Initial cleft size does not correlate with outcome in unilateral cleft lip and palate

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SUMMARY Clinical outcomes in children born with a cleft lip and palate (CLP) have been an area of interest for orthodontists for a number of years. Whilst tools for measurement of these outcomes are available, there is no widely accepted measure of initial cleft severity and no known quantitative indices. Therefore, the potential influence of initial severity remains unmeasured and largely ignored. The aim of this investigation was to determine the importance of initial cleft severity in determining patient outcome.

The longitudinal records of 49 children born with a unilateral cleft lip and palate (UCLP), and treated in a single centre were examined. An index of initial cleft severity was developed that categorizes the cleft area as a percentage of the total palate area. The dental arch relationships of the same patients at 6 years of age were also determined. The nature of the association between these was investigated for agreement and correlation by calculation of weighted Kappa and Spearman’s correlation coefficient, respectively. No evidence was found in this sample that the initial cleft area had any bearing on the quality of outcome at 6 years of age.

Introduction

Interest in factors associated with outcomes for patients born with a CLP is high. The challenge now is to provide these patients with improved care and optimal results. Inter-centre studies have demonstrated that factors which influence outcomes include patient load and organization of services (Shaw et al., 1992; Sandy et al., 1998). Orthodontists have developed a number of quantitative measurements of outcome (Bergland et al., 1986; Mars et al., 1987; Atack et al., 1997a,b), as well as qualitative indicators for patient satisfaction (Thomas et al., 1997; Turner et al., 1997). If the main influence on patient outcome is treatment received and, more specifically, primary surgical repair, is there also a potential role for intrinsic factors such as the disturbance of normal growth and development, or the severity of the initial cleft (Seckel et al., 1995)? There is some suggestion that the intrinsic growth potential of CLP patients is deficient (Ross, 1987), but investigations into the dental and facial characteristics of unoperated cleft subjects reveal relatively little restriction of facial growth (Mars and Houston, 1990; Spauwen et al., 1993). The severity of a cleft is usually evaluated from its appearance at birth. However, the size of a cleft will reflect differing contributions of tissue deficiency and/or tissue displacement. Evidence suggests that cleft lip is associated with distortions and displacements, rather than tissue deficiencies (Cosman and Crikeland, 1965; Lindsay and Farkas, 1972). The size of the alveolar cleft in UCLP is governed mainly by the degree of transverse segmental separation and only to a lesser extent by deficiency of alveolar tissue (Shaw, 1978). Cleft palates alone exhibit great variability in tissue deficiency (Athanasiou et al., 1988) and this may influence cleft severity in a small number of cases (Shaw, 1978).

The width of the cleft might affect the difficulty of surgical repair and, thus, indirectly outcome. A wider cleft may require considerable
movement of the palatal mucoperiosteum, resulting in larger areas of bone being denuded and the generation of more scar tissue, with subsequent contraction of the maxilla. The aim of this investigation was to determine the importance of initial cleft severity in determining patient outcome, by examining the severity of the initial cleft presentation and correlating this with an indicator of facial growth in the same patients at the age of 6 years.

**Subjects and methods**

**Sample identification**

The source of all subjects was the Cleft Lip and Palate Unit at Princess Margaret Hospital (PMH) in Perth, Western Australia. These patients received pre-surgical orthopaedic treatment from birth until lip repair, which comprised an intra-oral acrylic plate, plus extra-oral bonnet and strapping. At 3 months, the primary lip repair together with a Muir flap, an anterior palate repair and a primary rhinoplasty, were carried out. The palate was repaired at around 12 months using Wardill or VY push-back techniques. A single plastic surgeon undertook nearly all cleft surgery until his retirement in 1991, after which two surgeons were involved in providing this treatment.

A protocol for the collection of standardized records was introduced in 1985, which included study casts and photographs taken at birth, and ages 6, 9, 12, and 18 years. Thus, a comprehensive archive of standardized, longitudinal records existed for all cleft patients born since 1985.

The sample criteria were children born with a complete UCLP, including a soft tissue band of less than 5 mm, who had neonatal (before any intra-oral plate provision) and 6-year-old study casts. The sample was identified from the PMH database, and the nature of the cleft was verified by examination of birth study casts and photographs. Each subject was allocated an identification number.

Randomly generated numbers were used to assign models for measurement exercises. A total of 49 subjects who had neonatal, as well as their corresponding 6-year-old study casts were identified.

