Tooth Anomalies Associated with Congenital Sinuses of the Lower Lip and Cleft Lip/Palate

REIJO RANTA AND AARNE RINTALA

Dr. Ranta is orthodontist and head of the Department of Dentistry at the Finnish Red Cross Cleft Center and senior lecturer at the Department of Pedodontics and Orthodontics, Institute of Dentistry, University of Helsinke, Finland. He holds a D.D.S. degree and Odont. Dr. from the University of Helsinki.

Dr. Rintala is plastic surgeon and head of the Finnish Red Cross Cleft Center and senior lecturer in plastic surgery at the Medical Faculty, University of Helsinki, Finland. He is a medical graduate (M.D.) of the University of Helsinki.

Address:
Reijo Ranta, D.D.S.
F R C Cleft Center
Pohjoinen Hesperiankatu 17
00260 Helsinki 26
FINLAND

A tabulation and statistical evaluation of the incidence of tooth anomalies such as hypodontia, congenital absence and retarded or asymmetrical development. Congenital absence and hypodontia were found to be unusually prevalent in the presence of lower lip sinus syndrome.

The autosomal, dominantly inherited lower lip sinus syndrome is a very rare congenital malformation.[5,8,13,14,24,28] The frequency of lower lip sinus among cleft patients varies from 0.48%[16] to 2.0%.[24] The form of the sinuses varies from bilaterally symmetrical openings to atypical sinuses and microforms with only conical elevations. Microforms occur mainly in association with cleft palate.[24]

Numerous studies have focussed on the etiology, epidemiology, anatomical form and structure, and genetic transmission of the sinus syndrome. Other anomalies and developmental disorders associated with this syndrome have also received attention.[5,9,14,23,27] In contrast, only a few observations on dental development in sinus patients are reported, and these are based on isolated cases. Congenitally missing teeth have been reported in some cases.[28] A correlation between the sinus syndrome and congenitally missing lower second bicuspids was established in one family.[25]

Hypodontia is a very common developmental defect among children with cleft lip and/or palate, varying considerably in the different types of cleft.[4,22] Asymmetrical development of antimeric teeth is also common in children with cleft lip and palate.[19-21]

The purpose of the present study was to investigate the occurrence of congenital absence of permanent teeth and the asymmetrical development of antimeric teeth outside of the cleft region in association with the sinus syndrome. Third molars are excluded from consideration throughout this study. The results were further compared with the prevalence of hypodontia and the asymmetrical tooth formation in children with cleft lip and/or palate and in healthy noncleft children.

Material and Methods

The study was based on 42 patients with the sinus syndrome. These children were also studied in the epidemiological part of our research.[24] Ages ranged from 6 to 13 years. Table 1 shows the distribution by sex and type of cleft. In 26 cases the sinus was associated with cleft palate alone (CP). Cleft lip and palate (CLP) were present in 15 cases, and cleft lip and alveolus (CL) in one. In the analysis and comparison of the results, the subjects and the controls were classified in CP and CLP groups. The CL case was included in the CLP group.

TABLE 1

Distribution of subjects according to sex and type of cleft.

Type of cleft		*Boys*	*Girls*	*Total*
Cleft lip and alveolus,	bilat.	—	1	1
Cleft lip and palate	right	3	1	4
	left	2	1	3
	bilat.	3	5	8
CL(P) total		8	8	16
Cleft palate		9	17	26
Total		17	25	42

In the CLP group, the sinus was present as a classical bilateral or atypical cavity, and in the CP group as a microform of varying degree with conical elevations.

The subjects were undergoing orthodontic treatment at the Department of Dentistry of the Finnish Red Cross Cleft Center. Records on all subjects included a medical history, data on the dentition and occlusion recorded by one of the authors at the first clinical examination, and an orthopantomogram.

