CRANE Database

www.crane-database.org.uk

Annual Report on Cleft Lip and/or Palate 2017

On behalf of the Cleft Development Group

Royal College of Surgeons

ADVANCING SURGICAL CARE

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[1] Appendices 1 through 4 provide detail on the project team, the CDG Membership and Terms of Reference, and the project's Governance and funding arrangements.

Abbreviations

BCLP	Bilateral cleft lip and palate
CAPS-A	Cleft Audit Protocol for Speech—Augmented
CDG	Cleft Development Group
CEN	Clinical Excellence Network – previously referred to as Special Interest Group (SIG)
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
DfE	Department for Education
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
NPD	National Pupil Database
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
UCLP	Unilateral cleft lip and plate
VTCT	Vocational Training Charitable Trust
WHO	World Health Organization

Glossary

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.
Alveolus / alveolar	The part of the jaw that supports the teeth and contains the tooth sockets.
Caries (dental)	Dental caries are also known as tooth decay / dental decay or a cavity.
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 a region / 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.
Confidentiality Advisory Group (CAG)	An independent statutory body established to promote, improve and monitor information governance in health and adult social care. http://www.hra.nhs.uk/research-community/applying-for-approvals/confidentiality-advisory-group-cag/
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
Funnel Plot	A graph that identifies regions / units which are outliers, where the local situation might require closer inspection – either because an area is doing well or because there is some indication that it is performing poorly. In this report: • Each point on the funnel plot represents a region / unit. • Each funnel plot is for one outcome, with its values shown on the vertical/Y axis. • The size of the regions' /units' cohort is shown on the horizontal or X axis. • The benchmark value is shown as a horizontal line through the centre of the graph. The graph shows two funnels that lie on either side of the benchmark and are called the control limits – similar to confidence intervals. • The inner lines show 2 standard deviations or 95% control limits. The outer lines represent 3 standard deviations or 99.8% control limits. • The funnel shape is formed because the control limits get narrower as the population size increases. The outer funnel is used to decide if an area is significantly different to the benchmark with 99.8% confidence. If a point lies within the funnel then we conclude that it is not significantly different to the benchmark. If it falls outside the funnel then we can say the value is significantly 'better' or significantly 'worse' than the benchmark, depending on the direction of the indicator/outcome.

	Funnel Plot Source: David Spiegelhalter, Medical Research Council Biostatistics Unit - http://www.erpho.org.uk/Download/Public/6990/1/INPHO%204%20Quantifying%20performance.pdf
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 Pupil Database (NPD)	A database containing records on all pupils in England as they progress through primary and secondary education.
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

Annual Report 2017

CLEFT REGISTRY & AUDIT NETWORK

CRANE aims to audit, evaluate and report on the delivery of cleft services to children in England, Wales and Northern Ireland with the congenital abnormality of cleft lip and/or palate

17,840 children registered over the last seventeen years

Approximately 1050 new cases are registered in CRANE per year

2016 births

28.2% of children with a Cleft palate (CP) received a delayed diagnosis (after birth)

Key Findings

5-year olds

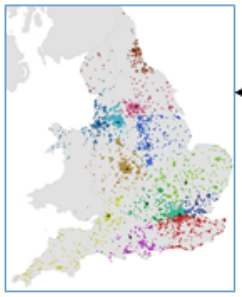
41% of children had at least one decayed, missing or filled tooth (>0 dmft), which is slightly higher than the rate in the general population (38.8%).

p > 0.05 No significant relationship found between children's facial growth and speech outcomes – for cases with UCLP

60% of children had speech scores that would suggest their speech was not significantly different from their non-cleft peers

All children treated in England

Most patients underwent their first surgical procedure in the surgical centre that was designated to provide cleft surgery for their region.



There are large differences in the distance that cleft patients and their families had to travel for their first surgical repair.

Recommendations

Clinical care

- Late diagnosis of cleft palate (CP) remains an important issue that must be addressed
- Increased preventative dental support for children with a cleft seems essential to reduce dental decay, particularly among those at greatest risk of caries

Outcome measures and reporting to CRANE

- Units' data submission to CRANE must improve, to allow the expansion of analyses and reporting of cleft outcomes
- Further work is required to explore the educational differences previously highlighted by our reports

Publications and future directions

Kate J Fitzsimons, Lynn P Copley, Efrosini Setakis, Susan C Charman, Scott A Deacon, Lorraine Dearden, Jan H van der Meulen. Early academic achievement in children with isolated clefts: a population-based study in England Archives of Disease in Childhood. **Published Online First: 02 November 2017.** doi: 10.1136/archdischild-2017-313777

Plan for 2018

Start work with our stakeholders to agree further outcomes for older children and young adults

1. Introduction

The Cleft Registry & Audit Network (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[2]. The geographical representation of the cleft regions / units is detailed in Appendix 5.

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 aims of the CRANE Database are:

1. 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; and
2. to record the treatment of children and adults with a cleft lip and/or palate and the outcome of such treatment.

This Annual Report presents findings from data submitted to CRANE[3] for children with a cleft lip and/or palate born in England, Wales and Northern Ireland between the 1 January 2000 and 31 December 2016. We describe:

- Analyses of data from Hospital Episode Statistic (HES), containing records on all admissions to NHS Hospitals in England, to report on the location of primary surgery in relation to place of residence;
- the proportion of babies born in 2016, and registered in the CRANE Database, who were diagnosed at birth, referred within 24 hours of birth, and contacted within 24 hours of referral;
- analyses of data from HES data linked to the CRANE Database at the individual level for consented children born from 2000 to 2012. We describe the results exploring factors impacting on diagnosis times among children with cleft palate alone;
- cleft-related outcomes at five years of age for children, registered in the CRANE Database, at five years of age (born 2004-2011).

This Annual Report aims to provide feedback to all stakeholders involved in cleft care, highlighting areas of success and areas requiring improvement in future reporting and in clinical practice.

[2] For further information on the background to the CRANE database please visit https://www.crane-database.org.uk/
[3] Registered in the CRANE Database by the 2 October 2017.

2. Methods

This report contains information on patterns of care and outcomes derived from two sources of data. These sources are (1) the CRANE Database, and (2) CRANE Database data linked to Hospital Episode Statistics (HES) data.

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 units that make up 10 Regional Cleft Centres / Managed Clinical Networks (as listed in Appendix 5). 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 units. Additional information, including 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 usually obtained by units 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 2 October 2017 pertaining to children born between 1 January 2000 and 31 December 2016 is included in the descriptions and analyses described in this Annual report. Patients whose parents have not consented to their data being used by CRANE have been excluded from the sections and tables in this report on five-year outcomes (as the data presented in these sections and tables are not collected for non-consenting cases).

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 have been defined according to growth charts published by the World Health Organisation (WHO)[4].

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 2011, 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 (10.2% of children born in 2016), cleft type is based on the type reported by the region/ 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 dental caries experience of 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 2010 (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[5]. CRANE collected both internal and external Five Year Old Index scores for consented children born between 2004 and 2010 with a complete UCLP (LAHSAL codes LAHS or HSAL). Some units 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

[4] World Health Organization. The WHO Child Growth Standards 2011. Available from: http://www.who.int/childgrowth/standards/en/.

[5] Johnson N, Williams AC, Singer S, Southall P, Atack N and Sandy JR. Dentoalveolar relations in children born with a unilateral cleft lip and palate (UCLP) in Western Australia. The Cleft Palate-Craniofacial Journal, 2000. 37 (1): p. 12-16.

analysed externally validated scores where available, where these were unavailable internal scores are included in the analysis.

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

CAPS-A scores collected at five years of age among children born between 2007 and 2010 were reported to CRANE for consented children only. The parameters of speech assessed include resonance (hypernasality and hyponasality), nasal airflow (audible nasal emission and nasal turbulence) and twelve Cleft Speech Characteristics (CSCs) scores[6] – including:

- Anterior oral CSCs – for dentalisation/interdentalisation, lateralisation/lateral, and palatalisation / Palatal characteristics;
- posterior oral CSCs – for double articulation and backed to velar/uvular characteristics;
- non-oral CSCs – for pharyngeal articulation, glottal articulation, active nasal fricatives, and double articulation characteristics; and
- passive CSCs – for weak and or nasalised consonants, nasal realisation of plosives, and gliding of fricatives.

Missing data

Missing data have been excluded from the denominators presented in all Tables and Appendices of this report. 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)

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.

The HES database 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. In addition to being able to identify and confirm cleft type in the CRANE Dataset, HES is used by CRANE to identify any additional anomalies for the CRANE cohort (see Appendix 6 for a list of the HES diagnosis and procedure codes used by CRANE).

[6] 2006 data is not included as only four Cleft Speech Categories (CSCs) summarising current 12 CSCs were collected for 2006 birth.

3. Mapping boundaries of Regional Cleft Networks in England

3.1. Background

The CSAG report recommended in 1998 that cleft services should be centralised into eight to 15 regional specialist cleft centres (hospitals providing cleft surgery) with surgeons performing repairs on at least 40 to 50 new cases per year and each hospital treating 100 to 120 new cases per year.[7]

The CRANE Database has demonstrated that since the publication of the recommendations to centralise cleft services in 1998, the number of NHS hospitals involved in providing primary cleft surgery in England has reduced from more than 40 to 13 specialist cleft centres in 2008.[8]

In this section, we briefly describe the impact of the current regional configuration of cleft services on the mobility pattern and distance between the residential addresses and the centres that provided the first primary surgical cleft repair among patients born between 2010 and 2014 in England.

3.2. Methods

We mapped the centroids[9] of the lower-layer super output area (LSOA) of the patients' residential addresss for the 5882 children that were identified in the Hospital Episode Statistics (HES) dataset as born with an oral cleft between 1 April 2007 and 31 March 2012 and having undergone major surgical cleft repair in England. LSOAs are small regions that include on average 1,500 people. The LSOA for 32 patients (0.5%) could not be mapped, because nine patients did not have a home LSOA recorded and a further 23 were from Scotland, Northern Ireland, the Isle of Man or the Channel Islands, which do not have an equivalent LSOA geography to England and Wales. As a result, the residential address of 5850 patients could be determined.

The specialist cleft centres were mapped according to postcode of the hospital in which they were located. We estimated the shortest road network distance in miles from each patient's residential address and the cleft centre in which they received their first primary surgical cleft repair.

A map of the geographical distribution of the addressed patients was drawn both for each individual cleft centre. On this map, one point represents a single patient. Each patient is randomly placed

[7] Clinical Standards Advisory Group. Clinical Standards Advisory Group: Report of a CSAG Committee on Cleft lip and/or palate. London: The Stationery Office; 1998.
[8] Fitzsimons KJ, Mukarram S, Copley LP, Deacon SC, Van der Meulen JH: Centralisation of services for children with cleft lip or patlate in England a study of hospital episode statistics. BMC Health Services Research 2012, 12:148.
[9] A centroid is the population-weighted "average coordinate" of the home addresses of the people leaving within an LSOA.

within their home LSOA in order that multiple patients resident in the same LSOA can be seen. This random placement within LSOA approach obscures the actual location of their residential address.

3.3. Results

The average distance between the patients' residential address and the hospital where they had their first surgical repair is 26.8 miles (median 20.6, 75th percentile % 35.8 and 90th percentile 54.5 miles).

In Figure 1 we present the residential address of all cleft patients born between 2010 and 2014 in England and Wales[10] and the location of the English surgical centres where they had their first major cleft repair, if undertaken in England rather than Wales, as is the model of delivery for patients born in North Wales. This figure shows that the majority of patients had surgical treatment in the surgical centre that was designated to provide primary surgical cleft surgery for the region they were living in. There is some "mix" of patients in the London area with some patients resident in the regions that are predominantly serviced by the Chelmsford and the Guy's and St Thomas' Trust having their treatment in Great Ormond Street Hospital.

It is also clear that the distance between the patients' residential address and the surgical centre that provided treatment varied across the country (Table 1). Mean distances for patients who had their first surgical repair in Manchester (12.5 miles) or North London (Great Ormond St; 14.9 miles) were considerably shorter than for those treated in Cambridge (41.4 miles) or Bristol (62.3 miles). The distribution of the travel distances is notably skewed for patients who had their first surgical treatment in Bristol with 25% having to travel at least 104.8 miles and 10% at least 144.1 miles.

[10] We are able to include information of patients from North Wales who were treated in England in the mapping work described in this chapter, because their information was captured in the Hospital Episode Statistics (HES) dataset. Unfortunately, we were unable to include information on patients from South Wales as we did not have access to Patient Episode Database for Wales (PEDW) data.

Figure 1. Map of the residential address of 5850 children that were born with a cleft between 2007 and 2012 in England and the location of the 13 specialist cleft centres (indicated with +) that carry out the first primary surgical repair. The colours differentiate the patients who were treated in each centre.

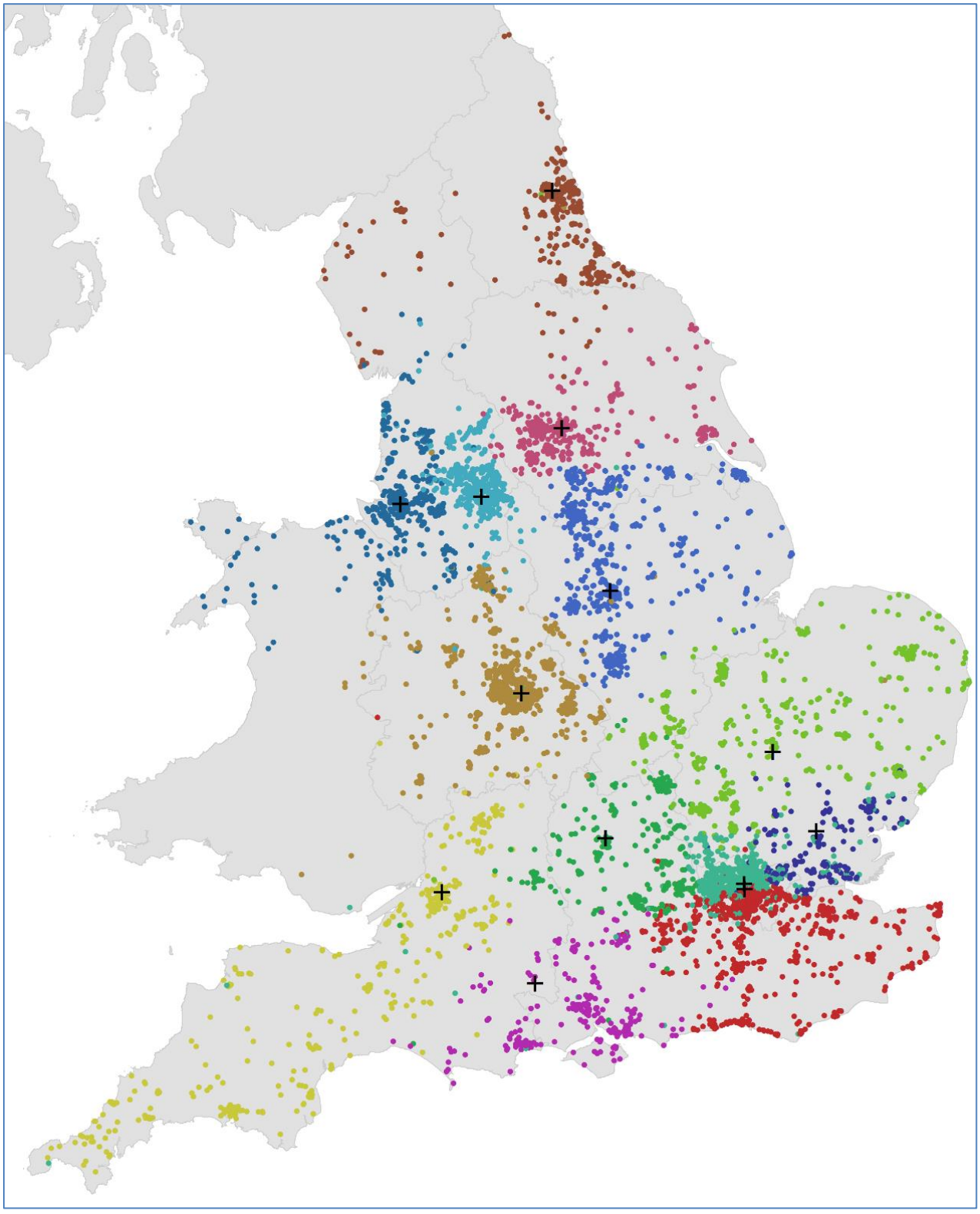


Table 1. Travel distance (miles) between the patients' residential address and the surgical centre that provided their first surgical repair for 5832 children born with a cleft between 2007 and 2012 in England. [§]

Regional Cleft Centre	Specialist Surgical Centre	Patients **N**	Mean distance (miles)	50th percentile	25th percentile	75th percentile	90th percentile
Northern &	Newcastle	346	27.4	22.3	8.9	37.0	57.5
Yorkshire	Leeds	384	18.5	12.7	8.2	21.4	53.9
North West &	Liverpool	434	23.1	16.0	7.5	33.8	48.8
North Wales	Manchester	440	12.5	9.7	6.2	15.6	26.1
Trent	Nottingham	545	30.6	29.2	17.8	39.9	54.0
West Midlands	Birmingham	664	20.5	15.5	7.2	29.2	44.7
East of England	Cambridge	445	41.1	39.3	28.3	52.0	64.9
North Thames	Grt Ormond St	658	14.9	9.2	5.7	14.6	25.4
	Chelmsford	217	20.7	21.2	16.3	25.1	30.9
The Spires	Oxford	306	27.1	29.6	20.9	33.8	37.4
	Salisbury	267	31.4	27.0	23.1	40.2	43.7
South West[¥]	Bristol	393	62.3	40.3	20.8	104.8	144.1
South Thames	GSTT	733	27.3	23.2	10.1	39.4	56.5

[§] 18 patients from the Isle of Wight were excluded; the road network distance could not be determined because they had to travel over water.
[¥] Patients from South Wales were not included; we did not have access to their information captured in Patient Episode Database for Wales (PEDW) data.

