	2.8 (2.2 to 3.4)	102 (52.0)	94 (48.0)	(40.9 to 55.0)	196
	Leeds[§¥]	2.8 (2.1 to 3.4)	95 (53.7)	82 (46.3)	(38.9 to 53.8)	177
North West & North Wales	Liverpool	2.2 (1.6 to 2.8)	85 (56.7)	65 (43.3)	(35.3 to 51.4)	150
	Manchester	2.1 (1.6 to 2.7)	102 (56.0)	80 (44.0)	(36.7 to 51.2)	182
Trent	Nottingham[¥]	5.3 (2.4 to 8.2)	9 (39.1)	14 (60.9)	(39.3 to 82.5)	23
West Midlands	Birmingham[§¥]	1.9 (1.5 to 2.3)	211 (59.8)	142 (40.2)	(35.1 to 45.4)	353
East	Cambridge	_	_	_	_	_
North Thames	Gt Ormond St[§¥]	2.1 (0.9 to 3.2)	22 (56.4)	17 (43.6)	(27.3 to 59.9)	39
	Chelmsford	1.4 (0.8 to 2.0)	37 (57.8)	27 (42.2)	(29.8 to 54.6)	64
The Spires	Oxford[§]	2.1 (1.4 to 2.7)	69 (58.5)	49 (41.5)	(32.5 to 50.6)	118
	Salisbury[§]	1.0 (0.6 to 1.4)	76 (70.4)	32 (29.6)	(20.9 to 38.4)	108
South Wales & South West	Swansea[§¥]	2.1 (1.5 to 2.7)	52 (54.7)	43 (45.3)	(35.1 to 55.5)	95
	Bristol[§¥]	1.2 (0.7 to 1.7)	57 (66.3)	29 (33.7)	(23.5 to 43.9)	86
South Thames	Guy's and St Thomas'[§¥]	1.7 (1.3 to 2.1)	105 (59.7)	71 (40.3)	(33.0 to 47.7)	176
Northern Ireland	Belfast	2.0 (1.2 to 2.8)	22 (43.1)	29 (56.9)	(42.8 to 70.9)	51
All	All	2.1 (1.9 to 2.2)	1044 (57.4)	774 (42.6)	(40.3 to 44.9)	1818

CRANE, 2004-2007 births

* 138/3708 (3.7%) children with submucous clefts excluded; 59/3570 (1.7%) children who died before the age of five excluded; 1693/3511 (48.2%) children with missing dmft data excluded; MCN, Managed Clinical Network; [§]BASCD calibrated assessor; [¥]Specialist paediatric dentist

3.6.4. Five Year Old Index (2004-2006 births)

Dental models of five-year old children with a complete UCLP were assessed using the Five Year Old Index to examine dental arch relationships. The index evaluates the effects of

primary surgery on the facial growth of children with UCLP before any other interventions, such as orthodontics or alveolar bone grafting, which may influence this growth further [15]. Dental arch relationships at five years are thought to predict treatment outcome in terms of facial growth on a population basis rather at the individual child level [16]. The Five Year Old Index may, therefore, also be used to compare treatment outcomes between centres and surgeons. Patients categorised as '1' and '2' on the index are considered to have the best possible outcome, while those categorised as '4' and '5' are thought to have very poor outcomes in terms of facial growth, and they may benefit from further surgery to correct their facial disproportion once facial growth is complete.

Table 12. Number (%) of CRANE-registered consented children born between 2004 and 2005 with a complete unilateral cleft lip and palate, according to Five Year Old Index scores and Administrative Unit

		Five Year Old Index n (%)					
Regional centre / MCN	**Administrative Unit**	1	2	3	4	5	**All***
Northern & Yorkshire	Newcastle	1 (14.3)	0 (0.0)	5 (71.4)	0 (0.0)	1 (14.3)	7
	Leeds	0 (0.0)	7 (30.4)	10 (43.5)	5 (21.7)	1 (4.3)	23
North West & North Wales	Liverpool	2 (8.3)	7 (29.2)	9 (37.5)	5 (20.8)	1 (4.2)	24
	Manchester	0 (0.0)	5 (45.5)	2 (18.2)	3 (27.3)	1 (9.1)	11
Trent	Nottingham	1 (11.1)	5 (55.6)	2 (22.2)	0 (0.0)	1 (11.1)	9
West Midlands	Birmingham	6 (12.8)	17 (36.2)	12 (25.5)	7 (14.9)	5 (10.6)	47
East	Cambridge	0 (0.0)	7 (38.9)	6 (33.3)	4 (22.2)	1 (5.6)	18
North Thames	Gt Ormond St	_	_	_	_	_	_
	Chelmsford	_	_	_	_	_	_
The Spires	Oxford	1 (5.6)	8 (44.4)	4 (22.2)	3 (16.7)	2 (11.1)	18
	Salisbury	3 (16.7)	6 (33.3)	3 (16.7)	6 (33.3)	0 (0.0)	18
South Wales & South West	Swansea	0 (0.0)	1 (12.5)	4 (50.0)	2 (25.0)	1 (12.5)	8
	Bristol	0 (0.0)	4 (30.8)	4 (30.8)	4 (30.8)	1 (7.7)	13
South Thames	Guy's and St Thomas'	4 (9.3)	16 (37.2)	15 (34.9)	5 (11.6)	3 (7.0)	43
Northern Ireland	Belfast	_	_	_	_	_	_
All	All	18 (7.5)	83 (34.7)	76 (31.8)	44 (18.4)	18 (7.5)	239

CRANE, 2004-2006 births

* 177/593(29.9%) children with an incomplete UCLP excluded; 7/423 (1.7%) children who died before the age of five are excluded; 177/416 (42.6%) children missing Five Year Old Index scores excluded; MCN, managed clinical network.

CRANE collected Five Year Old Index scores for children born between 2004 and 2006 with a complete UCLP. Seven (1.7%) children who died before their fifth birthday were excluded. Externally validated scores were provided for 239 (57.5%) eligible children by 12 of the 15 Administrative Units (*Table 12*).

Overall, 42.2% of complete UCLP patients born between 2004 and 2006 had Five Year Old Index scores in the two groups considered to have the best possible dental arch relationships (scores '1' or '2') while 25.9% of children had scores '4' or '5', reflecting poor dental arch relationships. This represents a significant (P=0.02) improvement compared to the CSAG findings that 36% (of 223 cleft children) had poor dental arch relationships at five years old in 1996 [5]. Comparisons between Units in five year old index scores are not appropriate because of the small number of children within each group.

