Cervical Vertebral Anomalies in Cleft Lip and Palate

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A survey was made of the upper cervical vertebrae in children with cleft lip and palate in order to determine the prevalence of cervical vertebral anomalies. The cleft sample consisted of 105 patients attending for orthodontic treatment. It was subdivided into cleft lip (CL), cleft palate (CP), unilateral cleft lip and palate (UCLP) and bilateral cleft lip and palate (BCLP) subgroups. A control was comprised of 120 orthodontic patients.

The cervical vertebral anomalies were classified into two types, posterior arch deficiency (PAD) and fusion anomalies (FUS).

The results confirmed that the cervical vertebral anomalies occurred significantly more often in the cleft sample (13%) than in the controls (0.8%), p < 0.001.

The total prevalence of cervical vertebral anomalies was similar in the four cleft sub-samples, but the occurrence of each of the two types of cervical vertebral anomalies showed a more differentiated pattern.

Posterior arch deficiency occurred significantly more often in cleft palate (CP), (16%) than in controls p < 0.001, but fusions did not occur more frequently in any cleft groups than in controls.

The neurological significance and diagnosis of abnormalities of the cervical spine has been described by Chamberlain (1939), McGregor (1948), McRae (1952, 1960), and Bull et al (1955). Their work was based on conventional nonstandardized radiographic techniques, so differences in magnification and positioning were involved.

The prevalence of cervical vertebral anomalies in a noncleft sample was investigated by Farman et al (1978). In 220 standardized lateral skull radiographs of referred orthodontic patients, 3.2 percent of the sample was found to exhibit posterior dehiscence of the atlas.

The radiographic appearance of anomalies of the cervical vertebrae on standardized lateral cephalometric films was described in detail by Farman et al (1978) and Farman and Escobar (1982). These anomalies include variations in the normal anatomy of the atlas, progressive degrees of lipping, supernumerary vertebrae, accessory ossicles, block vertebrae, and spina bifida.

The occurrence of cervical vertebral anomalies in subjects with cleft lip and palate has been reported by several authors. Cohn (1963) observed the occurrence of cleft palate in patients with Klippel-Feil syndrome, which includes block fusion of cervical vertebrae. Osborne et al (1971) investigated the prevalence of cervical vertebral anomalies in children with cleft lip and palate. The findings indicated that cervical vertebral anomalies were present more often in cleft palate than in other cleft categories or the control group.

On the other hand Minaba (1972) in a study of cervical vertebrae in patients with cleft lip and palate and in controls, found that the incidence of the abnormality, and especially fusion of cervical vertebrae, was higher in the group with cleft, but that no relation to the type of clefting was observed.

Since cleft lip and palate occurs in connection with a number of more generalized and less readily noticeable anomalies, it is important to draw attention at the initial diagnosis of clefting to the
possibility of other deformities (Pashayan, 1983). Although previous reports agree that patients with cleft lip and palate exhibit a higher prevalence of cervical vertebral anomalies than controls, the distribution of the type of anomaly in relation to the different cleft types has so far not been analyzed.

The aim of the present investigation was to examine the occurrence of the types of cervical vertebral anomalies in the various categories of cleft lip and palate and to compare this with the occurrence of cervical vertebral anomalies in a control group.

The Normal Cervical Spine

Difficulties in diagnosis of cervical vertebral anomalies can be overcome with knowledge of the normal anatomy and radiographic appearance of the cervical spine (Bailey, 1952; Farman and Escobar, 1982). The seven cervical vertebral units seen on a standardized lateral skull radiograph are separated by an intervertebral disc between the body of each unit. There are complex articulations between the atlas and the occipital condyles and between the atlas and axis (Hohland Baker, 1965).

The atlas, or C1, is a ring of bone with no body and no posterior spine; the anterior arch of the atlas has a tubercle which extends forward about 3 mm further than the front of the other vertebrae, the tip of which lies on a horizontal line drawn through the atlas (Huggare and Kylamarkula, 1985; Kylamarkula and Huggare, 1985). This anterior tubercle projects very close to the posterior wall of the nasopharynx. It has a role to play in the establishment of velopharyngeal competence, particularly in a child with cleft palate (Osborne et al, 1971). In normal head posture the posterior arch of the atlas should be clear of the base of the skull by 8 to 10 mm (Kylamarkula and Huggare, 1985), of the odontoid peg should be below basion (Chamberlain, 1939). Intrusion of the odontoid process of C2 into the foramen magnum is called basilar invagination (McGregor, 1948).

The posterior arch of C1 encircles the neural cord and is fused in the midline after 4 years of age (Last, 1978). A radiopaque shadow of the inner cortical plate of the posterior arch can be seen when this has taken place (Fig. 1).

