CLEFT REGISTRY & AUDIT NETWORK

Promoting excellence in cleft care

Cleft Registry and Audit NEtwork Database

Part of the Clinical Effectiveness Unit, of the Royal College of Surgeons of England

2020 Annual Report: Methods

Results of the audit in England, Wales and Northern Ireland for children born with a cleft between January 2000 and December 2019

On behalf of the Cleft Development Group

CRANE Annual Report Methods

1. Data sources and processing

Patient-level data on aspects of cleft care and outcomes are sourced from (1) the CRANE Database, and (2) Hospital Episode Statistics (HES) data linked to CRANE data at an individual level.

1.1. CRANE: 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. The CRANE Database collects data pertaining to a child's birth, demographics, type of cleft, time of diagnosis, time of referral to a cleft care team, and time of first contact between a patient and cleft care team. The CRANE Database also collects information about cleft-related treatment and outcomes. These data are reported to CRANE by the cleft care teams that make up 10 Regional Cleft Centres / Managed Clinical Networks (as listed in Appendix A). Each child born with a cleft in England, Wales and Northern Ireland should be referred to one of these cleft care teams shortly after having their cleft diagnosed.

Since January 2012, the CRANE Database 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 being treated by the specialist cleft care teams. 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 cleft care teams at some point between referral and the first primary repair. A coordinator within each cleft care team submits data to the CRANE Database on the children referred to them. Once a record has been created on the CRANE Database for a particular child, it can later be updated with further information.

CRANE 2020 Annual Report: Cohort

All data entered into the CRANE Database on children born up until 31 December 2019[1] is included in the descriptions and analyses reported. Children whose parents have not consented to their data being used by CRANE have been excluded from the sections and tables on: (1) gestation and weight at birth, (2) 5-year outcomes, and (3) HES analyses (as the data presented in these sections and tables are not collected for non-consenting cases).

The children and timeframes covered in each chapter, and sub-section, are indicated in the summary tables at the beginning of each section of the CRANE 2020 Annual Report (where relevant). Broadly, timeframes are the most recent ten years of available data:

- Registry Information is reported for children born between 1 January 2010 and 31 December 2019.
- Audit Outcomes at 5 years of age are reported for children born between 1 January 2004 and 31 December 2013 – with the exception of speech (2007-2013 births) and psychology (2011-2013 births) outcomes.
- CRANE Research is reported for children born between 1 January 2004 and 31 December 2019, as appropriate to the analysis in each of the three sub-sections.

[1] By the 13 July 2020.

1.2. Hospital Episode Statistics (HES): Data source

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

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 may be used to identify any additional congenital anomalies and syndromes diagnosed for the CRANE cohort (see Appendix B for a list of the HES diagnosis and procedure codes used by CRANE). We use HES to identify whether a child should be classed as 'non-syndromic' or 'syndromic' for CRANE-HES linked research only.

1.3. Missing data

Missing data have been excluded from the denominators presented in all Tables, Figures and Appendices of the CRANE report, with the exception of Tables and Figures relating to data completeness.

2. Outcomes at 5 years of age

2.1. Body Mass Index

In order for a child to be defined as underweight, normal, overweight or obese, a measurement is required of weight according to height. A widely accepted measure of weight according to height is the body mass index (BMI), calculated for this report using weight in kilograms divided by the height in metres squared (kg/m2), as per the definition of BMI by the Royal College of Paediatrics and Child Health (RCPCH)[2].

BMI was estimated for all consented children registered in CRANE with a recorded height (m) and weight (Kg) at five years. These estimates were then classified into the following BMI categories, according to the RCPCH BMI classification shown below[2].

Body Mass Index (BMI) categories, according to the Royal College of Paediatrics and Child Health BMI classification.

Description	BMI (Kg/m2)
Underweight	Less than 13.0
Healthy weight	13.0 to less than 17.5
Overweight	17.5 to less than 19.0
Obese	19.0 or more

2.2. Decayed, missing and filled teeth (dmft)

A dmft score describes the dental caries an individual has experienced and is a measure of oral health. It reflects the total number of teeth that are decayed, missing or filled. Analyses on dmft and DMFT data

[2]According to the Royal College of Paediatrics and Child Health - 2-20 years Body Mass Index (checked October 2020). Available from: https://www.rcpch.ac.uk/sites/default/files/2018-03/boys_and_girls_bmi_chart.pdf

were restricted to consented children (excluding children with an unspecified cleft and submucous cleft palate). The risk of dental caries is thought to be higher among children with a cleft lip and/or palate compared with children without an oral cleft[3].

- A dmft >0 indicates experience of dental decay.
- A dmft >5 indicates experience of extensive dental decay.

2.3. Dental health measures of intervention

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. dmft scores of 0 or scores for all three 'm', 'f' and 'dmft' data items are required for the calculation of Treatment Index. When calculated, treatment indices range from 0 to 1 and are usually expressed as a percentage[4]. 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 provides the child with a dentition where the disease is controlled and the child has a pain free mouth

Care Index reflects cases where children have experienced dental decay, identified at the earliest possible stage (which is preferable), and have been provided with care in the least invasive form possible, i.e. fillings. A dmft score of 0 or scores for both 'f' and 'dmft' data items are required for the calculation of the Care Index. When calculated, care indices also range from 0 to 1 and are usually expressed as a percentage[5]. Care indices with a value close to 1 (100%) indicate that there are high levels of care provided by fillings (not extraction or no treatment), which is the desired outcome. In instances where a tooth is very poorly formed extraction may be the treatment of choice.

