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## CEPHALOMETRIC AND MORPHOLOGIC CHANGES IN ADULT MALES WITH ISOLATED CLEFT PALATE

Z. ŠMAHEL

The present communication is aimed at a review of the extent and character of facial changes associated with isolated cleft palate in individual subgroups, according to the severity of the cleft. This study characterizes the final condition of deviations from normal after the use of an identical surgical technique. It is based on our earlier report dealing with the same problem in unilateral cleft lip and palate (Šmahel, 1982), with an identical selection of probands (according to age, sex and surgical technique). This provides the possibility of a comparison of the results obtained in both studies. Basic body characteristics in these patients are determined as well. The present communication deals with the results of cephalometric, somatometric and somatoscopic studies. Roentgenographic cephalometry used in these patients is described separately (Šmahel, in press).

### MATERIAL AND METHODS

The series examined included 90 males with isolated cleft palate (CP) ranging in age from 20 to 40 years. The patients were subdivided according to the severity of clefts in those with a complete cleft extending up to the Foramen incisivum (CP<sub>c</sub>, n = 33), those with an incomplete cleft involving at least one third of the hard palate (CP<sub>i</sub>, n = 37) and into those with cleft soft palate alone (CP<sub>s</sub>, n = 20) and therefore no assessment of the series as a whole was carried out (the series included no individuals with cleft soft palate with cleft hard palate involving less than one third of the latter or with submucuous clefts). All patients belonging into the assessed subgroups operated upon at the University Clinic for Plastic Surgery in Prague, where patients from all parts of Bohemia are treated, were requested to attend a check-up examination (with the exception of individuals from foreign ethnic groups and patients with associated anomalies and those subjected previously to maxillofacial osteotomy). The attendance rate was 84 per cent.

During the analysis of values obtained by measurements some individuals were excluded, i. e. in the first subgroup one patient because of the relatively advanced age at the time of the primary suture of the palate, in the second subgroup four patients for the same reason and one because of micrognathia (Pierre Robin syndrome) and in the third subgroup two patients because of delayed surgical intervention, another two because of submucous notch into the hard palate and one individual who was not yet treated by surgery (n=32, 32 and 15 resp.). However they were included into the assessment of somatoscopic characteristics. At the time of the check-up examination the mean age in individual subgroups amounted in CP<sub>c</sub> to 29.66 years (SD = 5.81), in CP<sub>i</sub> to 27.84 years (SD = 5.75) and in CP<sub>s</sub> to 30.13 years (SD = 4.83). Thus the

Tab. 1. Main body, head and face dimensions in adult males with isolated complete (CP<sub>c</sub>) and incomplete (CP<sub>i</sub>) cleft palate and cleft soft palate alone (CP<sub>s</sub>) in comparison with controls (the mean and standard error are given in mm)

	CP <sub>c</sub>		CP <sub>i</sub>		CP <sub>s</sub>		Control	
height (in cm)	175.03	1.27	174.47	0.96	175.40	1.82	176.88	0.90
weight (in kg)	77.16	2.38	71.62	2.08	75.40	3.40	77.22	1.73
chest circ. (cm)	95.81	1.51	91.09	1.02	95.13	2.08	93.46	0.98
chest width	291.25	4.11	280.63	3.28	285.00	5.92	292.00	2.91
chest depth	206.72	4.01	198.71	2.42	206.67	5.36	206.60	2.60
biacrom. width	392.19	3.69	388.75	3.32	386.00	3.60	391.70	2.22
bicrist. width	279.38	2.64	277.50	3.48	281.33	4.76	283.50	2.38
bitroch. width	326.88	2.84	321.72	2.88	325.67	3.78	326.12	2.55
Rohrer index	1.44	0.04	1.34	0.03	1.40	0.06	1.41	0.03
head circum.	573.44	2.83	570.16	3.12	569.67	4.27	573.60	2.08
g-op	188.91	1.07	187.94	1.32	187.00	1.89	188.42	0.76
eu-eu	157.69	1.14	159.25	0.98	159.13	1.20	159.40	0.91
ft-ft	109.59	0.76	109.69	0.89	108.87	1.04	109.92	0.57
t-t	144.16	1.10	142.38	0.83	141.47	1.29	144.22	0.72
zy-zy	142.56	0.99	142.47	0.78	141.47	1.25	142.82	0.70
go-go	107.81	0.78	107.88	1.02	106.80	1.10	108.46	0.78
en-en	32.19	0.42	32.13	0.52	31.67	0.73	32.60	0.36
ex-ex	90.72	0.52	91.03	0.74	88.93	1.16	90.30	0.47
al-al	34.38	0.44	34.25	0.38	34.47	0.78	35.62	0.31
ch-ch	49.12	0.58	48.44	0.58	48.53	0.86	53.16	0.40
n-gn	119.47	1.14	121.88	1.17	119.60	1.49	119.46	0.93
n-sto	71.91	0.65	73.88	0.76	72.07	0.90	72.92	0.56
n-sn	50.34	0.63	51.25	0.64	50.47	0.63	50.16	0.43
sto-gn <sup>+</sup>	48.94	0.72	49.97	0.69	47.73	0.67	49.04	0.52
t-g-t	311.25	1.92	311.72	2.28	307.67	3.26	313.10	1.57
t-sn-t	287.66	2.87	284.22	1.98	282.50	4.43	296.80	1.74
t-gn-t	319.84	2.66	312.97	2.07	312.14	5.19	322.10	2.02
i. cephalicus	83.53	0.60	84.75	0.60	85.20	0.81	84.58	0.49
i. facialis	83.84	1.05	85.56	0.79	84.73	0.92	83.66	0.62
i. fac. sup.	50.31	0.57	51.81	0.52	50.93	0.61	51.06	0.39
i. fac. inf.	34.38	0.62	35.12	0.47	33.93	0.47	34.38	0.36
i. mandib.	45.47	0.83	46.44	0.70	44.80	0.65	45.34	0.49
i. intercanthal.	35.56	0.41	35.28	0.41	35.53	0.50	35.98	0.29
i. nasalis	68.62	1.31	67.06	1.01	68.47	1.72	71.24	0.84
i. cheilozygomat.	34.38	0.36	33.97	0.40	34.33	0.67	37.14	0.23

+ significant difference between incomplete clefts and soft palate clefts at  $p < 0.05$



Tab. 2. Dimensions of lateral facial regions, oronasal region and ear lobes in adult males with individual types of isolated cleft palate (CP<sub>c</sub>, CP<sub>i</sub>, CP<sub>s</sub>) in comparison with controls (the mean and standard error are given, in mm or degrees)

	CP <sub>c</sub>		CP <sub>i</sub>		CP <sub>s</sub>		Control	
t-n dx	122.16	0.77	122.28	0.87	120.33	1.24	122.50	0.63
t-n sin	122.22	0.77	122.13	0.95	120.73	1.39	122.62	0.58
t-sn dx <sup>(*)</sup>	128.75	0.95	126.88	0.93	124.80	1.51	129.74	0.57
t-sn sin	128.31	1.01	128.56	0.94	125.93	1.65	130.68	0.66
t-gn dx <sup>x</sup>	144.44	1.11	141.44	0.91	139.67	1.67	144.30	0.78
t-gn sin <sup>x</sup>	143.41	1.19	141.91	0.95	138.67	1.80	143.80	0.79
t-go dx	70.59	0.99	67.25	0.77	68.27	1.10	72.74	0.69
t-go sin	69.22	1.06	65.59	0.81	66.80	1.29	71.94	0.70
t-ex dx <sup>x</sup>	80.34	0.74	79.63	0.63	78.00	0.81	79.98	0.53
t-ex sin <sup>x</sup>	80.62	0.74	79.59	0.61	78.13	0.93	80.06	0.49
t-ch dx <sup>x</sup>	111.88	1.02	109.69	0.85	108.00	1.41	111.98	0.68
t-ch sin <sup>(x)</sup>	111.56	1.01	110.06	0.85	107.87	1.71	111.94	0.72
ex-ch dx	73.38	0.63	73.69	0.69	72.73	0.76	72.44	0.49
ex-ch sin	73.00	0.59	73.41	0.65	72.07	0.77	72.00	0.48
go-ex dx	98.47	1.01	97.91	0.88	98.53	1.15	100.80	0.65
go-ex sin	98.34	1.05	97.22	0.87	98.40	1.37	100.90	0.63
go-ch dx	78.12	0.87	77.00	0.78	77.80	1.25	81.76	0.65
go-ch sin	78.31	0.90	77.13	0.81	77.87	1.45	81.70	0.65
go-gn dx	89.59	0.84	88.16	0.63	89.13	1.54	92.16	0.64
go-gn sin	89.88	0.82	88.13	0.67	88.20	1.45	92.14	0.63
obs-n dx <sup>(x)+</sup>	118.97	0.92	119.03	0.80	116.27	1.22	120.34	0.61
obs-n sin	118.72	0.91	119.19	0.91	116.80	1.39	120.20	0.62
obi-sn dx	116.28	1.04	113.88	0.98	113.20	1.86	118.24	0.64
obi-sn sin	115.62	1.10	114.19	1.00	113.53	1.81	118.62	0.66
sa-sba dx	66.41	0.70	65.38	0.79	66.13	0.84	66.38	0.53
sa-sba sin	66.16	0.73	65.22	0.77	65.93	0.78	66.10	0.58
pra-pa dx	36.69	0.50	35.59	0.35	35.80	0.48	36.88	0.32
pra-pa sin	36.47	0.53	35.56	0.35	35.33	0.57	36.72	0.30
protrusion dx	17.72	1.81	19.13	1.28	18.60	1.82	21.54	1.00
protrusion sin	20.81	1.44	23.91	1.27	24.40	2.29	21.66	0.99
inclination dx	16.62	0.71	15.66	0.68	14.13	1.06	14.08	0.49
inclination sin	16.97	0.71	16.19	0.61	14.80	1.05	14.30	0.51
prn-sn	21.31	0.30	21.44	0.41	21.20	0.63	21.82	0.27
prn-sbal dx	30.97	0.19	31.28	0.40	31.47	0.53	32.20	0.31
prn-sbal sin	31.12	0.22	31.69	0.40	31.73	0.52	32.48	0.31
sn-sbal dx	14.75	0.23	14.44	0.20	14.33	0.45	15.36	0.18
sn-sbal sin	14.22	0.21	14.34	0.16	14.73	0.40	15.08	0.16
sn-ls	16.66	0.34	18.13	0.50	17.40	0.85	18.18	0.35
sbal-ll dx	18.25	0.30	19.66	0.40	18.67	0.81	20.64	0.32
sbal-ll sin	18.34	0.32	19.66	0.40	18.80	0.75	20.52	0.31
nose deviat. <sup>0</sup>	-0.38	0.47	+0.69	0.53	+0.47	0.52	+0.40	0.22
prn deviat. <sup>0</sup>	+0.09	0.32	+0.81	0.50	+0.60	0.38	+0.52	0.19
columel. dev. <sup>0</sup>	+0.06	0.45	-0.16	0.40	-0.87	0.56	+0.04	0.19
m-prn	+0.09	0.21	+0.38	0.25	+0.40	0.26	+0.34	0.13
m-sn	-0.16	0.14	+0.19	0.20	+0.40	0.24	+0.22	0.10
Topinard dx	1.91	0.07	1.91	0.07	1.93	0.12	2.00	0.05
Topinard sin	1.91	0.05	1.88	0.09	1.93	0.07	2.00	0.05

+ deviation to the right, - deviation to the left (<sup>0</sup>in degrees)

x significant differences between complete clefts and soft palate clefts at  $p < 0.05$ ; (x) at  $p < 0.1$

+ significant difference between incomplete clefts and soft palate clefts at  $p < 0.05$

! significant difference between right and left side at  $p < 0.05$

difference from the group of controls (27.18 years, SD = 5.76) was not significant ( $t = 1.87, 0.50$  and  $1.77$  resp.). The differences between individual subgroups were not significant as well ( $t = 1.24, 1.31$ , and  $0.27$  resp.). According to the results of the F-test the proportional age distribution showed in no subgroup significant differences. The series of controls consisted of volunteers subjected to hospital treatment for injuries and of normal students. Their body height and weight were representative of our general population.