3.4. Summary

The mobility pattern for children born with a cleft in England follows closely the regional configuration of cleft services that was completed in 2008. Most patients were treated in the surgical centre that was designated to provide cleft surgery for their region. The boundaries in the London region are less strictly followed than elsewhere. There are large differences in the distance that cleft patients and their families have to travel for their first surgical repair. Distances tend to be the longest for patients who had their first treatment in Bristol, serving the South-West of England or Cambridge, serving the East of England.

3.5. Further work

In 2018, the CRANE Project Team will carry out further work, aiming to answer more detailed questions, including an analysis of the impact of travel distance on the timing of treatment, an assessment of the extent to which overall travel time can be reduced by changing regional boundaries (i.e. location allocation analysis), and an exploration of the factors that have an impact on whether or not patients travel to other centres than the one serving their own region.

4. CRANE

In this chapter, we present findings on children with a cleft lip and/or palate, born between 1 January 2000 and 31 December 2016 in England, Wales and Northern Ireland. These data have been analysed to assess registration patterns, the timing of diagnosis, referral and contact with units around the time of birth, and cleft-related outcomes at five years of age.

4.1. Registrations and contact with cleft teams

Of the total 17,840 children born and registered in the CRANE Database over the last seventeen years[11], **1,056 children have been registered in 2016.** Of these 1,056 children:

- Cleft palate (CP) continues to be the most common of the four cleft types[12], comprising 39.4% of 2016 registrations.
- Bilateral cleft lip and palate (BCLP) is the least common type with 9% of 2016 registrations; with Cleft Lip (CL) registrations at 21.6%.
- 18.6% of 2016 registrations had unilateral cleft lip and palate (UCLP) of which 71.4% had complete UCLP (defined by either 'LAHS..' or '..HSAL' LAHSAL codes).
- 11.4% of registrations did not have their type of cleft specified (either by LAHSAL codes or by the units).
- Visit the CRANE database website https://www.crane-database.org.uk/ to review the Tables on registrations over the last 10 years, by **cleft type** and **year of birth**, according to region / unit.

With regards to families being referred to cleft teams:

- Overall in 2016, 36.3% of children were missing data on referral time.
- Of the 673 children with a reported referral time, 79.5% were referred to a Cleft Unit within 24 hours of birth.
- The proportion of referrals within 24 hours of birth varied significantly according to cleft type ($p<0.001$), with CP patients having the lowest proportion; consistent with later diagnosis times for these children.
- The proportion of referrals within 24 hours of birth also varied significantly according to cleft /administrative unit ($p<0.001$) Despite this statistically significant variation between units, overall rates of referral within 24 hours remain high (as for previous reporting years).
- The patterns of referral according to time of diagnosis were consistent with patterns described in past years[13].

[11] 1 January 2000 and 31 December 2016.
[12] Cleft type is defined according to reported LAHSAL codes or, where LAHSAL has not been reported (for 10.2% of children registered in 2016), it is based on the cleft type reported by the region / unit registering the child.
[13] For past CRANE Database Annual Reports please visit https://www.crane-database.org.uk

With regards to families first being contacted by cleft teams:

- Overall in 2016, 18.9% were missing the first contact time between units.
- Of the 856 children with a reported contact time, units established contact with 96.9% of parents within 24 hours of referral.
- Patterns of variation in referrals according to **cleft type** and the **cleft /administrative unit** did not vary significantly ($p>0.05$), and were consistent with patterns described in past years[14].
- All units contacted at least 93.3% of their patients within 24 hours of receiving the referral. This demonstrates the commitment of units to ensure a timely response to new referrals of babies born with a cleft, to help support these babies and their families in the important initial stages.

With regards to families being approached for consent to collect data into childhood (beyond registration and diagnosis):

- The parents/carers of 69.4% of children born in 2016 had been approached for consent, and 99.1% of these had provided consent – which is extremely positive.
- It had not been possible to obtain consent (verification) for 6.2% of all children born in 2016.
- The proportions of children consented, varied across the regions / units submitting data to CRANE – but remain consistent with proportions reported in past years (for detail on this please consult previous CRANE Database Annual Reports).
- As highlighted in previous Annual Reports, there is still a relatively high proportion of children whose parents have yet to be approached for consent (30.6% in 2016). Units with high levels of patients yet to be consented are encouraged to review their process for approaching parents for consent, as consent is essential for the collection of a full dataset and the linkage to other datasets.

4.2. Characteristics of children born with a cleft lip and/or palate in 2016

Concerning all children born in 2016:

- 56% of children born in 2016 were boys[15]. These boys were significantly more likely to have a CL, UCLP or BCLP than their female counterparts ($p<0.001$)[16].
- CP was significantly more prevalent among females (55% vs. 45% in males, $p<0.001$).
- Among all the children born in 2016, six (0.6%) deaths were reported to CRANE. All of which occurred between one month and one year of age. It is not known from CRANE whether these children had additional anomalies or syndromes.

[14] For past CRANE Database Annual Reports please visit https://www.crane-database.org.uk
[15] Twenty-three children did not have their sex reported to CRANE (2% of the total children registered)
[16] Males comprised 58% of CL cases, 69% of UCLP cases, and 74% of BCLP cases

With regards to only consented children[17]:

- The mean gestation for those born in 2016 was 38.6 weeks (95% CI 38.4 to 38.8 weeks) and ranged from 29 to 42 weeks[18].
- Forty-seven (9.9%) babies were premature (born before 37 weeks' gestation), which is higher than the seven per cent national average in England and Wales[19], 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 41.5% of consented children were missing this information.
- As for gestational age, a valid birth weight was reported for 379 (52.2%) consented babies born in 2016. The mean birth weight was 3.2kg (95% CI 3.1 to 3.2kg), which is consistent with the national average in England.

4.3. Timing of diagnosis

The majority of all babies with a cleft diagnosed in 2016 were antenatally (42.9%) or at birth (42.9%). The proportion of children diagnosed antenatally varies according to cleft type ($p<0.001$), with only 1.3% of CP patients diagnosed antenatally in 2016 compared to rates of 67%, 82.4% and 88.3%% for CL, BCLP and UCLP respectively. See Appendix 7 for detail on cleft types and timing of diagnoses for all 2016 births.

The 2012 our Annual Report highlighted the issue of late diagnosis among children with CP, reporting that 1.1% were diagnosed during antenatal screening and 66.8% were diagnosed at birth, leaving 32.1% who were diagnosed late according to the National Standard[20]. Because of this, we continue to investigate factors associated with a late CP diagnosis, through analyses of (a) CRANE data alone (in Section 4.3.1), and (b) CRANE data linked with HES data (in Section 4.4).

4.3.1. Diagnosis times among CRANE children with a cleft palate alone, 2012-2016 births

This year (as for previous years), we have examined diagnosis time among CP patients born over the last five years, between 1 January 2012 and 31 December 2016. No statistically significant differences were found between birth years ($p=0.45$), indicating diagnosis times have not improved in recent years.

Table 2 shows the CP diagnosis times according to the region / unit. The proportion of CPs diagnosed at birth ranged from 56.1% (South Thames) to over 77.4% (Manchester). This wide and significant variation ($p<0.001$) suggests that practice varies considerably between maternity units, with some

[17] As these data are not collected for non-consenting cases.
[18] Gestational age was reported for 379 (52%) of the consented babies born in 2016. Therefore, further improvements in data completeness are required.
[19] Office for National Statistics. Gestation-specific infant mortality. Part of Gestation-specific infant mortality in England and Wales, 2013. Published 14 October 2015 (this is the latest release – checked November 2017). Available from: http://www.ons.gov.uk/ons/rel/child-health/gestation-specific-infant-mortality-in-england-and-wales/2013/stb-gestation-specific-infant-mortality.html.
[20] Bannister P. Management of infants born with a cleft lip and palate. Part 1. Infant, 2008. 4(1): p. 5-8.

better than others at identifying a cleft of the palate during the newborn examination or due to problems with feeding.

Overall, 13.3% of children with a CP were not diagnosed until they were more than one week old, which is concerning given that the National Standard 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. Cleft services are advised to encourage their referring maternity units to identify all clefts as promptly as possible.

Table 2. Number (%) of CRANE-registered children born between 2012 and 2016 with a cleft palate, according to the timing of diagnosis and region / unit.

Regional Cleft Centre / MCN	Administrative Unit	Time of diagnosis in relation to birth[a] n (%)						
		Antenatal	At birth	≤1 week[b]	≤1 month	≤6 months	>6	**All**
Northern &	Newcastle	2 (1.4)	92 (63.4)	16 (5.6)	13 (9)	14 (9.7)	8 (5.5)	145
Yorkshire	Leeds	0 (0)	102 (71.3)	21 (7.3)	8 (5.6)	6 (4.2)	6 (4.2)	143
North West &	Liverpool	4 (3.3)	88 (73.3)	16 (6.2)	4 (3.3)	6 (5)	2 (1.7)	120
North Wales	Manchester	1 (0.7)	106 (77.4)	17 (6.6)	4 (2.9)	4 (2.9)	5 (3.6)	137
Trent	Nottingham	1 (0.5)	129 (64.2)	47 (23.4)	8 (4)	12 (6)	4 (2)	201
West Midls.	Birmingham	1 (0.4)	186 (74.4)	36 (14.4)	7 (2.8)	14 (5.6)	6 (2.4)	250
East	Cambridge	1 (0.8)	93 (71)	22 (16.8)	9 (6.9)	5 (3.8)	1 (0.8)	131
North Thames	GOSH/Chelms	8 (2.8)	174 (61.5)	68 (24)	11 (3.9)	16 (5.7)	6 (2.1)	283
The Spires	Oxford/Salisbury	2 (1.1)	134 (72.4)	25 (13.5)	9 (4.9)	10 (5.4)	5 (2.7)	185
South Wales &	Swansea	0 (0)	56 (73.7)	12 (5.9)	6 (7.9)	1 (1.3)	1 (1.3)	76
South West	Bristol	4 (3.1)	86 (67.2)	14 (6.9)	14 (10.9)	8 (6.3)	2 (1.6)	128
South Thames	GSTT	3 (1.1)	156 (56.1)	77 (27.7)	16 (5.8)	20 (7.2)	6 (2.2)	278
N. Ireland	Belfast	0 (0)	55 (75.3)	8 (11)	1 (1.4)	5 (6.8)	4 (5.5)	73
All	All	27 (1.3)	1,457 (67.8)	379 (17.6)	110 (5.1)	121 (5.6)	56 (2.6)	2,150

Note: MCN - Managed Clinical Network.

[a] 142/2,292 (6.2%) missing diagnosis time and excluded from 'All' values.

[b] Recording of 'timing of diagnosis' within 72 hours commenced in May 2014 to align CRANE data collection with NIPE standards[21]. With only small numbers having been recorded using this timing, we report '≤72 hours' cases within the '≤1 week' timing (until recording of this timing is well established).

This year, we also conducted an exploration of the impact of different types of cleft palate based on the presentation (as recorded when reporting LAHSAL codes, as described in Chapter 2), on diagnosis times among children with cleft palate alone, born in the last five years between 1 January 2012 and 31 December 2016.

Table 3 shows that the completeness of the hard and soft palate impact on the timing of the CP diagnosis. Specifically:

- CP cases with any type of hard palate were significantly more likely to be identified at birth (by almost 20%) than CP cases where there was no hard palate involvement ($p<0.001$). Where

[21] UK National Screening Committee Newborn and Infant Physical Examination (NIPE) Standards and Competencies 1 document (2008) – setting out the standard for 95% newborn to be screened by 72 hours after birth (page 13 of the document found at http://newbornphysical.screening.nhs.uk/getdata.php?id=10639).

there was no hard palate involvement, 23.9% of CP cases remain undiagnosed until after a week had elapsed.

- Children with CP including complete hard palates were significantly more likely to be identified at birth than those with incomplete hard palates (p<0.001). This delay is addressed within the next week, by which point around 90% of CP cases have had this identified.
- CP cases with complete soft palates were significantly more likely to be identified at birth (almost twice as likely) than incomplete soft palates (p<0.001). With an incomplete soft palate, 35.5% of these CP cases remain undiagnosed until after a week had elapsed.

Table 3. Number (%) of CRANE-registered children born between 2012 and 2016 with a cleft palate, according to complete / incomplete hard and soft palates.

Palate type	Status	Time of diagnosis in relation to birth n (%)						
		Antenatal	At birth	≤1 week	≤1 month	≤6 months	>6 months	**All***
Hard palate	Incomplete (h)	8 (1.1)	500 (70.1)	131 (18.4)	34 (4.8)	32 (4.5)	8 (1.1)	713
	Complete (H)	14 (2.1)	535 (80.6)	87 (13.1)	17 (2.6)	10 (1.5)	1 (0.2)	664
All	All	22 (1.6)	1035 (75.2)	218 (15.8)	51 (3.7)	42 (3.1)	9 (0.7)	1377
Hard palate	No 'h' or 'H'	5 (0.6)	422 (54.6)	161 (20.8)	59 (7.6)	79 (10.	47 (6.1)	773
	Either 'h' or 'H' present	22 (1.6)	1035 (75.2)	218 (15.8)	51 (3.7)	42 (3.1)	9 (0.7)	1377
All	All	27 (1.3)	1457 (67.8)	379 (17.6)	110 (5.1)	121 (5.6)	56 (2.6)	2150
Soft Palate	Incomplete (s)	1 (0.4)	116 (41.6)	63 (22.6)	28 (10)	41 (14.	30 (10.8)	279
	Complete (S)	22 (1.2)	1320 (71.8)	306 (16.6)	79 (4.3)	77 (4.2)	34 (1.8)	1838
All	All	23 (1.1)	1436 (67.8)	369 (17.4)	107 (5.1)	118 (5.6)	64 (3)	2117

*Totals for the sections of this table were based on where the hard and soft palate information had been reported as part of the LAHSAL code(s). Missing data have resulted in the variation in denominator.

4.4. Diagnosis times among children with a cleft palate alone, using CRANE data linked to HES data, 2007-2012 births

This year, we used HES data linked to the CRANE database at the individual level for consented children born between 1 January 2000 and 31 December 2012 to conduct an exploration of the impact of ethnicity, syndromic status[22] and deprivation on diagnosis times among children with cleft palate alone.

Considering missing data, at least three quarters of data on timing of diagnoses for children with CP was missing between 2000 (94.6%) and 2006 (73.4%). From 2007 we have seen a drastic improvement in data quality (only 15% of data was missing in 2007). Because of this we conducted our analyses for this section on data from 2007 to 2012.

Table 4 below shows the variation in timing of diagnoses for CP patients according to syndromic status, ethnicity and deprivation. Specifically:

[22] 'Syndromic' cleft patients are defined as such if they have received specific diagnoses of syndromes and anomalies, in addition to their cleft diagnoses, as specified in Appendix 6.