**Development of the Birth Severity Index**

The neonatal study casts were placed on a scanner with the alveolar crest resting against the surface. All the models were scanned on the same occasion, and the images were stored in Paint Shop Pro software (Microsoft Corporation, Redmond, WA, USA) as ‘TIF’ files. The Windows version of Scion Image PC (Scion Corporation) software, in conjunction with a digitizing tablet, were used for study cast analysis.

A single operator (NJ), under standard conditions, viewed the study cast images in random order. First, the outline of the palate was traced, following the alveolar crest, across the alveolar cleft anteriorly, and posteriorly the inter-tuberosity line. Secondly, the outline of the cleft itself was traced, again following the line of the alveolar crest anteriorly and the inter-tuberosity line posteriorly. Subsequently, the cleft area was calculated as a percentage of the palate area. Two weeks later, the study casts were re-randomized and retraced by the same operator under the same conditions. The average percentage value for each subject was calculated from the duplicate measurements.

The cleft area/palate area percentages were plotted to determine the nature of the sample distribution. Subsequently, the boundaries for five categories of the index were identified. This was calculated using parametric (mean and standard deviation) and non-parametric criteria (percentiles). The subjects were then categorized into one of the five index grades.

**Method error estimation**

Method error was investigated by analysis of the replicated measurements. Calculation of paired t-tests, mean difference, and standard deviation of the difference was necessary to determine and differentiate random and systematic errors.

**Assessment of 6-year-old models**

The 6-year-old study casts were laid out in random order and graded individually by two
examiners (NJ and AW) trained in the use of the 5-year-old index for dental arch relationship analysis (Atack et al., 1997a,b). After a suitable delay, the study casts were re-randomized, re-assigned on a work bench, and re-graded. Intra- and inter-examiner agreement for the 5-year-old index was analysed by calculation of Kappa statistic, with application of linear weighting.

**Testing the association between the severity at birth, and the dental arch relationship at 6 years old**

The relationship between the birth severity index grade and the 6-year-old dental arch relationship grade using the 5-year-old index, for each subject, was analysed. Agreement was tested with the Kappa statistic and correlation determined by Spearman’s correlation coefficient.

**Results**

**Development of birth severity index**

The mean cleft area/palate area proportion was 36.9 per cent, median 37.2 per cent, and standard deviation 5.3 per cent. The range was from 26.6 to 50.7 per cent. Since the nature of the distribution was ambiguous (Figure 1), it was decided to calculate the boundaries for the five grades of the categorical index using parametric and non-parametric criteria.

For the parametric criteria, mean and standard deviation were used such that the boundaries were:

1. mean minus 2 SD;
2. mean minus 1 SD;
3. mean plus 1 SD;
4. mean plus 2 SD.

For the non-parametric criteria, the 20th, 40th, 60th, and 80th percentiles were used. The distribution of the sample within the birth severity index, using these criteria are given in Table 1.

The relationship between the ‘parametric criteria’ index grades and the ‘non-parametric criteria’ index grades was investigated for agreement and correlation, using weighted Kappa and Spearman’s correlation coefficient, respectively. The agreement was found to be moderate, with a Kappa value of 0.55, and the correlation was significant \((P < 0.01)\) with a coefficient value of 0.80.

**Table 1** Birth severity index distribution.

<table>
<thead>
<tr>
<th>Index grade</th>
<th>Parametric criteria</th>
<th>Non-parametric criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Frequency</td>
<td>Percentage</td>
</tr>
<tr>
<td>1</td>
<td>0</td>
<td>9</td>
</tr>
<tr>
<td>2</td>
<td>9</td>
<td>19</td>
</tr>
<tr>
<td>3</td>
<td>34</td>
<td>69</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>5</td>
<td>2</td>
<td>49</td>
</tr>
</tbody>
</table>
Error estimation

The results are presented in Table 2. The paired t-tests demonstrated no significant differences between observations. Although the systematic errors were small, the random errors were larger. The standard deviation was 22 per cent of the mean total palate area and 18 per cent of the mean cleft area. Replication of measurements and the averaging of the results will reduce these random errors (Houston, 1983).

Assessment of 6-year-old models

This sample of 49 subjects with initial birth records also had study casts, from around 6 years of age, available. The mean age of the subjects at impression taking was 6 years 3 months and the standard deviation was 5 months, with a range 5 years 1 month to 7 years 8 months.

Intra-examiner agreement for the 5-year-old index was found to be very good to good, with weighted Kappa values of 0.85 and 0.76. Inter-examiner agreement was found to be good, with weighted Kappa values of 0.70 and 0.65.

The results of the grading of study casts using the 5-year-old index are shown in Figure 2.