Primary palatoplasty was carried out between the age of two and seven years by a push-back usually (69 cases) including pharyngeal fixation (secondary pharyngeal fixation in another eight cases). This was similar as in unilateral cleft lip and palate (CLP<sub>uni</sub>) reported in an earlier study (Šmahel, 1982). There were further no significant age differences at the time of surgery (in CP<sub>c</sub> 4.54 years, in CP<sub>i</sub> 5.11 years and in CP<sub>s</sub> 5.17 years) as compared to CLP<sub>uni</sub> (4.91 years,  $t = 0.99, 0.59$  and  $0.67$  resp.), as well as between individual subgroups ( $t = 1.40, 0.15$  and  $1.25$  resp.). The same held true for the distribution determined with the F-test. The age at the time of surgery could not account for the differences in facial configuration between individual subgroups. Thus these subgroups, as well as the group of CLP<sub>uni</sub> were fully comparable.

The method of our study is based on anthropometric principles and described in detail in one of our previous papers (Šmahel, 1982). The results are presented in tabellar form (Tab. 1—2) yet for clearer illustration the differences from controls are presented separately (Tab. 3—4) for individual subgroups and are compared with corresponding values for the series of CLP<sub>uni</sub>. Rohrer's index characterizes the height-weight proportions, the other indexes denote facial proportions (cheilozygomatic index, the width of the oral slot in proportion to the width of the face, i. e.  $100 \times \text{ch-ch} : \text{zy-zy}$ ).

Basic statistical characteristics were calculated and the F-test and the t-test were used for all comparisons (the latter also for the determination of the frequency of somatoscopic characteristics). For the analysis, according to Topinard's classification of the form and position of the nares a quantitative determination of qualitative characteristics was used. The exclusion of individuals with a relatively delayed primary palatoplasty was carried out by Dixon's test of extreme deviations (the requested significance level  $p < 0.01$ ). Details were presented in the above mentioned earlier report (Šmahel, 1982).

## RESULTS

The results are described for all subgroups simultaneously with statements of data relative to all occurring differences.

**Somatic development:** Body height and weight were slightly below the average, but for the subgroup of incomplete cleft palate with a significant reduction of body weight ( $p < 0.05$ ) and body height close to this level ( $p < 0.1$ ). The difference of body height from controls was always smaller than in CLP<sub>uni</sub> and in the series of 90 males as a whole amounted to 2.4 cm ( $p < 0.05$ ). Of the other somatometric characteristics a significant reduction occurred only in values of the width and depth of the chest in incomplete cleft palate in which the other parameters were slightly below the average as well.

Tab. 3. Differences of main body, head and face dimensions in adult males with isolated complete (CP<sub>c</sub>) and incomplete (CP<sub>i</sub>) cleft palate and cleft soft palate alone (CP<sub>s</sub>) from controls as compared to the differences in unilateral cleft lip and palate (in mm)

	CP <sub>c</sub>	CP <sub>i</sub>	CP <sub>s</sub>	CL.P <sub>uni</sub>
height (in cm)	-1.85	-2.41( )	-1.48	-3.60
weight (in kg)(+)	-0.06	-5.60	-1.82	-4.65
chest circum. (cm)+	+2.35	-2.37	+1.67	-0.80
chest width+	-0.75	-11.37	-7.00	-1.14
chest depth(+)	+0.12	-7.89	+0.07	-4.19
biacromial width	+0.49	-2.95	-5.70	-3.42
bicristal width	-4.12	-6.00	-2.17	-4.29
bitrochanteric w.	+0.76	-4.40	-0.45	-5.78
Rohrer index+	+0.03	-0.07	-0.01	-0.01
head circum	-0.16	-3.44	-3.93	-6.06
g-op	+0.49	-0.48	-1.42	-1.16
eu-eu	-1.71	-0.15	-0.27	-0.95
ft-ft	-0.33	-0.23	-1.05	+1.55( )
t-t	-0.06	-1.84	-2.75( )	-2.56
zy-zy	-0.26	-0.35	-1.35	+0.34
go-go	-0.65	-0.58	-1.66	+0.32
en-en	-0.41	-0.47	-0.93	+1.38
ex-ex	+0.42	+0.73	-1.37	+1.49×
al-al	-1.24	-1.37	-1.15	+0.83( )
ch-ch	-4.04	-4.72	-4.63	-2.18
n-gn	+0.01	+2.42	+0.14	+2.73
n-sto(+)	-1.01	+0.96	-0.85	-2.26
n-sn	+0.18	+1.09	+0.31	-1.64
sto-gn	-0.10	+0.93	-1.31	+4.05***
t-g-t	-1.85	-1.38	-5.43	+5.35×
t-sn-t	-9.14	-12.58	-14.30	-10.08
t-gn-t+	-2.26	-9.13	-9.96( )	-5.72
index cephalicus	-1.05	+0.17	+0.62	+0.11
i. facialis	+0.18	+1.90( )	+1.07	+1.79
i. fac. sup.(+)	-0.75	+0.75	-0.13	-1.68
i. fac. inf.	-0.00	+0.74	-0.45	+2.76***
i. mandibularis	+0.13	+1.10	-0.54	+3.54
i. intercanthalis	-0.42	-0.70	-0.45	+0.99
i. nasalis	-2.62( )	-4.18	-2.77	+4.23
i. cheilozygomat.	-2.76	-3.17	-2.81	-1.55

(×)p<0.1    p<0.05    \*\*p<0.01    \*\*\*p<0.001

+significant differences between complete and incomplete clefts at p<0.05; (+)at p<0.1

**Neurocranium:** The length {g-op} and the width {eu-eu} of the neurocranium showed no changes, similarly as the frontal width {ft-ft, t-g-t}. The circumference was insignificantly reduced in incomplete cleft palate and in cleft soft palate.

**Cranial base:** Its width {t-t} was not changed.

**Upper face:** Neither height {n-sto, n-sn}, nor width {zy-zy} dimensions of the upper face showed any changes, inclusive of the interocular distance {en-en} and the distance between the outer canthi {ex-ex}. The reduction of the subnasal arch {t-sn-t} was related to maxillary retrusion and

Tab. 4. Differences of the dimensions of lateral facial regions, oronasal region and of ear lobes in individual subgroups of isolated cleft palate from controls as compared to those in unilateral cleft lip and palate (in mm or degrees)

	CP <sub>c</sub>	CP <sub>i</sub>	CP <sub>s</sub>	CLP <sub>uni</sub>
t-n dx	-0.34	-0.22	-2.17	+0.88
t-n sin	-0.40	-0.49	-1.89	-0.21
t-sn dx	-0.99	-2.86 <sup>x</sup>	-4.94 <sup>xx</sup>	-3.50 <sup>xxx</sup>
t-sn sin	-2.37 <sup>x</sup>	-2.12 <sup>(x)</sup>	-4.75 <sup>xx</sup>	-6.37 <sup>xxx</sup>
t-gn dx <sup>+</sup>	+0.14	-2.86 <sup>x</sup>	-4.63 <sup>x</sup>	-1.58
t-gn sin	-0.39	-1.89 <sup>x</sup>	-5.13 <sup>x</sup>	-1.70
t-go dx <sup>++</sup>	-2.15 <sup>(x)</sup>	-5.49 <sup>xxx</sup>	-4.47 <sup>xx</sup>	-0.77
t-go sin <sup>++</sup>	-2.72 <sup>x</sup>	-6.35 <sup>xxx</sup>	-5.14 <sup>xx</sup>	-0.36
t-ex dx	+0.36	-0.35	-1.98 <sup>x</sup>	0.00
t-ex sin	+0.56	-0.47	-1.93 <sup>(x)</sup>	-0.23
t-ch dx	-0.10	-2.29 <sup>x</sup>	-3.98 <sup>x</sup>	-2.89 <sup>xx</sup>
t-ch sin	-0.38	-1.88 <sup>(x)</sup>	-4.07 <sup>x</sup>	-3.47 <sup>xxx</sup>
ex-ch dx	+0.94	+1.25	+0.29	-0.80
ex-ch sin	+1.00	+1.41 <sup>(x)</sup>	+0.07	+0.10
go-ex dx	-2.33 <sup>x</sup>	-2.89 <sup>x</sup>	-2.27 <sup>(x)</sup>	-1.06
go-ex sin	-2.56 <sup>x</sup>	-3.68 <sup>xx</sup>	-2.50	-1.81 <sup>x</sup>
go-ch dx	-3.64 <sup>xx</sup>	-4.76 <sup>xxx</sup>	-3.96 <sup>xx</sup>	-2.76 <sup>xx</sup>
go-ch sin	-3.39 <sup>xx</sup>	-4.57 <sup>xxx</sup>	-3.83 <sup>x</sup>	-3.44 <sup>xxx</sup>
go-gn dx	-2.57 <sup>x</sup>	-4.00 <sup>xxx</sup>	-3.03 <sup>(x)</sup>	-1.76 <sup>x</sup>
go-gn sin	-2.26 <sup>x</sup>	-4.01 <sup>xxx</sup>	-3.94 <sup>x</sup>	-1.31
obs-n dx	-1.37	-1.31	-4.07 <sup>xx</sup>	+1.54
obs-n sin	-1.48	-1.01	-3.40 <sup>x</sup>	+1.35
obi-sn dx <sup>(+)</sup>	-1.96 <sup>(x)</sup>	-4.36 <sup>xxx</sup>	-5.04 <sup>x</sup>	-3.96 <sup>xxx</sup>
obi-sn sin	-3.00 <sup>x</sup>	-4.43 <sup>xxx</sup>	-5.09 <sup>x</sup>	-6.15 <sup>xxx</sup>
sa-sba dx	+0.03	-1.00	-0.25	-0.90
sa-sba sin	+0.06	-0.88	-0.17	-1.07
pra-pa dx <sup>(+)</sup>	-0.19	-1.29 <sup>xx</sup>	-1.08	-0.52
pra-pa sin	-0.25	-1.16 <sup>x</sup>	-1.39	-0.84 <sup>x</sup>
protrusion dx	-3.82	-2.41	-2.94	-1.26
protrusion sin	-0.85	+2.25	+2.74	+1.05
inclination dx	+2.54 <sup>xx</sup>	+1.58 <sup>(x)</sup>	+0.05	+0.42
inclination sin	+2.67 <sup>xx</sup>	+1.89 <sup>x</sup>	+0.50	+0.10
prn-sn	-0.51	-0.38	-0.62	-0.11
prn-sbal dx	-1.23 <sup>xx</sup>	-0.92 <sup>(x)</sup>	-0.73	-1.82 <sup>xxx</sup>
prn-sbal sin	-1.36 <sup>xxx</sup>	-0.79	-0.75	-3.48 <sup>xxx</sup>
sn-sbal dx	-0.61 <sup>x</sup>	-0.92 <sup>xx</sup>	-1.03 <sup>x</sup>	+0.55 <sup>(x)</sup>
sn-sbal sin	-0.86 <sup>xx</sup>	-0.74 <sup>xx</sup>	-0.35	-0.94 <sup>xxx</sup>
sn-ls <sup>+</sup>	-1.52 <sup>xx</sup>	-0.05	-0.78	-4.20 <sup>xxx</sup>
sbal-ll dx <sup>++</sup>	-2.39 <sup>xxx</sup>	-0.98 <sup>(x)</sup>	-1.97 <sup>x</sup>	-1.40 <sup>xx</sup>
sbal-ll sin <sup>+</sup>	-2.18 <sup>xxx</sup>	-0.86 <sup>(x)</sup>	-1.72 <sup>x</sup>	-0.59
nose deviation <sup>0</sup>	0.78	0.29	0.07	2.83 <sup>xxx</sup>
prn deviation <sup>0</sup>	0.43	0.29	0.08	2.64 <sup>xxx</sup>
columel. deviat. <sup>0</sup>	0.02	0.20	0.91	0.91
m-prn	0.25	0.04	0.06	1.74 <sup>xxx</sup>
m-sn	0.38 <sup>x</sup>	0.03	0.18	2.01 <sup>xxx</sup>
Topinard dx	-0.09	-0.09	-0.07	+0.71 <sup>xxx</sup>
Topinard sin	-0.09	-0.12	-0.07	-0.05

(<sup>x</sup>)p<0.1    <sup>x</sup>p<0.05    <sup>xx</sup>p<0.01    <sup>xxx</sup>p<0.001    <sup>0</sup>in degrees

<sup>+</sup>significant differences between complete and incomplete clefts at p<0.05, <sup>++</sup>at p<0.01,

(<sup>+</sup>)at p<0.1



was in individual subgroups the more marked the smaller the extent of the cleft palate.