Congenitally missing permanent teeth (excluding the third molars) were identified from the orthopantomogram. The results were compared with the data recorded at the clinical examinations for detection of previously extracted teeth. Asymmetrically developing tooth pairs, peg-shaped upper lateral incisors and teeth with a very severe delay in development were also identified. In determining asymmetry, twelve grades were used to classify dental development.[11] A tooth pair was regarded as developing asymmetrically if one tooth was at least one grade in development behind the antimeric tooth.

The frequency of hypodontia and incidence of congenitally missing teeth outside the cleft region in the sinus groups were compared with the corresponding results on children with only cleft lip and palate. Children with cleft lip and palate seen at the Finnish Red Cross Cleft Center[22] served as controls. These controls consisted of 841 children with cleft lip and palate of varying degrees, ranging in age from 7 to 12 years.

TABLE 2

Presence of hypodontia (excluding the third molars), asymmetrical toothpairs, peg-shaped teeth and teeth with a severe delay in development in childrn with lower lip sinuses. (International tooth numbers.)

Case	*Boy*	*Girl*	*Type of cleft*	*Congenitally missing teeth* *upper jaw*	*lower jaw*	*Asymmetrical tooth pairs*	*Peg-shaped*	*Severe delay in development*
1		X	CLPb	15,12,22,25	45,35	14/24		
2		X	CLPb	15,12,22,25	47,45,35			
3	X		CLPb	15,12,22,23,24,25	47,35			
4		X	CLPb	15,12,22,24,25				
5	X		CLPb	15,12,22	35			
6		X	CLPb	15,12,22				
7		X	CLP	15,14,12,22				
8		X	CLPb	12,11,22				
9		X	CL b	12,11,22				
10	X		CLPlu	22,25				
11	X		CLPlu	22		17/27,15/25		
12		X	CLPlu	22		13/23,15/25		
13	X		CLPru	15,25		11/21		
14	X		CLPru	12				
15	X		CLPru	15	47,45,42			25,37
16		X	CLPru			11/21		
17	X		CP	12			22	
18	X		CP		45,34,35			
19	X		CP	25	35			15
20	X		CP	25	45			
21	X		CP	25	45			
22	X		CP			35/45		
23-25	X		CP					
26	X		CP	12,22,25		14/25		15
27		X	CP	25				
28		X	CP	15,12			22	
29		X	CP	22				
30		X	CP	22			12	
31		X	CP		45			
32		X	CP	22			12	
33		X	CP	12,22				
34		X	CP		45			
35		X	CP	12			22	
36		X	CP	15,12,22,24,25	45,42,41,31,32,25	11/21		
37		X	CP	25	45			15,35
38-42		X	CP					

Symbols: CLP = cleft lip and palate, b = bilateral, r = right unilateral, l = left unilateral, CL = cleft lip and alveolus, CP = cleft palate.

Among the Cps (17-24) the sinus was micoform with conical elevations.

Among the CLPs (1-16) the sinus was bilateral or with atypical openings.

The weighted arithmetic mean of percentage value by sex and type of cleft, calculated from the sinus group, were used in the comparison. The frequency of asymmetrical development of antimeric teeth was compared with the corresponding groups of children with cleft lip and palate seen at this Center.[19,21] The hypodontia of the lateral incisor in the cleft region was also compared with the corresponding figures on these controls.[18]

Hypodontia was compared with the data on hypodontia in healthy non-cleft Finnish children.[12] In the statistical analysis of hypodontia, the CP and CLP groups of the sinus syndrome were compared with the data on the analogous cleft groups, and both sinus groups were also compared to each other.

The significance of the results was determined by Student's t-test. In the analysis, the frequency of hypodontia was assumed to be the same for the sinus groups and for the data on the control groups.

Results

Table 2 is a case by case listing of the congenitally missing teeth, asymmetrically developing tooth pairs, peg-shaped upper lateral incisors and teeth with a very severe delay in development. Sex and type of cleft, as well as the interrelationship of the type of cleft and the type of sinus, are also noted in each case.