3.6.5. Cleft Audit Protocol for Speech – Augmented scores (2006 births)

For the first time, we are reporting speech outcomes assessed at five years of age. The Cleft Audit Protocol for Speech – Augmented (CAPS-A) score has been used to assess speech among children with a cleft affecting the palate (CP, UCLP and BCLP). Several parameters of speech are assessed, including resonance (hypernasality and hyponasality) and nasal airflow (audible nasal emission and nasal turbulence), which reflect structurally-related speech, for example, the ability of the palate to close off the nasal airway during speech. Four cleft speech categories (anterior oral, posterior oral, non-oral and passive) are also assessed. These are summaries of the 12 cleft speech characteristics (CSCs), which reflect speech sound difficulties that can affect the clarity and intelligibility of a child's speech.

A total of 341 (48.4%) out of 704 consented children born with a CP, UCLP or BCLP in 2006 had at least one speech score reported. The scores for each assessed speech parameter can be seen in *Tables 13* and *14*. In *Table 13*, scores colour-coded as green indicate that the child's palate is functioning well in terms of the assessed parameter. No action, either speech therapy or surgery, would be required with green scores. Amber for hyponasality is indicative of nasal obstruction, while amber or red for hypernasality, nasal emission or nasal turbulence are indicative of structurally-related speech difficulties that may involve palate function and/or palatal fistulae. These difficulties may require surgical treatment.

In terms of resonance, 6.3% of children had moderate or severe hypernasality (nasal sounding speech) (*Table 13*), indicative of velopharyngeal insufficiency (VPI), when the palate is unable to close off the nasal airway during speech. Results of the cleft speech summary categories showed that 8.3% of children had passive articulation errors affecting three or

more consonants (*Table 14*), which are likely to be the consequence of VPI and is consistent with the hypernasality scorings. The most frequent nasal airflow error was nasal turbulence; however, this was mainly mild in nature, affecting less than 10% of speech in one fifth of the children.

Table 13. Number (%) of CRANE-registered consented children born in 2006 with a cleft affecting the palate, according to CAPS-A scores for resonance and nasal airflow

Description	Score	N	(%)
RESONANCE – HYPERNASALITY			
Absent	0	247	(74.8)
Borderline – minimal	1	40	(12.1)
Mild – evident on close vowels	2	22	(6.7)
Moderate – evident on open and close vowels	3	13	(3.9)
Severe – evident on vowels and voiced consonants	4	8	(2.4)
Total		330	(100.0)
RESONANCE – HYPONASALITY			
Absent	0	285	(84.1)
Mild – partial denasalization of nasal consonants and adjacent vowels	1	49	(14.5)
Marked – denasalization of nasal consonants and adjacent vowels	2	5	(1.5)
Total		339	(100.0)
NASAL AIRFLOW – AUDIBLE NASAL EMISSION			
Absent on pressure consonants	0	311	(91.5)
Occasional: pressure consonants affected <10% of the sample	1	23	(6.8)
Frequent: pressure consonants affected >10% of the sample	2	6	(1.8)
Total		340	(100.0)
NASAL AIRFLOW – NASAL TURBULENCE			
Absent on pressure consonants	0	258	(75.9)
Occasional: pressure consonants affected <10% of the sample	1	67	(19.7)
Frequent: pressure consonants affected >10% of the sample	2	15	(4.4)
Total		340	(100.0)

CRANE, 2006 births

11/715 (1.5%) children who died before the age of five are excluded; 374/704 (53.1%) children missing hypernasality scores excluded; 365/704 (51.9%) children missing hyponasality scores excluded; 364/704 (51.7%) children missing nasal airflow scores excluded.

Out of the 326 (46.5% of eligible children and 95.9% of those with at least one speech score reported) children with a reported score for each of the four parameters in *Table 13*, 271 (83.1%) had all green scores, indicating that no structural problems existed in relation to these parameters.

Table 14 presents the cleft speech summary categories. Children with just one or two affected consonants might require further monitoring and/or therapy, while those with three or more consonants affected might require therapy as well as further investigation of the palate and possible surgery.

Anterior oral CSCs were the most commonly occurring, affecting 49.1% of children; however, these may only have a minor effect on speech intelligibility, and, if treatment is indicated, this would involve speech therapy only and not further surgery. The more significant categories are posterior and passive errors, which are more likely to affect a child's intelligibility. Therapy would often be indicated for these children, and/or further investigation of structure and possible surgery.

Of the 323 (45.9% of eligible children and 94.7% of those with at least one speech score reported) children with a summary score for each of the four CSC categories, 175 (54.2%) did not exhibit features of CSCs[2].

Table 14. Number (%) of CRANE-registered consented children born in 2006 with a cleft affecting the palate, according to CAPS-A scores for cleft speech summary categories

	Cleft Speech Summary Categories N (%)			
Description/Score	Anterior Oral	Posterior Oral	Non-oral	Passive
Absent (A)	169 (50.9)	274 (82.8)	268 (81.2)	285 (87.7)
1 or 2 consonants affected (B)	65 (19.6)	28 (8.5)	27 (8.2)	13 (4.0)
3 or more consonants affected (C)	98 (29.5)	29 (8.8)	35 (10.6)	27 (8.3)
Total	332 (100.0)	331 (100.0)	330 (100.0)	325 (100.0)

CRANE, 2006 births

11/715 (1.5%) children who died before the age of five are excluded; 372/704 (52.8%) children missing Anterior oral scores excluded; 373/704 (53.0%) children missing Posterior oral scores excluded; 374/704 (53.1%) children missing Non-oral scores excluded; 379/704 (53.8%) children missing Passive scores excluded.

Overall, out of the 315 (44.7% of all eligible children and 92.4% of those with at least one speech score reported) children with scores across all eight assessed speech parameters, 153 (48.6%) had speech scores that would suggest their speech is within the normal range and not significantly different to their non-cleft peer group[3], while 23.2% of children received at least one score indicating a possible structural problem with the palate that may require

[2] Scores A or B for anterior oral and A for posterior oral, non-oral and passive categories.
[3] Scores 0 or 1 for hypernasality, hyponasality, audible nasal emission and nasal turbulence, and A or B for anterior oral and A for posterior oral, non-oral and passive categories.

further surgery[4]. It should be noted that these figures may be an underestimate and overestimate, respectively, because of limitations to the way that CSC data were collected for the first time this year. The method for collecting the CSC dataset will be discussed with the Lead Speech and Language Therapy group to improve our data for reporting this area of speech in the future.