2.4 Five Year Old Index

Dental models obtained from 5-year old children with complete 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 complete UCLP before any other interventions are performed, such as orthodontics or alveolar bone grafting, which may influence this growth further[6]. The CRANE Database collected both internal and external Five Year Old Index scores for consented children 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 this report we have analysed externally validated scores where available; where these were unavailable, internal scores are included in the analyses.

[3] (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.

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

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

[6] 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.

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

CAPS-A ratings collected at five years of age among children born with a cleft affecting the palate are reported to the CRANE Database for consented children only. The parameters of assessed speech include resonance (hypernasality and hyponasality), nasal airflow (audible nasal emission and nasal turbulence) and twelve Cleft Speech Characteristics (CSCs) scores, 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;
- passive CSCs – for weak and or nasalised consonants, nasal realisation of plosives, and gliding of fricatives.

Speech outcomes are reported for non-syndromic children, as defined according to syndrome information entered into the CRANE Database, who have had all 16 CAPS-A ratings reported.

2.5. Psychology

Children are screened by psychologists at 5 years of age (and sometimes prior to that) using the **Tiers of Involvement Measure (TIM)** and the **Strengths and Difficulties Questionnaire (SDQ)**. The CRANE Database collected TIM and SDQ scores, as well as dates of psychological screening, for consented children with all cleft types.

The **Tiers of Involvement Measure (TIM)** is used to record the tier (level) of involvement when a Psychologist sees a patient/family in a Cleft Multi-Disciplinary Team (MDT) Clinic. The tiers are as follows:

0. Patient not seen by Psychologist.
1. Patient seen and psychosocial screen completed.
    a. No psychological concerns requiring cleft psychological input.
    b. Psychological support and/or needs met by other services e.g. Child and Adult Mental Health services (CAMHs).
2. Psychological input provided in clinic.
    a. Preventative input only.
    b. Input in response to a problem/concern raised by family/child.
    c. Both preventative input and input in response to problem/concern raised by family/child.
3. Further action required by Psychologist but appointment not necessary (e.g. liaison with school, written information sent to family).
4. Psychologist appointment necessary (for the purposes of this report, TIM scores 4, 5 and 6 are included in this category[7]).

The **Strengths and Difficulties Questionnaire (SDQ)** is a brief behavioural screening questionnaire designed for use with 3-16 year olds. The SDQ asks about 25 attributes, some positive and others negative, which are divided between the following scales:

[7] A score of 5 refers to a psychology appointment deemed as needed but resources do not allow for this to be offered in a timely way. A score of 6 refers to families who are already receiving psychology appointments when they are seen at age 5 years.

1. emotional symptoms (5 items)
2. conduct problems (5 items)
3. hyperactivity/inattention (5 items)
4. peer relationship problems (5 items)
5. prosocial behaviour (5 items)
6. scales 1 to 4 are added together to generate a 'Total difficulties' score (based on 20 items).

The CRANE Database collects the 'Total difficulties' score as well as the final scores for subscales 1 to 5, resulting from questionnaires completed by the parents of CRANE-registered children at 5 years of age[8].

Exploration of the data collected, using the six SDQ scores, has been conducted according to their categorisation into the following 4 bands: (1) Close to average, (2) slightly raised, (3) high, and (4) very high.

3. Analyses

3.1. Year of birth

Data have been analysed according to year of birth, unless otherwise stated. Reporting of each five-year outcome was restricted to children born in the birth years specified in the summary table at the beginning of each chapter. Children dying before five years of age and those with a submucous cleft palate were excluded from these analyses and from the denominator when calculating data completeness.

3.2. 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. L, A, H and/or S), an incomplete cleft (lower case letter, e.g. l, a, h and/or s), or no cleft (left blank). Where LAHSAL has not been reported (in approximately 6% of cases across the last 10 years), 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.

3.3. Statistical analyses

Proportions describing categorical outcomes (e.g. good, fair and poor 5-year old index scores) were compared across the categories of the primary explanatory variable of interest , such as year of birth and cleft type, using Chi-Square Tests. The means of normally distributed continuous outcomes (e.g. birth weight) were compared across categories using t-tests (when no more than two categories, e.g. boys and girls) and Analysis of Variance (ANOVA) (when there were three or more groups, e.g. cleft type). For non-parametric outcome data (e.g. Treatment Index and Care Index), a Kruskall-Wallis test was used to compare differences between categories.

[8] Using the parent version for 4-16 year olds. Goodman R (1997) The Strengths and Difficulties Questionnaire: A Research Note. Journal of Child Psychology and Psychiatry, 38, 581-586. For more information visit www.sdqinfo.com

Appendix A: Regional Cleft Centres / Managed Clinical Networks and associated cleft care teams

The CRANE Database covers England, Wales and Northern Ireland. Cleft care is currently delivered by eight Regional Cleft Centres and two Managed Clinical Networks (MCN). Several of the Regional Cleft Centres are split between two hospitals, where the primary surgery is usually undertaken, and therefore Hospitals / cleft care teams in a region may submit data separately to the CRANE Database, as shown in the Table below.

Regional Cleft Centre / MCN	**Hospitals / Cleft care teams**
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 cleft care teams combined upon request by the Spires' Clinical Director (January 2017).
**Data for Oxford and Salisbury cleft care teams combined upon request by the Spires' Clinical Director (June 2016).
***Frenchay Hospital, Bristol service moved to University Hospitals Bristol during 2014.

Appendix B: 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 malformations used to identify 'syndromic' cleft patients in CRANE-HES linked research work. A patient was 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 their HES episodes.

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