- The likelihood of antenatal diagnosis of a CP was significantly higher for syndromic children ($p<0.001$), this is probably due to the checks these children receive because of their additional anomalies.
- We found no significant association between timing of diagnoses for CP patients and their ethnicity ($p=5$), which is positive. Nevertheless, we must bear in mind the small numbers of patients from other and unknown ethnic groups reported.
- We also found no significant relationship between timing of diagnoses for CP patients and their deprivation ($p=0.6$). This too is a positive finding.

These initial findings should be interpreted with caution due to the smaller sample size. Analyses of data from a greater number of children are necessary to examine true differences that may exist between these groupings.

The analyses should be revisited in future with a larger sample. A larger sample will become available once a new linkage 'look-up' file is made available from the NHS Digital in 2018. This will allow us to include in our analyses those births that have been registered in CRANE since the last linkage exercise to HES which was conducted in March 2013.

Table 4. Number (%) of children born between 2007 and 2012 with a cleft palate, according to syndromic status, ethnicity and deprivation.

		Time of diagnosis in relation to birth n (%)						
Factors	**Status**	Antenatal	At birth	≤1 week	≤1 month	≤6 months	>6	**All**
Syndromic Status	Syndromic	72 (6.2)	690 (59.8)	214 (18.6)	73 (6.3)	73 (6.3)	31 (2.7)	1153
	Non-syndromic	24 (2.5)	681 (71)	153 (16)	62 (6.5)	32 (3.3)	7 (0.7)	959
Ethnicity	White	68 (4.4)	1016 (65.7)	264 (17.1)	98 (6.3)	72 (4.7)	29 (1.9)	1547
	Other	21 (5.8)	219 (60)	73 (20)	27 (7.4)	20 (5.5)	5 (1.4)	365
	Unknown	7 (3.5)	136 (68)	30 (15)	10 (5)	13 (6.5)	4 (2)	200
All	All	96 (4.5)	1371 (64.9)	367 (17.4)	135 (6.4)	105 (5)	38 (1.8)	2112
Deprivation	Q1 – Most	16 (4.5)	236 (66.7)	54 (15.3)	22 (6.2)	17 (4.8)	9 (2.5)	354
	Q2	17 (4.4)	249 (65)	79 (20.6)	20 (5.2)	15 (3.9)	3 (0.8)	383
	Q3	19 (4.5)	282 (66.4)	72 (16.9)	27 (6.4)	20 (4.7)	5 (1.2)	425
	Q4	23 (5.2)	271 (61)	82 (18.5)	30 (6.8)	30 (6.8)	8 (1.8)	444
	Q5 – Least	18 (3.9)	305 (65.3)	75 (16.1)	35 (7.5)	22 (4.7)	12 (2.6)	467
All	All	93 (4.5)	1343 (64.8)	362 (17.5)	134 (6.5)	104 (5)	37 (1.8)	2073

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

Five year outcomes include height, weight, decayed missing and filled teeth (dmft), the Five Year Old Index, and the Cleft Audit Protocol for Speech – Augmented (CAPS-A) scores. These are reported for only consented children born between 2004 and 2011 (excluding children with submucous cleft palates)[23]. Information and analyses of these data are presented in the next subsections.

4.5.1. Reporting of outcomes, 2004-2011 births

We describe the data completeness for outcomes at five years of age, according to region/unit[24], below:

- There is a high proportion of missing data for five-year old weight and height (for eligible children 63% and 63.5% missing respectively), this is despite continued improvements year-on-year; with wide variation in reporting weight and height data across regions. Variation in reporting ranged from 80% for both weight & height (Leeds), to less than 3% for both weight & height (Northern Ireland). This suggests that this data is not routinely collected in some regions.
- The proportion of eligible children with reported decayed, missing, filled teeth (dmft) index scores varied across regions from 20.6% (East) to 87.3% (West Midlands). The East of England will always lag behind because the first 5-year-old data was not collected until 2009 births, prior to this date there was no paediatric dentist in this region[25].
- The proportion of children with reported Five Year Old Index scores continues to increase year-on-year, which is encouraging. Nevertheless, there was wide variation in reporting of Five Year Old Index data across the regions/units from 9.8% (Northern Ireland[26]) to 88.5% (The Spires).
- CRANE is encouraged by the fact that regions/units have shown increased rates in reported speech data year-on-year since the expanded 16 CAPS-A speech outcome scores were first requested 4 years ago. The proportion of eligible children with Speech outcome scores ranged from 47.1% (Liverpool) to 84.3% (Swansea).

It is acknowledged that sometimes there are reasons outside the units' control as to why outcome(s) data cannot be collected, and we encourage centres to report these. Nevertheless, it is positive to note that reporting has increased for some outcomes at five years of age[27] since this time last year; and it is hoped that this trend will continue over the next few years.

[23] Submucous cleft palate patients excluded from all five year outcomes as all/most teams do not audit these patients.
[24] See Report number 1. 'Outcomes' behind the CRANE Database log-in for further detail- https://www.crane-database.org.uk/
[25] Recent appointment of a paediatric dentist to examine children (and determine dmft) has resulted in a small improvement in data completeness for the East (2%).
[26] Although Northern Ireland submitted data for only 9.8% of their eligible patients, they have collected Five Year Old Index data for only 3 years – and therefore are likely to show improved data completion rates year-on-year.
[27] 1% for weight, 2% for height, -0.4% for dmft, -0.5% for 5 year index and -1.9% for speech.

4.5.2. Decayed missing and filled teeth (dmft), 2004-2010 births

The dmft describes the dental caries an individual has experienced 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[28]. We collect dmft data on CRANE-registered consented children at five years of age.

Among children with a reported dmft outcome[29] , 41.4% 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, with scores ranging from 0 to 20. Four hundred and seventy-five children (17.1%) had a dmft score greater than 5.

Dental caries according to cleft type

Table 5 shows the prevalence of dental caries according to cleft type, with the mean dmft and the proportion of children with at least one dmft (>0 dmft) varying significantly according to cleft type ($p<0.001$).

The dmft data, obtained in 2005, available for five-year old children in the general population in England and Wales shows that 38.8% of five-year olds had at least one dmft, with a mean number of 1.5[30]. The comparable figure of 41.4% among CRANE-registered children (shown in Tables 5 and 6) is close (although slightly higher) to that of the general population. This is likely to be due to the fact that the number of dmft among children with a CL was lower than the general population (33.5% versus 38.8%). Despite this, the mean dmft was higher among CP, UCLP and BCLP patients (2.3, 1.9 and 2.8, respectively) compared to the general population's mean (of 1.5).

[28] (1) Al-Dajani M. Comparison of dental caries prevalence in patients with cleft lip and/or palate and their sibling controls. The Cleft Palate-Craniofacial Journal, 2009. 46(5): p. 529-531. (2) Britton, KF and Welbury, RR, Dental caries prevalence in children with cleft lip/palate aged between 6 months and 6 years in the West of Scotland. European Archives of Paediatric Dentistry, 2010. 11 (5): p. 236-241.

[29] Submucous cleft palate patients excluded from all five year outcomes as all/most teams do not audit these patients.

[30] Dental Health Services Research Unit from National Health Service - British Society for the Study of Community Dentistry. Dental caries experience of 5-year-old children in Great Britain 2005/2006. 2011, Available from: http://www.dundee.ac.uk/tuith/search/bdsearch.html.

Table 5. Number (%) of CRANE-registered consented children born 2004-2010 with a cleft lip and/or palate according to the number of decayed, missing or filled teeth (dmft) at 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[a]**
CL	1.3 (1.1 to 1.5)	544 (66.5)	274 (33.5)	(30.3 to 36.7)	818
CP	2.3 (2.1 to 2.5)	941 (59.9)	629 (40.1)	(37.6 to 42.5)	1570
UCLP	1.9 (1.8 to 2.1)	568 (54.4)	476 (45.6)	(42.6 to 48.6)	1044
BCLP	2.8 (2.4 to 3.2)	207 (48.6)	219 (51.4)	(46.6 to 56.2)	426
Not specified	2.1 (0.6 to 3.6)	18 (58.1)	13 (41.9)	(23.6 to 60.3)	31
All	2.0 (1.9 to 2.2)	2,278 (58.6)	1,611 (41.4)	(39.9 to 42.9)	3,889

[a] Exclusions from 'All' values (not mutually exclusive) include children with missing dmft data, with submucous clefts[31], and children who died before the age of five.
Note: CL - Cleft Lip, CP - Cleft Palate, UCLP - Unilateral cleft lip and palate, and BCLP - Bilateral cleft lip and palate.

The fact that dmft were submitted for only 59.5% of children means that these data should be interpreted with caution, as it is possible that the overall findings from the limited data made available to CRANE may not be representative of the entire 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.

Dental caries according to region / unit

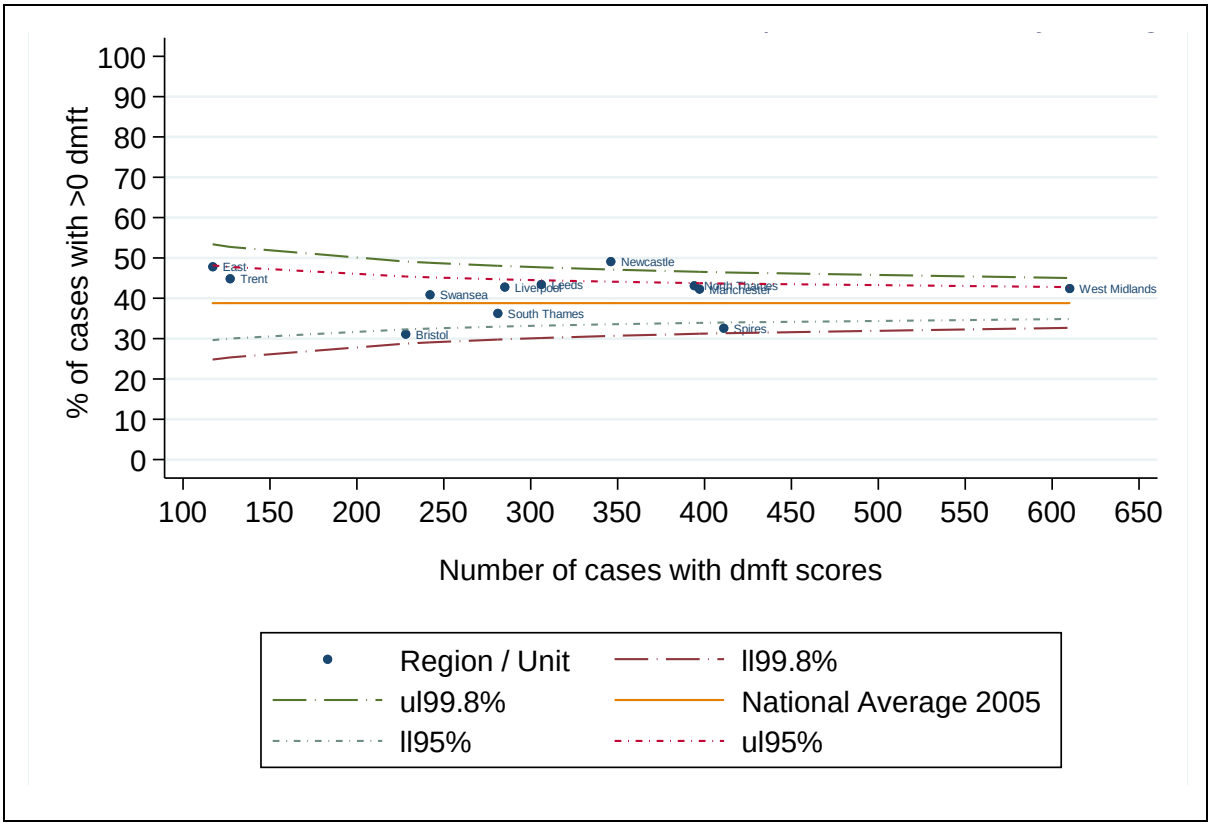
Table 6 shows the prevalence of dental caries according to region / unit[32]. There was a significant variation in dmft scores across Units ($p<0.001$). Children registered in the Northern & Yorkshire region had the highest numbers of mean dmft, which were significantly different to the overall mean (2.0). Bristol and the Spires region had mean dmft values that were significantly lower than the overall mean.

In terms of the proportion of cleft children with at least one dmft (>0 dmft), Bristol had the lowest proportion (31.1%), which was significantly different to the overall proportion among cleft children. Whilst the proportion of cleft children with at least one dmft varied between regions, for the majority of regions their rate does not seem to differ substantially from their region's total population rate[33].

[31] Submucous cleft palate patients excluded from all five year outcomes as all/most teams do not audit these patients.
[32] Not all units had a dentist who was currently calibrated for collection of cleft data.
[33] Dental Health Services Research Unit from National Health Service - British Society for the Study of Community Dentistry data. Dental Caries Experience of 5-year-old Children in Great Britain 2005 / 2006. Available from: http://www.app.dundee.ac.uk/tuith/search/tables/tab2005_6.htm.

The funnel plot[34] in Figure 2 further demonstrates the proportion of five-year olds with at least one decayed missing or filled tooth (>0 dmft) according to the number of children with valid dmft scores at each region/ unit. This funnel plot is centred on the national average of 38.8% (with at least one dmft), obtained in 2005, for five-year old children in the general population in England and Wales[35].

Figure 2. Funnel plot of five-year olds (born between 2004 and 2010) with at least one dmft (>0 dmft), according to the number of children at each region / unit with dmft scores.



Note: Funnel plot centred on the national average of 38.8% (with at least one dmft), obtained in 2005, for five-year old children in the general population in England and Wales (only). Therefore only 12 units shown as Northern Ireland data excluded.

Figure 2 shows that most regions' / units' rates of at least one dmft (>0 dmft) fall within the expected range given the number of children with valid dmft scores at their region / unit. No site has a >0 dmft rate below the lower 99.8% control limit, and one unit (Newcastle) has a >0 dmft rate above the upper 99.8% control limit. This means they have significantly high rates of children with >0 dmft – which is unlikely to be as a result of chance[36] and is worth investigating (more information on funnel plots can be found in the Glossary at the front of this report).

[34] This funnel plot is calculated using valid data as denominators (not considering missing data), subject to the same inclusions and exclusions as data in Table 5. In addition, it is not adjusted (or risk adjusted) in any way.
[35] Dental Health Services Research Unit from National Health Service - British Society for the Study of Community Dentistry. Dental caries experience of 5-year-old children in Great Britain 2005/2006. 2011. Available from: http://www.dundee.ac.uk/tuith/search/bdsearch.html.
[36] Regional differences in the levels of dental disease are not only be affected by the dental care received by children. Oral health is also affected by deprivation, ethnicity, cultural differences in attitudes to dental health and water fluoridation levels.

Table 6. Number (%) of CRANE-registered consented children born between 2004 and 2010 with a cleft lip and/or palate – according to the number of decayed, missing or filled teeth (dmft) and the average treatment index at age five years by region / unit.

