The Association of Submucous Cleft Palate and Clefting of The Primary Palate

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478 records of patients with cleft palate were reviewed to determine the prevalence and significance of submucous cleft palate associated with clefting of the primary palate. The prevalence of submucous cleft palate in the 71 patients with clefts of the primary palate (SMCP-CL) was 13 per cent. This is two to three times greater than the prevalence of isolated submucous cleft palate found in cleft palate clinic patients. Patients with SMCP-CL often had the symptoms of velopharyngeal incompetence (VPI) and middle ear disease. The increased prevalence of SMCP and frequent symptomatology of patients with clefting of the primary palate make it essential that patients with cleft lip have early, thorough evaluation for SMCP. Early detection of SMCP associated with cleft lip and close follow-up, permits the prevention of ear problems and the proper management of VPI should it develop.

KEY WORDS: cleft palate, cleft lip, submucous cleft palate, primary palate, velopharyngeal incompetence, middle ear disease

Submucous clefting of the palate (SMCP) was first described by Roux in 1825. Subsequent reports were added by Kelly (1910), Dorrance (1930), and Veau (1931). The classic diagnostic triad of bifid uvula, midline diastasis of the palatal muscles, and notching of the posterior border of the hard palate was described by Calnan in 1954. The prevalence, the nature of the defect, problems associated with SMCP, and treatment have subsequently been described (Bergstrom, 1971; Burdi, 1967; Caldarelli, 1978; Conway, 1951; Crikelair, 1970; Dellon, 1973; Dibbell, 1965; Dorrance, 1930; Fara, 1971; Kaplan, 1977; Massengill, 1966; Stark, 1954; and Weatherley-White, 1976). One of us, Barbel Holtmann, noted an apparent increased prevalence of SMCP in patients with clefts of the primary palate. Since information in the literature concerning the prevalence of SMCP in patients with clefting of the primary palate (SMCP-CL) was unclear, we reviewed our cleft palate patients to clarify and determine the significance of this association.

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Method

The records of 478 active patients of the Washington University Cleft Palate Clinic were reviewed for age, sex, type of cleft, other problems associated with the cleft, and the presence or absence of SMCP. Calnan's criteria of bifid uvula, midline diastasis of the palatal muscles, and notching of the posterior border of the hard palate were used to establish the diagnosis of SMCP. If some but not all criteria were present, the case was considered to represent a microform and was not counted as SMCP. Cleft-related problems included velopharyngeal insufficiency (VPI), feeding problems, and middle ear disease.

Each patient was completely evaluated by the cleft palate team consisting of a pediatrician, an audiologist, an otolaryngologist, a speech pathologist, a pedodontist, an orthodontist, a prosthedontist, an oral surgeon, and a plastic surgeon.

Three groups of patients were identified. Group I consisted of 374 patients with overt clefts of the secondary palate with or without clefting of the primary palate. Group II con-

sisted of 33 patients with submucous clefts of the palate without clefting of the primary palate. Group III consisted of 71 patients with clefts of the primary palate without overt clefting of the secondary palate.

Results

The patients in Group III were the focus of this study. Thirteen per cent (9/71) of these patients had SMCP in addition to clefts of the primary palate (SMCP-CL). There were six males and three females, ranging in age from 1 month to 4-8/12 years.

Age at the time of diagnosis of SMCP ranged from 19 days to 4 years. In four cases the diagnosis was not made until the child was noted to have abnormal speech.

Feeding problems, including frequent emesis and nasal regurgitation of formula, were found in 33 per cent of the patients with SMCP-CL. None of the patients with isolated clefting of the primary palate or isolated SMCP had had feeding problems.

Evidence of middle ear disease was present in 78 per cent of SMCP-CL patients. Thirteen per cent of patients with isolated clefting of the primary palate and 45 per cent of patients with isolated SMCP had evidence of middle ear problems (Table 1). Fifty per cent of SMCP-CL patients required myringotomy as opposed to 6.5 per cent of the patients with clefts of the primary palate, but no abnormality of the secondary palate.

Velopharyngeal insufficiency, diagnosed by speech performance in patients at least three years of age, was present in five per cent of cleft lip patients, 44 per cent of SMCP-CL patients, and 88 per cent of patients with isolated SMCP (Table 2).

Discussion

Stewart et al. (1970, 1971) found the prevalence of SMCP in a random population of 10,836 Denver school children to be one in 1200 students.

TABLE 1. Significant Middle Ear Disease

	CL	*SMCP-CL*	*SMCP*
Present	13%	78%	45%
Absent	87%	22%	55%

The incidence of significant middle ear disease among patients with CL, SMCP-CL, and isolated SMCP.

TABLE 2. Speech

VPI	*CL*	*SMCP-CL*	*SMCP*
Present	5%	44%	88%
Absent	53%	12%	6%
Too early to evaluate	42%	44%	6%

The incidence of velopharyngeal incompetence among patients with CL, SMCP-CL, and isolated SMCP.

The prevalence of isolated SMCP among patients in cleft palate clinics has been reported to range from three to five per cent (Meskin et al., 1964; Porterfield and Trabue, 1965; Gylling and Soivio, 1965; Crikelair et al., 1970). The prevalence of isolated SMCP among our cleft palate patients was seven per cent. However, the prevalence of SMCP in patients with clefts of the primary palate was 13 per cent. This 13 per cent prevalence of SMCP in patients with clefting of the primary palate seems too high for the chance association assumed in the past. The sex distribution of 2/3 male to 1/3 female was the same for SMCP-CL, isolated SMCP, and cleft lip-cleft palate in our study group. This may mean that SMCP-CL and isolated SMCP are microforms of cleft lip-cleft palate. Such microforms could result from either less severe interference with mesodermal migration or decreased individual susceptibility.

Random, isolated, undiagnosed cases of SMCP tend to be asymptomatic. This was confirmed by the fact that only two of nine Denver school children found to have submucous cleft palates were symptomatic (Weatherley-White et al., 1972). One symptomatic child had a speech defect (articulation errors without hypernasality), and the other had recurrent episodes of serous otitis media.

Patients with isolated SMCP seen at cleft palate clinics are, on the other hand, far more symptomatic. Middle ear problems were noted in 79 per cent of these patients by Kelly (1910), in 44 per cent by Stewart (1970, 1971) and in 39.6 per cent by Bergstrom and Hemenway (1971). Weatherley-White (1976) found VPI in 25 per cent of 44 patients with SMCP. Massengill (1966) observed nasal speech and lack of complete velopharyngeal closure in 100 per cent of 20 patients with isolated SMCP. This increased symptomatology is not surprising since these patients were

referred to cleft palate clinics because symptoms drew attention to their defects.

In our study group with SMCP-CL, we found middle ear disease in 78 per cent of cases. VPI was present in 44 per cent of the cases, and 44 per cent were too young to evaluate speech. Interestingly, the diagnosis of SMCP was frequently missed until the child began to speak. At that time, VPI drew attention to the defects in the secondary palate.

Summary

Four hundred and seventy-eight records of patients with cleft palate were reviewed to determine the prevalence of SMCP associated with clefting of the primary palate. The prevalence of SMCP-CL in 71 patients with clefting of the primary palate was 13 per cent. Many patients with SMCP-CL were symptomatic with VPI and middle ear disease. The increased prevalence of SMCP and frequent associated symptomatology in patients with clefts of the primary palate makes evaluation of these patients for SMCP essential. Early detection of SMCP and close follow-up helps to prevent hearing problems and to provide proper management of VPI should it develop.

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