**Mandibular region:** Mandibular width (go-go) or the height of the lower face (sto-gn) were changed in no subgroup, while there was a significant reduction of the mandibular body length (go-gn) as well as of the dimension which was in good correlation with the length of the mandibular branch (t-go). The shortening of both dimensions conspicuously was substantially smaller in complete clefts ( $p < 0.05$ ) than in incomplete clefts or in clefts of the soft palate alone. Therefore these two subgroups alone showed a significant reduction of the submandibular arch (t-gn-t).

**Oronasal region:** The nose was narrowed (al-al) as well as the width of nostrils thresholds (sn-sbal) and the length of nasal wings (prn-sbal), even though the differences in the latter characteristic did not attain regularly the significance level. The narrowing was reflected by lower values of the nasal index. Nasal depth was unchanged (prn-sn) and we failed to reveal within this region any deviations of the nasal structures or differences between both sides (inclusive of the nares shape according to Topinard). The oral slot was highly significantly narrowed in all subgroups (ch-ch,  $p < 0.001$ ), the difference from controls was twofold as compared to CLP<sub>uni</sub> (the cheilozygomatic index:  $p < 0.001$ ). The upper lip was somewhat shortened below the insertion of nasal wings (sbal-l), especially in complete clefts, where the difference was significant at the median plane as well (sn-ls). However the latter observation was not confirmed by X-ray cephalometry and represented the single discrepant result obtained by both methods.

**Laterofacial regions:** Among the laterofacial dimensions were reduced in the first place those which showed some relation to the retrusion of the middle face (t-sn, obi-sn, t-ch). Similarly as in the case of the subnasal arch they were the more markedly shortened the slighter the cleft palate. A reduction was present also in characteristics which were in some relations to the shortening of the mandibular branch (go-ex), or to the shortening of the mandibular body and the retrusion of the maxilla (go-ch), without any differences between individual subgroups. Similarly as changes of the submandibular arch the distance between the chin and the tragion (t-gn) was reduced only in incomplete clefts and in cleft soft palate. However, in incomplete clefts the difference was substantially smaller because of the slightly elongated face (n-gn) and since the chin was thus situated at a lower level (in spite of its slight retrognathia). From the above mentioned followed that deviations from normal were regularly most marked in cleft soft palate alone with a significant reduction of some other characteristics as well (t-ex, obs-n). This was in agreement with facial width dimensions (t-t, zy-zy, go-go a. o.) reduced most markedly in this very subgroup.

**Ear lobes:** No deviations from normal were ascertained in the size (sa-sba, pra-pa), protrusion or localization of ear lobes, the width (pra-pa) was slightly reduced in incomplete clefts alone. The inclination of the ear lobe was more marked in incomplete and complete clefts. This was related to the

increased frequency of ear lobe retroinclination, which was ascertained in 12 individuals within our series as a whole (i. e. in 13.3 %,  $p < 0.01$ ). Out of these five had unilateral retroinclination only. The ear lobe was situated bilaterally at a low level in three patients (3.3 % insign.); it was always associated with retroinclination. An ear lobe protrusion was present in five individuals (5.6 %,  $p < 0.05$ ), however only one patient had bilateral protrusions.

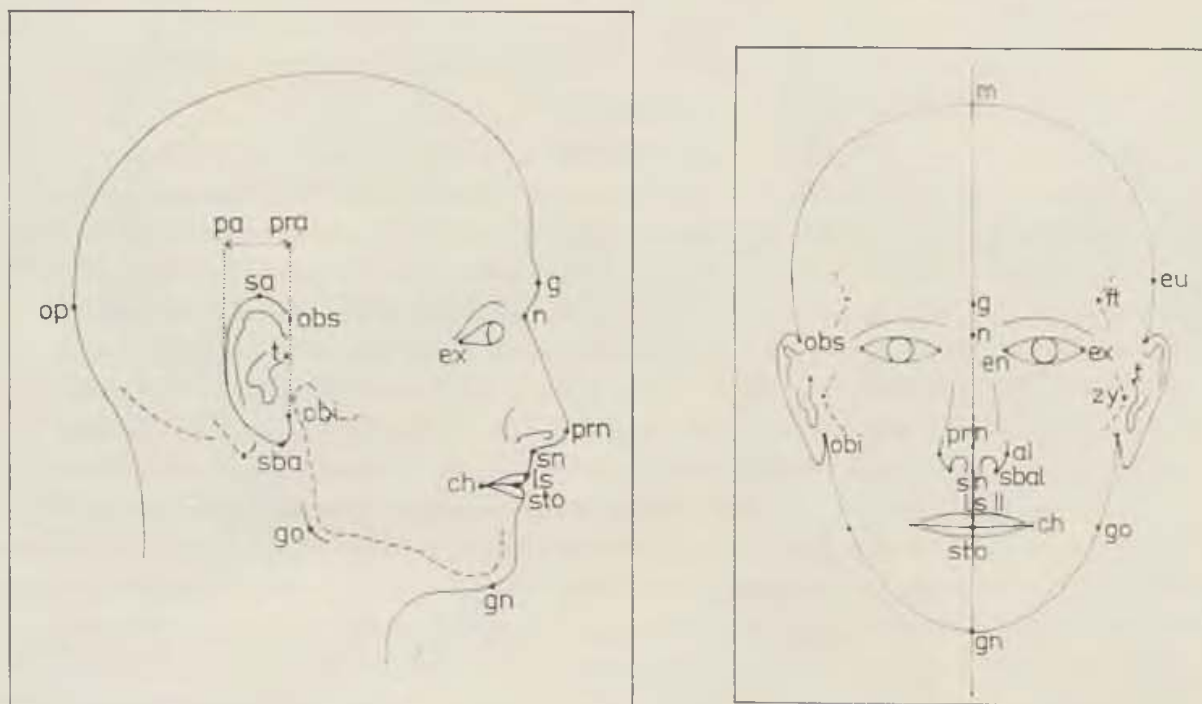


Fig. 1, 2. Cephalometric points used in the study.

**The face as a whole:** The total height of the face [n-gn] showed significant changes in none of our subgroups, but was slightly increased in incomplete clefts which was in agreement with the teleroentgenographic patterns which showed significant elongation. The investigated indexes confirmed that facial proportions were not altered, but for the width of the nose and oral slot (according to the values of nasal and cheilozygomatic indexes).

**Facial asymmetry:** No significant differences were disclosed between the dimensions of the right and left side, inclusive of the oronasal region. Some asymmetries were recorded in characteristics estimated by visual inspection (see below).

**Somatoscopic characteristics:** Visual inspection of the ear lobe features failed to disclose significant differences as compared to those in controls, however, anomalies of the outer ear were more frequent (in 24 individuals, i. e. 26.7 % as compared to 4 % in the group of controls i. e.  $p < 0.001$ ). They included both changes of position in 17 individuals (i. e. 18.9 %,  $p < 0.001$ ), and atypical shape in 10 individuals (i. e. 11.1 %,  $p < 0.01$ ).

Among facial somatoscopic features the vertical difference between the position of the inner eye canthi occurred more frequently as compared to con-

trols (30 % and 10 % resp.  $p < 0.001$ ). Vertical dislocation in the insertion of nasal wings occurred in 41.1 % of our series as a whole, as compared to 32 % in controls. Their depth showed changes in 22.2 % (as compared to 16 % in controls). The deviation of the septum within the nasal cavity was present in 3.3 %. All these differences were not significant. Retrocheilia was recorded in a single case, yet in further eight individuals examined (8.9 %,  $p < 0.01$ ) the upper lip was situated in anteroposterior direction at the level of the lower lip and thus showed a good prominence in 90 % of the patients, as compared to all controls. Other significant deviations from normal were not disclosed (asymmetric nares in 13.3 % as compared to 8 % in controls, epicanthus in 3.3 % and 4 % resp., antimongoloid palpebral slits in 1.1 % etc.).

**Variability:** As compared to controls the variability was increased only very rarely (in each subgroup in two different characteristics). It was reduced only in the length of nasal wings in the subgroup of complete clefts. Because of the large numbers of investigated characteristics these deviations from normal were considered solely as accidental. The similarity of variability in the two series confirmed that they were actually homogeneous and fully comparable.

#### DISCUSSION

In our series of adult males with isolated cleft palate the reduction of body height actually was very small ( $-0.3$  SD) and thus did not allow an interpretation of etiopathogenetic factors involved. Experimental results (Peterka and Jelínek, 1981) suggested that isolated cleft palate developed later than cleft lip and palate under a more potent action of teratogenic agents. This observation could explain a possibly more marked global involvement of the organism in CP than in CLP which should be the more extensive, the smaller the isolated cleft palate (because of a relatively late development a small cleft palate requires the action of a highly potent teratogen which would result in most severe repercussions). This relationship was suggested by somatometric characteristics in incomplete cleft palate as compared to those in complete clefts. The numbers of patients with cleft soft palate alone were too small to allow a similar comparison. However, the deviations of body height were always smaller in the CP subgroups than in CLP<sub>uni</sub> and their secondary origin (Smahel, 1982) so far represented the most satisfactory explanation. The extent of participation of primary factors could be determined only on the basis of comprehensive studies, including the whole period of growth. However, the situation would be somewhat different in primarily involved structures (i. e. the jaws) where the above described relationship could be anticipated.

The reported cephalometric patterns were in good agreement with the results of roentgenographic measurements described in another paper (Smahel, in press). These results showed that contrary to CLP and isolated cleft palate was not associated with a reduction of vertical growth of the upper face and with an increased interocular distance and therefore also of the minimum frontal width. These deviations from normal, as well as some others (there was no increase of anterior mandibular height and thus also of the lower



face) were confirmed by cephalometric measurements. The absence of a disorder of the vertical growth of the upper face was probably due to the normal continuity of the maxilla and thus of its normal interaction with the nasal septum. This was in agreement with the pattern in incomplete CLP<sub>uni</sub> (Šmahel, 1982), where the continuity was partially maintained as well. Normal interocular distance was confirmed in CP also by other authors[ see below].