Regional centre / MCN	Administrative Unit	Number of decayed, missing or filled teeth (dmft)					Treatment Index		Care Index	
			0	>0		All[a]		All[b]		All[b]
		Mean (95% CI)	n (%)	n (%)	95% CI	(N)	Average (%)	(N)	Average (%)	(N)
Northern & Yorkshire	Newcastle§¥	2.7 (2.3 to 3.2)	176 (50.9)	170 (49.1)	(43.8 to 54.4)	346	(69.2)	317	(59.2)	319
	Leeds§¥	2.5 (2.1 to 3)	173 (56.5)	133 (43.5)	(39.4 to 49)	306	(70.9)	306	(64.9)	306
North West & North Wales	Liverpool	1.9 (1.5 to 2.3)	163 (57.2)	122 (42.8)	(37 to 48.5)	285	(68.9)	280	(62.2)	280
	Manchester	2.1 (1.7 to 2.5)	229 (57.7)	168 (42.3)	(37.4 to 47.2)	397	(71.2)	395	(65.1)	396
Trent	Nottingham¥*	2.9 (2 to 3.7)	70 (55.1)	57 (44.9)	(36.1 to 53.6)	127	(78.3)	124	(63.4)	125
West Midlands	Birmingham§¥	2.1 (1.8 to 2.4)	351 (57.5)	259 (42.5)	(38.5 to 46.4)	610	(69.4)	608	(62.8)	609
East	Cambridge*	2.6 (1.9 to 3.4)	61.0 (52.1)	56.0 (47.9)	(38.7 to 57)	117	(71.8)	112	(65.3)	112
North Thames	GOSH/Chelms.¥	2.0 (1.7 to 2.4)	224 (56.9)	170 (43.1)	(38.2 to 48)	394	(77.9)	370	(67.6)	370
The Spires	Oxford/Salis.	1.5 (1.2 to 1.8)	277 (67.4)	134 (32.6)	(28 to 37.1)	411	(90)	326	(87.5)	326
South Wales & South West	Swansea§¥	1.8 (1.4 to 2.1)	143 (59.1)	99 (40.9)	(34.7 to 47.1)	242	(78.3)	240	(68.1)	242
	Bristol§¥	1.1 (0.8 to 1.4)	157 (68.9)	71 (31.1)	(25.1 to 37.2)	228	(82.5)	224	(77)	224
South Thames	GSTT§¥	1.6 (1.3 to 2)	179 (63.7)	102 (36.3)	(30.6 to 41.9)	281	(83.9)	277	(78.3)	278
Nrthn. Ireland	Belfast	2.6 (1.9 to 3.3)	75 (51.7)	70 (48.3)	(40 to 56.5)	145	(76.5)	144	(62.6)	144
All	All	2.0 (1.9 to 2.2)	2,278 (58.6)	1,611 (41.4)	(39.9 to 42.9)	3,889	(75.5)	3,723	(67.9)	3,731

Note: MCN – Managed Clinical Network.

[a] Exclusions from 'All' values (not mutually exclusive) include children with missing dmft data, with submucous clefts[37], and children who died before the age of five.

[b] Exclusions from Treatment and Care Index (not mutually exclusive): Children who died before the age of five, children with submucous clefts, and cases without a dmft score of 0[38] or all relevant dmft data items (to allow calculation of treatment and care index scores).

Individual unit considerations: §Cleft calibrated assessor. *dmft data not been submitted in years past as no paediatric dentist in place to examine children (determining dmft) or no administrative support in place to submit data to CRANE – it is anticipated these units' data completeness will show improvement in future reports. ¥Specialist paediatric dentist.

[37] Submucous cleft palate patients excluded from all five year outcomes as all/most teams do not audit these patients.

[38] If a dmft score for an individual is 0 then the treatment index and care index = 1 (100%) as there is no untreated dental disease.

Dental Treatment and Care Indices

Table 6 also shows the average treatment index and care index (reported for the second time) for children according to region / unit. Both indices are calculated from the dmft[39], as raw dmft scores give a figure for dental disease experienced but do not distinguish if there is active disease present at the time or not (treatment index) or the proportion of children who have received care in the form of fillings (care index).

The treatment index reflects whether the mouth is dentally fit at that moment in time. i.e. If dental disease has occurred, the treatment index indicates the extent to which it has been dealt with and the degree to which the child has been rendered free from active decay. When calculated, treatment indices range from 0 to 1 and are usually expressed as a percentage[40]. Treatment indices with a value of 1 (100%) indicate that there is no untreated disease, which is the desired outcome. Furthermore, average treatment indices of 100% can be indicators of having mechanisms in place to deal with any disease occurring, and thereby providing the child with a dentition where the disease is controlled and the child has a pain free mouth.

For the 3,723 children with dmft scores of 0 or scores for all three 'm', 'f' and 'dmft' data items – to allow calculation of the treatment index – there was significant variation in treatment index scores across units ($p<0.001$). Children registered by Liverpool, Newcastle and in the West Midlands had the lowest average proportion of treated dental disease (68.9. 69.2 and 69.4 respectively; approximately 6% less than the national average), while the Spires region had highest average proportion of treated dental disease (90%, approximately 15% more than the national average).

The care index reflects cases where children have experience dental decay, which has been identified at the earliest possible stage (which is preferable), and have been provided with care in the least invasive form possible – in the form of fillings. When calculated, care indices also range from 0 to 1 and are usually expressed as a percentage[41]. Care indices with a values close to 1 (100%) indicate that there are high levels of care provided by filling (not extraction or no treatment), which is the desired outcome. Conversely in situations where levels of care low (and decay could be addressed by filling but has not) the care index is close to 0%. Furthermore, average care indices of 100% can be indicators of having mechanisms in place to increase levels of care in relation to fillings.

For the 3,731 children with dmft scores of 0 or scores for both 'f' and 'dmft' data items, to allow calculation of the care index, there was significant variation in care index scores across Units ($p<0.001$). Children registered by Newcastle had the lowest average care index (59.2%, approximately 8% less than the national average), which means decay that might be treated by fillings has remained untreated or decay was so severe extraction was the treatment of choice. Meanwhile the Spires region had the highest average proportion of children receiving care by fillings (87.5%, approximately 20% more than the national average).

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[39] Treatment Index calculated as = (Total number of missing teeth in primary dentition (m) + Total number of filled teeth in primary dentition (f)) / 'Total number of decayed, missing or filled teeth in primary dentition (dmft).
Care Index calculated as = Total number of filled teeth in primary dentition (f) / 'Total number of decayed, missing or filled teeth in primary dentition (dmft).

[40] If a dmft score for an individual is 0 then the treatment index is 1 (100%) as there is no untreated dental disease.

[41] If a dmft score for an individual is 0 then the care index is 1 (100%) as there is no dental disease.

It is also worth considering that 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, ethnicity, 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[42]. 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, while the North East had the highest proportion (50%)[43]. Accurate water fluoridation data will be useful for interpreting dmft regional differences and allowing for risk adjustment in the long term.

4.5.3. Five Year Old Index, 2004-2010 births

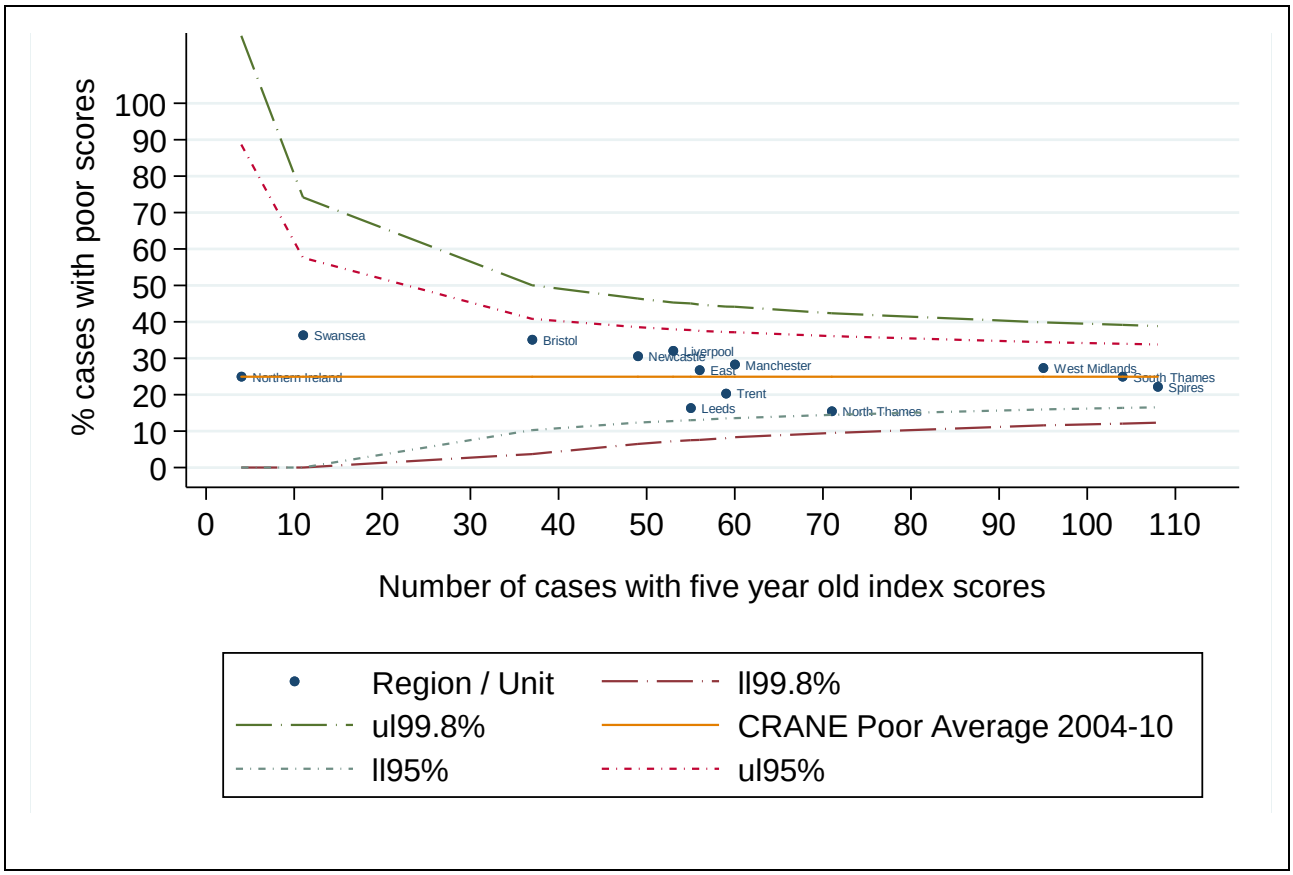
Dental models of five-year old children with a complete UCLP were assessed using the Five Year Old Index to examine dental arch relationships. The index evaluates the effects of primary surgery on the facial growth of children with UCLP before any other interventions, such as orthodontics or alveolar bone grafting, which may influence this growth further[44]. Dental arch relationships at five years are thought to predict treatment outcome in terms of facial growth on a population basis rather than at the individual child level[45]. The Five Year Old Index may, therefore, also be used to compare treatment outcomes between centres and surgeons. Patients scoring '1' and '2' on the index are considered to have the best possible outcomes, while those scoring '4' and '5' are thought to have poor outcomes in terms of facial growth, and they may benefit from further surgery to correct their facial disproportion once facial growth is complete.

The majority of Five Year Old Index scores provided by all regions/units were externally validated (in 660/762 (86.6%) of eligible cases), and where externally validated scores were unavailable, internal scores were included in the analysis. Overall, 44.8% of complete UCLP patients born between 2004 and 2010 had Five Year Old Index scores in the two groups considered to have the best possible dental arch relationships (scores '1' or '2') while 24.9% of children had scores '4' or '5', reflecting poor dental arch relationships. This represents an improvement, compared to the CSAG findings that 36% (of 223 cleft children) had poor dental arch relationships at five years old in 1998[46] (see Appendix 8 for detail on children born between 2004 and 2009 with a complete unilateral cleft lip and palate, according to Five Year Old Index scores and region / unit).

[42] McDonagh M, Whiting P, Bradley M, Cooper J, Sutton A, Chestnutt I, et al. A systematic review of public water fluoridation. BMJ, 2000. 321: p. 855-859.
[43] Dental Health Services Research Unit from National Health Service - British Society for the Study of Community Dentistry data. Dental Caries Experience of 5-year-old Children in Great Britain 2005 / 2006. Available from: http://www.app.dundee.ac.uk/tuith/search/tables/tab2005_6.htm.
[44] Johnson N, Williams AC, Singer S, Southall P, Atack N and Sandy JR. Dentoalveolar relations in children born with a unilateral cleft lip and palate (UCLP) in Western Australia. The Cleft Palate-Craniofacial Journal, 2000. 37 (1): p. 12-16.
[45] Atack N, Hathorn IS, Semb G, Dowell T and Sandy JR. A new index for assessing surgical outcome in unilateral cleft lip and palate subjects aged five: reproducibility and validity. The Cleft Palate-Craniofacial Journal, 1997. 34 (3): p. 242-246.
[46] Clinical Standards Advisory Group, Clinical Standards Advisory Group. Report of a CSAG Committee on cleft lip and/or palate, 1998, The Stationery Office, London.

The funnel plot[47] in Figure 3 further demonstrates the proportion of CRANE five-year olds with the poorest (4 and 5) Five Year Old Index outcome scores according to the number of children at each region / unit with index scores. It is centred on the national average of poor Five Year Old Index scores for CRANE five-year olds across all units of 24.9%[48]. It also shows that all regions' / units' rates of poor index scores fall within the expected range given the number of children (born between 2004-2009) with valid index scores at their unit. i.e. No unit has a 'poor index score rate' below the lower 99.8% control limit or above the upper 99.8% control limit (more information on funnel plots can be found in the Glossary at the front of this report).

Figure 3. Funnel plot of five-year olds (born between 2004 and 2010) with poor Five Year Old Index scores according to the number of children at each region / unit with index scores.



Note: Funnel plot centred on national average (for 2004-2010 births reported in CRANE) of poor Five Year Old Index scores across all units of 24.9%.

The fact that Five Year Old Index scores were submitted for only 70.1% of children, and the wide variation in the number of children within each region / unit (ranging from 4 to 108), means that the data presented in this section should be interpreted with caution, as 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.

[47] This funnel plot is calculated using valid data as denominators (not considering missing data), subject to the same inclusions and exclusions as data in Appendix 8. In addition, it is not adjusted (or risk adjusted) in any way.
[48] Versus the 36% national average identified by Clinical Standards Advisory Group (CSAG). Report of a CSAG Committee on cleft lip and/or palate, 1998, The Stationery Office, London.

Relationship between facial growth and speech, 2007-2010 births

We describe below analyses exploring the relationship between children's facial growth and speech outcomes for UCLP children. This is to establish whether or not there is a relationship between these children achieving good or poor outcomes in terms of facial growth and good / normal speech outcomes. Good outcomes for facial growth and speech have been defined as follows:

- Patients scoring '1' and '2' on the Five Year Old Index are considered to have the best possible outcome (good outcomes), while those scoring '4' and '5' are thought to have poor outcomes in terms of facial growth (detailed in section 4.5.4 on Five Year Old Index scores).
- Normal speech is achieved in cases where patients score 'normal' (green) scores across all 16 Cleft Audit Protocol for Speech – Augmented (CAPS-A) speech parameters (detailed further in the next section 4.4.4 on CAPS-A scores).

Table 7 shows that the proportion of children with good Five Year Index scores who have/have not achieved normal speech do not differ significantly from the proportion of children with poor scores (p=0.85). Overall, 32.8% (95/259) of consented children born between 2007 and 2010 had achieved scores indicating good facial growth and normal speech; while 18.5% (48/259) had scores indicating poor facial growth and not achieving normal speech.

Table 7. Number (%) of CRANE-registered[a] consented children born between 2007 and 2010[b], with good or poor Five Year Old Index scores at five years of age, by those achieving/not achieving normal speech.

Five Year Old Index scores	Normal Speech					
	Achieved		Not Achieved		Total	
	N	(%)	N	(%)	N	(%)
Good scores	85	(32.8)	84	(32.4)	169	(65.3)
Poor scores	42	(16.2)	48	(18.5)	90	(34.7)
Total	**127**	**(49)**	**132**	**(51)**	**259**	**(100)**

[a] Registered in CRANE by 2 October 2017.
[b] Excluding children who died before the age of 5 years, children with an incomplete UCLP, children with submucous cleft palates, syndromic children, children missing Five Year Old Index scores data, and children missing one or more of all 16 CAPS-A data items.

We also explored the relationship between facial growth and children's scores for the following 6 individual structurally related CAPS-A speech parameters – whose poor scores are indicative of structural issues of the palate / poor surgery:

1. Resonance: Hypernasality.
2. Nasal Airflow: Audible Nasal Emission.
3. Nasal Airflow: Nasal Turbulence.
4. Passive CSCs: Weak and or nasalised consonants.
5. Passive CSCs: Nasal realisation of plosives.
6. Passive CSCs: Gliding of fricatives.

The proportion of children with good Five Year Index scores who had achieved good (green) scores for the above 6 CAPS-A speech parameters did not differ significantly from the proportion of children with poor (amber or red) CAPS-A scores.

Therefore there currently is no evidence of positive or negative correlation between facial growth and normal speech scores.

These initial findings should be interpreted with caution due to the small sample size. The analyses should be revisited in future with a larger sample. A larger sample will become available as the collection of the full 16 CAPS-A outcome scores (which started 4 years ago) expands for births after 2010; along with the yearly expansion of the collection of Five Year Index scores. In addition, as the data completeness of Five Year Old Index and CAPS-A scores continues to improve year-on-year; so will the sample size available for analyses.

