Cleft-affected children in Mayo: 1999–2007

Abstract
The aims of this study were to investigate the medical and dental care of children born in Mayo with cleft lip and/or palate in the period 1999–2007. Thirteen subjects were identified – nine males and four females. Galway was the main locus for cleft surgical referral and care, with 10 subjects receiving treatment by the same surgeon. A total of 54% of subjects required ENT care, with 62% receiving speech and language therapy. DMFT and dmft were zero and 0.037, respectively. Mean age at first dental visit (to a private or community dental clinic, specialist paediatric dental practice or Western Health Board orthodontic service) was 21 months. This represents progress in the objective of early access and provision of dental care for this special care dentistry needs group.

Introduction
Cleft lip and/or palate (CL/P) malformations are the most common congenital abnormalities of the craniofacial region.1-5 The incidence of CL/P varies according to race/ethnicity, gender, family history, socio-economic status and cleft type.2,6 It is more common among Indian and Oriental populations (1.7-2.3:1,000), and least common among Afro-Caribbeans (0.3-0.6:1,000).2,7 In Caucasians the reported incidence varies from 1:1,000 to 1:600 live births.1,3,5,7 The aetiology of cleft lip (CL) with or without cleft palate (CP) is complex.4 It is known that both genetic and environmental factors, acting either independently or in combination, are responsible for facial clefting.5,4

Aim
The objectives of this study were to identify and classify all cleft-affected children born in Co. Mayo, Ireland, between January 1, 1999, and December 31, 2007, and to investigate their access to and receipt of essential medical and dental services.

Subjects and methods
Dental records centralised at the Mayo General Hospital’s Dental Department were reviewed. The first aim was to identify all children born in Mayo with CL/P during the period January 1, 1999, to December 31, 2007. Two clinicians calibrated in WHO dmft/DMFT index protocols carried out the study. Clinical examination of the CL/P children comprised extra-oral and intra-oral examination. This included assessment of...
Results
Fourteen CL/P children were identified, of which 13 were born and resident in Co. Mayo in the period January 1, 1999 to December 31, 2007. All were Caucasian – nine males (69%) and four females (31%). The mean age at examination was five years and 10 months. The age range was two years and one month to nine years and four months. Four (31%) subjects had CL/P, seven (54%) had isolated CP and two (15%) had isolated CL (Figures 1 and 2).

In the isolated CP group (n=7), four patients were female and three were male. A submucous CP was identified in one female subject. In the CL/P (n=4) group, all were unilateral (UCL/P) and male. In this subgroup, there was an equal right-side/left-side involvement. Two subjects, both male, presented with isolated CL. Both were unilateral and left-sided. Three cases were syndromic, of which two were isolated CP and one was CL/P. Three syndromes, namely DiGeorge, Van der Woude and Goldenhar syndromes, had been definitively identified prior to their dental assessment. Systemic abnormalities diagnosed in these cases included hydrocephalus, cranial cyst, septo-optic dysplasia, nystagmus, absent corpus callosum, partial loss of sight, insignificant heart murmur as an infant, pre-auricular skin tags, cervical vertebral anomalies, fused ribs on right side and macrostomia. A positive family history was found in 31%. Seven children (54%) required ENT treatment, with placement of ventilation tubes on at least one occasion. Four (31%) have continued hearing problems requiring further ENT intervention. Eight children (62%) had received speech therapy in the past.

All cleft surgical care had been provided outside Co. Mayo. The plastic surgeon responsible for surgical repair was identified in 12 cases. One surgeon based in Galway was responsible for the operative repair of 10 children. A second surgeon based in Dublin was responsible for the care of the remaining two patients. The age at lip repair varied from three to seven months, with an average age at repair of 4.16 months. The mean age of palatal repair was 11 months, with a range of seven months to 16 months.