The interpretation of X-ray films disclosed simultaneously that the most severe skeletal deviations generally associated with clefts, i. e. shortening of maxillary depth and deficient mandibular growth occurred both in isolated cleft palate and in cleft lip and palate. They resulted in retrusion of both jaws, especially of the maxilla. There were however some exceptions in individual CP subgroups. In complete cleft palate the mandibular body showed only a slight shortening resulting in minimum retrognathia. In cleft soft palate alone, on the contrary, the retrusion of the maxilla was slighter (by one half) as compared to the other two subgroups. Generally the global skeletal anomalies were most marked in incomplete cleft palate, where changes of the skeletal facial profile corresponded to the situation in CLP<sub>uni</sub> with an increase of the total facial height. The soft profile presented however a different situation. In complete and incomplete cleft palate maxillary retrusion was to a marked degree masked by soft tissues (and therefore no retrochelia occurred), while this was not present in cleft soft palate alone. Thus the soft facial profile showed the largest deviations from controls (i. e. retrusion) in cleft soft palate and the slightest changes in complete cleft palate (Šmahel, in press). The background of this pattern provided an explanation of findings obtained by cephalometric measurements including the soft profile. Dimensions reflecting the retrusion of the middle face (t-sn-t, t-sn, obi-sn, t-ch) were the more markedly reduced, the lesser the cleft palate. The dimensions reflecting mandibular retrognathia (t-gn-t, t-gn) were not reduced in complete cleft palate. The smallest shortening of the mandible in complete clefts is important in etiopathogenetic view since this type of isolated CP could not be due to mandibular growth retardation during embryogenesis as it is the case in the other types of cleft palate. This question is discussed in the second part of our study (Šmahel, in press).

Generally cleft soft palate was associated with smallest facial dimensions which could be due to the more potent teratogenic impulse. Visual inspection resulted in reports of certain changes in facial physiognomy in isolated cleft palate and thus it could be possible that they were most conspicuous in soft cleft palate alone. This also suggested the above described deviations of the soft profile and the smaller face in this type of CP. In addition to these characteristics some specific characteristics within the oronasal region were described, inclusive of the reduced dimensions of the nose, of the upper lip and especially of the width of the oral slot. These changes were conspicuous during visual inspection in children and could be due partially to respiratory particularities (differing air circulation and maxillary retrusion could result in a smaller nose), though it failed to provide an explanation for the trend towards microstomia. These changes were found in all types of isolated cleft



palate, but there was no relation between the degree of the cleft and the extent of the deviations (however, in CPs the numbers of cases were too small).

The increased frequency of ear lobe anomalies in our series was in agreement with the important role of exogenous factors in the etiology of these malformations. An almost twofold occurrence as compared to CLP<sub>uni</sub> corresponded to the higher proportions of nongenetic factors involved and to the reported higher frequency of associated anomalies (Klásková, 1971 a. o.). Similarly as in CLP<sub>uni</sub> a retroinclination of the ear lobe was the most frequent deviation, which was in agreement with reports by other authors quoted by Šmahel (1982). This resulted also in the more obtuse angle of ear lobe inclination. According to the results of our studies no other substantial deviations from normal of the other parameters of the outer ear could be expected.

With the exception of some smaller irregularities of somatoscopic characteristics we failed to disclose any facial asymmetries inclusive of those within the oronasal region. Because of the character of this malformation there were no mechanical or some other reasons which could account for their development. The facial proportions were unchanged as well.

There are relatively only a few anthropometric studies on patients with isolated cleft palate. Dahl (1970) ascertained a reduction of body height by 6.7 cm in adult males with this type of cleft as compared to a series of controls including university students. However, in comparison to body height in conscripts this difference amounted only to 2.3 cm, which was in agreement with our findings. The smaller stature in children with clefts was underlined already by Ross and Coupe (1965), a retarded maturation by Menius et al. (1966). Januszewska (1968) mentioned smaller body length and weight from three to six months of age. Ranalli and Mazaheri (1975) failed to disclose any changes in the growth rate of children with clefts up to six years of age, and Hunter and Dijkman (1977) mentioned a certain height-weight deficiency only from ten years upwards. Malinowski et al. (1973) recorded in children and adolescents with clefts deviations from normal in body weight, shoulder width and chest depth, but failed to disclose any changes of body height, circumference and width of the chest, pelvic width or length of the lower extremities and of the trunk. Deviations in the depth of the chest could be related to the differing type of respiration. Virtually no pertinent data on the neurocranium were found in the literature, only Dahl (1970) mentioned in males a smaller circumference, length and to a lesser degree also the width of the neurocranium. The difference could be due partially to the higher than average body height in his control series of students (see above).

Smaller facial width was mentioned regularly at a low significance level by Dahl (1970) and by Farkas and Lindsay (1972a) which was in agreement with the more gracile face. Unchanged biauricular width (t-t) was ascertained in both of these reports. The latter two authors investigated in more detail the face in adult individuals with isolated cleft palate and equally found reduced subnasal and submandibular arches and a reduction of nasal width and especially of the oral slot (by 8 mm). The height of the upper face was identical with that in controls. Yet these authors failed to observe a reduction

of the height of the upper lip. Somatoscopic features were assessed with a differing procedure and thus no definite comparison was possible (they mentioned the more vertical position of the nares; an asymmetry of the nares did not occur more frequently — in 9.6 %).

The absence of changes in the interocular distance in isolated cleft palate was confirmed by the studies of Farkas and Lindsay (1972b) in adults and by Figalová et al. (1974) in children prior to surgery. Both reports showed, at the same time, that the distance between the outer eye canthi could be reduced, though the differences were not always significant. The narrowing was due to smaller facial width and was not of an extent which could result in adults in changes of proportions within the ocular region (see intercanthal index).

Ear lobe configuration was studied also in isolated cleft palate by Farkas and Lindsay (1973). They found a narrowing of the ear lobe and a more obtuse inclination angle. In our series the width of the ear lobe was reduced in the subgroup with an incomplete cleft and its inclination was increased in both subgroups with cleft hard palate. The above quoted authors mentioned that the ear lobe was situated at a lower level, due to the smaller distance between the chin and the lower margin of ear lobe insertion (gn-obi). However this dimension characterized retrognathia. In their earlier study Farkas and Lindsay (1972a) determined further an increased frequency of ear lobe deformities.

We failed to disclose in the literature any similar data relating the deviations in isolated cleft palate to the extent of cleft.

#### SUMMARY

Anthropometric and visual studies were carried out in 90 adult males with isolated cleft palate operated upon with an identical method. The results obtained were assessed in individual subgroups according to the extent of the cleft (complete, incomplete and soft palate alone).

Body characteristics in our patients were only slightly below the average and also the neurocranium failed to show any significant deviations from normal. Contrary to cleft lip and palate there was no reduction of the height of upper face or an increase of the height of the lower face, or of the interocular distance, while there was a retrusion of the middle face and a shortened mandible, especially in slighter degrees of clefts. Cleft soft palate was associated with the most marked reduction of facial dimensions. The oronasal region was more gracile, the nasal width, the height of the upper lip and in particular the width of the oral slot were all of them reduced (there was a tendency towards microstomia). The facial proportions were not disturbed, an asymmetry was absent even within the oronasal region. No deviations were found in the size and configuration of the ear lobe, there was only a more obtuse inclination angle and a marked increase of the frequency of anomalies (26.7 %). There were some differences between individual subgroups. Some particularities were explained on the basis of the roentgencephalometric pattern (e. g. the reduction of the subnasal arch was the more marked the lesser the extent of the cleft). Retrocheilia did not occur.

## RESUME

### **Les modifications céphalométriques et morphologiques chez les hommes adultes avec la division palatine isolée**

Šmahel, Z.

Du point de vue d'anthropologie, on a examiné 90 hommes adultes avec la division palatine isolée qui ont été opérés par la même méthode. Ils sont évalués dans les sous-groupes selon le degré de la malformation (division totale, partielle ou seulement division du voile du palais).

Des caractéristiques corporelles sont d'une légère médiocrité, le crâne ne montre pas de déviations significatives. Contrairement au bec-de-lièvre, la hauteur du visage supérieur et du visage inférieur n'est pas diminuée, la distance entre les yeux n'est pas aussi diminuée. La partie moyenne du visage est retroussée, la mandibule est raccourcie, surtout dans les cas des divisions moins graves. Les dimensions du visage sont diminuées au maximum s'il s'agit de la division du voile palatin. La région oralo-nasale est plus gracile, la largeur du nez est réduite aussi que la hauteur du lèvre supérieur et surtout la largeur de la fissure orale (tendance à la microstomie). La proportionnalité du visage n'est pas rompue, les asymétries ne se manifestent ni dans la région oralonasale. La grandeur et la formation du pavillon de l'oreille ne montrent pas de déviations, mais l'angle d'inclination est augmenté aussi que la fréquence des anomalies (26,7 %).

Parmi les sous-groupes il y a quelques différences, les particularités sont expliquées sur la base de radiocéphalométrie. Rétrochéilie n'était pas trouvée.

## ZUSAMMENFASSUNG

### **Kephalometrische und morphologische Veränderungen bei erwachsenen Männern mit isolierter Gaumenspaltung**

Šmahel, Z.

Antropologisch wurden 90 erwachsene Männer untersucht, die eine mit gleicher Methode operierte isolierte Gaumenspaltung aufweisen. Sie wurden in Untergruppen je nach dem Umfang der Spaltung (vollständige, unvollständige Spaltung und Spaltung nur des weichen Gaumens) eingeschätzt.

Die körperlichen Charakteristiken sind nur schwach unter dem Durchschnitt und auch der Gehirnkasten weist keine bedeutsamen Abweichungen auf. Im Gegensatz zu Lippen- und Gaumenspalten ist die Höhe des Gesichtsrückenteils nicht kleiner und auch die Höhe des Gesichtsvorderteils oder die Entfernung zwischen den Augen nicht grösser, dagegen ist eine Retrusion der Mitteletage des Gesichts vorhanden und der Unterkiefer ist kürzer, besonders bei leichtem Grad der Spalte. Die Abmessungen des Gesichts sind am meisten verkleinert bei Spaltungen des weichen Gaumens. Das oronasale Gebiet ist graciler, die Breite der Nase, die Höhe der Oberlippe und vor allem die Breite der Mundspalte ist kleiner (Tendenz zu Mikrostomie). Die Proportionalität des Gesichts ist nicht gestört, und auch im oronasalen Gebiet gibt es keine Asymmetrie. In der Grösse und Form der Ohrklappen wurden keine Abweichungen gefunden, jedoch der Inklinationswinkel ist grösser und die Frequenz der Abnormalitäten häufiger (26,7 %). Zwischen den einzelnen Untergruppen existieren einige Unterschiede, und die Besonderheiten werden auf Grund der Befunde der Roentgenkephalometrie erklärt. Retrocheilien traten nicht auf.



## RESUMEN

### Cambios morfológicos y cefalométricos en hombres adultos con fisura paladial aislada

Šmahel, Z.

Fueron sometidos a exámenes antropológicos 90 hombres adultos, todos con fisura paladial aislada y operados por el mismo métodos. Se los evalúa en grupos correspondientes a la envergadura de la fisura (completa, incompleta y sólo la del velo).

Las características del cuerpo son sólo ligeramente inferiores al promedio, tampoco el cráneo cerebral presenta anomalías significantes. En comparación con las escisiones labiopaladiales no se detecta acortamiento de la altura de la parte superior de la cara ni una excesiva altura de la parte inferior de la cara o una mayor distancia entre los ojos, sin embargo se puede comprobar la existencia del arremango de la parte media de la cara, y la mandíbula presenta acortamiento particularmente en caso de escisiones menos graves. La zona oronasal resulta más grácil y se observa una reducción del ancho de la nariz, de la altura del labio superior y ante todo del ancho de la boca (tendencia a la microstomia). La proporcionalidad de la cara no resulta alterada, no hay asimetrías ni en la zona oronasal. No se han detectado diferencias en cuanto al tamaño y la formación del pabellón, sin embargo es más grande el ángulo de inclinación y mucho mayor resulta la frecuencia de las anormalidades (26,7 %). Entre los individuales grupos existen ciertas diferencias, las particularidades se explican con ayuda de lo detectado por radicefalometría. La retroquellía no existe.