4.5.4. Cleft Audit Protocol for Speech – Augmented scores, 2007-2010 births

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). The 16 CAPS-A speech outcome scores assessed[49] include:

- Resonance (hypernasality and hyponasality) and nasal airflow (audible nasal emission and nasal turbulence). These are structurally related speech characteristics reflecting aspects such as the ability of the palate to close off the nasal airway during speech.
- 12 individual cleft speech characteristics (CSCs) grouped into four categories of CSCs – anterior oral, posterior oral, non-oral and passive – are also assessed. These reflect articulation patterns which can affect the clarity and intelligibility of a child's speech.

Table 8 shows the distribution of proportions of reported speech outcomes (or reasons why not collected) by region / unit. 64% of consented children born between 2007 and 2010 had reported speech outcomes for all 16 CAPS-A parameters, and 24% had reported reasons why speech data was not collected (e.g. Patient deceased or emigrated, transferred in or out of area, etc[50]).

Table 8. Number (%) of CRANE-registered[a] consented children born with a cleft palate (2007-2010), with speech outcome data or reasons this outcome was not collected at five years of age, according to region / unit.

Regional centre / MCN	Administrative Unit	Speech[b]					
		Eligible Consented cases	Outcome Reported		Reason outcome not collected provided		Total cases acc. for
		N	n	(%)	n	(%)	(%)
Northern	Newcastle	182	122	67%	37	20%	87%
& Yorkshire	Leeds	179	106	59%	64	36%	95%
North West	Liverpool	223	105	47%	99	44%	91%
& North Wales	Manchester	229	154	67%	60	26%	93%
Trent	Nottingham	267	178	67%	67	25%	92%
West Midlands	Birmingham	295	205	69%	44	15%	84%
East	Cambridge	217	136	63%	66	30%	93%
North Thames	GOSH & Chelmsford	361	212	59%	84	23%	82%
The Spires	Oxford & Salisbury	265	172	65%	58	22%	87%
South Wales	Swansea	134	113	84%	17	13%	97%
& South West	Bristol	187	115	61%	24	13%	74%
South Thames	Guy's and St Thomas'	256	162	63%	74	29%	92%
Northern Ireland	Belfast	115	84	73%	12	10%	83%
All	**All**	**2,910**	**1,864**	**64%**	**706**	**24%**	**88%**

[a] Registered in CRANE by 2 October 2017. Note: MCN - Managed Clinical Network.
[b] Exclusions (not mutually exclusive): children who died before the age of 5 years, with submucous cleft palates, missing one or more of all 16 CAPS-A data items, born with either a CL or a non-specified cleft type, and syndromic children.

[49] Four Cleft Speech Categories (CSCs) summarising the all 12 CSCs were collected for births prior to 2007. Because of this, speech data collected before 2007 is not included in this report. Changes to expand data collection to 12 CSCs were made in 2014.
[50] Plus: Syndromic Diagnosis; Clinically contraindicated (other than syndromic) - this record type for this patient; Lack of staff / facilities / equipment; Patient DNA / cancelled / did not consent / cooperate; or Other reason.

Overall, rates of data completeness ranged from 82% of children being accounted for by North Thames to 97% of children being accounted for in Swansea (see Appendix 9 for detail of missing data).

The scores for each of the individual 16 CAPS-A assessed are presented in Appendix 11.

Resonance and Nasal Airflow

In terms of resonance, 4.9% of children had moderate or severe hypernasality i.e. nasal sounding speech[51]. This is indicative of velopharyngeal dysfunction (VPD), which is when the palate is unable to close off the nasal airway during speech. In addition, results of the Cleft Speech Characteristics show that 2.2% of children had 'weak and or nasalised consonants' and 1.5% of children had 'nasal realisation of plosives' (passive articulation errors) affecting three or more consonants, which are likely to be the consequence of VPD and is consistent with the hypernasality scorings.

It should be noted that, in order to achieve these outcomes, 19.4% of the children with reported surgical data[52] have had surgery for speech purposes (secondary speech surgery) before the age of five years.

84.1% children with reported scores for all four resonance and nasal airflow parameters had scores indicating that no structural problems existed in relation to these parameters[53].

Cleft Speech Characteristics (CSCs)

'Palatalisation / Palatal' anterior oral CSCs were the most commonly occurring CSC, affecting 23.2% of children (11.9% with scores of B and 11.3% with scores of C). However, these may only have a minor effect on speech intelligibility, and, if treatment is indicated, this would probably involve speech therapy only. The more significant characteristics are the posterior, non-oral and passive CSCs, 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.

In addition, out of the children with reported scores for all 12 CSC parameters, 66.8% had scores indicating they did not exhibit cleft speech characteristics[54].

Nationally agreed Speech Outcome Standards

Further to reporting on the 16 CAPS-A speech parameters separately, we report on the proportion of five-year olds meeting each of the following three nationally agreed Speech Outcome Standards[55]:

1. The achievement of *normal speech* (speech outcome standard #1): This standard is achieved in cases where patients have normal (green) scores across all 16 CAPS-A speech parameters.
2. The absence of speech difficulties as a result of existing or previous structural anomalies (speech outcome standard #2a): This standard is achieved in cases where patients have no reported history of

[51] With a hypernasality score of '3' or '4' (red scores).
[52] VP surgery/fistula repair data was reported for 99.4% of eligible children.
[53] All green scores of '0' or '1'.
[54] All green scores of 'A' and in selected cases of 'B' – as per Appendix 11.
[55] Britton L, Albery L, Bowden M, Harding-Bell A, Phippen G, and Sell D(2014) A Cross-Sectional Cohort Study of Speech in Five-Year-Olds With Cleft Palate ± Lip to Support Development of National Audit Standards: Benchmarking Speech Standards in the United Kingdom. The Cleft Palate-Craniofacial Journal: July 2014, Vol. 51, No. 4, pp. 431-451.

surgery for speech purposes and have normal (green) scores across the following six CAPS-A speech parameters: Hypernasal resonance, both nasal airflow parameters (audible nasal emission and nasal turbulence), and all three Passive CSCs.

3. The absence of cleft-related articulation difficulties (speech outcome standard #3): This standard is achieved in cases where patients have normal (green) scores across the following 10 CSCs: All three Anterior Oral CSCs, both Posterior Oral CSCs, all four Non Oral CSCs, and gliding of fricatives (a Passive CSC).

Normal speech

Out of the 1,760 children (born 2007-2010) with reported scores for all 16 CAPS-A speech parameters, 60.1% of children across all units had normal (green) scores across all 16 parameters. This means that the national speech outcome standard #1 target of 55%[56] was met and exceeded on average by the CRANE cohort born in 2007-2010.

Closer examination of the proportion of children achieving *normal speech* across the cleft types audited for speech (CP, UCLP & BCLP) highlighted a statistically significant difference between rates of *normal speech* reported by cleft type ($p<0.01$). This meant that significantly more children with a CP achieved *normal speech* (71.3% of children with CP – with reported speech score – achieved *normal speech*) and significantly fewer children with BCLP achieved *normal speech* (only 35.8% of children with BCLP had achieved *normal speech* by age 5).

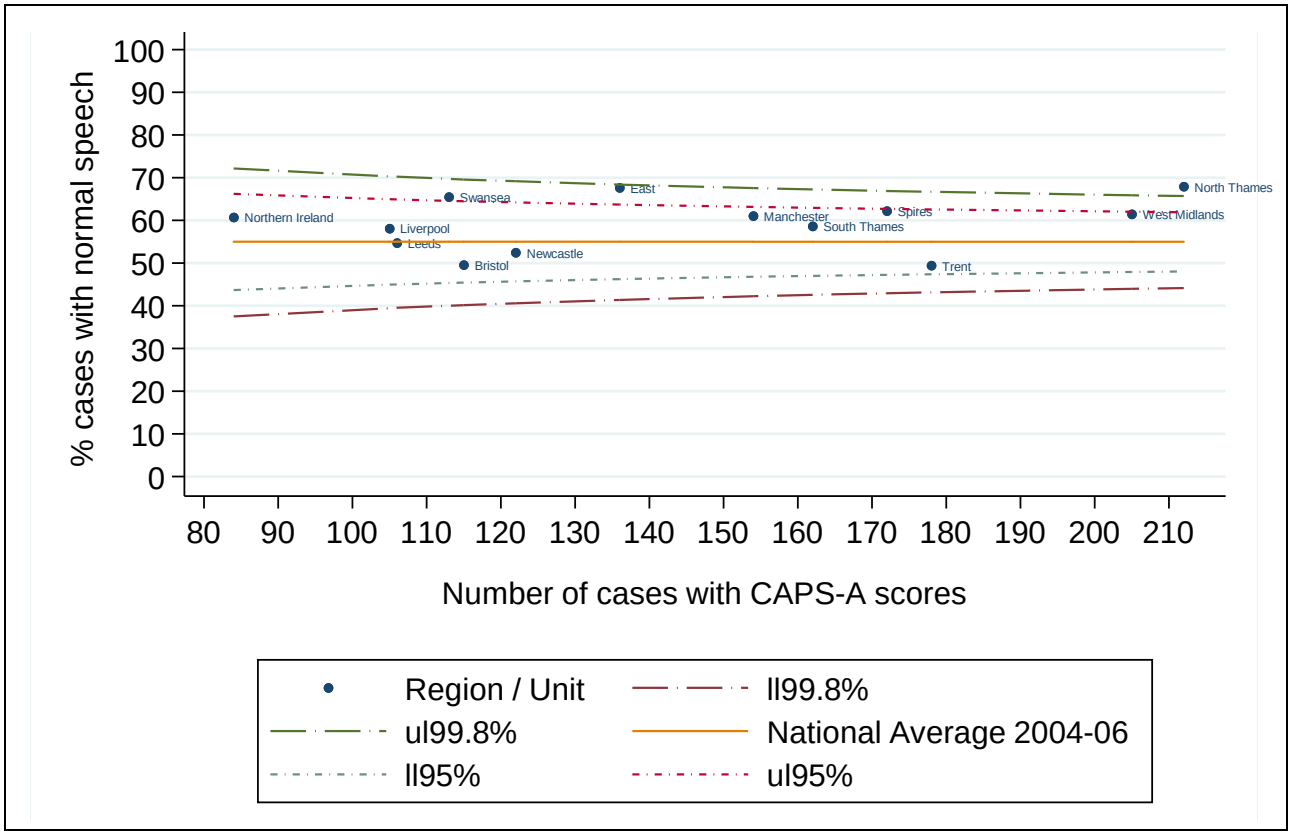
The funnel plot[57] in Figure 4 shows the proportion of children (born in 2007-2010) achieving *normal speech* according to the number of auditable children at each region / unit with scores for all 16 CAPS-A speech parameters (more information on funnel plots can be found in the Glossary at the front of this report). Figure 4 shows that rates of *normal speech*, for most regions / units, fell within the expected range given it is centred on the agreed national average of 55% and the number of children with valid speech scores at each unit. I.e. no unit has *normal speech* rates below the lower 99.8% control limit, which is positive.

As for last year's report, the North Thames region has significantly high rates of children with normal speech (with rates above the upper 99.8% control limit), which is not as a result of chance and worth investigating.

[56] Based on the national outcome mean resulting from statistical analysis on 2004-06 Speech Outcome data – completed by the Lead Speech and Language Therapy Group, with statistical support from the Cleft Collective in Manchester, and presented in April 2014 to the Leads group.

[57] This funnel plot is calculated using valid data as denominators (not considering missing data), subject to the same inclusions and exclusions as data in Table 8. In addition, it is not adjusted (or risk adjusted) in any way.

Figure 4. Funnel plot of five-year olds (born 2007-2010) with scores suggesting normal speech, according to the number of children at each region / unit with CAPS-A scores.



Note: Funnel plot centred on national average identified as the national outcome mean of 2004-06 speech outcome data[58] of 55%.

Nevertheless, this graphical representation of the data showing high levels of *normal speech* achieved at North Thames (67.9%) and lower levels of *normal speech* achieved in the Trent region (49.4%) – should be interpreted with caution. This is because there is wide variation in rates of missing data between regions/units (as detailed in Appendix 9), and speech outcomes at 5 years of age are indicative of historical rather than current service provision.

When considering the most recent 3 years of data, of the 1,405 children born in 2008, 2009 and 2010, the proportion of children achieving *normal speech* fell within the expected range for all regions/units with slightly less variation; which is positive (see Appendix 10).

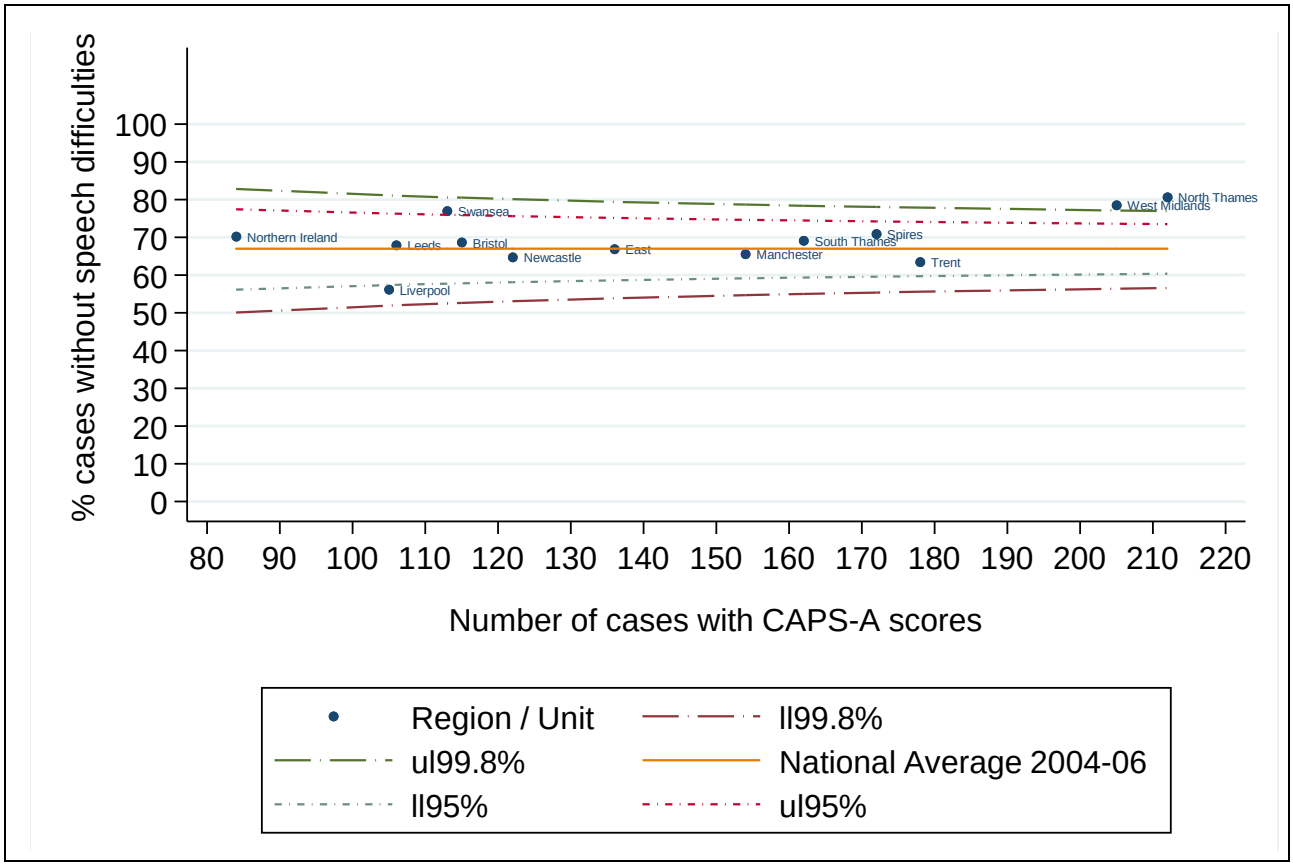
Children with no evidence or history of a structurally related speech problem

Figure 5 shows the proportion of five-year olds with speech scores that suggest they do not have structurally related speech difficulties [59], according to the number of children at each region / unit with CAPS-A scores. It is centred on the national average of 67%, identified as the national outcome mean of 2004-06 speech outcome data[60]; and shows that all but two regions' / units' rates of *no structurally related*

[58] Resulting from statistical analysis on 2004-06 Speech Outcome data completed by the Lead Speech and Language Therapy Group, with statistical support from the Cleft Collective in Manchester, and presented in April 2014 to the Leads group.
[59] As a result of existing or previous structural anomalies – specifically there is no evidence of a structurally related problem and they have not had VP surgery or fistula repair for speech.
[60] Resulting from statistical analysis on 2004-06 Speech Outcome data completed by the Lead Speech and Language Therapy Group, with statistical support from the Cleft Collective in Manchester, and presented in April 2014 to the Leads group.

speech difficulties fall within the expected range given the number of children with CAPS-A scores at their region / unit.