Testing the association between the severity at birth and the dental arch relationship at 6 years old

The association between the birth severity index grade and the 6-year-old dental arch relationship grade was investigated for agreement and correlation, using weighted Kappa and Spearman’s correlation coefficient, respectively. The results are presented in Table 3 and suggest that there is no association between them.

Discussion

This study demonstrates, albeit in a sample from one unit, that the severity of the initial cleft (measured as a proportion of the total palate) does not correlate with the outcome in terms of dental arch relationship. Several issues arise from this study.

<table>
<thead>
<tr>
<th>Paired t-test</th>
<th>Total palate area</th>
<th>Cleft area</th>
</tr>
</thead>
<tbody>
<tr>
<td>t = 0.53 (NS)</td>
<td>t = 1.21 (NS)</td>
<td></td>
</tr>
</tbody>
</table>

| Mean difference | 0.02 | 0.03 |
| SD of difference| 0.28 | 0.17 |

Table 2 Method error estimation for birth severity index.

<table>
<thead>
<tr>
<th> </th>
<th>‘Parametric criteria’ index</th>
<th>‘Non-parametric criteria index</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weighted Kappa</td>
<td>-0.10</td>
<td>-0.01</td>
</tr>
<tr>
<td>Spearman’s rho</td>
<td>0.03</td>
<td>-0.05</td>
</tr>
</tbody>
</table>

Table 3 Analysis of the relationship between the birth severity index grades and the 6-year-old dental arch relationship grade.
In order to investigate the role of initial cleft severity in determining patient outcome, methodology for assessment of severity is necessary. Several measurement systems have been designed for the quantitative analysis of palatal dimensions of the newborn’s palate. A series of landmarks have been developed and are widely accepted (Sillman, 1964; Stöckli, 1971; Robertson et al., 1977). However, there is no gold standard for validity testing of landmark positions (Seckel et al., 1995). These are often quoted as reproducible with no statistical evaluation. A well-constructed investigation of landmark precision suggests that errors can be substantial, up to 10 per cent, and that experience of the operator is important (Seckel et al., 1995).

The common medium for quantitative assessment of palatal dimensions is dental casts. These may be combined with facial casts to enable three-dimensional (3D) analysis of the maxilla (Wada et al., 1984; Bacher et al., 1998). Graphic reproduction of dental casts is also common. This involves photocopying techniques to generate a two-dimensional (2D) image (Stöckli, 1971; Mazaheri et al., 1971; Krischer et al., 1975; Schwartz et al., 1984). Generation of a simulated 3D computer image has now become possible using stereophotogrammetry (Berkowitz and Pruzansky, 1968; Berkowitz, 1971; Berkowitz et al., 1974), computer controlled 3D co-ordinate digitizing apparatus (Mishima et al., 1996) and a combination of precision camera with Moire light illumination (Bacher et al., 1998). Technological advances have also yielded devices to assist in measurement, including the Reflex microscope (Kriens, 1991; Heidbuchel et al., 1998), Optocom (Van der Linden et al., 1972; Heidbuchel and Kuijpers-Jagtman, 1997), digital sliding callipers (Honda et al., 1995), and a 3D digitizer system (Kramer et al., 1994, 1996a,b).

Methods are also available for calculation of palate surface area. Techniques have evolved from the measurement of 2D photocopies of study casts (Huddart, 1970) to more complex 3D images. Stereophotogrammetry enables division of the surface into a number of plane triangles, and the area of each triangle is then computed and summed to yield the total area (Berkowitz et al., 1974). The smaller the number of triangles, the greater the accuracy, but the greater the number of co-ordinates necessary. Even with 200 triangles, a 0.3 per cent error exists, although this is very consistent. It is a costly and lengthy procedure, which requires elaborate equipment and a specialized operator (Newton and Evans, 1976). More ingeniously, Huddart et al. (1978) used a vacuum moulded plastic template of the alveolar segment area, which was then cut and flattened under a piece of glass before being measured with a planimeter. The error is reported as 2.1 per cent for cleft models, but 7.1 per cent for intact palate measurement. Cooling shrinkage from the vault of the palate may in part explain this error.

Despite considerable literature in this area, relatively few studies report measurement of the actual cleft defect. Cleft dimensions are described in terms of cleft widths. Anterior cleft width is the alveolar cleft along the crest of the ridge, middle cleft width is at the intersection of the inter-canine point line and posterior cleft width is at the intersection of the inter-tuberosity line (Mazaheri et al., 1971; Berkowitz et al., 1974; Krischer et al., 1975; Kramer et al., 1994; Honda...
et al., 1995). This may be supplemented by the oblique cleft width, which is the distance from the tip of the lesser segment to the margin of the cleft along a line parallel to the inter-tuberosity line (Mazaheri et al., 1971; Shaw, 1978; Schwartz et al., 1984).