Table 3 and Fig. 1 show the frequency of hypodontia in the permanent dentition outside the cleft region for girls and boys in the control cleft group and for the CP and CLP groups among the 42 children with lower lip sinus. In the control cleft group, hypodontia and missing teeth were more prevalent in girls than in boys. In the CP and CLP groups, however, no sta-

TABLE 3

Frequency of hypodontia outside the cleft region (excluding the third molars) among 42 children with lower lip sinuses in association with cleft lip (palate), number of congenitally missing teeth and means of missing teeth per child with hypodontia by sex and type of cleft.

	Cleft palate (N = 26)			*Cleft lip and palate (N = 16)*			*Combined groups and sexes (N = 42)*
	boys	*girls*	*combined*	*boys*	*girls*	*combined*	
No. of children	9	17	26	8	8	16	42
No. of children with hypodontia	5	12	17	6	6	12	29
percent	55.7 ± 16.6	70.6 ± 11.1	65.4 ± 9.3	75.0 ± 15.3	75.0 ± 15.3	75.0 ± 10.8	69.1 ± 7.1
No. of missing teeth							
mean of missing teeth per child with	10	27	37	18	17	35	72
hypodontia	2.0	2.3	2.2	3.0	2.8	2.9	2.5
Ranta (22)[x]			33.3 ± 2.3			58.1 ± 2.6	42.8 ± 1.8

[x] Weighted arithmetic mean of percentage values by sex and type of cleft in cleft-affected children for comparison.

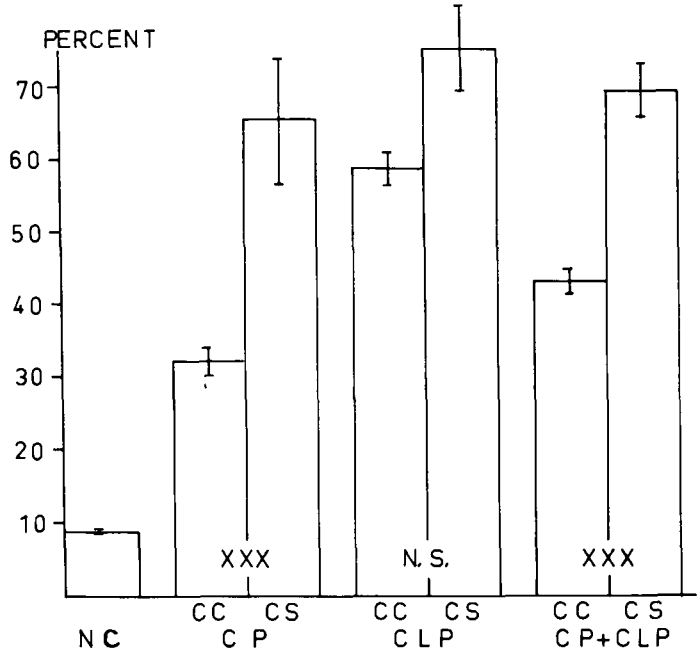

Fig. 1 Comparison of prevalence of hypodontia (Table 3) outside the cleft region in permanent dentition (excluding the third molar). CC = weighted arithmetic mean of percentage values by sex and type of cleft of cleft-affected children (22), CS = present samples, cleft + sinus, CP = cleft palate, CLP = cleft lip (palate), NC = noncleft, healthy ychildren, Haavikko (12), 8.0%.
noncleft, healthy children, Haavikko (12), 8.0%.
XXX = $p < 0.001$, N.S. = not significant (t test)

tistically significant differences at the level of $P < .05$ were found between the sexes. Table 3 also shows the number of missing teeth for each of the children in whom one or more permanent tooth germs were congenitally missing. In the different groups the mean number varies from 2 to 3, so that in the CLP group more teeth were missing per child than in the CP group. For both of these groups this number was always higher than that for the corresponding control groups.

The distribution of hypodontia in percent by jaws and type of cleft (Table 4 and Fig. 2) demonstrates that congenitally missing teeth in the region outside the cleft were more frequent in the sinus groups.