Figure 5. Funnel plot of five-year olds (born 2007-2010) with scores suggesting no structurally related speech difficulties, according to the number of children at each region / unit with CAPS-A scores.



Note: Funnel plot centred on national average identified as the national outcome mean of 2004-06 speech outcome data[61] of 67%.

Two regions –the West Midlands and North Thames – have significantly high rates of children without structurally related speech difficulties (with rates above the upper 99.8% control limit), which is not as a result of chance and worth investigating.

When considering the most recent 3 years of data the children born in 2008 to 2010 (n=1,405), the proportion of children achieving *no structurally related speech difficulties* also fell within the expected range for most regions/units; with North Thames still having significantly high rates of children without speech difficulties (with rates above the upper 99.8% control limit) (see Appendix 10).

Children without cleft-related articulation difficulties

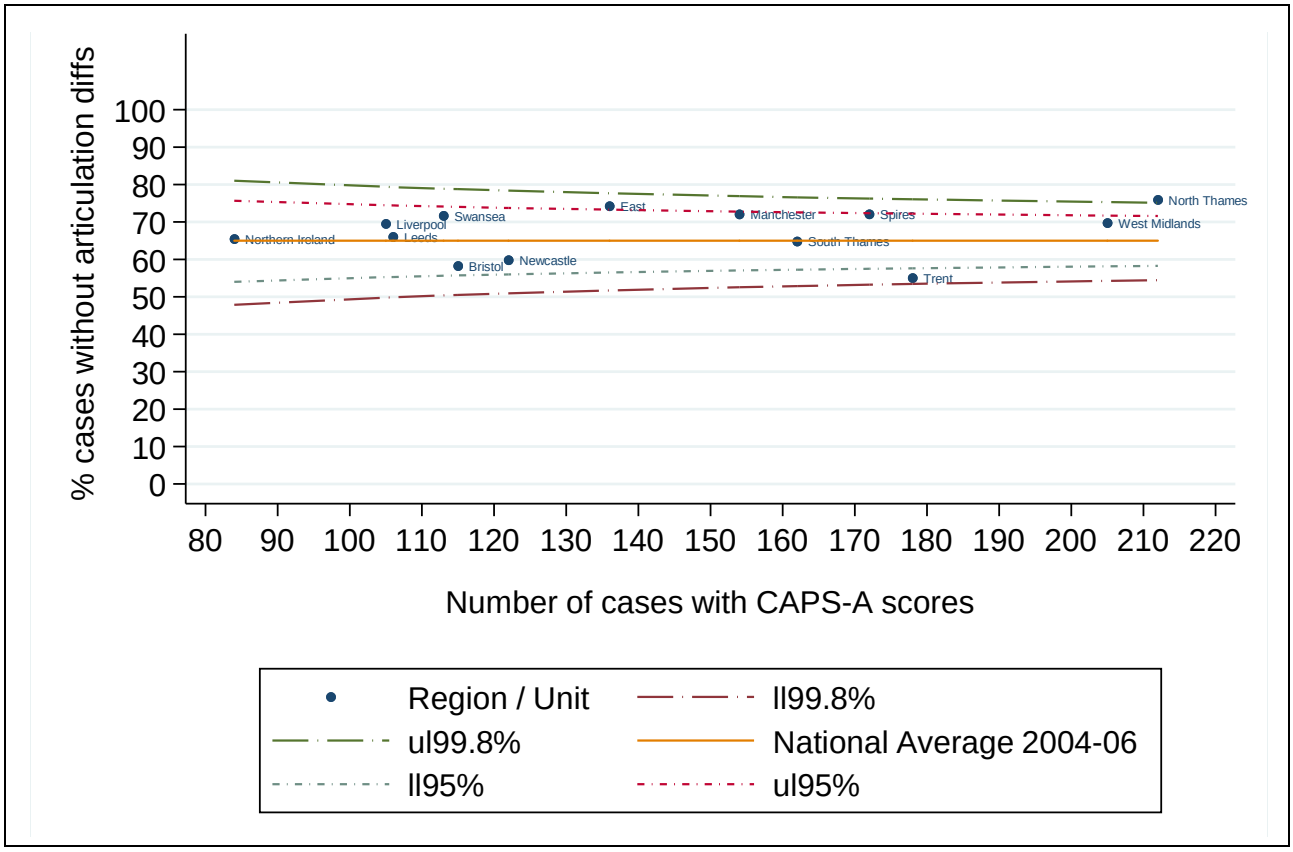
Figure 6 shows the proportion of five-year olds with speech scores that suggest they do not have cleft-related articulation difficulties[62], according to the number of children at each region / unit with CAPS-A scores. It is centred on the national average of 65%, identified as the national outcome mean of 2004-06

[61] Resulting from statistical analysis on 2004-06 Speech Outcome data completed by the Lead Speech and Language Therapy Group, with statistical support from the Cleft Collective in Manchester, and presented in April 2014 to the Leads group.
[62] No cleft type articulation difficulties requiring SLT and/or surgery

speech outcome data[9]; and it shows that most regions' / units' rates of 'no articulation difficulties' fall within the expected range given the number of children with valid CAPS-A scores at their region / unit.

Nevertheless, the North Thames region has a *no articulation difficulties* rate above the higher 99.8% control limit. This means they have significantly high rates of children without articulation difficulties – which is not as a result of chance and worth investigating.

Figure 6. Funnel plot of five-year olds (born 2007-2010) with scores suggesting no cleft-related articulation difficulties, according to the number of children at each region / unit with CAPS-A scores.



Note: Funnel plot centred on national average identified as the national outcome mean of 2004-06 speech outcome data[63] of 65%.

Despite the considerations around missing data described above, presenting the data in Figures 4, 5 and 6 as funnel plots centred on national averages[64] is the most conservative method (at this time[65]) of checking whether or not any units deviate significantly from the expected standards.

When considering the most recent 3 years of data the children born in 2008 to 2010 (n=1,405), the proportion of children achieving *no articulation difficulties* fell within the expected range for all regions/units; which is positive (see Appendix 10).

[63] Resulting from statistical analysis on 2004-06 Speech Outcome data completed by the Lead Speech and Language Therapy Group, with statistical support from the Cleft Collective in Manchester, and presented in April 2014 to the Leads group.
[64] Resulting from statistical analysis on 2004-06 Speech Outcome data completed by the Lead Speech and Language Therapy Group, with statistical support from the Cleft Collective in Manchester, and presented in April 2014 to the Leads group.
[65] No consensus has been reached on the factors that should be incorporated into an adjustment (or risk adjustment) of this data.

5. Development of CRANE Database and future directions

5.1. Future development of the CRANE Database and website

The development and introduction of data downloads and real time reports, accessible to cleft teams through the web based platform for the database, has allowed the CRANE project team to (1) reduce the quantity and length of reporting undertaken[66], (2) conduct consultations and feasibility studies aimed at informing the next developments of the CRANE Database, and (3) continue to devote time into analysis of data for commissioners and teams and subsequent submission of peer reviewed publications.

In the last year, the real time reports accessible to teams (behind the log-in) – on 5-year outcomes, consent, diagnosis time by cleft type and missing identifiers (such as NHS No.) – have come into full use.

These reports have been joined by two accessible reports (outside of the log-in) on registered births by region and cleft type data over the last ten years.

In the next year, the project aims to scope the following online reports:

- Real time reporting of delayed CP diagnosis – available on the website to the public.
- Access to funnel plots of key outcome data – for cleft teams.

We also intend to continue work on specifying data collection of the following sections for the database, as proposed by our stakeholders:

- LAHSAL data collection items changed to collect LAHSHAL data to increase the phenotypic data available for analysis and linkage to other projects.
- Surgical section / items (allowing collection of data on multiple surgeries).
- Dental Defects of Enamel (DDE) section/items (at 5 and 10yrs) as proposed by the Paediatric Dental CEN of CFSGBI.

We aim to review the impact of these changes after 12 months with feedback from both clinical teams and commissioners.

Furthermore, we intend to produce a patient and parent friendly reports of key data alongside this report (to be published in early 2018).

[66] Originally we were contracted to deliver a written annual report and a progress report 6 months apart. This concept was based on delivery before real time reporting was available. We have also developed other methods of communication with teams and stakeholders in recent years. These include patient and parent friendly reports of key data and eNewsletters to clinical teams and managers.

5.2. Outcome measures

Entry of data for the additional outcomes collected at 5 and 10 years of age (since May 2016) is underway – specifically for Psychology at 5 years of age and Paediatric Dentistry at 10 years of age. We aim to report on Psychology at 5 years for 2011 births in the 2018 Annual Report.

Despite the progress made, 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. We have been asked to see if outcomes can be agreed for older children and young adults by CFSGBI. We aim to run a multiphase consultation using different methods of engagement to develop consensus and identify valid and robust measures. Also, our strategy will continue to involve linkage to other data sources to reduce the burden of data collection from teams, where possible:

5.2.1. Newborn screening

The CRANE Dataset was expanded in May 2014 on request to allow recording of 'timing of diagnosis' within 72 hours to align CRANE data collection with Newborn and Infant Physical Examination (NIPE) standards [67]. Analysis including data on this timing was not conducted for 2016 births (for this Annual Report) as only small numbers were recorded using the '≤72 hours' timing (n=48, 5.1%)[68]. We anticipate being able to report this in future.

5.2.2. Older children and young adults

As suggested above we will aim to run a multiple stage consultation process with members of CFSGBI and other stakeholders to identify if any current measures for 10 years and above can be identified. These will need to be assessed for validity and robustness before establishing if national consensus exists with any proposed measures.

5.2.3. Patient and Parent Reported Experience

The Cleft Psychology Clinical Excellence Network (CEN), upon request by the Craniofacial Society of Great Britain and Ireland (CFSGBI) Council and the Cleft Development Group (CDG), identified and piloted measurements[69] to evaluate patient (and parent) reported experience. These measures were combined into one Patient (and Parent) Reported Experience Measure (PREM) questionnaire and, through piloting[70] by the Cleft Psychology CEN, adjusted to apply to cleft services.

[67] In line with a statement of the UK National Screening Committee Newborn and Infant Physical Examination (NIPE) Standards and Competencies 1 document (2008) – setting out the standard for 95% newborn to be screened by 72 hours after birth (page 13 of the document found at http://newbornphysical.screening.nhs.uk/getdata.php?id=10639).

[68] Therefore we included '≤72 hours' cases within the '≤1 week' timing for this report and will report on '≤72 hours' referrals once this data item has come fully into use.

[69] (1) The Friends and Family Test (FFT) – developed by the Department of Health, and (2) the Experience of Service Questionnaire (CHI-ESQ) satisfaction assessment scales – developed by the Commission for Health Improvement (CHI).

[70] For full copies of documentation around the Cleft Psychology CEN review and pilot please contact Vanessa Hammond, Chair of Cleft Psychology CEN on vanessa.hammond@wales.nhs.uk.

The CRANE project team and the Cleft Psychology CEN collaborated to conduct a 14-month feasibility study to test PREM data collection, analysis and reporting, with a view to developing a method to implement this nationally.

The final report approved by the CDG was submitted to the CFSGBI Council for consideration at their 9th November meeting.

5.3. Data sources and future analyses

5.3.1. National Pupil Database (NPD)

The improved linkage between NPD and CRANE resulted from improving the quality of the postcode data held by CRANE. We are continuing our work with NPD and CRANE-HES-linked data. Future analyses will involve exploring in more detail children's educational attainment at Key Stage 2, when children are 11 years of age. Tracking children's educational attainment across different assessments as they age will allow us to study whether attainment gaps persist and to what extent, or whether children with a cleft do catch up with their peers in the general population.

Future analyses will also aim to examine the correlation between educational outcomes and treatment outcomes recorded in CRANE, such as speech quality, and we are interested in exploring whether there are aspects of the cleft treatment pathway, such as timing of repair, that may explain observed differences in educational outcomes within the cleft cohort.

We are collaborating with the Institute of Education research group, who have substantial previous experience in analysing NPD data and adjusting for the multiple factors that affect educational achievement.

5.3.2. Hospital Episode Statistics (HES)

The Clinical Effectiveness Unit is expecting to receive an updated HES – containing hospital data up to March 2017, early in 2018. This will allow us to refresh our analyses involving HES data, such as those involving the NPD, mortality, secondary speech surgery, and grommets. We are in the process of applying to NHS Digital to link these recent HES Data to CRANE to include in our analyses those births registered in CRANE since the last linkage exercise was conducted in March 2013.

5.3.3. Newborn Hearing Screening Programme (NHSP)

We are continuing to explore the option of requesting linkage between our CRANE Database and the Newborn Hearing Screening Programme (NHSP)[71] data – via Public Health England (PHE) – with the purpose of looking at the relationship between clefts and Permanent Childhood Hearing Impairment (PCHI) and the effect of PCHI on children's outcomes.

[71] http://hearing.screening.nhs.uk/

5.3.4. Dental data

We are seeking permission for linkage between our CRANE Database and NHS Dental data for England and Wales– with NHS Business Services Authority. With this linkage, we would aim to look at the quality of dental care and access for children with clefts of the lip and palate. We will be reporting on this linkage exercise in the 2018 Annual Report.

5.4. Quality Dashboard

The CRANE project team have submitted data for the 2016/17 and 2017/18 quality dashboards. This was done for the following five out of the six items requested – two of which were new requests (CLP01 and CLP02):

- Measure Number CLP00: The number of CRANE-registered children born within a specified quarter of the calendar year (refreshed every quarter).
- Measure Number CLP01: The number of Parents contacted by a Cleft team Clinical Nurse Specialist (CNS) within 24 hours of referral with an antenatal diagnosis of Cleft Lip and/or Palate – born within a specified quarter of the calendar year (refreshed every quarter).
- Measure Number CLP02: The number of Parents receiving visit from a Cleft team CNS within 24 hours of first referral (provided the child has not reached the age of one year) – born within a specified quarter of the calendar year (refreshed every quarter).
- Measure Number CLP06: The number of 5 year old children with a decayed, missing and filled teeth (dmft) index score, as a percentage of all 5 year old children (refreshed annually).
- Measure Number CLP09: The number of five year old children with 5 year old index scores 1 or 2 (as indicator of maxillary growth in patients with complete UCLP[72]) – as a percentage of the number of 5 year old children with a 5 year old index score (refreshed annually) [previously numbered CLP08].

The sixth item requested by Methods – the speech data – was once again provided directly by the centres. Specifically:

- Measure Number CLP07: The number of 5 year old children with green Cleft Audit Protocol for Speech – Augmented CAPS-A scores – (who have speech within normal range) as a percentage of the number of 5 year old children with a CAPS-A score (refreshed annually).

Future productions of Quality Dashboard CRANE tables have been confirmed – potentially including speech data (future dates have yet to be agreed).

[72] Atack NE, Hathorn IS, Semb G, Dowell T and Sandy JR. A new index for assessing surgical outcome in unilateral cleft lip and palate subjects aged five: reproducibility and validity. Cleft Palate Craniofac J. 1997 May;34(3):242-6.

5.5. Collaboration

- Work with the Cleft Collective to provide phenotype data, to allow the Early Career Researcher group the opportunity to report on this area in the UK.
- We are looking at data sharing with the National Congenital Anomaly and Rare Disease Registration Service (NCARDRS) which has replaced the previous BINOCAR project.
- We are collaborating with the UCL Institute of Education, research group, who have substantial previous experience in analysing NPD data and adjusting for the multiple factors that affect educational achievement.
- We are exploring how the Cleft Collective could undertake a linkage exercise to the CRANE data. Estimates of costs have been provided for the initial exercise and detailed proposal of the intention of the linkage has been drafted.

5.6. CRANE Communications

5.6.1. Dissemination of 2017 findings

- Publication of the Annual Report will be announced via our regular eNewsletter, which will be circulated in December, and it will be available on our website.

- We will also work with our close collaborators – such as the CFSGBI and CLAPA – to expand the reach of our eNewsletter (and the report).