The axis, or C2, is the largest cervical vertebral unit and is identified by the presence of the odontoid process, a projection which extends from the body of C2 through the neural arch of the atlas. The normal radiographic appearance of this process is a peak with a rounded summit having a steeply inclined anterior surface and a

![FIGURE 1 Radiographic appearance of the normal cervical spine as seen in the lateral cephalometric radiographic projection, with accompanying trace.](image-url)
vertical posterior edge (Fig. 1). A space exists between the vertical posterior edge of the dens and the inner surface of the anterior arch of the atlas. This is called the atlas-dens interval (ADI). A survey of lateral radiographic views of the cervical spine of young subjects aged 3 years to 15 years (Locke et al, 1966) showed this interval to be no longer than 4 mm. The axis has the largest spinous process which can be palpated in the neck as the first bulge encountered when the finger is drawn down the neck from the occipital protuberance. The lateral masses have superior and inferior articulations, and fusions can occur at these points (Fig. 2A).

The cervical vertebral units C3 to C6 are all similar with well outlined bodies (Fig. 1). The lateral masses have upper and lower articular facets, and the posterior arch has a bifid spinous process.

**MATERIAL AND METHOD**

The sample with cleft was comprised of 105 children (53 males, 52 females) chosen at random from the hospital records of patients with cleft lip and palate deformity. Those below the age of 6 years were excluded, because failure of upper cervical vertebral components to de-

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![Figure 2 A](image1.png)  
**FIGURE 2 A.** Spina bifida at C1 (PAD), B. Dehiscence at C1 (PAD). Block fusion of C2,3,4 and C4,5,6 is also present.
TABLE 1 Survey of Sample

<table>
<thead>
<tr>
<th>Diagnostic Group</th>
<th>Male</th>
<th></th>
<th>Female</th>
<th></th>
<th>Total</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N</td>
<td>%</td>
<td>N</td>
<td>%</td>
<td>N</td>
<td>%</td>
</tr>
<tr>
<td>Cleft lip</td>
<td>6</td>
<td>11.3</td>
<td>4</td>
<td>7.7</td>
<td>10</td>
<td>9.5</td>
</tr>
<tr>
<td>Cleft palate</td>
<td>16</td>
<td>30.2</td>
<td>22</td>
<td>42.3</td>
<td>38</td>
<td>36.2</td>
</tr>
<tr>
<td>Unilateral cleft lip and palate</td>
<td>21</td>
<td>39.6</td>
<td>20</td>
<td>38.5</td>
<td>41</td>
<td>39.0</td>
</tr>
<tr>
<td>Bilateral cleft lip and palate</td>
<td>10</td>
<td>18.9</td>
<td>6</td>
<td>11.5</td>
<td>16</td>
<td>15.2</td>
</tr>
<tr>
<td>Total cleft sample</td>
<td>53</td>
<td>52</td>
<td>105</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Control</td>
<td>60</td>
<td>50</td>
<td>120</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

velop or fuse can only be determined after the usual time for complete development and fusion has passed. The sample was classified according to cleft type: cleft lip (CL), cleft palate (CP), unilateral cleft lip and palate (UCLP), and bilateral cleft lip and palate (BCLP).

The total occurrence of the various types of clefts (Table 1) included 10 patients with cleft lip (CL) (9.5%), 38 patients with cleft palate (CP) (36.2%), 41 patients with unilateral cleft lip and palate (UCLP) (39.0%), and 16 patients with bilateral cleft lip and palate (BCLP) (15.2%). The occurrence did not differ between sexes.

The control sample consisted of 120 children (60 males, 60 females) sequentially referred for orthodontic treatment, and data collection involved the tracing of standardized lateral skull radiographs.

The radiographs were taken in a Morita Panelipse machine with attached cephalostat. The focus median plane distance was 150 cm, and the median plane distance to the film was 15 cm. Initially the radiographs were examined for the cervical vertebral anomalies listed in Table 2. In the sample with cleft each subject chosen for the study had a series of films available for scrutiny. The radiograph selected was the one that most clearly demonstrated the anomaly.

For the purposes of analysis the cervical vertebral anomalies were categorized into two main groups: (1) posterior arch deficiency (PAD): spina bifida (Fig. 2A); dehiscence (Fig. 2B), and (2) fusions (FUS): fusion of two components (Fig. 3A); block fusion (Fig. 3B); occipitalization (Fig. 3C).