[4] Scores 3 or 4 for hypernasality, 3 for audible nasal emission and nasal turbulence, and C for posterior oral, non-oral and passive categories.

4. Hospital Episode Statistics

In this section, we present data on children who have at least one HES record of an English NHS hospital admission with a diagnosis code for cleft lip and/or palate.

4.1. Mortality among children with a cleft lip and/or palate in England

Only a handful of studies have examined mortality in the first year of life among those with a cleft. A recent meta-analysis included nine studies from developed countries and found that children with a cleft had an infant mortality rate (IMR) nine times greater than the general population [17]. Even children without additional anomalies or syndromes were found to have an IMR twice as high as the general population. The studies included in the meta-analysis were mostly historical, with the largest study taking place between 1956 and 1965. Furthermore, only two UK studies were included, comprising a total of 760 cleft cases and 23 deaths. With medical advances, infant mortality in the general population has declined substantially in recent years; however, recent and current infant mortality levels among children with a cleft are unknown.

Our aim was to examine national neonatal (<28 days of life) and infant (<1 year of life) mortality rates among children with a cleft born in England between 2001 and 2010. We have presented mortality according to the absence and presence of additional anomalies or syndromes and by cleft type classification.

4.1.1. All cleft children

We identified 12,589 children, born between 1 January 2001 and 31 December 2010, who were diagnosed with a cleft lip and/or palate. Among these children, 388 died within the first year of life. This represents an infant mortality rate (IMR) of 30.8 (95% confidence interval [CI] 27.8 to 34.0) per 1,000 live births (*Table 15*). Early neonatal deaths (<7 days after birth) accounted for 28.6% of the deaths, while a further 20.1% occurred in the late neonatal period (7-27 days after birth). The remaining 51.6% of deaths occurred in the post-neonatal period, between 28 and 364 days after birth. A total of 10 (2.6%) children had undergone a primary cleft repair before their death.

4.1.2. Children with additional anomalies or syndromes

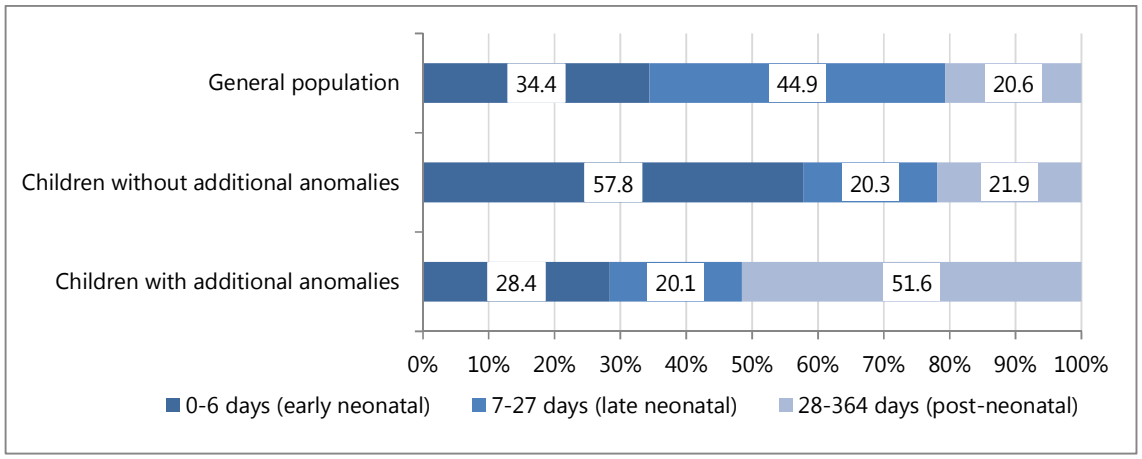
3,397 (27.0%) children were identified as having additional anomalies or syndromes. Of these, 324 died within the first year, representing 83.5% of all children who died. The IMR for cleft children with additional anomalies was 95.4 (95% CI 85.3 to 106.3) per 1,000 live births. In comparison, 64 out of the 9,192 children without additional anomalies died, corresponding to an IMR of 7.0 (95% CI 5.4 to 8.9) per 1,000 live births (*Table 15*).

Table 15. Deaths per 1,000 live births among children with a cleft, according to the absence or presence of additional anomalies and cleft type, 2001-2010 births

		Deaths per 1,000 live births (95% CI)		
Group	**Births**	0-27 days	28-364 days	0-364 days
Clefts without additional anomalies				
Cleft lip	2,420	0.8 (0.1 to 3.0)	0.8 (0.1 to 3.0)	1.7 (0.4 to 4.3)
Cleft palate	3,783	6.6 (4.3 to 9.8)	1.6 (0.6 to 3.5)	8.2 (5.6 to 11.6)
Cleft lip and palate	3,989	5.8 (3.7 to 8.7)	1.5 (0.5 to 3.3)	7.3 (4.9 to 10.4)
Total	9,192	5.4 (4.0 to 7.2)	1.5 (0.8 to 2.6)	7.0 (5.4 to 8.9)
Clefts with additional anomalies				
Cleft lip	281	53.4 (29.8 to 88.1)	42.7 (22.0 to 74.6)	96.1 (63.3 to 139.8)
Cleft palate	2,476	27.9 (21.7 to 35.3)	41.6 (34.0 to 50.4)	69.5 (59.5 to 80.7)
Cleft lip and palate	640	114.1 (89.4 to 143.4)	81.3 (60.7 to 106.5)	195.3 (162.6 to 232.7)
Total	3,397	46.2 (39.3 to 54.0)	49.2 (42.0 to 57.2)	95.4 (85.3 to 106.3)
All clefts	12,589	16.4 (14.3 to 18.8)	14.4 (12.4 to 16.6)	30.8 (27.8 to 34.0)
General population[a]	6,595,519	3.4 (3.3 to 3.4)	1.5 (1.5 to 1.6)	4.9 (4.8 to 4.9)

The age of death varied according to the absence or presence of additional anomalies. The greatest risk of death for children with a cleft alone occurred in the early neonatal period, with 57.8% of infant deaths occurring within the first six days of life (*Figure* 1). The corresponding rate among children with additional anomalies was 28.4%. A large difference between groups was also observed in post-neonatal deaths, which accounted for 20.6% and 51.6% of deaths in those without and those with additional anomalies, respectively. The proportion of late neonatal deaths was similar in both groups (*Figure 1*). A greater proportion of children with a cleft alone than children with additional anomalies underwent a primary cleft repair before dying (6.3% vs. 1.9%).