There is some evidence that measurements in the cleft region are subject to greater error than those taken elsewhere, especially the posterior cleft width as it is constructed (Shaw, 1978). However, in a study of landmark and measurement precision, alveolar cleft width was the second best of those analysed (Seckel et al., 1995).

No reports of surface area measurements of the cleft defect are known. Stereophotogrammetry is probably the most accurate technique for palatal surface area measurement, but it is costly and lengthy, and requires specialized training and complex equipment. In contrast, the technique used in this study is simple and flexible with no special equipment or training necessary.

There has been no consideration of the relative proportion of the defect to the palatal segments. Furthermore, no categorical indices of initial cleft severity currently exist. To determine the importance of initial cleft severity, it is necessary to investigate the relationship between initial severity assessment and patient outcome. There are few other longitudinal studies that report both palatal cleft dimensions and measure subsequent arch development (Mazaheri et al., 1971; Krischer et al., 1975; Schwartz et al., 1984; Friede et al., 1988; Kramer et al., 1994, 1996a; Honda et al., 1995). One publication investigated arch dimensional changes from birth to 5 years old for a variety of cleft types (Mazaheri et al., 1971). They found a wide variation in initial arch form and a variable pattern of arch development. Surgical closure of the lip had a significant effect upon reduction in alveolar and palatal cleft width, and on segmental relationships. Later reduction in cleft width was attributed to segmental repositioning and growth of the palatal shelves. A pattern of reduced antero-posterior and lateral dimension development compared with normal subjects was identified. At 5 years the maxillary width was reduced, but the maxillary length was similar to normal subjects. For UCLP subjects, 26 per cent had a complete buccal crossbite, 3 per cent had an anterior crossbite, and 6 per cent had anterior and buccal crossbites.

The data from this investigation suggests that the initial presentation of UCLP is probably a poor predictor of outcome. Only a few studies have been published on prediction of palatal growth and development in cleft lip and palate children (Krischer et al., 1975; Schwartz et al., 1984; Friede et al., 1988; Kramer et al., 1996b). Most of these were retrospective and restricted to the prediction of crossbites based on presurgical palatal measurements. Using probability trees, Krischer et al. (1975) investigated alveolar cleft widths prior to lip surgery and arch form following lip surgery, as predictors of crossbites. For subjects who had received pre-surgical orthopaedics neither was a predictor, but for subjects treated with surgery only, both variables were predictors of crossbite. Schwartz et al. (1984) used discriminant analysis to identify four pre-lip surgery variables (width of alveolar process and palatal shelf on the lesser segment; total antero-posterior length of the cleft segment; maxillary inter-canine width; and mandibular retromolar width). When combined and weighted these could correctly predict crossbites in 90 per cent of subjects. However, multiple linear regression analysis techniques were less successful, enabling the correct prediction of only 50 per cent of crossbites (Friede et al., 1988). A subsequent study, involving multilevel modelling of longitudinal data, failed to find any predictive measurements for UCLP palatal growth and development, but detected a relationship with growth velocity (Kramer et al., 1996b).

Methods exist for more comprehensive analysis of occlusal relationships in UCLP patients than simply crossbite presence or absence. Dental arch relationship indices for assessment at 5 (Atack et al., 1997a,b) and 10 years of age (Mars et al., 1987) are now well established as outcome measures (Sandy et al., 1998). These both place importance on antero-posterior incisor relationship in addition to transverse relationships. No previous study has compared initial palatal cleft severity with these widely accepted indices.

The present investigation was only possible because of a farsighted record policy at PMH, which was established 15 years ago. Elsewhere,
similar strategies have yielded invaluable information providing sufficient evidence to design more challenging and powerful studies (Roberts et al., 1991; Semb, 1991). Constancy of operators and technique is a further variable that was controlled, to some extent, in this investigation. Further sampling from other centres with similar archives is probably needed to determine the reliability of the observations. The relatively normal distribution of the initial cleft area and the 6-year-old outcomes led us to believe that the sample examined was representative. The lack of correlation between the initial defect and the 6-year-old outcome was surprising, but suggests surgeons may develop their own techniques for coping with these varied presentations.

The lack of any detectable relationship between initial severity and outcome implies that the severity of the cleft at birth does not play a significant role in UCLP patient outcome and that severity may be outweighed by other factors.

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