- A *Summary of Findings for Patients and Parents/Carers* from this 2017 Annual Report will be produced in collaboration with CLAPA, and edited by the Plain English Campaign[73], with the aim of publishing it in early 2018.

5.6.2. Publications and presentations related to the CRANE Database

Publication(s)

- Kate Jane Fitzsimons, Lynn P Copley, Efrosini Setakis, Susan C Charman, Scott A Deacon, Lorraine Dearden, Jan H van der Meulen. Early academic achievement in children with isolated clefts: a population-based study in England Archives of Disease in Childhood Published Online First: 02 November 2017. doi: 10.1136/archdischild-2017-313777

Oral presentation(s)

- International Consortium Health Outcome Measures (ICHOM) Attending ICHOM Global Cleft Lip and Palate meeting May 2016 GOSH, London and subsequent conference calls including March 2017
- CFSGBI Annual Scientific Conference April 2017, Newcastle
- Toronto Pierre Robin Sequence, May 2017. We presented data at the 2nd International Robin sequence Consensus Meeting, Peter Gilgan Centre for Research and Learning, Toronto, May 7-8, 2017 (http://robin-sequence.com/). This will help develop our strategy on the data collection that will be proposed on PRS babies. We heard from Marie Wright about the RCPCH surveillance project on PRS at

[73] http://www.plainenglish.co.uk/

the last meeting and we await their data collection completion before working on a potential joint paper.

- Dutch Cleft meeting November 2017 presented the historical and current situation with national data collection in the UK at the Dutch Cleft meeting, Amsterdam, November 2017

5.7. Scotland

CRANE attended a meeting in December 2015 and presented a case to Scottish commissioners/clinicians with a view to securing their commitment to add their cleft care data to CRANE. Scotland service managers asked for an estimate of the costs for the annual levy and subsequently confirmed their intention to join the project. We are currently engaged in the process of setting up the necessary permissions for sharing data with Scotland

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 1,056 children born with a cleft in 2016 had been registered on CRANE at the time of preparing this report. This equates to an incidence of approximately one in every 682 live births in England, Wales and Northern Ireland[74].

Children can now be registered with CRANE prior to obtaining parental consent. However, consent must still be obtained so that complete outcomes data 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 third of the children born in 2016 had not been approached for consent at the time of preparing this report. Units with a high proportion of unconsented patients are encouraged to review their consent-taking process, to obtain consent in a timely fashion to enable the reporting of complete data[75].

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[76]. CRANE will continue to explore ways to improve communication and links with units to improve the submission of data in the future. Continued development of the CRANE Database and Website is expected to play a key role in facilitating this (see Chapter 5 for further detail on this).

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 should continue to 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 still falling below the NHS Fetal Anomaly Screening Programme target detection rate of 75%[77].

[74] 696,271 Births in England & Wales and 24,076 Births in Northern Ireland in 2016 (Office for National Statistics, https://www.ons.gov.uk/peoplepopulationandcommunity/birthsdeathsandmarriages/livebirths/datasets/birthsummarytables // Northern Ireland Statistics & Research Agency, http://www.nisra.gov.uk/demography/default.asp8.htm)

[75] See Report number 2. 'All consent' behind the CRANE Database log-in for further detail- https://www.crane-database.org.uk/

[76] See Report number 1. 'Outcomes' behind the CRANE Database log-in for further detail- https://www.crane-database.org.uk/

[77] Donna Kirwan and NHS Fetal Anomaly Screening Programme in collaboration with the Royal College of Obstetricians and Gynaecologists (RCOG), British Maternal and Fetal Medicine Society (BMFMS) and the Society and College of Radiographers (SCoR),

2. Less than a third of children with a cleft palate alone (28.2%) are being diagnosed late according to the national standard, whereby clefts should be diagnosed within 24 hours of birth to enable immediate referral to a specialist hospital[78].

3. 79.5% of the children born in 2016 with a cleft were referred by a maternity unit to a Cleft Unit within 24 hours of birth. This proportion varied significantly according to cleft type and the unit receiving the referrals.

4. Units established contact with 96.9% of parents within 24 hours of their child's referral; with no statistically significant difference according to cleft type, or between units receiving the referrals. This is positive and demonstrates the commitment of units to ensure timely response to new referrals of babies born with a cleft, to help support these babies and their families in the important initial stages.

5. Despite these positive figures reporting for the two points above, the proportion of children missing data for both referral time (36.3%) and first contact time (18.9%) have increased by 8% in 2016. This needs to addressed to return to a trend improvement year-on-year in this area. Prompt referral is recommended to ensure that the baby and their family receive appropriate care and support as soon as possible.

Cleft-related outcomes at five years

6. 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. Nevertheless, average treatment and care indices (of 75.5% and 67.9% respectively) across units indicate that, in the majority of cases, units have mechanisms in place to deal with any dental disease occurring.

7. Approximately one quarter of children with a complete UCLP have poor dental arch relationships (24.9%) that may benefit from further surgery to correct facial disproportion. While there is room for improvement, this proportion is substantially 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[79].

8. Close to three quarters of children (60.1%) with a complete speech assessment had speech scores that would suggest their speech is not significantly different from their non-cleft peer group, with scores suggesting they have achieved *normal speech*. This means that the national speech outcome standard #1 target of 55%[80] was not only met, but it was exceeded (on average) by the CRANE cohort born in 2007-2010.

NHS Fetal Anomaly Screening Programme. 18+0 to 20+6 Weeks Fetal Anomaly Scan National Standards and Guidance for England, 2010, NHS Fetal Anomaly Screening Programme: Exeter.

[78] Bannister P. Management of infants born with a cleft lip and palate. Part 1. Infant, 2008. 4(1): p. 5-8.

[79] Clinical Standards Advisory Group, Clinical Standards Advisory Group. Report of a CSAG Committee on cleft lip and/or palate, 1998, The Stationery Office, London.

[80] Based on the national outcome mean resulting from statistical analysis on 2004-06 Speech Outcome data – completed by the Lead Speech and Language Therapy Group, with statistical support from the Cleft Collective in Manchester, and presented in April 2014 to the Leads group.

Appendices

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, University Hospital Bristol NHS Foundation Trust University of Bristol
Jibby Medina	Research Fellow	Clinical Effectiveness Unit
Kate Fitzsimons	Research Fellow (On maternity leave in 2017)	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 (Until the 5 October 2017)	Clinical Effectiveness Unit
Christian Brand	Lecturer	Clinical Effectiveness Unit
Efrosini Setakis	Research Data Scientist	Clinical Effectiveness Unit / University College London (UCL) Institute of Education

Appendix 2: Members of the Cleft Development Group

Members of the Cleft Development Group (CDG)

Simon van Eeden	Chair, CDG
Victoria Beale	Clinical Director, North West, IoM & North Wales Cleft Network
Lorraine Britton	Lead Speech and Language Therapist, Trent Regional Cleft Lip & Palate Service
Alec Cash	Clinical Lead, South Thames Cleft Service
Chris Coslett	Specialised Planner, Women and Children's Services, Welsh Health Specialised Services
Sinead Davis	Chair, CEN for Cleft ENT and Hearing and Consultant ENT Surgeon
Scott Deacon	CRANE Clinical Project Leader
Mark Devlin	Lead Clinician of Cleft Care Scotland
David Drake	Cleft Surgery Training Interface Group
Yvette Edwards	Joint Representative of CDs and Managers Group
Norman Hay	Clinical Lead, North Thames Cleft Service
Chris Hill	Northern Ireland Clinicians
Peter Hodgkinson	Clinical Lead, Newcastle Site, Northern and Yorkshire Cleft Service & Chair Cleft Centres
Nichola Hudson	Lead Clinical Nurse Specialist
David Landes	Public Health Consultant
Karine Latter	Nursing deputy for Nichola Hudson
Kate le Marechal	Clinical Psychologists CEN
Sian Lewis	Medical Director - Welsh Health Specialised Services Committee
Kanwalraj Moar	Consultant cleft surgeon, Addenbrookes
Ailbhe McMullin	Specialist and Consultant Orthodontist, Royal Manchester Childrens' Hospital
Jason Neil-Dwyer	Clinical Director, Trent Cleft Service
David Orr	Cleft Services in the Republic of Ireland
Susan Parekh	Paediatric Dentistry CEN
Marie Pinkstone	Chair of the Lead Speech & Language Therapists (SLT) Group; and Lead SLT for North Thames Managed Cleft Network
Sandip Popat	Restorative Dentistry CEN
Jonathan Sandy	Lead of the Cleft Collective
Ian Sharp	Deputy Chair , CDG; CRG for Paediatric Services representative; and Clinical Director, West Midlands Cleft Centre
Bill Shaw	Lead at Manchester Clinical Trials Centre
Jackie Smallridge	Consultant Paediatric Dentist, CleftNetEast
Alistair Smyth	Clinical Lead for Leads Service
David Steel	Chair Programme Director, National Services Division, NHS Scotland
David Stokes	CLAPA Chief Executive
Imogen Underwood	President, Craniofacial Society, Principal Speech and Language Therapist, Birmingham Children's Hospital
Jan van der Meulen	Senior Epidemiologist, Clinical Effectiveness Unit
Jennifer Williams	Deputy for Per Hall and Lead Clinical Nurse Specialist, CleftNetEast
Mike Winter	Medical Director, National Services Division, Scotland

Appendix 3: NHS UK Cleft Development Group – Terms of Reference

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 (the Craniofacial Society Anomalies Register) and then CRANE (Cleft Registry and Audit Network). 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 Implementation 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 Implementation Monitoring Group which were 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 the funding of the CRANE Register and is responsible for ensuring that there is a contact in place with NHS England and that monies are paid annually to the hosting Clinical Effectiveness Unit at the Royal College of Surgeons. 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 and demographic 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, and 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 member's ex-officio who will be members during their terms of that office whether it is 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. The Group may decide to re-elect the holders or extend the period of office.

The composition will be:

1. Commissioners of Cleft Care. These should include a commissioner from NHS England, one from Wales, one from Scotland and one from Northern Ireland (each nominated by their equivalent national specialist commissioning body).
2. Public Health Consultants. These should include a representative of commissioning areas who are actively involved in cleft commissioning, and will normally be Consultants in Dental Public Health.
3. A Lay representative from a Parent Support Group (1) (to be nominated by CLAPA).
4. Cleft surgeons (2) (nominated by the surgical CEN).
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 Clinical Excellence Network).
9. A Clinical Nurse Specialist (1) (to be nominated by the lead Clinical Excellence Network).
10. A Psychologist (1) (to be nominated by the Cleft Psychologists Clinical Excellence Network).
11. A Paediatric Dentist (1) (to be nominated by the Cleft Paediatric Dentist Clinical Excellence Network).
12. The Co-ordinator/Chair of the UK Cleft Centres Clinical Directors' Group (1).
13. A Service Manager/Cleft Co-ordinator (1) (to be nominated by the Cleft Coordinators Clinical Excellence Network).
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 one or more 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. paediatricians, anaesthetists and genetics) as and when those disciplines have formally established national clinical excellence networks 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 October 2017

Appendix 4: 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, the Wales Specialised Health Services Committee, and the Northern Ireland Specialist Services Commissioning Team. Funds are raised through a levy calculated on a weighted per capita basis from the commissioning bodies in England, Wales and Northern Ireland. The levy is currently collected by Specialised Commissioning (East Midlands).

Appendix 5: Regional Cleft Centres and Managed Clinical Network and their associated regions / 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. Several of the Regional Cleft Centres are split between two hospitals, where the primary surgery is usually undertaken, and therefore Hospitals/ Administrative Units in a region may submit data separately to the CRANE Database , as shown in the Table below.

Regional Cleft Centre / MCN	Administrative Unit
Northern & Yorkshire	Royal Victoria Infirmary, 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 (GOSH), London Broomfield Hospital, Chelmsford
The Spires**	John Radcliffe Hospital, Oxford & Salisbury District Hospital, Salisbury
South Wales & South West	Morriston Hospital, Swansea University Hospitals Bristol***
South Thames	Guy's and St Thomas' Trust (GSTT), London
Northern Ireland	Royal Belfast Hospital for Sick Children, Belfast

Notes:
MCN – Managed Clinical Network.
*Data for GOSH and Broomfield units combined upon request by the Spires' Clinical Director (January1804 2017).
**Data for Oxford and Salisbury units combined upon request by the Spires' Clinical Director (June 2016).
***Frenchay Hospital, Bristol service moved to University Hospitals Bristol during 2014.

Appendix 6: Diagnosis and Procedure Codes, Hospital Episode Statistics (HES)

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) codes for cleft lip and cleft palate repairs.

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

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

Continued on next page...

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Code	Description
	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

Appendix 7: Timing of diagnosis detail

Number (%) of CRANE-registered children born in 2016 with a cleft lip and/or palate according to the timing of diagnosis and cleft type.

	Time of diagnosis in relation to birth[a] n (%)						
Cleft type	Antenatal	At birth	≤1 week[b]	≤1 month	≤6 months	>6 months	**All**
CL	148 (69.8)	58 (27.4)	2 (0.9)	0 (0)	4 (1.9)	0 (0)	212
CP	5 (1.3)	275 (70.5)	59 (15.1)	20 (5.1)	26 (6.7)	5 (1.3)	390
UCLP	158 (88.3)	21 (11.7)	0 (0)	0 (0)	0 (0)	0 (0)	179
BCLP	75 (82.4)	16 (17.6)	0 (0)	0 (0)	0 (0)	0 (0)	91
Not specified	20 (27.4)	35 (47.9)	8 (11)	4 (5.5)	3 (4.1)	3 (4.1)	73
All	**406 (43)**	**405 (42.9)**	**69 (7.3)**	**24 (2.5)**	**33 (3.5)**	**8 (0.8)**	**945**

Notes:

CL - Cleft Lip, CP - Cleft Palate, UCLP - Unilateral cleft lip and palate, and BCLP - Bilateral cleft lip and palate.

[a] 111/1,056 (10.5%) missing diagnosis time and excluded from 'All' values.

[b] Recording of 'timing of diagnosis' within 72 hours commenced in May 2014 to align CRANE data collection with NIPE standards[81]. With only small numbers having been recorded using this timing (n=47, 4.7%), we report '≤72 hours' cases within the '≤1 week' timing (until recording of this timing is well established).

[81] UK National Screening Committee Newborn and Infant Physical Examination (NIPE) Standards and Competencies 1 document (2008) – setting out the standard for 95% newborn to be screened by 72 hours after birth (page 13 of the document found at http://newbornphysical.screening.nhs.uk/getdata.php?id=10639).

Appendix 8: Five Year Old Index scores detail

Number (%) of CRANE-registered consented children born between 2004 and 2010 with a complete unilateral cleft lip and palate[82], according to Five Year Old Index scores and region / unit.

Regional centre / MCN	Administrative Unit	Five Year Old Index n (%) 1	2	3	4	5	All[a]
Northern & Yorkshire	Newcastle	8 (16.3)	7 (14.3)	19 (38.8)	10 (20.4)	5 (10.2)	49
	Leeds	6 (10.9)	25 (45.5)	15 (27.3)	8 (14.5)	1 (1.8)	55
North West & North Wales	Liverpool	4 (7.5)	16 (30.2)	16 (30.2)	12 (22.6)	5 (9.4)	53
	Manchester	3 (5)	24 (40)	16 (26.7)	9 (15)	8 (13.3)	60
Trent	Nottingham	5 (8.5)	23 (39)	19 (32.2)	6 (10.2)	6 (10.2)	59
West Midlands	Birmingham	8 (8.4)	33 (34.7)	28 (29.5)	21 (22.1)	5 (5.3)	95
East	Cambridge	4 (7.1)	20 (35.7)	17 (30.4)	13 (23.2)	2 (3.6)	56
North Thames	GOSH & Chelms.	6 (8.5)	30 (42.3)	24 (33.8)	11 (15.5)	0 (0)	71
The Spires	Oxford & Salisbury	13 (12)	37 (34.3)	34 (31.5)	19 (17.6)	5 (4.6)	108
South Wales & South West	Swansea	0 (0)	2 (18.2)	5 (45.5)	2 (18.2)	2 (18.2)	11
	Bristol	1 (2.7)	16 (43.2)	7 (18.9)	8 (21.6)	5 (13.5)	37
South Thames	Guy's and St Thomas'	10 (9.6)	39 (37.5)	29 (27.9)	17 (16.3)	9 (8.7)	104
Northern Ireland	Belfast	0 (0)	1 (25)	2 (50)	1 (25)	0 (0)	4
All	All	68 (8.9)	273 (35.8)	231 (30.3)	137 (18)	53 (7)	762

[a] Exclusions from 'All' values (not mutually exclusive): Children with an incomplete UCLP, children who died before the age of five, and 272/924 (29.4%) children missing Five Year Old Index scores data.