## REFERENCES

1. Farkas, L. G., Lindsay, W. K.: Morphology of Adult Face after Repair of Isolated Cleft Palate in Childhood. *Cleft Palate J.*, 9 : 132, 1972a.
  2. Farkas, L. G., Lindsay, W. K.: Morphology of the Orbital Region in Adults Following the Cleft Lip/Palate Repair in Childhood. *Am. J. Phys. Anthropol.*, 37 : 65, 1972b.
  3. Farkas, L. G., Lindsay, W. K.: Ear Morphology in Cleft Lip and Palate Anomaly. *Arch. Oto-Rhino-Laryng.*, 206 : 57, 1973.
  4. Januszewska, W.: Rola lekarza pediatrii w leczeniu dzieci z rozszczepem wargi górnej i podniebienia. *Czas. Stomat.*, 21 : 641, 1968.
  5. Klásková, O.: Epidemiologický průzkum rozštěpových vad. *Čs. Pediat.*, 26 : 584, 1971.
  6. Malinowski, A., Cieslik, J., Prusiewicz, A., Obrebowski, A.: Budowa somatyczna dziewcząt i chłopców s rozszczepem podniebienia w zależności od dojrzałości morfologicznej. *Przegl. Antrop.*, 39 : 133, 1973.
  7. Menius, J. A., Largent, M. D., Vincent, Ch. J.: Skeletal development of Cleft Palate Children as Determined by Hand-Wrist Roentgenographs: A Preliminary Study. *Cleft Palate J.*, 3 : 67, 1966.
  8. Peterka, M., Jelínek, R.: Cleft Palate — a Morphogenetically Heterogeneous Congenital Defect. *Acta Chir. plast.*, 23 : 152, 1981.
  9. Ranalli, D. N., Mazaheri, M.: Height-Weight Growth of Cleft Children, Birth to Six Years. *Cleft Palate J.*, 12 : 400, 1975.
  10. Ross, R. B., Coupe, T. B.: Craniofacial Morphology in Six Pairs of Monozygotic Twins Discordant for Cleft Lip and Palate. *J. Can. Dent. Ass.*, 31 : 149, 1965.
  11. Šmahel, Z.: Cephalometric and Morphologic Changes Associated with Unilateral Cleft Lip and Palate in Adults. *Acta Chir. plast.*, 24 : 1, 1982.
  12. Šmahel, Z.: Craniofacial Morphology in Isolated Cleft Palate in Adults in Relation to the Extent of the Cleft. In press.
- The other references are quoted in Šmahel (1982).

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## THE LONG-TERMED RESULTS OF THE PLASTIC SURGERY OF THE EYELIDS AND ORBITAL REGION DEFECTS RESULTING FROM WARTIME FIREARMS WOUNDS

M. V. ZAJKOVA, E. V. KOROLEVA

A lot of studies concerned with the covering of the defects of the eyelids after injuries caused by wartime firearms. Most authors always used only one technique of blepharoplasty (Kolen, 1943, 1950; Belokrinkin, 1969; and others). The combination of individual techniques including allografting of the bone and cartilage was employed rather rarely (Tomasjevskaja, 1947; Zajkova, 1969; and others). To remove broad defects of the eyelids combined with defects of the orbital and other facial regions also the tubed flaps has been used successfully (Tomasjevskaja, 1948; Muchin, 1951; Chitrov, 1954, 1960; Zajkova, 1981; and others).

However, no data concerning the late postoperative conditions of the patients with the defects resulting from firearms wounds are found in the literature. Neither indications nor contraindications for different techniques of blepharoplasty have been studied.

We have studied specific features and long-termed results of the eyelid and orbit plasties in 107 patients wounded in 1941—1945 (587 surgical interventions were performed altogether).

Simple defects occurred in 27 % of patients, in 73 % the eyelid defects were complex, i. e., combined with defects of other anatomical regions of the face. The clinical manifestations of complex defects varied considerably. The broad through and through defects of the eyelids usually combined with defects of several walls of the orbit, and in many patients the adjacent regions were destroyed (the nose, cheeks, forehead and temple). In all cases the globe of the wounded side was destroyed (anophtalmos — 104, atrophy — 3 cases).

The surgical technique was selected with respect to the size and localization of the defect and conditions of its margins and adjacent regions as well. The individual operations differed considerably but one of the techniques of the plasty performed in several stages was usually considered as principal.

A simple plasty was performed in 13 patients: in 5 of them with simple defects, in 8 with complex ones. The eyelid deformations from scars were removed using local tissue and the pedicle flaps from the adjacent skin. The

flaps were formed and shifted in one stage. The tubed flap was used as the only technique in 11 patients (54 operations) suffering from complete penetrating defects of the eyelids combined with nasoorbital defects as a result of a shell-splinter wound. The operations were performed at maxillofacial and ophthalmology departments. The defects were removed at the total absence of the adjacent tissue reserve with no resulting scars in the face.



Fig. 1. Patient C., wounded by an explosive shot. A complete defect of the lower eyelid, defect of the left inferior external orbital wall. Before the operation.

A combined one-stage blepharoplasty, i. e., combination of the skin plasty with transplantation of the skin autografts and allografts of the bone and cartilage was performed in 18 patients (54 operations). The combination of the above mentioned surgical approaches enabled us to use reserves of the healthy tissues and to remove scars of the eyelids and of orbit margins or walls as well. The combined plasty with the pedicle flap to remove penetrating defects of one or both eyelids was performed in 7 patients. In cases of the anophthalmos the inner plate of the eyelid was formed by turning over the skin flap elevated on a broad base from adjacent tissues. An eye prosthesis was inserted. The pedicle flap was sutured in the margin of the eyelid defect. The flap was formed from the upper eyelid, forehead, temple and nasolabial fold with respect to the extent of the eyelid defect. Defects of the bony walls of the orbit were covered by transplanting allografts of the cartilage and bone.

In 5 patients the plasty using the pointed tubed microflap, i. e., the skin flap on the tubed pedicle (Zajkova, 1969) was used. The pointed tubed microflap was cut out of the region of the upper eyelid, and the pointed tubed flap out of the temple region. Both these flaps were employed under the same indications as those for the pedicle flap.

The multistage combined plasty using the tubed flap was applied in 6 patients (47 operations). The indications for this type of the plasty, combined

with various techniques using local tissues, were complex defects of the eyelid and orbits.

To remove broad intricate defects of the eyelids (both simple and complex ones) a multistage combined blepharoplasty was used (354 surgical interventions in 76 patients). All plasties involved transplantations of the skin autografts and allografts of the cartilage and bone.



Fig. 2. The same patient after the second stage of the treatment. The lower pedicle of the tubed flap formed on the left aspect of the chest was transferred.

The severe scar deformation of the eyelids in 3 patients wounded by a shell-splinter were removed using the plasty with adjacent tissue (according Imre), Z-plasty (Linberg) and the lateral advancement successively.

The pedicle flaps were used in 28 patients (113 operations). Altogether 46 pedicle flaps were formed, of this number 28 at the first stage of the treatment. Transplantation of the cartilage was used for covering of extensive defects of the bony walls of the orbit in combination with the pedicle flaps and tubed flaps.

Plasty using the tubed flap was performed in 41 patients (342 surgical interventions) in cases of broad defects of the eyelids and orbital region. The tubed flaps were formed on the temple (4), shoulder (5), neck (2) and chest

(36). Covering of nasoorbital defects required two or three flaps. Variants of the tubed flap plasty were dependent on the extent and localization of the defect, and were always combined with all previously mentioned techniques, including the grafts.

The healing without complications occurred in 96.6 %, and in 3,4 % of cases complications occurred. Immediately unsatisfactory results occurred in 3 patients due to complete necrosis of the pedicle flaps caused by heavy scars of the defect margins.



Fig. 3. The same patient. Plasty with the tubed flap.

The long-termed observation (from 6 months to 37 years) showed satisfactory results in 94.9 % of patients.

To demonstrate good functional and cosmetic results we present abbreviated case history of two patients.

1. Patient C., aged 28. Admitted on August 28, 1949. Diagnosis: atrophy of the left globe, complete penetrating defect of the lower eyelid and defect of the lower external wall of the orbit as the result of explosive shot wound in 1942.

At admission only small clod was left of the left globe. The upper lid was lowered, the lower lid completely destroyed. A deep scar and the defects of the lower and external orbital walls were grown together (figure 1). August 26, 1949. The remains of the left globe were removed.

September 10, 1949. The tubed flap (20 X 8 cm) was formed on the left anterior aspect of the chest.

October 11, 1949. The lower pedicle of the flap was transferred to the left zygomatic region (figure 2).

December 29, 1949. The complete plasty of the lower lid and of lower and external orbital walls by the tubed flap was performed. The scars were excised



along the lower external margin of the orbit. The flap, formed in this way, was turned over and fixed at the level of the eye corner. The upper pedicle of the tubed flap was transected, spread out and implanted into the margins of the defect. An eye prosthesis was inserted (figure 3). December 15, 1959. The piece of cartilage ( $2 \times 1 \times 1$  cm), fixed in ethanol, was transplanted.



Fig. 4. The same patient after 20 years. — Fig. 5. Patient F., wounded by a mine-splinter. A complete penetrating defect of the lower eyelid, defect of the inner and lower walls of the orbit, defect of the nose, scars in the left face. The situation after transfer of the lower pedicle of the tubed flap.

The patient was observed for 20 years. The lower eyelid and left orbital region were restored completely. The position of the left eye prosthesis corresponded to the position of the right healthy eye (figure 4).

2. Patient F., born in 1921. Admitted on May 12, 1945. Diagnosis: Anophthalmos, a complete through and through defect of the lower eyelid, defect of the inner and inferior walls of the orbit, defect of the nose, scars on the left face. The lower eyelid completely absent. Wounded by a mine splinter in 1945. Without any previous surgical interventions.

May 21, 1945. The first stage of the surgery — the tubed flap was formed on the left aspect of the chest ( $22 \times 8$  cm).

August 2, 1945. The second stage — the lower pedicle of the flap was transferred to the left cheek (figure 5).

September 2, 1945. The third stage — the upper pedicle was transferred to the glabellar region (figure 6).

October 26, 1945. The fourth stage — the rhinoplasty was performed by maxillo-facial surgeons.



Fig. 6. The same patient. The transfer of the upper pedicle.



Fig. 7. The same patient. Plasty of the lower eyelid and nose using the tubed flap. —

December 21, 1945. The fifth stage — the complete plasty of the lower eyelid. The inner plate of the eyelid was formed by turning over the skin flap. The skin of the lid was formed from a part of the tubed flap transferred to the cheek region.



Fig. 8. The same patient after 35 years.

February 2, 1946. The sixth stage — the complete reconstruction of the lower eyelid by the tubed flap. For the lower arch of the cavity the flap cut out of the implanted tubed flap was turned over. After cantoplasty an eye prosthesis was inserted. The lid skin was replaced by material of the tubed flap (figure 7).

July 7, 1947. The seventh stage — the reconstruction of the lower lid using local tissues.

After 35 years (on December 3, 1981) the lids were completely restored and the eye prosthesis in correct position. The colour of the transplanted skin did not differ from that of adjacent skin (figure 8).