Figure 1. Age at infant death among children with a cleft, according to the absence or presence of additional anomalies



4.1.3. Cleft type classification

Among children with a cleft alone, those with a CL had the lowest neonatal and infant mortality rates, while children with CP and CLP had the highest rates. Conversely, among those with additional anomalies or syndromes, the lowest mortality rates were observed with CP. Children with CLP had the highest risk of mortality (*Table 15*).

The age of death did not differ substantially between cleft types among those with a cleft alone. Among those with additional anomalies, 59.8% of children with CP who died did so in the post-neonatal period. This compares to 44.4% among those with CL and 41.6% among those with CLP.

4.1.4. Summary

Compared with the general population IMR of 4.9 per 1,000 live births between 2001 and 2010 [18], this study found that infants with a cleft were 6.3 times more likely to die in the first year of life. There was a substantial difference in mortality rates depending on the absence or presence of additional anomalies or syndromes. Children with a cleft alone were 1.4 times more likely to die in the first year of life than the general population. The increased risk of death was restricted to the early neonatal period only. Children with a cleft and additional anomalies or syndromes were found to be 19.5 times more likely to die in the first year of life than the general population, and the increased risk of death was most pronounced in the post-neonatal period. It should be noted that it is possible that some of

the children with a cleft alone who died may have had undiagnosed or unreported additional anomalies.

It is important to emphasise that these findings are unlikely to have implications for cleft care, since the majority of deaths occurred prior to cleft surgery. CRANE is planning to extend this work in the future by examining the causes of deaths among children with a cleft.

5. Development of the CRANE Database and future directions

5.1. National Pupil Database

Currently little is understood about the impact of facial clefting on non-health outcomes such as educational achievement. This has been identified as a priority for cleft research from patient, carer and clinical perspectives [19].

The National Pupil Database (NPD), held by the Department for Education, holds a wide range of information about pupils who attend schools and colleges in England [20]. CRANE is seeking permission to link the CRANE Database to the National Pupil Database at the individual pupil level in order to describe the educational outcomes for a cohort of individuals born with a cleft lip and/or palate in England and to compare these outcomes with those of the non-cleft cohort. It is anticipated that this pilot project will provide evidence of the feasibility of linkage between CRANE and the NPD and inform future research using the linked data.

5.2. Quality Dashboard

In the current pilot for 2013/14, CRANE will provide Methods Consulting Limited, who has been tasked by NHS England with delivering the platform and analysis of the data for the dashboards, with data for three out of the 12 key performance indicators (KPIs) on the current draft of the agreed Quality Dashboard, developed by the Clinical Reference Group (CRG). CRANE will be able to provide data for a further two KPIs next year, once small changes to the data collection screens on the database have been made. It is envisaged that the number of KPIs collected through CRANE will increase once the data collection process has been reviewed following the pilot stage. These additional data items will require approval by the CDG prior to inclusion on the database.

5.3. Public Health England

CRANE has been involved in a scoping exercise across existing congenital anomaly registers and disease specific registers by Public Health England in 2013. This is to examine the

feasibility of developing a national congenital anomaly register that will capture all congenital anomalies in England. We await the decision from Public Health England but have expressed our intention to comply with any data submission requirements to this project.

5.4. Clinical Reference Group (CRG)

The inclusion of submitting data to CRANE as a requirement to the D07/S/a National Service Specification (Cleft Lip and or Palate Services including Non-cleft Velopharyngeal Dysfunction (VPD) (All Ages)), developed by the CRG, is a welcome advance in helping to improve the quality and completeness of data held in the CRANE database. CRANE is exploring ways to develop communication and links with cleft teams that should also help to improve data submission.

The CRANE team is committed to working with commissioners to make sure that its outputs are consistent with current and future commissioning requirements, which may in the future include areas such as performance reporting.

5.5. Future analyses

Surgical care for hearing

We are currently working on analyses using HES data to examine the trends and current delivery of surgical interventions for resolving otitis media with effusion in children with cleft lip and/or palate. We will assess whether changes in practice occurred after the publication in 2008 of NICE guidelines on this surgical procedure [21].

Mortality

We are currently using HES data linked with the ONS mortality dataset to examine neonatal and infant mortality rates among children with a cleft lip and/or palate. We are examining deaths according to the absence or presence of additional anomalies, cleft type and socio-economic deprivation. We hope to extend this work by investigating the causes of deaths occurring among children with clefts. This will require an application to HES requesting the causes of deaths occurring in the neonatal period.

Equity and treatment and outcomes

We plan to explore possible associations between socio-demographic factors (index of multiple deprivation and ethnicity) and burden of care and outcomes using linked CRANE-HES data.

Educational outcomes

If CRANE is successful in its application to access the National Pupil Database, we will work on linking this to CRANE and HES data. This linkage will offer huge potential for examining educational outcomes among children with a cleft lip and/or palate.

5.6. Collaboration

CRANE is collaborating with a number of individuals and organisations:

- Since the publication of our annual report last year, which highlighted the problem of late diagnosis of CP, the Royal College of Paediatrics and Child Health (RCPCH) has set up a working group to develop a best practice guide and an e-learning module on the palate examination in the neonate. The overall aim is to increase the proportion of timely detections of CP by promoting a visual technique of examination of the mouth and palate, supplemented by palpation where appropriate, as well as to aid clinical awareness by alerting all health care professionals responsible for the newborn examination to symptoms associated with cleft palate. CRANE has been invited to act as a stakeholder in this project, which is due to commence in November 2013.
- CRANE has agreed to share data with other registers affiliated with the British Isles Network of Congenital Anomaly Registers (BINOCAR), with the aim of improving the completeness of anomaly reporting.
- CRANE is currently collaborating with an ENT specialist on a paper focusing on the use of grommets among children with a cleft in England.
- CRANE will be involved with a multidisciplinary group from the CFSGBI evaluating previously collected national speech data to identify possible risk adjustment factors for the speech outcome data which could be utilised when reporting surgeon- or team-specific data in the future.
- The Healing Foundation Cleft Gene Bank and Cohort Study supported by the Vocational Training Charitable Trust (VTCT) called the Cleft Collective (www.cleftcollective.org.uk) will be the world's largest cleft lip and palate research programme, which is taking place in the UK. Up to 5,000 children and their families

are being recruited to the Birth Cohort Study hosted by the University of Bristol and many are being invited to take part in clinical trials and other studies coordinated by the Clinical Trials Unit, at the University of Manchester and the Royal Manchester Children's Hospital. The Centre for Appearance Research at the University of the West of England will be working on the psychological issues associated with cleft lip and palate and the support needed by families and children. We are currently working with the Cleft Collective team to establish whether CRANE could share data with this research project.