When comparing the prevalence of missing teeth, the CLP sinus group was found to have more hypodontia than the CP sinus group ($P < 0.05$). Likewise, comparison of the sinus group (6.3%) to the cleft controls (3.4%) showed significantly more teeth missing in the sinus group ($P < 0.001$). For the healthy children this percentage was 0.5%.

Table 5 shows a greater incidence of hypodontia in the sinus groups ($P < 0.05$), with the highest incidence for the lower second bicuspids ($P < 0.01$).

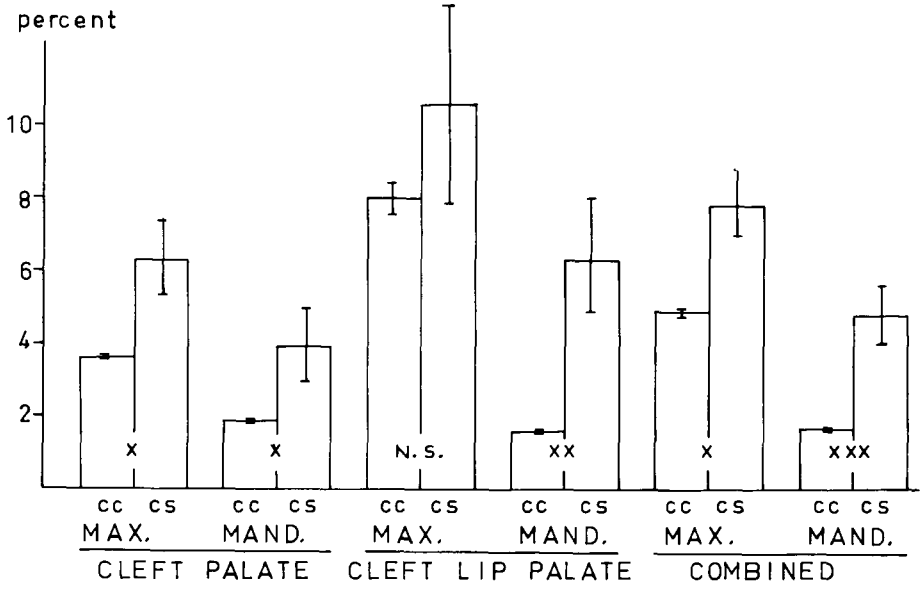

Fig. 2 Comparison of the prevalence of congenitally missing teeth among all teeth examined (Table 4) in maxilla and mandible between the groups of cleft + sinus (CS) and the weighted arithmetic means of percentage values by sex and type of cleft of cleft-affected children (CC) (22).

X = $p < 0.05$, XX = $p < 0.01$, XXX = $p < 0.001$, N.S. = not significant (t test)

Among the subjects there were 25 alveolar clefts, in 22 of which the bud of the lateral incisors at the cleft was congenitally missing (88.0%). For the controls this percentage was 39.3%. Of the 59 teeth outside the cleft region, lateral incisors were missing in 12 cases (20.3%). Of these, 6 teeth were missing symmetrically and 5 asymmetrically. Five of the upper lateral incisors were peg-shaped, and in each of these cases the antimeric tooth was congenitally missing.

A very severe delay in development was noted in 6 teeth. Of these, 4 were upper second bicuspids, 1 lower second bicuspid and one lower second molar. The delayed development was always associated with congenital absence of the antimeric tooth.

Discussion

In the cleft lip and palate cases, the frequency as well as distribution of hypodontia vary greatly in the different cleft groups.[4,22] Therefore, when comparing the differences of the sinus groups and the cleft-affected children, the results for the controls were changed to accord with sex and type of cleft in the sinus groups, using weighted arithmetic means of percentage values by sex and type of cleft. Considering the small number of subjects in the sinus groups, this comparison method appeared to be the most reliable one.