To summarize, the complex defect of the eyelid and of the orbital region was completely covered using one tubed flap and local tissues.

The main cause of uncomplete removal of the defects in 5.13 % of the patients seemed to be inappropriate technique, extremely extensive defects of the orbit and deep scars of the defect margins.

#### CONCLUSIONS

1. Defects of the eyelids and orbital region resulting from wartime firearm wounds are extremely variable, complicated and — in most cases — extensive.

2. In these defects, the main condition of the successful treatment is the choice of proper surgical technique for the blepharoplasty. Therefore, the degree of impairment of the anatomical relations at the eyelid margins as well as in surrounding tissues must be considered.

3. Choice of the technique for the blepharoplasty cannot be inflexible. Each technique has indications and contraindications of its own. The extent of the eyelid defect, its localization, conditions of its margins as well as of surrounding anatomical regions have to be taken into account.

4. What follows are the main principles of the plastic reconstruction of the eyelid and orbital regions in defects resulting from firearms wounds: a) excision of cutaneous and subcutaneous scars at the margins of the defect; b) the complete reconstruction of the eyelids and orbital region using local tissues as well as allografts of the cartilage and bone (in case of orbital walls defects); c) immediate insertion of an eye prosthesis; d) the plasty should proceed from periphery to the centre, i. e., it has to start with covering of orbital and facial defects and proceed to formation of the eyelids.

5. In cases of wartime firearms injuries, the simple technique can be used successfully only for covering of the superficial defects of the eyelids. The principal technique for complex deep defects of the eyelids is the combined blepharoplasty performed in several stages increases effectiveness of the reconstructive treatment of complex deformations and broad defects of the eyelids. The categorical indications for the tubed flap are nasoorbital complications of the eyelid defects as well as extensive injuries of the orbit and of adjacent regions.

M .D.

#### S U M M A R Y

The eyelid defects resulting from wartime firearms wounds are extremely diverse, the anophtalmos being the rule. In our patients, penetrating defects of the eyelids, combined with defects of the bony orbital walls and of surrounding anatomical regions, predominated. Choice of proper surgical technique was governed by the size and localization of the defect. The simple defects of the eyelids were removed by a simple plasty using local tissues and the pedicle flaps. More complicated deformities and penetrating defects (both solitary and complex ones) were treated by combined plasty in one stage. Extensive deformities and complex penetrating defects of the eyelids and orbital walls were treated by a series of surgical interventions using different techniques of the skin plasty and tissue transplantation. The most extensive defects of the eyelids, orbital walls and surrounding structures were covered using the tubed flap in combination with previously mentioned techniques. The results were evaluated in the period up to 37 years and were satisfactory in 94.9 % of patients.

#### R E S U M E

**Les résultats des opérations plastiques sur les paupières et dans la région orbitaire en traitant les traumatismes causés par des armes à feu**

Zajkova, M. V., Koroleva, E. V.

Les défauts des paupières, causés par les traumatismes des armes à feu, se distinguent par sa multiplicité et sa complexité et, dans la plupart des cas, sont accompagnés par l'anophtalmie. Chez nos malades, les défauts pénétrants des paupières ont dominé,



souvent accompagnés par les défauts pénétrants des murs osseux de l'orbite ou d'autres structures anatomiques. Il faut que le choix du procédé opératoire soit individuel et dépendant de l'étendue et de localisation du défaut. Des déformités ulcéraives des paupières non compliquées ont été abolies à l'aide d'une plastie simple, en utilisant des tissus voisins et des lambeaux pédiculés. Les déformations plus compliquées et les défauts pénétrants des paupières (compliqués plus ou moins) étaient abolies par une plastie combinée, dans une étape unique. Quand il s'agissait de grandes déformités et des défauts pénétrants des paupières et d'orbite, on les a restreint par une plastie successive qui combine des techniques variées, les homogreffes incluses. Ces techniques-ci étaient enrichies des lambeaux cylindriques tubulés en cas où il s'agissait des défauts les plus vastes et les plus compliqués des paupières, de l'orbite et des structures voisines.

Les résultats du traitement étaient suivis pendant presque 37 ans et dans 94,9 % des cas étaient considérés comme satisfaisant.

#### ZUSAMMENFASSUNG

##### **Langfristige Ergebnisse einer Plastik der Augenlider und des Orbitalgebiets bei Verletzungen durch Kampfschüsse**

Zajkova, M. V., Koroleva, E. V.

Durch Schiesswaffen verursachte Verletzungen der Augenlider zeichnen sich durch aussergewöhnliche Vielseitigkeit und Kompliziertheit aus und werden in den meisten Fällen von Anophthalmie begleitet. Bei den von uns behandelten Patienten überwogen penetrierende Defekte der Augenlider, begleitet von penetrierenden Defekten der Knochenwände der Augenhöhle und der umliegenden anatomischen Strukturen. Die Wahl der Operationstechnik ist individuell und hängt vom Umfang und der Lokalisierung des Defekts ab. Unkomplizierte narbenartige Deformierungen der Augenlider wurden durch einfache Plastik unter Verwendung des umgebenden Gewebes und der stielartigen Lappen beseitigt. Komplizierte Deformierung und penetrierende Defekte der Augenlider (mit oder ohne Komplikationen) wurden durch kombinierte einmalige Plastik beseitigt. Bei komplizierten penetrierenden Defekten der Augenlider und der Augenhöhle wurden ausgedehnte Deformierungen durch stufenweise Plastik unter Anwendung einer Kombination verschiedener Techniken einschliesslich Transplantation von Allopfpfropfen beseitigt. Bei den umfangreichsten komplizierten Defekten der Augenlider, der Augenhöhle und der umgebenden Strukturen wurden in Kombination mit den angeführten Techniken und der Transplantation von Allopfpfropfen auch walzenförmige tubulierte Lappen angewendet. Die Ergebnisse der Behandlung wurden bis zu 37 Jahren beobachtet und zufriedenstellende Funktionsergebnisse wurden bei 94,9 % der Patienten erzielt.

#### RESUMEN

##### **Resultados duraderos de la plástica de los párpados y la zona orbital en caso de defectos producto de heridas por armas de fuego**

Zajkova, M. V., Koroleva, E. V.

Los defectos de los párpados causados por heridas por armas de fuego de combate tienen un carácter particularmente multiforme y complejo y son acompañados de anoftalmia en la mayoría de los casos. En los pacientes sometidos a nuestro tratamiento lo que predominó fueron los defectos penetrantes de los párpados acompañados de defectos penetrantes de las paredes óseas de la órbita y las estructuras anatómicas aledañas. La táctica de operación es diferente en los casos individuales en dependen-

cia de la localización y el tamaño del defecto. Deformaciones cicatrizales no complicadas fueron eliminadas por plásticas simples aprovechándose los tejidos circundantes y lóbulos con pecíolo. Deformaciones más complicadas y defectos penetrantes de los párpados (con y sin complicaciones) fueron eliminados por plásticas combinadas aisladas. En caso de complicados defectos penetrantes de los párpados y la órbita se eliminaron las deformaciones amplias con plásticas sucesivas combinándose diferentes técnicas incluyendo los alotransplantes. En defectos complicadísimos se utilizaron también los lóbulos tubulados cilíndricos combinándose con las técnicas de alotransplantes. Los resultados del tratamiento fueron controlados durante, al máximo, 37 años lográndose resultados funcionales satisfactorios en un 94,9 % de los pacientes.

#### REFERENCES

1. **Belokrinkin, D. A.:** Reconstruction of the Eyelid Defects. In: *Reconstructive Surgery of Disabled Soldiers of the Second World War*. Gorkij, 1960.
2. **Zajkova, M. V.:** Plastic Operations in Ophthalmology. M.: Medicina, 1969.
3. **Zajkova, M. V.:** The Tubed Flap „Filatov“ in Ophtalmic Surgery. Izjevsk, 1981.
4. **Kolen, A. A.:** Techniques of the Eyelid Plasty in War-Time Wounds. *Vest. Ophtalmol.*, 6 : 3, 1943.
5. **Kolen, A. A.:** Handbook of Plastic Surgery of the Eye Region. M., 1950.
6. **Muchin, M. V.:** The Tubed Flap (Filatov) in Reconstructive Surgery. In: *Problems of Orthopaedics, Traumatology and Prosthetics*. Charkov, 1951.
7. **Tomasjevskaja, A. G.:** Plastic Operation on Defects of the Eyelids Resulting from Fire-Arms Wounds. Thesis, Sverdlovsk, 1947.
8. **Tomasjevskaja, A. G.:** The Tubed Flap in Plastic Surgery of the Eyelid Defects Resulting from the Fire-Arms Wounds. In: *Reconstructive Surgery of the Injury Consequences*. Sverdlovsk, 1948.
9. **Chitrov, F. M.:** The Tubed Flap in Plastic Surgery of Face and Neck Defects. M.: Medicina, 1954.
10. **Chitrov, F. M.:** Reconstructive Surgery of Extensive Complicated Defects of the Face. *Chirurgija*, 6 : 68, 1960.

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## SELECTIVE OSTEOTOMY AND GRADUAL DISTRACTION IN TREATMENT OF PATIENTS SUFFERING FROM DEFORMITIES OF THE CENTRAL PART OF THE FACE

U. T. TAIROV

To manage both congenital and acquired deformities of the central third of the face is a great problem of present-day stomatological surgery, as occurrence of the maxillofacial anomalies, as well as of head traumatism, which is the cause of 12—15 % of these deformities, is relatively very frequent. Until lately treatment of micrognathia and retrognathism of the maxilla has been regarded as difficult and their surgical correction considered problematic. In most cases, a surgical intervention was performed on seemingly larger mandible, so that the normal jaw was adjusted to the abnormal one.

Recent progress in surgery and anaesthesiology made more frequent reconstructive operations of the facial skeleton possible (Bezrukov et al., 1977; Tessier, 1971; Epker and Fish, 1977; Bell, 1977).

The methods used have the following features in common. Osteotomy of the facial bones is performed at the so called weak spots with regard to the type of both the deformity and occlusion. The maxillary segment is immediately displaced and fixed in the anatomically normal position. The fixation is accomplished by means of metal sutures and intraoral as well as external apparatuses. Diastases between the osseous segments are filled by auto- and homografts.

However, such surgical interventions are rather traumatic, they cause a considerable loss of blood, and the filling of the diastases by grafts is extremely difficult and sometimes (with regard to anatomical relations) even impossible. Sutures of metal wire, bone grafts and intensive inflammatory reaction as well slow down the progress of regeneration of the osseous tissues. Consequently, the maxilla is mobile for a long time and remissions are frequent. Other possible complications are discussed by Westwood and Tilson (1975).

Therefore we tried to elaborate a considerate technique of surgical treatment of congenital and acquired deformities of the middle third of the face (that of the retrognathism and micrognathia of the upper jaw) enabling us to establish normal relations between the upper and lower jaws.



Research and surgery were carried out at the Department of Stomatology of the Central Research Institute, Moscow, and the Department of Surgical Stomatology of the Tadjik Medical Institute. Our approach is based on the results of operations moving the upper jaw bone both in the sagittal and transversal direction (Kalamkarov, 1967; Dellinger, 1973; Kambara, 1977; Jackson et al., 1979) and on the experience of traumatologists in lengthening of the extremities by means of the gradual distraction (Ilizarov, 1977; Lavrischeva and Shtin, 1976).

We had to solve two principal problems:

1. construction of an apparatus suitable for the gradual distraction, 2. elaboration of a considerate technique for selective osteotomy of the maxilla that would maintain the continuity of the periosteum and of the oral mucous membrane as well.