Appendix 9: Cleft Audit Protocol for Speech: Augmented scores detail

Number (%) of CRANE-registered consented children born with a cleft palate in 2007-2010, with reported speech outcomes, exclusion reasons and missing data at five years of age, according to region / unit.

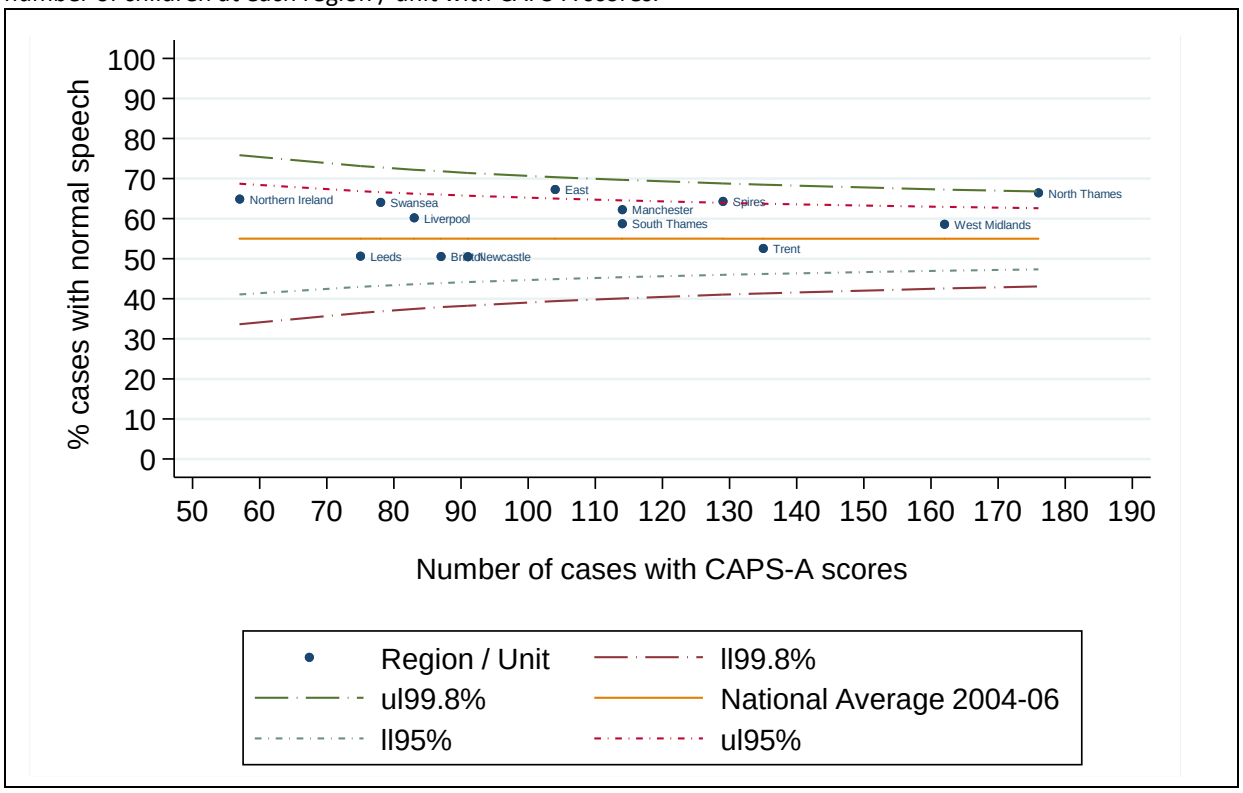
Regional centre / MCN	Administrative Unit	Speech* N	Reported n	Reported (%)	Reason outcome not collected n	Reason outcome not collected (%)	Total cases acc. for (%)	Missing Data n	Missing Data (%)
Northern & Yorkshire	Newcastle	182	122	67%	37	20%	87%	23	13%
	Leeds	179	106	59%	64	36%	95%	9	5%
North West & North Wales	Liverpool	223	105	47%	99	44%	91%	19	9%
	Manchester	229	154	67%	60	26%	93%	15	7%
Trent	Nottingham	267	178	67%	67	25%	92%	22	8%
West Midlands	Birmingham	295	205	69%	44	15%	84%	46	16%
East	Cambridge	217	136	63%	66	30%	93%	15	7%
North Thames	GOSH & Chelms.	361	212	59%	84	23%	82%	65	18%
The Spires	Oxford & Salisbury	265	172	65%	58	22%	87%	35	13%
South Wales & South West	Swansea	134	113	84%	17	13%	97%	4	3%
	Bristol	187	115	61%	24	13%	74%	48	26%
South Thames	Guy's and St Thomas'	256	162	63%	74	29%	92%	20	8%
Northern Ireland	Belfast	115	84	73%	12	10%	83%	19	17%
All	**All**	**2,910**	**1,864**	**64%**	**706**	**24%**	**88%**	**340**	**12%**

*Excluding 588/2,013 (29.2%) children with submucous cleft palates (2.6%), missing one or more of all 16 CAPS-A data items (1.4%), or born with either a CL (24%) or a non-specified cleft type (1.2%) are excluded from speech data.

[82] Submucous cleft palate patients excluded from all five year outcomes as all/most teams do not audit these patients.

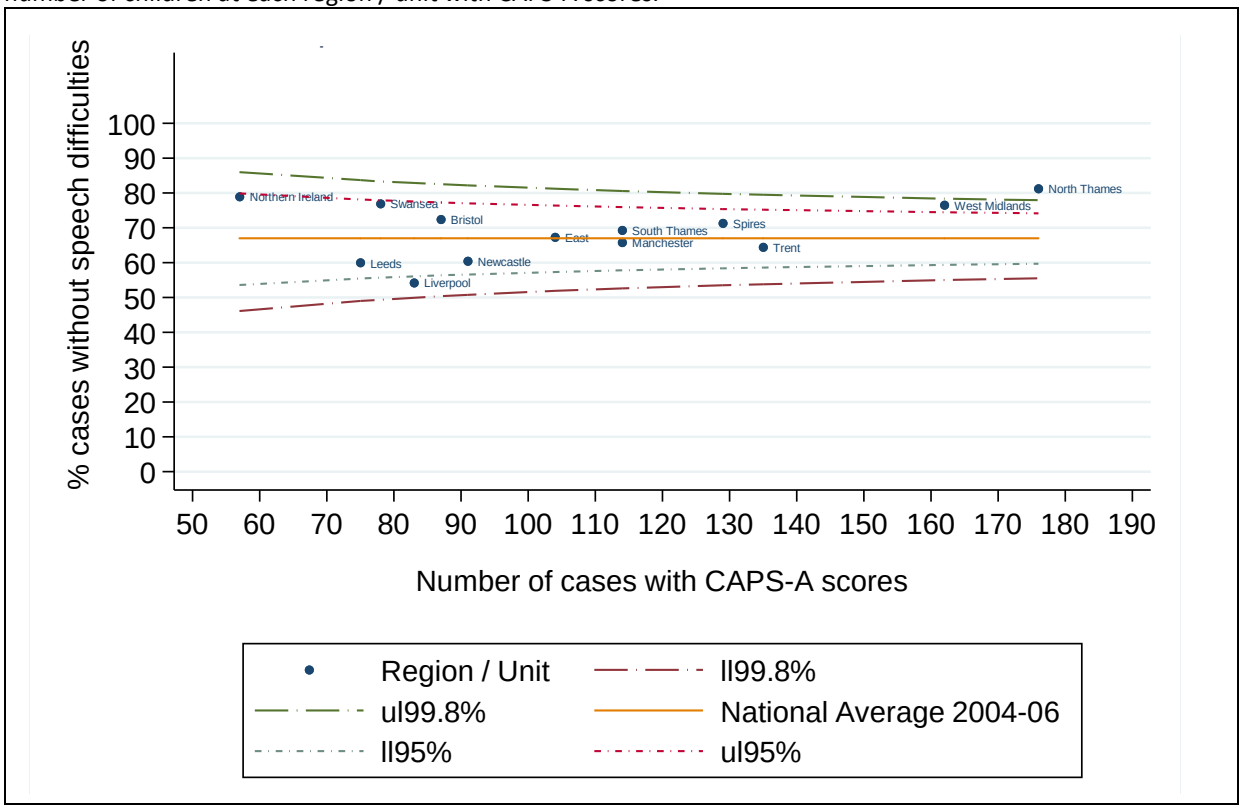
Appendix 10: Cleft Audit Protocol for Speech: Funnel plots for 2008-2010 births

Figure A. Funnel plot of five-year olds (born 2008-2010) with scores suggesting normal speech, according to the number of children at each region / unit with CAPS-A scores.



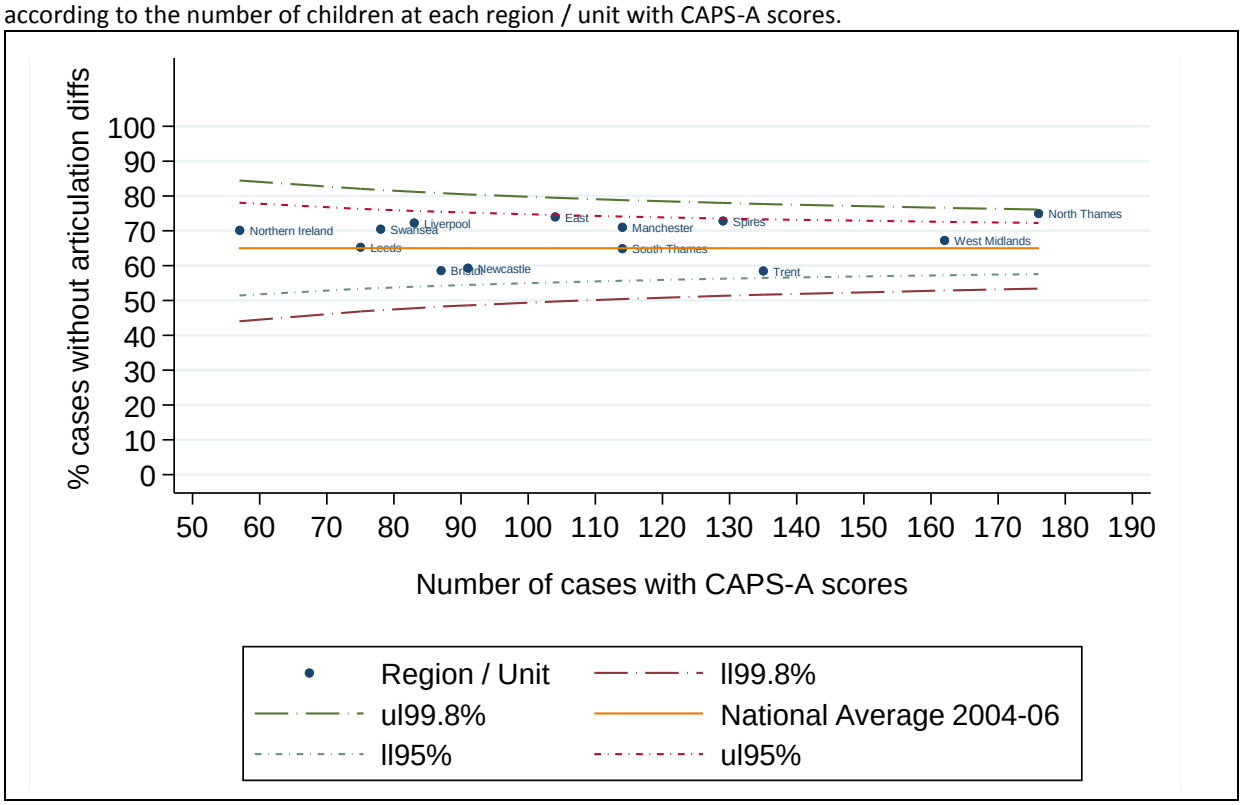
Note: Funnel plot centred on national average identified as the national outcome mean of 2004-06 speech outcome data of 55%.

Figure B. Funnel plot of five-year olds (born 2008-2010) with scores suggesting no speech difficulties, according to the number of children at each region / unit with CAPS-A scores.



Note: Funnel plot centred on national average identified as the national outcome mean of 2004-06 speech outcome data of 67%.

Figure C. Funnel plot of five-year olds (born 2008-2010) with scores suggesting no cleft-related articulation difficulties, according to the number of children at each region / unit with CAPS-A scores.



Note: Funnel plot centred on national average identified as the national outcome mean of 2004-06 speech outcome data of 65%.

Appendix 11: Cleft Audit Protocol for Speech: Scores for individual 16 CAPS-A parameters

Resonance and Nasal Airflow

In Table A, scores are colour-coded as green when 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.

Table A. Number (%) of CRANE-registered [a] consented children born with a cleft palate in 2007-2010, according to the four parameters for resonance and nasal airflow

Description	Score	N	(%)
RESONANCE – HYPERNASALITY			
Absent	0	1,356	(77)
Borderline – minimal	1	204	(11.6)
Mild – evident on close vowels	2	114	(6.5)
Moderate – evident on open and close vowels	3	51	(2.9)
Severe – evident on vowels and voiced consonants	4	35	(2)
RESONANCE – HYPONASALITY			
Absent	0	1,473	(83.7)
Mild – partial dentalization of nasal consonants and adjacent vowels	1	262	(14.9)
Marked – dentalization of nasal consonants and adjacent vowels	2	25	(1.4)
NASAL AIRFLOW – AUDIBLE NASAL EMISSION			
Absent on pressure consonants	0	1,617	(91.9)
Occasional: pressure consonants affected <10% of the sample	1	104	(5.9)
Frequent: pressure consonants affected >10% of the sample	2	39	(2.2)
NASAL AIRFLOW – NASAL TURBULENCE			
Absent on pressure consonants	0	1,382	(78.5)
Occasional: pressure consonants affected <10% of the sample	1	318	(18.1)
Frequent: pressure consonants affected >10% of the sample	2	60	(3.4)
TOTAL		**1,760**	**(100)**

[a] Registered in CRANE by 2 October 2017.

Cleft Speech Characteristics (CSCs)

Table B presents the cleft speech characteristics (CSCs). A colour coding of green indicates the CSC is absent or considered to be a minor speech characteristic unlikely to require intervention. A colour coding of amber or red indicates the CSC is affecting one or more consonants to the extent that therapy and / or surgery may be required.

Table B. Number (%) of CRANE-registered[a] consented children born with a cleft palate in 2007-2009, according to the twelve Cleft Speech Characteristics (CSCs) parameters.

Cleft Speech Characteristics (CSCs)		Score	N	(%)
ANTERIOR ORAL CSCs	1. Dentalisation / Interdentalisation	A	1406	(79.9)
		B	354	(20.1)
	2. Lateralisation / Lateral	A	1,643	(93.4)
		B	68	(3.9)
		C	49	(2.8)
	3 Palatalisation / Palatal	A	1,352	(76.8)
		B	210	(11.9)
		C	198	(11.3)
POSTERIOR ORAL CSCs	4. Double Articulation	A	1,709	(97.1)
		B	46	(2.6)
		C	5	(0.3)
	5. Backed to Velar / Uvular	A	1,508	(85.7)
		C	102	(5.8)
		D	150	(8.5)
NON ORAL CSCs	6. Pharyngeal Articulation	A	1,729	(98.2)
		C	20	(1.1)
		D	11	(0.6)
	7. Glottal Articulation	A	1,648	(93.6)
		C	58	(3.3)
		D	54	(3.1)
	8. Active Nasal Fricatives	A	1,601	(91)
		C	98	(5.6)
		D	61	(3.5)
	9. Double Articulation	A	1,727	(98.1)
		C	21	(1.2)
		D	12	(0.7)
PASSIVE CSCs	10. Weak and or nasalised consonants	A	1,661	(94.4)
		C	42	(2.4)
		D	57	(3.2)
	11. Nasal realisation of plosives	A	1,709	(97.1)
		C	25	(1.4)
		D	26	(1.5)
	12. Gliding of fricatives	A	1,725	(98)
		C	26	(1.5)
		D	9	(0.5)
		TOTAL	**1,760**	**(100)**

[a] Registered in CRANE by 2 October 2017.