- We are also exploring ways to support feasibility studies conducted by The Healing Foundation Cleft and Craniofacial Clinical Research Centre supported by VTCT based at the University of Manchester

5.7. Outcome measures

Currently the outcome section of the Database is hampered by the lack of agreed measures which have been shown to be valid and reliable in assessing the outcome of cleft care.

Speech

The Cleft Audit Protocol for Speech—Augmented (CAPS-A) tool, a valid and reliable measure of speech outcome [22], has been piloted against agreed national speech standards derived by the Lead Speech and Language Therapy group in the last 24 months. Administrative Units are now submitting CAPS-A data at five years of age. Our original aim was to extend this to report speech data at 10 years of age, with agreement from the Lead Speech and Language Therapy Group. At a recent national workshop of the CFSGBI for a minimum dataset it was decided not to add 10 year data at this stage, partly due to the resources involved in consensus listening as part of submitting speech data for audit purposes. Consequently, 10-year speech data will not be collected by CRANE at this stage.

Hearing

The current draft of the Quality Dashboard aims to measure the process of hearing assessment by five years of age in children with a cleft. The aim of the CRANE Database would be to report outcomes for the children relevant to the care they receive, as outlined in our original contract with Specialist Commissioning. Therefore, our aim would be to assess the progress of the Quality Dashboard for hearing outcomes and work with ENT and audiologists with a cleft-related interest to identify valid and reliable outcome measures that could be agreed for the database.

Psychology

Psychological measures and patient/parent satisfaction should be explored. The CFSGBI have tasked the Psychology Special Interest Group (SIG) with reviewing potential tools or measures that could be adapted or developed for the cleft population. Discussions need to be on-going with the appropriate SIG. Outcomes should reflect the proposed measures within the Quality Dashboard, as long as these are long-term and are based on valid and reliable measures of outcome. Furthermore, not all Units have access to psychologists currently or they have limited access, which will hamper data collection.

5.8. CRANE Database meeting with users

CRANE organises meetings with representatives of the Administrative Units approximately once a year. The last meeting took place on 19 February 2013. Key points from the meeting are listed below.

Consent and NIGB approvals

- An updated version of the information leaflet will shortly be issued for use.
- CRANE will consider having the information leaflet and consent form translated into Welsh, Urdu and Punjabi, as some patients decline consent because they cannot read the information provided.
- CRANE will write to teams requesting a progress update with respect to the exercise to re-verify consent for patients registered between 2000 and 2006. It is important to complete this exercise as the NIGB approval that allows submission of outcomes data for existing registered patients where consent has not yet been verified will not continue indefinitely.

Data collection

- Teams are requested to submit notification data of new births including details of the cleft type within 2-weeks of birth.
- Teams are encouraged to submit any outstanding speech outcomes data (2006 birth year – 5-year outcomes). Teams will be contacted in due course with any proposed changes to speech data submission before data is due to be collected for the next cohort.

- Teams are reminded that additional 'audit users' can be set-up to assist with entry of outcomes data for their specialty.
- Teams are reminded that downloads of outcomes data are available from the system.

Quality Dashboard

- CRANE is working with the Methods team who is delivering the Quality Dashboard pilot on behalf of NHS England. We are aiming to have the information available centrally from the database, avoiding the need for separate data capture. To facilitate this, changes to the capture of 'first contact' information by the database will be made.

Press release / delayed diagnosis of cleft palate

- Following on from the press release at the time of last year's Annual Report publication, CRANE is working with CLAPA to send a pack to all maternity units highlighting the issue of delayed detection of cleft palates. The pack will hopefully include a spatula/tongue depresser and torch to guide maternity units about the proper visualisation of the palate in the newborn exam.

The next CRANE Database Users' meeting is scheduled for Spring 2014.

5.9. Publications and presentations related to the CRANE Database

Publications

The following papers have recently been published:

Fitzsimons, KJ, Copley, LP, Deacon, SA and van der Meulen, JH, Hospital care of children with a cleft in England. *Archives of Disease in Childhood*, 2013.

Fitzsimons, KJ, Copley, LP, Smallridge, JA, Clark, VJ, van der Meulen, JH and Deacon, SA, Hospital admissions for dental treatment among children with cleft lip and/or palate born between 1997 and 2003: an analysis of Hospital Episode Statistics in England. *International Journal of Paediatric Dentistry*, 2013.

The following papers are being prepared for peer review:

Fitzsimons KJ, Copley LP, van der Meulen JH and Deacon SA. Mortality among children with a cleft lip and/or palate in England, 2001-2010

Fitzsimons KJ, Panagamuwa C, Copley LP, van der Meulen JH and Deacon SA. Surgical management of otitis media with effusion in children with cleft lip and/or palate born in England between 1997 and 2005

Poster presentations

Copley L, Fitzsimons K, Deacon S, van der Meulen J. "Enhancing the potential of anomaly registers using linked Hospital Episode Statistics data" *Craniofacial Society of Great Britain and Ireland (CFSGBI) Annual Conference* (April 2013)

Copley L, Fitzsimons K, Deacon S, van der Meulen J. "Mortality among children born with a cleft lip and/or palate: An analysis of English Hospital Episode Statistics linked to Office for National Statistics mortality data" *Craniofacial Society of Great Britain and Ireland (CFSGBI) Annual Conference* (April 2013)

6. Conclusions

This Annual Report presents national-level data on children born with a cleft lip and/or palate in England, Wales and Northern Ireland.

A total of 1,127 children born with a cleft in 2012 had been registered on CRANE at the time of preparing this report. This represents the highest number of annual registrations CRANE has received since it was established in 2000. This high number should not be interpreted as an increase in clefting incidence, but, instead, it represents the improved function of the database as a national register of cleft births. The number of registered cleft births in 2012 equates to an incidence of approximately one in every 670 live births in England, Wales and Northern Ireland [23, 24].