To fulfil these tasks we elaborated the new method<sup>1)</sup> whose nature rests in the following points. Under endotracheal anaesthesia the mucous membrane and periosteum are incised horizontally at the level 2—3 mm under the alveolar sulcus. The frontal and lateral aspects of the maxilla are exposed as far as the pterygoid processes of the sphenoid bone. Z-like incision is made in the area of the upper lip frenulum so that the region of the apertura nasi and of the nasal cavity floor is exposed. Bridges of intact mucous membrane and periosteum are left bilaterally at the region of the canine and the first praemolar. Osteotomy is performed with a thin fissure bur. The osteotomy cut begins at the level of the juncture of the nasal bones and it is carried horizontally from the apertura piriformis under the infraorbital foramen and then prolonged to and along the zygomaticomaxillaris suture. The lateral and posterior walls of the maxilla are transected at the level of the pterygoid processes of the sphenoid bone with a thin chisel. The pterygoid plates are detached from the tuber maxillae with a special hooked chisel and the nasal septum is transected. In order to increase the mobility of the cartilaginous nasal skeleton the quadrilateral nasal cartilages are transected subcutaneously (according to Bezrukov). When checking the mobility of the segment the wound is closed by cutgut sutures.

Modifications in character and line of osteotomy depended on the age of the patient, density of his bones and other specific features of the deformities. If the bony walls of the upper jaw are rather thin, the technique of incomplete osteotomy is used (see Fig. 1).

10—12 days after the operation a cast-metal cap with hooks is placed on the maxillary teeth. The displacement of the maxillary segment into the normal position is accomplished by an apparatus of our own construction<sup>2)</sup>.

The apparatus, supported by a thoracocervical corset [2], consists of a metal arch [4] with multiple notches, in which the distractors [6] are fixed.

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<sup>2)</sup> Author's certificate No. 876124 (coauthors: V. A. Sukatchevi and V. I. Gunko).

The arch is connected in the hinge-like manner with a vertical bar (3), the second end of which is attached to the corset. The distractors have apertures (9) for the hooks of the cap (1) and are provided with screws (8) for regulation of the pulling force. The proper position of the distractors — i. e., the proper direction of the pulling force — is secured by means of nuts and fixators (10, 11). The pulling force is generated by a spring (9) located at the axis of the distractor. The force can be decreased or increased by changing the length of the spring. The force produced at particular length of the spring is given in kilograms on the shaft of the distractor.

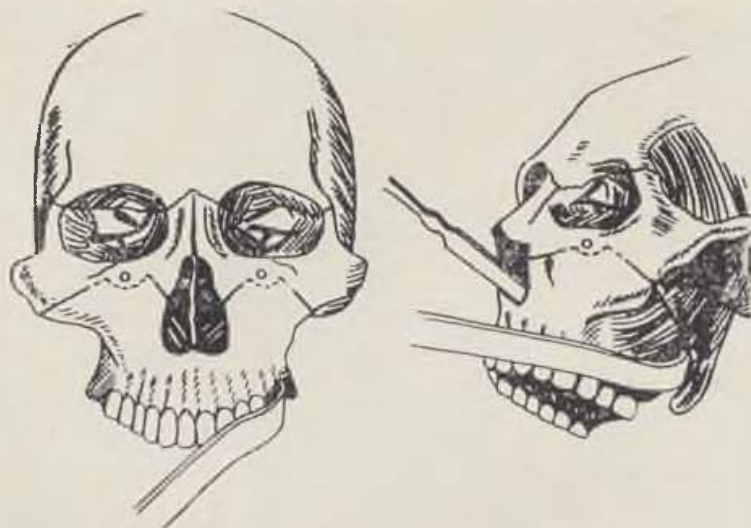


Fig. 1. The scheme of the technique of osteotomy of the maxilla. The line of the incomplete osteotomy is dashed.

As the first step, the corset is made and the supporting bar fixed to it. When the corset is completely dry (within a day) the arch with the distractors is installed at the free end of the supporting bar. The cast-metal cap is applied to the maxillary teeth and its hooks are connected with the distractors. The required direction of the pulling force is reached by changing the position of the arch on the supporting bar and of the distractors on the arch. The maxilla is moved downward and forward, i. e., in the direction of its normal growth. Intensity of the force pulling the cap is controlled and indicated at the distractor axis (Figures 2 and 3).

The present paper is based on the treatment of 42 patients (23 males, 19 females) suffering from deformities of the bones of the central third of the face of various aetiology. The details (the age of the patients and types of deformities) are given in the table. We evaluated the laboratory data, photographs, teleroentgenograms, electroodontometric data and the functional ability of the teeth.

In micrognathia and retrognathism the upper jaw bone was moved into the normal position in one stage by the method described. The surgery was completed by mandibular osteotomy (performed after a week by a classic tech-

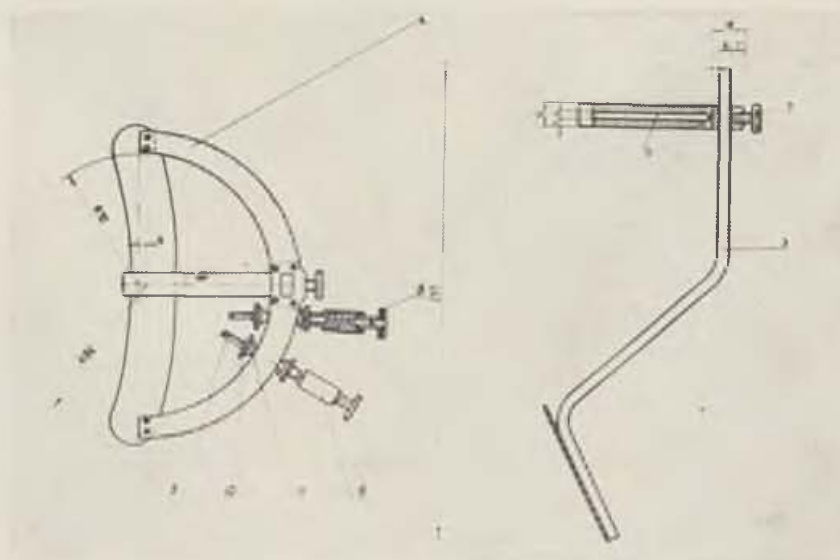


Fig. 2. The scheme of the distraction apparatus. The support is provided by a thoracocervical corset. — Fig. 3. The scheme of the distraction apparatus.



nique) in combinations of micrognathia or retrognathism of the maxilla with mandibular prognathism (Figures 4—8).



Fig. 4. Patient C. in profile. Occlusal relations before and after the treatment.



Fig. 5. Patient B., front face before and after the surgery.

Displacement of the maxillary complex was executed within the period from 22 to 60 days by the force varying from 1.5 to 5.0 kgm. The force used and duration of the displacement varied according to the age of the patient and severity of the deformity. Greater force for a longer period of time had to be used in patients with the cleft palate owing to extensive scars resulting from previous surgical interventions.

To summarize, osteotomy of the maxilla is performed at the frontomaxillar, zygomaticomaxillar and pterygoideomaxillar buttresses, osteotomy of the nasal septum at its bottom. Lateral walls of the nasal cavity are left intact. In dependence on the patient's age, the radical or incomplete osteotomy of the anterior wall of the maxilla is chosen.



Fig. 6. Patient B. in profile. Occlusal relations before, during and after the treatment.



Fig. 7. Patient E. in profile. Occlusal relations before, during and after the treatment.

Connection of the maxilla with the skull skeleton is secured by maintained continuity at the medial and posterior aspects of the maxilla, as well as by means of nourishing mucoperiosteal bridges in the regions of the canine and the first praemolar. This results in a more adequate blood supply to the moved maxillary segment and in a better postoperative regeneration.

The above described considerate method of osteotomy decreases considerably the duration and traumatism of the surgical intervention and of negative effects of other factors as well. It does not produce wide diastases between the bones, dispenses with a metal sutures and bone grafts, and the inflammation and lymphostasis are less pronounced.

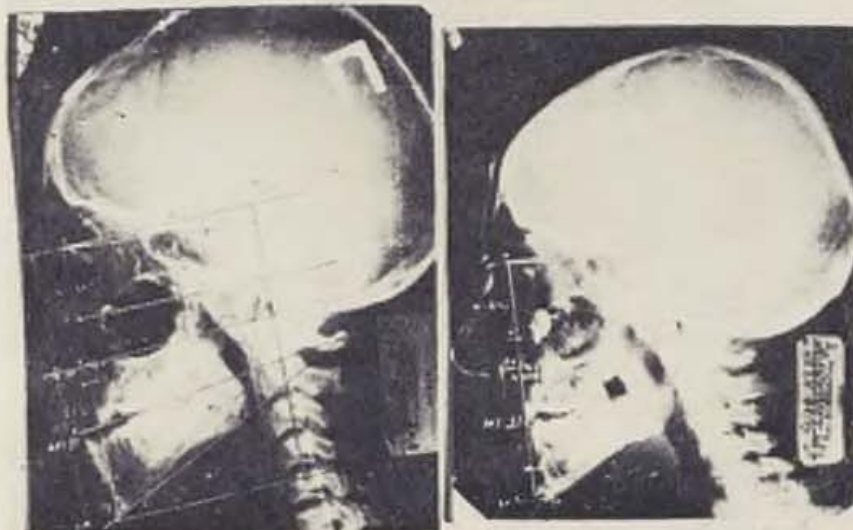


Fig. 8. Patient E. Tele X-ray before and after the treatment.

It is generally known that under a gradual distraction the new bone is formed by the direct angiogenic endosteo-periosteal ossification. Simultaneously, the bone architecture is rebuilt, so that a full bodies callus is formed and remissions prevented.

The method of selective osteotomy of the bones of the middle third of the face followed by gradual distraction of the maxilla is used for surgery

Table I. Number and types of deformities of the central third of the face

Age in years	Deformity			
	upper jaw		upper + lower	total
	micrognathia	retrognathism	jaw	
From 16 to 20	7	8	4	19
21 to 25	8	2	5	15
26 to 30 and more	1	5	2	8
Total	16	15	11	42



of micrognathia and retrognathism of the upper jaw. It allows the gradual displacement of the maxillary complex into normal anatomical position, decreases traumatism of the operation and secures permanent therapeutical effects. Satisfactory functional and cosmetic results in 24 patients followed for the period of 4 months up to 3 years demonstrate good perspective of this method.

M. D.

#### SUMMARY

The technique of selective osteotomy of the bones of the central third of the face followed by the gradual distraction is proposed for surgical treatment of the patients with retrognathism and micrognathia of the upper jaw. The technique permits the gradual displacement of the maxilla into the anatomically normal position, it is less traumatic and secures permanent therapeutical effects. Our clinical results, followed for the period of 4 months up to 3 years, demonstrate good perspective of this method.

#### RESUME

**L'ostéotomie sélective avec la distraction dosée au traitement des déformations du squelette de la moyenne partie de la face**

Tajrov, U. T.

C'est l'ostéotomie sélective qui est proposée pour le traitement opératoire de la micrognathie et de la rétrognathie du maxillaire supérieur. Il s'agit de l'ostéotomie sélective des os de la moyenne partie du visage qui est suivie par la distraction dosée du maxillaire supérieur.

Cette méthode permet la transposition successive du maxillaire supérieur dans la position juste du point de vue d'anatomie, elle est moins traumatique et — en plus — elle garantit la stabilité d'effet obtenu par le traitement. Les résultats cliniques aussi que les observations prolongées (4 mois jusqu'à 3 ans) prouvent une bonne perspective de cette méthode.

#### ZUSAMMENFASSUNG

**Selektive Osteotomie mit dosierter Distraction bei der Behandlung von Deformierungen des Skeletts des Mittelteils des Gesichts**

Tajrov, U. T.