The age of first dental visit was identified in 11 (85%) subjects. Patient age at first dental visit varied from three months to seven years and one month, with a mean age of 21 months. With two infant subjects awaiting the commencement of dental eruption, the presence of decayed, missing and filled teeth was recorded in 11 children (85%). The mean recorded dmft was 0.037, with six having a zero dmft. Six children were in the mixed dentition. The DMFT for these children was found to be zero. Seven children (54%) had received fissure sealants, three on the permanent molars and the remaining four on their primary molars. Two children presented with an absent congenital permanent maxillary lateral incisor associated with the cleft-affected site. A talon cusp was identified in a maxillary lateral incisor in one child and another child presented with a supernumerary primary maxillary lateral incisor in the cleft-affected site. Incisor classification was used to determine malocclusion. Eleven children had a malocclusion. Five patients were Class III and the remainder Class I. Of the five children with a Class III malocclusion, three had isolated CP and two had CL/P.

Discussion
Facial clefts can be classified according to type, site, side and extent. CL/P infants born in Co. Mayo in the period under investigation (1999-2007) showed no significant difference in cleft type from previously reported international studies. In relation to gender, males are more commonly born with CL or a combination of CL/P, while for isolated CP, females are more commonly affected. While the number of subjects in this study is small, more males had an isolated CL/P, while more females had an isolated CP. In a previous
study of cleft-affected children born in the Western Health Board region, males predominated in the CL/P group.\textsuperscript{19-21} A variety of systemic anomalies, which may involve the cardiovascular, musculoskeletal, craniofacial and/or genitourinary systems, have been reported to affect 21-37\% of cleft patients.\textsuperscript{6,12} Three children (23\%) in this study had associated systemic anomalies. Middle ear disease and ENT problems in cleft-affected children have been extensively documented.\textsuperscript{6} Howevar-Boltezar\textsuperscript{11} demonstrated that ENT aural pathology appeared in almost 66\% of children with isolated CP or unilateral CL/P. Early ENT specialist evaluation for cleft-affected children is important.\textsuperscript{17} Auditory problems, though minor and confined to otitis media, were found in 54\% of subjects, and 31\% needed continued ENT care. Cleft-affected children are at risk of developing abnormal speech patterns.\textsuperscript{6} In this study, 62\% required and received speech therapy to assist with verbal communication.

Dental anomalies are common in cleft-affected patients. These comprise variations in the number of teeth, malformed roots and/or crowns,\textsuperscript{5,12} enamel hypoplasia and hypocalcification,\textsuperscript{6,14,15} eruption disturbances, natal and neonatal teeth, and ectopic position.\textsuperscript{6,14} Teeth in the cleft-affected site may show marked rotations, or retroclination giving rise to cleftside malocclusion.\textsuperscript{11} Hypodontia is reported to be significantly higher in cleft patients, with the maxillary permanent lateral incisor the most commonly missing tooth.\textsuperscript{16,17} Some 15\% of subjects in this study presented with hypodontia, and in all cases this involved the maxillary lateral incisor in the cleft-side. There is an increased tendency towards a Class III incisal relationship in cleft patients.\textsuperscript{1,13,19,20} In this study, 38\% presented with a Class III malocclusion.

Mayo has one maternity and paediatric centre. In 1999 protocols were established in the Paediatric Department of Mayo General Hospital to ensure that the Community Dental Service is informed of any CL/P patients.\textsuperscript{10} Mayo has one maternity and paediatric centre. In 1999 protocols were established in the Paediatric Department of Mayo General Hospital to ensure that the Community Dental Service is informed of any CL/P children born in Mayo between 1999 and 2007. Dental access and dental care for the patient with a cleft lip and palate. Part 2: From mixed dentition to adolescence and young adulthood. \textit{Br Dent J} 2000; 188 (3): 131-134.

\textbf{Conclusions}

Isolated CP was the most common cleft anomaly found in children born in Mayo between 1999 and 2007. Dental access and dental treatment were available to patients in Mayo from early childhood. Given a patient mean age of 21 months for the first dental visit, further investigation is desirable to establish the reason for the wide range of age for the first dental visit.

\textbf{References}