Although children can now be registered with CRANE prior to obtaining parental consent, consent must still be obtained so that complete data, including outcomes, can be collected and reported by CRANE. The consent rate is very high among patients who have been through the consent process, which is encouraging. However, almost one fifth of the children born in 2012 had not been consented at the time of preparing this report. Further, this proportion ranged from 0% to 49% between Units. Units with a high proportion of unconsented patients are encouraged to review their consent-taking process, with the aim of obtaining consent in a timely fashion to enable the reporting of complete data.

The majority of Units collect all the data items requested by CRANE; however, the reporting of some data, in particular outcomes at five years of age, is variable between Units. A few Units have provided outcome data for more than 75% of their eligible patients, suggesting that the reporting of outcomes is feasible. CRANE is exploring ways to improve communication and links with Units to improve the submission of data in the future.

Collecting and reporting outcomes among children with a cleft is important for evaluating treatment, drawing comparisons between different groups of patients, providing information to patients and parents, and for planning future services. The inclusion of submitting data to CRANE as a requirement in the National Service Specification for cleft lip and/or palate services will improve the quality and completeness of data held in the CRANE database.

Based on the data reported to CRANE, we have highlighted some areas that should be addressed by maternity, paediatric, cleft and dental services to improve care and outcomes:

Diagnosis, Referral and Contact

1. Antenatal diagnosis rates of cleft lip, with or without cleft palate, are falling below the NHS Fetal Anomaly Screening Programme target detection rate of 75% [25].

2. One third of children with a cleft palate alone are being diagnosed late according to the national standard, which states that clefts should be diagnosed within 24 hours of birth to enable immediate referral to a specialist hospital [4]. This proportion has increased since the previous year.

3. Only half of the children born in 2012 with a cleft were referred to a Cleft Unit within 24 hours of birth. This proportion varied substantially according to the Unit receiving the referrals. Prompt referral is recommended to ensure that the baby and their family receive appropriate care and support as soon as possible. Once referred, Cleft Units established contact with the family within 24 hours, which is encouraging.

Cleft-related outcomes at five years

4. Children with a cleft are at increased risk of poor oral health. Children with a cleft affecting both the lip and palate are at the greatest risk of caries and may benefit from targeted preventive intervention.

5. One quarter of children with a complete UCLP have poor dental arch relationships that may benefit from further surgery to correct facial disproportion. While there is room for improvement, this proportion is significantly lower than the 36% of five year old children with a cleft who were reported by CSAG to have poor dental arch relationships in 1996.

6. Almost one quarter of children with a complete speech assessment received at least one score indicating a possible structural problem with the palate that may require further surgery.

As a result of our analyses of HES data linked with the Office for National Statistics Mortality dataset, we have been able to report national infant mortality rates among different groups of children with a cleft. Although these data have limited implications for clinical practice in terms of cleft care, as the majority (97.4%) of deaths occurred prior to cleft treatment, they should be of interest to those involved in the care of children with a cleft lip and/or palate. The key findings were:

- Children with a cleft alone have an infant mortality (<1 year of life) rate that is 1.4 times higher than the background rate. The increased risk of death appears to be restricted to the early neonatal (<7 days of life) period only. It is possible that some of the deaths

occurred among children with additional anomalies that were not diagnosed or reported.

- Children with a cleft and additional anomalies or syndromes have an infant mortality rate that is 19.5 times higher than the background rate. The increased risk of death is most pronounced in the post-neonatal (28-364 days of life) period.

Cleft Units should review the findings in this report and identify areas in which local improvements are required to help ensure the provision of high quality care for children with a cleft.

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Appendix 1: CRANE Project Team

Members of CRANE project team

Scott Deacon	Clinical Project Lead, Lead Consultant Orthodontist	Clinical Effectiveness Unit; South West Cleft Unit North Bristol NHS Trust; University of Bristol
Kate Fitzsimons	Research Fellow	Clinical Effectiveness Unit
Lynn Copley	Data Manager	Clinical Effectiveness Unit
Jan van der Meulen	Clinical Epidemiologist	Clinical Effectiveness Unit; London School of Hygiene and Tropical Medicine
Jackie Horrocks	CRANE Administrator	Clinical Effectiveness Unit

Appendix 2: Governance and funding

Ownership

It has been agreed that the "ownership" of the CRANE Database lies with the Craniofacial Society of Great Britain and Ireland (CFSGBI) as it represents the multidisciplinary group of professionals involved in the care of patients with a cleft lip and/or palate.

Cleft Development Group

The Cleft Development Group is a body with two distinct roles. Firstly, it is responsible for making arrangements for the running and commissioning of the CRANE Database.
Secondly, it is responsible for providing guidance on all aspects of the delivery of cleft care in England and Wales. It includes representatives from all the stakeholders in cleft care in England and Wales, including commissioners, public health consultants/regional cleft leads, specialists in the provision of cleft care, and parents and patients. It also has representatives from the health services in Wales, Scotland and Northern Ireland, as well as a representative from the Republic of Ireland cleft service.

Funding

Funding of the CRANE Database is currently coordinated and agreed by representatives of the national Specialised Commissioning Group for England and the Wales Specialised Health Services Committee. Funds are raised through a levy calculated on a weighted per capita basis from the commissioning bodies in England and Wales. The levy is currently collected by Derbyshire County PCT.

Appendix 3: Members of the Cleft Development Group

Members of Cleft Development Group

Stephen Robinson	Chair/Orthodontics (SIG CFSGBI)
Liz Albery	Leads Group of the Speech and Language Therapy SIG CFSGBI
Geoffrey Carroll	Medical Director, Wales Health Specialised Services Committee
Michelle Collard	Paediatric Dentistry (Special Interest Group (SIG) CFSGBI)
Scott Deacon	CRANE Clinical Project Lead
Mark Devlin	Scotland Clinicians
David Drake	Cleft Surgery Interface Committee
Mandy Elder	East of England Specialised Commissioning Group
Sue Gregory	Department of Health (Dept. CDO England)
Per Hall	Cleft Surgeon (British Association of Plastic, Reconstructive and Aesthetic Surgeons (BAPRAS))
Chris Hill	Northern Ireland Clinicians
Nichola Hudson	Specialist Cleft Nurses (SIG CFSGBI)
David Landes	North of England Dental Public Health Consultant
Fiona Mackison	South East Coast SCG
Fiona Marley	National Specialised Commissioning Group
Kate Le Marechal	Clinical Psychologists (SIG CFSGBI)
Jan van der Meulen	Clinical Epidemiologist
David Orr	Ireland Clinicians
Alison Sims	Cleft Co-ordinators and Managers (SIG)
Rona Slator	President of the CFSGBI
Adrian Sugar	Wales Clinicians
Peter Hodgkinson	Chair, Cleft Centres Clinical Directors/Managers Group;
Alistair Smyth	Cleft Surgeon (British Association of Oral and Maxillofacial Surgeons)
Mike Winter	Medical Director, National Services Division, Scotland
Ken Wragg	East Midlands Dental Public Health Consultant
Christopher Allen	Deputy for Ken Wragg and David Landes
Jackie Horrocks	Minutes Secretary, CRANE/Clinical Effectiveness Unit