The frequency of hypodontia was greater than in the controls in both the CP and the CLP sinus groups,

TABLE 4

Distribution of hypodontia in percent by type of cleft in both jaws. Percentage values of cleft-affected children (22) given in the bottom line for comparison; weighted arithmentic mean of the percentage values by sex and type of cleft.

	Cleft palate (N = 26)		*Cleft lip and palate (N = 16)*		*Both groups (N = 42)*	
	Maxilla	*Mandible*	*Maxilla*	*Mandible*	*Maxilla*	*Mandible*
No. of examined teeth	364	364	199	224	563	588
No. of missing teeth ..	23	14	21	14	44	28
percent	6.3 ± 1.3	3.9 ± 1.0	10.6 ± 2.2	6.3 ± 1.6	7.8 ± 1.1	4.8 ± 0.9
Ranta (22)	3.06 ± 0.2	1.82 ± 0.2	8.10 ± 0.4	1.60 ± 0.2	4.9 ± 0.2	1.73 ± 0.1

with the greater difference in the CP group ($P < 0.001$). For the difference between the sinus groups, the level of significance was only $P < 0.05$. One explanation for the lower level of association with cleft type in the sinus syndrome cases may be due the common autosomal dominant genetic etiology of the sinus syndrome. It may also indicate that the microsinus forms included in the CP group belong to the sinus syndrome, and that even the mild cases diagnosed really belong to the sinus syndrome. This observation is supported by the conclusion of Hirth et al.[13,14] that the lower lip sinuses and paracentral whorles may belong to the same graduation scheme. Nor can the possibility be excluded that some other kind of genetic factor influencing hypodontia may be involved. The concept that the etiology of hypodontia is genetic and polygenic, quasi-continuous type is very widely accepted.[2,6,26] A dominant autosomal gene with incomplete penetrance and variable expressivity has also often been stated to be the etiological factor of hypodontia.[10] External risk factors, exerting their effect primarily at the embryonic stage, have been found to influence the development of tooth buds.[2,15,17]

The frequency of congenitally missing teeth being higher in both the upper and lower jaw and in the different tooth groups than among the controls, shows that the increased hypodontia associated with sinus syndrome involves all permanent teeth. Furthermore, hypodontia appears to be more frequent in the teeth of the lower jaw. This observation is very well in accord with Butler's Field Theory.[3,7] According to this theory, hypodontia involves the lower and upper second bicuspids in particular and the upper lateral incisors, but also the other teeth which are less exposed to disturbing factors.

Schneider's[25] observations of a substantial number of congenitally missing permanent teeth in the members of one family demonstrated a highly significant correlation of lower lip sinus and hypodontia. In our series (Table 2, Cases No. 1, 2, 3), there were three siblings, one a boy and two girls, all of whom had bilateral CLP and bilateral symmetrical sinuses. In all of them the upper lateral incisors, upper and lower second bicuspids, and additionally in Case No. 3 one upper cuspid and one first bicuspid were missing. In Case No. 1 the upper first premolars developed asymmetrically.

The high frequency of hypodontia in our subjects, the three cases mentioned above, and Schnieder's[25] observations show that hypodontia is a feature of the lower lip sinus syndrome. The theoretical possibility of "two separate, dominantly inherited dis-

orders, the syndrome of lower lip sinus and a syndrome of congenitally missing teeth,"[25] is hardly acceptable in the light of the present investigation.

The frequency of the development of asymmetrical tooth pairs was slightly less in our subjects than in the controls. According to Adams and Niswander,[1] this asymmetry (developmental 'noise') occurring in the development of organ pairs is due to hazardous external factors excerting their effect at the embryonic stage. Both of the paired organs are affected by the genetic factors in the same way.

Considering the multifactorial etiology of cleft lip and palate, the asymmetrical development of tooth pairs may be due to external factors. Among our subjects with sinus syndrome the asymmetrical development of teeth was slightly less than among the controls, although the frequency of hypodontia was considerably higher. Thus, the increased hypodontia would be due to genetic factors, the influence of which was also indicated by the asymmetrical development of teeth.