For each subject in the control group and the cleft sample, the first four cervical vertebrae and the atlanto-occipital articulation were traced on matte acetate tracing paper. A radiograph demonstrating the most normal appearance of the cervical spine was chosen from the control series of standardized lateral skull radiographs of the patients without cleft. A tracing was made and used for comparative purposes (Fig. 1). Normal appearance was determined when the first four cervical vertebrae of each subject could be traced as separate entities, with all anatomical components present (Fig. 1). The criteria used for inclusion in the anomaly category relied on careful scrutiny of the radiograph and tracing to enable the following diagnoses to be made:

1. **Diagnosis of posterior arch deficiency (PAD)**. Figure 1 shows the radiograph selected from the control group as most representative of the normal cervical spine, together with a tracing of this normal appearance. The diagnosis of spina bifida occulta was made when lack of internal cortical outline of the posterior arch of the vertebral unit was a consistent finding throughout the series of radiographs taken for that patient after the age of 6 years when fusion of the posterior arch is expected to be completed (Fig. 2A). More extensive absence of posterior arch (dehiscence) is shown in Figure 2B.

2. **Diagnosis of fusion anomalies (FUS)**. Fusion was diagnosed when bony continuity was demonstrated at the articular facets, neural arch, or transverse processes of adjacent vertebrae and when this continuity was a consistent finding throughout the whole series of radiographs available for that patient (Fig. 3A). More extensive fusion involving the vertebral bodies was diagnosed when continuity existed between one vertebral unit and the next, and no complete intervertebral disc space was identified. This block fusion is shown in Figure 3B.

Occipitalization of the atlas may be a complete
FIGURE 3  A. Fusion of C2 to C3 at the articulatory facets, B. Block fusion of C2,3,4 at the vertebral bodies, C. Occipitalization of the atlas. The atlas is fused to the occipital bone. Facet and posterior arch fusion at C2,3,4 is also shown.
TABLE 2 Definitions of Cervical Vertebrae Anomalies (CVA)

<table>
<thead>
<tr>
<th>CVA</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dehiscence</td>
<td>Failure of part of a vertebral unit to develop</td>
</tr>
<tr>
<td>Spina Bifida</td>
<td>Failure of posterior part of the neural arch to fuse</td>
</tr>
<tr>
<td>Fusion</td>
<td>Fusion of one unit with another at the articulation facets, neural arch or transverse processes.</td>
</tr>
<tr>
<td>Block fusion</td>
<td>Fusion of one unit with another but including the vertebral bodies.</td>
</tr>
<tr>
<td>Occipitalization</td>
<td>Assimilation either partial or complete of the atlas with the occipital bone.</td>
</tr>
</tbody>
</table>

... or partial assimilation of the atlas with the base of the skull. Basilar impression usually accompanies this anomaly (McCrae and Barnum, 1953), and if intrusion of odontoid process into foramen magnum occurs (basilar invagination), it may or may not be accompanied by neurological disturbances (McCrae, 1952, 1960). No patients in this study showed this anomaly, but Figure 3C demonstrates the radiological appearance.

RESULTS

Findings for the control group (N=120) and the cleft sample (N=105) are presented in Table 3 and Figures 4 and 5. The statistical analyses of the data were carried out using Fisher’s exact test. The probability values reported are for two-tailed tests.

In the control group, one patient had cervical vertebral anomalies (0.8%), and this was a posterior arch deficiency (PAD). In the cleft sample, 13.3 percent of the patients had anomalies of the cervical vertebrae. This was a more frequent occurrence than in the control group and was statistically significant (p <0.001). Eleven patients exhibited posterior arch deficiency (PAD) (10.5%), and three patients (2.9%) had fusion anomalies.

The results of analysis of the occurrence of cervical vertebral anomalies in the four types of clefting is presented in Figures 4 and 5. Posterior arch deficiency (Fig. 4) occurred in 15.8 percent of the cleft palate (CP) subsample. This was significantly more frequent than in the control group (p <0.001). In the cleft lip (CL) category, the occurrence of posterior arch deficiency was 10 percent; this did not differ from the control group value to a statistically significant extent. In the unilateral cleft lip and palate category (UCLP), the occurrence was 9.8 percent, a value that differed significantly from that of the control group (p <0.05). Posterior arch deficiency did not occur in the bilateral cleft lip and palate (BCLP) subgroup.

Fusion anomalies (Fig. 5) did not occur in the control group nor in the cleft lip or bilateral cleft lip and palate subsamples. Fusion anomalies did occur in 5.3 percent of the cleft palate sample and in 2.4 percent of the unilateral cleft lip and palate sample. These values did not differ from those of control group subjects to a statistically significant extent.