Appendix 4: Terms of Reference for the Cleft Development Group

The Origins of the Cleft Development Group (CDG)

The NHS Cleft Development Group was formed in November 2004 out of the previous CRANE/Cleft Levy Board, the CRANE Management Group and their Advisory bodies. These groups and bodies had been responsible for the national cleft database, CARE and then CRANE. The implementation of the DoH's guidance regarding the re-organisation of cleft services in the UK which stemmed from the DoH Clinical Standards Advisory Group report into the care of patients with Clefts of the Lip and/or Palate (1998) was the responsibility of the Cleft Implementation Group (CIG). When this group was terminated by the DoH, a new body took over its role, the Cleft Monitoring Group. When that body was terminated, the Cleft Development Group (CDG) was asked to take over its role too.

The Roles of the CDG

The CDG has two distinct roles which arise from its origins.

1. The CDG is responsible for guidance on all aspects of the delivery of re-organised cleft care in England and Wales and, when asked, by Scotland and Northern Ireland. It gives advice to the cleft centres, to health authorities, trusts, boards, commissioning groups and consortia and to the Departments of Health in England and the devolved administrations. It represents all stakeholders in cleft care and works with all to ensure the highest quality of cleft care in the UK to all patients who need it. It inherits the responsibilities of the Cleft Implementation Group and the Cleft Monitoring Group which were largely advisory.

2. The CDG is responsible for the commissioning of, the strategic governance of and is ultimately responsible for the national cleft database which used to be called CARE and is now called CRANE. It must negotiate and agree a contract for the running of CRANE and have operational oversight of the implementation of that contract. It is responsible for funding of the CRANE Register and is responsible for ensuring that the agreed levy is collected annually through the NHS Specialist Commissioners. It will approve an annual budget and business plan for CRANE drawn up with the contract holders and will review income and expenditure and ensure that the terms of reference are implemented. It will determine the location of the register and will appoint the Clinical Director/Project Leader who will be accountable to the Group.

   The CDG's responsibility stems from Health Services Circular 1998/238 which states that "A CARE Register, with which all patients should be registered, will be maintained by the Craniofacial Society of Great Britain – this will form the basis for national audit".

   The database was UK wide when run by the Craniofacial Society of Great Britain and Ireland and before it became the responsibility of the CRANE Levy Board. Devolution of government in the UK resulted in 4 distinct health services and as a result CDG came to be responsible for a national database for the recording of all children with clefts of the lip and/or palate born and treated in England and Wales, as the health service in Wales indicated its support for this development at an early stage. It has since then successfully sought to include in its work strong relationships also with the cleft services in Scotland, Northern Ireland and the Irish Republic.

   The CDG is responsible for providing data for cleft births and cleft treatment for England and Wales and it also endeavours, with the cooperation of the health services in Scotland and Northern Ireland, to do so for the whole of the UK.

The national CRANE database has two primary functions:-

a. the recording of all birth, demographic and epidemiological data related to children born in England and Wales with the congenital abnormality of clefting of the lip and/or palate, and where possible extending this to the whole of the UK and Ireland
b. the recording of all treatment of children and adults in England and Wales with clefts of the lip and/or palate and the outcome of such treatment, and where possible extending this to the whole of the UK and Ireland

The data from (a) will provide the same kind of information as other congenital anomaly registers and will be the basis for reports, audit and research in that area. The data from (b) will provide the basis for national cleft audit which is intended to be a major and integral role of CRANE.

The relationships between the bodies involved in the national cleft database, CRANE, are defined by a Tripartite Agreement (2007) between the Cleft Development Group, the NHS Specialist Commissioners and the Craniofacial Society of Great Britain and Ireland.

Composition of the Cleft Development Group

The composition of the Cleft Development Group should reflect all stakeholders involved in cleft care. Consequently its composition (and consequently these Terms of Reference) will need to be changed from time to time. The Members of the Cleft Development Group will normally and primarily be active clinical members of a designated Cleft Team, public health consultants, commissioners of cleft care and representatives of parent/patient organisations. Membership of the **Group** will be for a term of three years which can be extended at the behest of the nominating organisation, except for members ex-officio who will be members during their terms of that office whether it be less or more than 3 years. The **Group** will elect its own Chair, who will remain in office for 3 years. The Group will also elect a Vice Chair. Either the Chair or the Vice Chair should be a Specialist Commissioner. The Group may decide to re-elect the holders of these offices.
The composition will be:

1. Commissioners of Cleft Care. These should include at least **two** commissioners from Specialist Commissioning Groups in England *(nominated by the National Specialist Commissioning Group for England)*, **one** from Wales, **one** from Scotland and **one** from Northern Ireland *(each nominated by their equivalent national specialist commissioning body)*. It is intended that there should be no more than six specialist commissioners in total to be agreed and appointed by the bodies which contribute data to the database (in the case of Scotland by sharing its data with CDG). Only those commissioning groups which pay the levy may vote on issues relating to CRANE.
2. Public Health Consultants. These should include representatives of commissioning areas who are actively involved in cleft commissioning, and will normally be Consultants in Dental Public Health. There should be at least **two** *(to be nominated by the BASCD Consultants in Dental Public Health Group)*.
3. A Lay representative from a Parent Support Group (1) *(to be nominated by CLAPA)*
4. Cleft surgeons (2) *(presently one nominated by BAOMS and one by BAPRAS)*
5. The President of the Craniofacial Society of Great Britain and Ireland
6. The Chair of the Cleft Surgery Training Interface Group
7. A Speech & language therapist (1) *(to be nominated by the Lead Cleft Speech and Language Therapy Group)*
8. An Orthodontist (1) *(to be nominated by the Cleft Orthodontists Special Interest Group)*.
9. A Specialist Cleft nurse (1) (*to be nominated by the Cleft Nurses Special Interest Group)*
10. A Psychologist (1) *(to be nominated by the Cleft Psychologists Special Interest Group)*
11. A Paediatric Dentist (1) *(to be nominated by the Cleft Paediatric Dentist Special Interest Group)*