Summary and Conclusions

Tooth anomalies were clinically and radiographically investigated in the permanent dentition of 42 children with the syndrome of lower lip sinus and cleft lip and/or palate. 26 of these children had cleft palate alone associated with sinus microforms (conical elevations). In the remaining 16 children cleft lip and palate was associated with bilateral symmetrical or atypical sinuses.

The prevalence and distribution of congenitally missing permanent teeth and of asymmetrically developing tooth pairs (excluding the third mo-

TABLE 5

Frequency of congenitally missing upper and lower second bicuspids, upper lateral incisors outside the cleft region and the other teeth (excluding the third molars) by type of cleft and for both groups combined.

Weighted arithmetic mean of percentage values by sex and type of cleft of cleft affected children (22) given in the last line for comparison.

	Cleft palate (N = 26)			*Cleft lip and palate (N = 16)*			*Both groups (N = 42)*			
	No. of examined teeth	*No.*	*Missing teeth percent*	*No. of examined teeth*	*No.*	*Missing teeth percent*	*No. of examined teeth*	*No.*	*Missing teeth percent*	*Ranta (22) percent*
Upper 2nd premolar ...	52	10	19.2 ± 5.5	32	16	50.0 ± 8.8	84	26	31.0 ± 5.1	20.1 ± 1.0
Lower 2nd premolar ...	52	9	17.3 ± 5.3	32	11	34.0 ± 8.4	84	20	23.8 ± 4.7	9.5 ± 0.7
Upper lateral incisor ...	52	12	23.1 ± 5.8	7	—	—	59	12	20.3 ± 5.3	9.61 ± 0.9
The other teeth	572	6	1.1 ± 0.4	352	8	2.3 ± 0.8	924	14	1.52 ± 0.4	0.68 ± 0.1
Total	728	37	5.08 ± 0.8	423	35	8.27 ± 1.3	1151	72	6.26 ± 0.7	3.42 ± 0.1

lars and the upper lateral incisors in the region of alveolar cleft) were compared with the analogous data on cleft-affected children[18,19,21,22] and non-cleft children.[12]

1. The prevalence of hypodontia was 65.4% of cases in the CP group, 75.0% in the CLP group and 69.1% in the entire sample. For the controls the corresponding percentages were 33.3%, 58.1% and 42.8%. Among the CLP subjects with lower lip sinus, the incidence of hypodontia was significantly higher than among the controls ($P < 0.001$). The difference was also demonstrable between the CP microsinus group and the CP control group. No differences was found between the sexes.

2. In the sinus subjects the number of congenitally missing teeth was 6.3% of the teeth examined and in the controls 3.4%. In the CP and CLP sinus groups, the number of teeth missing in the upper jaw was almost double the number in the corresponding control groups. In the entire sinus group, significantly more teeth were missing in the upper jaw than in the controls ($P < 0.05$) and in the lower jaw the significance was even greater ($P < 0.001$).

3. In the sinus group, the incidence of hypodontia was greater for all teeth. The upper second bicuspids were most frequently congenitally missing (31.0%), followed by the lower second bicuspids (23.8%) and the upper lateral incisors (20.3%). Other teeth accounted for only 1.52%. The corresponding percentages for the controls were 20.1%, 9.5%, 9.6% and 0.72%. In the cleft region, 88% of the upper lateral incisors were missing.

4. Asymmetrical development of tooth pairs occured slightly less in the sinus subjects than in the controls.

On the basis of these results, it can be concluded that the sinus forms present in the CP group and even the mild cases diagnosed with all probability belong to the sinus syndrome. Lower lip sinus syndrome is obviously not only a local developmental disorder, because the dentition demonstrates considerably increased hypodontia. The increased hypodontia and slightly decreased asymmetrical development of the teeth indicate that the etiological part played by the genetic factors is clearly more decisive in sinus syndrome than in cleft patients in general or, indirectly, this may also indicate a multifactorial etiology of "ordinary clefts."

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