DISCUSSION

The present sample of patients with cleft lip and palate was drawn at random from the hospital records of children in the Edinburgh area and is probably representative of this type of anomaly in the cleft population. The distribution of the different types of clefting corresponds to that

TABLE 3 Cervical Vertebrae Anomalies in the Cleft and Control Samples

<table>
<thead>
<tr>
<th>Sample</th>
<th>N</th>
<th>PAD</th>
<th>Fusion</th>
<th>Number With Anomaly</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N</td>
<td>%</td>
<td>N %</td>
<td>N %</td>
</tr>
<tr>
<td>CL</td>
<td>10</td>
<td>10.0</td>
<td>0.0</td>
<td>1 10.0</td>
</tr>
<tr>
<td>CP</td>
<td>38</td>
<td>15.8</td>
<td>5.3</td>
<td>6* 15.8</td>
</tr>
<tr>
<td>UCLP</td>
<td>41</td>
<td>9.8</td>
<td>2.4</td>
<td>5 12.2</td>
</tr>
<tr>
<td>BCLP</td>
<td>16</td>
<td>0.0</td>
<td>0.0</td>
<td>0 0.0</td>
</tr>
<tr>
<td>Total</td>
<td>105</td>
<td>10.5</td>
<td>2.9</td>
<td>12 13.3</td>
</tr>
<tr>
<td>Control</td>
<td>120</td>
<td>0.8</td>
<td>0.0</td>
<td>1 0.8</td>
</tr>
</tbody>
</table>

* Two subjects with cleft palate exhibited more than one anomaly of the cervical vertebrae.
FIGURE 4 Percentage of subjects with posterior arch deficiency (PAD) in the cleft and control samples.

FIGURE 5 Percentage of subjects with fusion anomalies in the cleft and control samples.
reported by Fogh-Andersen (1942), and Osborne (1968).

The prevalence of cervical vertebral anomalies in the cleft sample was 13.3 percent and was similar to that reported by Osborne (1968) (14.1%).

The prevalence of cervical vertebral anomalies in cleft lip (CL), cleft palate (CP), and unilateral cleft lip and palate (UCLP) was found to be similar in the present study. This is in agreement with the findings of Minaba (1972) but differs from those of Osborne (1968) and Ross and Lindsay (1965) who found a higher prevalence of cervical vertebral anomalies in the cleft palate (CP) subsample.

Since the groups of cervical vertebral anomalies are comprised of deficient as well as excessive osseous development, it was felt that a subclassification of these anomalies might provide more detailed information of cervical vertebral anomalies in the various types of clefting.

The posterior arch deficiency (PAD) group of cervical vertebral anomalies was subdivided into those in whom failure of fusion of the posterior arch had occurred (spina bifida) and those in whom all or part of the posterior arch had failed to develop (dehiscence).

The fusion (FUS) group of cervical vertebral anomalies was subdivided into those in whom fusion was identified at the articulatory facets, neural arch, or transverse process and those in whom block fusion that involved the vertebral bodies had taken place. A third group in whom occipitalization, involving the atlas with the base of the skull, had taken place was added for completion, but no patients in the study had this cervical vertebral deformity.

Posterior arch deficiency (PAD) occurred most frequently in the group with cleft palate (16%), but fusions did not occur significantly more often in any cleft group than in the controls.

The occurrence of the different types of cervical vertebral anomalies in the groups with cleft does not follow a similar pattern but suggests a specific relationship between isolated cleft palate (CP) and posterior arch deficiency. The differentiated pattern of occurrence of the different types of cervical vertebral anomalies suggests different etiological mechanisms.

The primitive embryonic cellular origins of upper cervical vertebrae and the basilar and condylar parts of the occipital bone are similar. Both develop from parachordal cartilage that arises from the cranial end of the notochord and incorporates the sclerotomes of the four occipital and upper cervical somites (Bosma, 1976). It has been further postulated that the nasal capsule and the nasal septum have cellular origins in the sclerotomes of this area (Wilson, 1973).

Ross et al (1965) suggested that severe vertebral anomalies are an early developmental fault of mesenchyme, and they have shown that in patients with severe fusion anomalies, there is a decrease in size of the occipital bone, the embryonic origin of both the cervical vertebrae and the occipital bone being similar.

The present study suggests a specific relationship between cleft palate (CP) and posterior arch deficiency (PAD). The possibility exists that the mechanism involved in palatal shelf fusion during embryonic development may also have an effect on the development and fusion of the posterior arch of the first cervical vertebrae, particularly as the anomaly is in an area developed from the same or similar paraxial mesoderm. The results do however confirm that a child born with cleft lip and palate deformity is more likely than a normal control group to have associated cervical vertebral anomalies.

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