12. The Co-ordinator/Chair of the UK Cleft Centres Clinical Directors' Group (1)
13. A Cleft Co-ordinator (1) (*to be nominated by the Cleft Coordinators Special Interest Group).*
14. A Representative from the group of 'other' specialities involved in cleft care (1) *(to be nominated by CFS Council).*
15. A Clinical representative from Northern Ireland (1) / Scotland (1) / Wales (1) / England (as appropriate, if not already represented) *(to be nominated by those countries)*
16. There may be representation, as determined by CDG to be appropriate, of any national bodies representative of Audit (1) and Research (1)
17. Clinical Directors/Clinical Leads of UK Cleft Centres not otherwise represented on CDG shall be invited to attend and become voting members so that all centres will be represented.
18. The Clinical Director/Project Leader of the CRANE service will be in attendance at Group meetings to which he/she will report, except when required to be absent because their own position is being discussed/decided. This individual will not be a voting member of the Group unless in another capacity and will not be eligible to become Chair.
19. The Director of the body which holds the contract for CRANE will be in attendance at Group meetings to which he/she will report, except when required to be absent because their own position is being discussed/decided. The Director will not be a voting member of the Board and will not be eligible to become the Chair.
20. A representative of the DH will always be invited to meetings and will receive minutes but will not be a voting member of the Board and will not be eligible to become the Chair.
21. Such other people who from time to time would serve the interests of the Cleft Development Group may be co-opted for a period of one year at a time.

Deputies for members may be appointed from time to time provided they are done so formally in writing by the nominating body to the CDG Chair. Where an individual comes to represent two positions on CDG, that person will continue to fulfil those roles and no additional person will be elected.
Additional representation will be considered (e.g. cleft paediatricians, cleft anaesthetists, cleft ENT and Audiology, cleft genetics) as and when those disciplines have formally established national special interest groups which genuinely represent those disciplines.

Meetings

Meetings will normally be held three times per year but must be held at least twice yearly with administrative support provided by the body which holds the CRANE contract, or the DoH or NHS bodies.

Amended May 2012

Appendix 5: Diagnosis and procedure codes, Hospital Episode Statistics

International classification of Disease 10th Revision (ICD-10) diagnostic codes for cleft lip and/or palate.

Code	Description
Q35	Cleft palate
Q36	Cleft lip
Q37	Cleft palate with cleft lip

Classification of Surgical Operations and Procedures 4th Revision (OPCS-4) procedure codes for cleft lip and cleft palate repairs

Code	Description
F031	Correction of deformity to lip
F291	Correction of deformity to palate

Classification of Surgical Operations and Procedures 4th Revision (OPCS-4) codes used to define a 'cleft-related procedure' admission for patients with a cleft diagnosis and history of a primary surgical cleft repair

OPCS 3-char code	Description
Chapter D:	**Ear**
D14	Repair of eardrum
D15	Drainage of middle ear
D20	Other operations on middle ear
Chapter E:	**Respiratory tract**
E02	Plastic operations on nose
E03	Plastic operations on nose
E07	Other plastic operations on nose
E08	Other operations on internal nose
E09	Operations on external nose
E10	Other operations on nose
E21	Repair of pharynx
Chapter F:	**Mouth**
F01	Partial excision of lip
F02	Extirpation of lesion of lip
F03	Correction of deformity of lip
F04	Other reconstruction of lip
F05	Other repair of lip
F06	Other operations on lip
F09	Surgical removal of tooth
F10	Simple extraction of tooth
F11	Preprosthetic oral surgery
F14	Orthodontic operations
F29	Correction of deformity of palate
F30	Other repair of palate
F32	Other operations on palate
F42	Other operations on mouth

International classification of Disease 10th Revision (ICD-10) diagnostic codes for syndromes and anomalies used to identify 'syndromic' cleft patients. Patients were defined as 'syndromic' if there was a record of any of the following codes in any of the fourteen diagnosis code fields for any of that patient's HES episodes.

Code	Description
D821	Di George's syndrome
	Congenital malformations of the nervous system (Q00-Q07)
Q00	Anencephaly and similar malformations
Q01	Encephalocele
Q02	Microcephaly
Q03	Congenital hydrocephalus
Q04	Other congenital malformations of brain
Q05	Spina bifida
Q06	Other congenital malformations of spinal cord
Q07	Other congenital malformations of nervous system
Q16	Congenital malformations of ear causing impairment of hearing
Q18	Other congenital malformations of face and neck
	Congenital malformations of the circulatory system (Q20-Q28)
Q20	Congenital malformations of cardiac chambers and connections
Q21	Congenital malformations of cardiac septa
Q22	Congenital malformations of pulmonary and tricuspid valves
Q23	Congenital malformations of aortic and mitral valves
Q24	Other congenital malformations of heart
Q25	Congenital malformations of great arteries
Q26	Congenital malformations of great veins
Q27	Other congenital malformations of peripheral vascular system
Q28	Other congenital malformations of circulatory system
Q380	Congenital malformations of lips, not elsewhere classified
Q75	Other congenital malformations of skull and face bones
Q86	Congenital malformation syndromes due to known exogenous causes, not elsewhere classified
Q87	Other specified congenital malformation syndromes affecting multiple systems
	Chromosomal abnormalities, not elsewhere classified (Q90-99)
Q90	Down's syndrome
Q91	Edwards' syndrome and Patau's syndrome
Q92	Other trisomies and partial trisomies of the autosomes, not elsewhere classified
Q93	Monosomies and deletions from the autosomes, not elsewhere classified
Q95	Balanced rearrangements and structural markers, not elsewhere classified
Q96	Turner's syndrome
Q97	Other sex chromosome abnormalities, female phenotype, not elsewhere classified
Q98	Other sex chromosome abnormalities, male phenotype, not elsewhere classified
Q99	Other chromosome abnormalities, not elsewhere classified