Für die operative Behandlung der Mikrognathie und Retrognathie des Oberkiefers wird selektive Osteotomie der Knochen des Mittelteils des Gesichts vorgeschlagen unter darauffolgender dosierter Distraction des Oberkiefers. Diese Methode, die eine allmähliche Umstellung des Oberkiefers in die anatomisch korrekte Position gestattet, ist weniger traumatisch und gewährleistet die Beständigkeit des erzielten Behandlungseffekts. Die klinischen Ergebnisse sowie die langfristige Beobachtung der Patienten (von 4 Monaten bis zu 3 Jahren) beweisen die aussichtsreiche Perspektive dieser Methode.

#### RESUMEN

**Osteotomia selectiva con distracción dosificada en el tratamiento de las deformaciones del esqueleto de la parte central de la cara**

Tajrov, U. T.

Para el tratamiento operativo de la micrognatia y retrognatia del maxilar superior se plantea la osteotomia selectiva de los huesos de la parte central de la cara seguida

CONTRIBUTION TO THE CRYOTHERAPY  
OF MALIGNANT MELANOMA

J. Měšťák, J. Faltýn, J. Stříteský

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Fig. 6 — Histological sample of malignant melanoma after cryofixation

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CHONDROECTODERMAL DYSPLASIA (ELLIS-van CREVELD  
SYNDROME) REPORT OF A CASE

I. Horák, Z. Šmahel



Fig. 2. Left forearm at the age of 15 years. Shortening and hypoplasia of forearm bones, curved radius with luxation of its markedly hypoplastic head, dislocation of ulnar head, larger olecranon, missing apophysis of lateral humeral epicondyle





Fig. 3. Right elbow joint at the age of 15 years. Deformed and dislocated radial head, humeral apophysis visualized on medial epicondyle only.

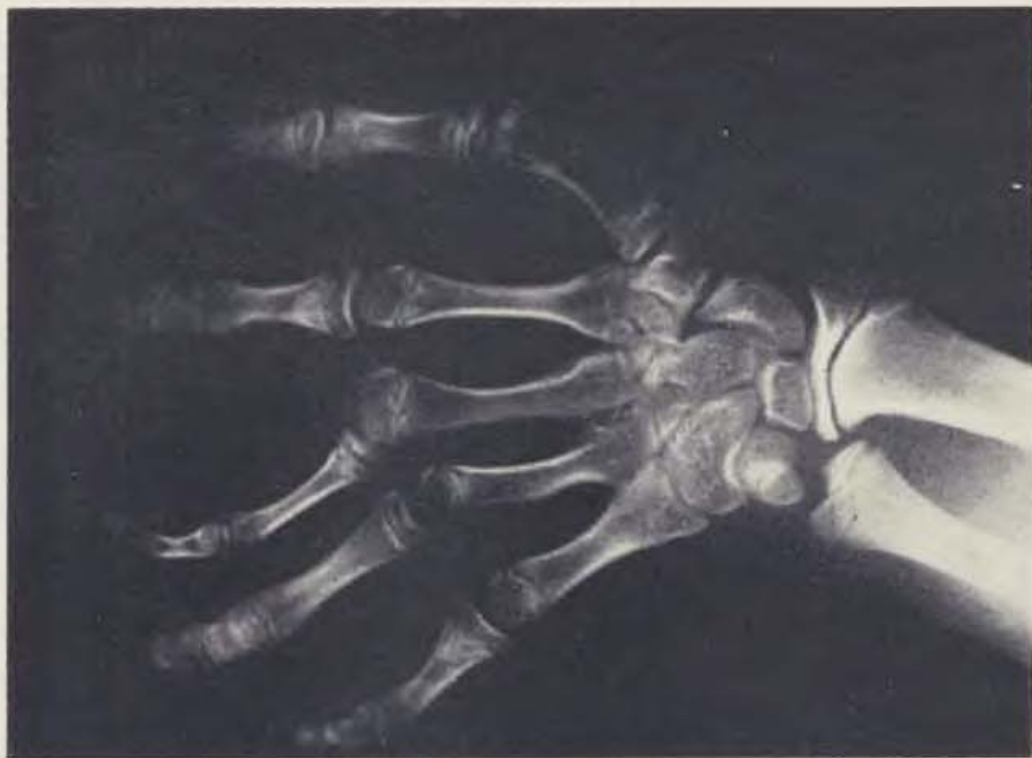
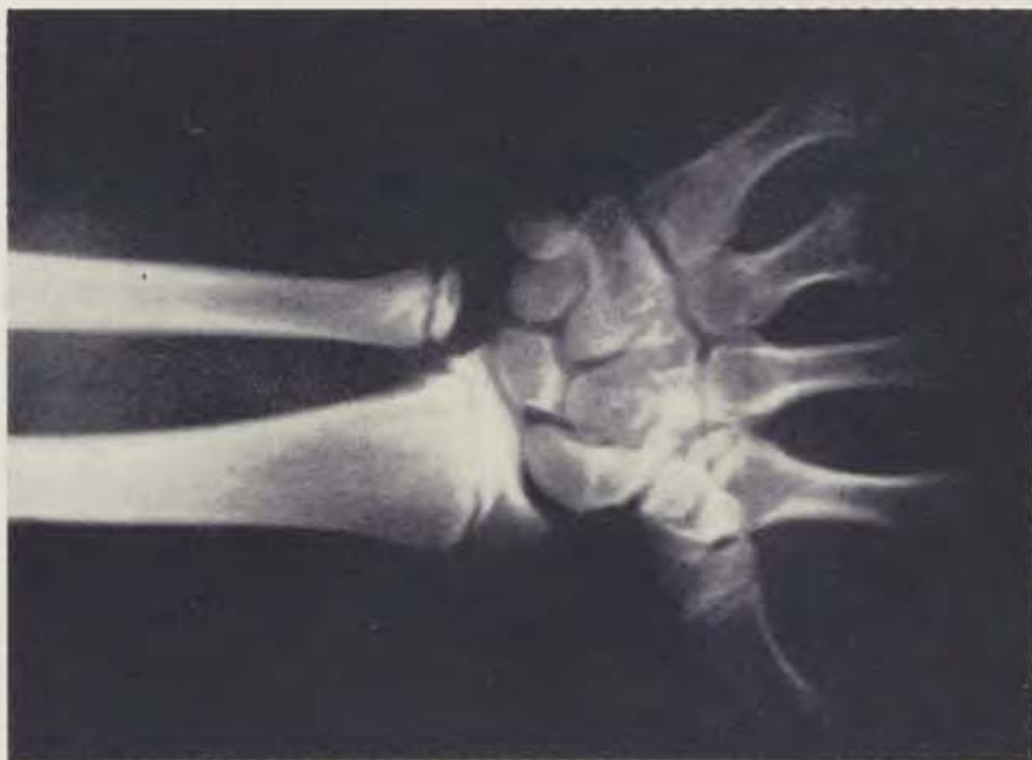


Fig. 4. Right wrist at the age of 15 years. Irregular epiphyseal growth zones, hypoplastic ulnar head, very narrow radial epiphysis on ulnar side, enlarged os hamatum. — Fig. 5. Left hand at the age of 15 years. Irregular contour of radiocarpal joint, on ulnar side a narrow radial epiphysis, deformed os hamatum, cone-shaped epiphyses of most proximal phalanges and metacarpal bones, shortening of middle and terminal phalanges, subluxation of 3rd finger towards the ulnar side, bifurcation of 4th metacarpal bone was resected, epiphyses on some middle and terminal phalanges are missing.

por la distracción dosificada del maxilar superior. Este método, que posibilita trasladar paulatinamente el maxilar superior en la posición anatómicamente adecuada, es menos traumático garantizando la estabilidad del efecto logrado por el tratamiento. Los habidos resultados clínicos así como las revisiones a largo plazo de los pacientes (de 4 meses a 3 años) prueban lo perspectivo de dicho método.

#### REFERENCES

1. **Bezrukov, V. M., Ospanova, B. B., Rudko, V. V., Stepanova, I. G.:** Clinical Picture, Diagnosis and Treatment of Complicated Deformities of the Jaws. *Stomatologia*, 1 : 47, 1977.
2. **Ilizarov, G. A.:** Clinical and Theoretical Aspects of the Compensatory and Distractive Osteosynthesis. In: *Theoretical and Practical Aspects of Intraosseous Distractive Osteosynthesis*, Moscow, 1977, p. 14.
3. **Chalamkaraov, Ch. A.:** Morphological Rebuilding of the Maxillofacial System during Widening of the Maxilla in the Period of the Decidual Dentition. *Stomatologia*, 5 : 73, 1967.
4. **Lavritsheva, G. I., Schit, V. P.:** Special Features of Reparative Processes during Distractive Osteosynthesis. In: *Proceedings of the Federal Traumatological and Orthopedical Congress*, Moscow, 1976, p. 170.
5. **Bell, W. H.:** Correction of the Short-Face Syndrome — Vertical Maxillary Deficiency; A Preliminary Report. *J. oral Surg.*, 35, 2 : 110, 1977.
6. **Dellinger, E. I.:** A Preliminary Study of Anterior Maxillary Displacement. *Amer. J. Orthodont.*, 63, 5 : 509, 1973.
7. **Epker, B. N., Fish, H. C.:** Surgical Orthodontic Correction of Open-Bite Deformity. *Amer. J. Orthodont.*, 71 : 278, 1977.
8. **Jackson, G. W., Kokich, V. G., Shapiro, P. A.:** Experimental and Postexperimental Response to Anteriorly Directed Extraoral Force in Young Macaca Nemestrina. *Amer. J. Orthodont.*, 75, 3 : 318, 1979.
9. **Kambara, T.:** Dentofacial Changes Produced by Extraoral Forward Force in the Macaca Irus. *Amer. J. Orthodont.*, 71, 3 : 249, 1977.
10. **Tessier, P.:** Total Osteotomy of the Middle Third of the Face for Faciostenosis and Sequaelae of Le Fort — III Fractures. *Plastic reconstr. Surg.*, 48, 6 : 533, 1971.
11. **Westwood, R. M., Tilson, H.:** Complications Associated with Maxillary Osteotomies. *J. oral. Surg.*, 33, 2 : 104, 1975.

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#### Correction

K. Malinský is the correct version of the name of the co-author of the report "Scope for Ultrasound Diagnosis of the Depth of Thermal Damage" which appeared in *Acta Chir. plast.*, 24, 4 : 235, 1982.



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## SPECIFICITIES OF SKIN PLASTIC OPERATIONS IN THE PERIANAL, PERINEAL AND SURROUNDING REGIONS

T. TEMELKOV, K. TROSHEV

It would seem at first that plastic operations in the perianal, perineal and surrounding regions ought to be no different from any other type of plastic operation so long as the principles of physiological surgery are observed. In fact, there are considerable differences there as regards the local anatomical specificities, indications for surgery and post-operative care. This may perhaps account for the lack of popularity enjoyed by skin plastic operations in that particular region. Examining literature from the fields of plastic surgery, proctology and general surgery available to us for the past 15 years we failed to find many publications about or references to this particular problem (1—1). That was why we decided to summarize and review it.

### Functional Topographic Anatomy.

The perianal, perineal and surrounding regions are all marked by characteristic variability of shape in different positions of the body. In the gynaecological position, the perineum exhibits the shape of an irregular rectangle with an uneven relief — oval in shape and of different diameter depending on whether it is viewed from the side or from above. In the female, the relief is complicated by the anal opening and by the genitalia. The males have a crest-like elevation on the raphe perinei.

In an erect body standing at attention the perineum with its tissues takes a rectangular form extending along an antero-posterior axis and a crest-like elevation running along the raphe perinei.

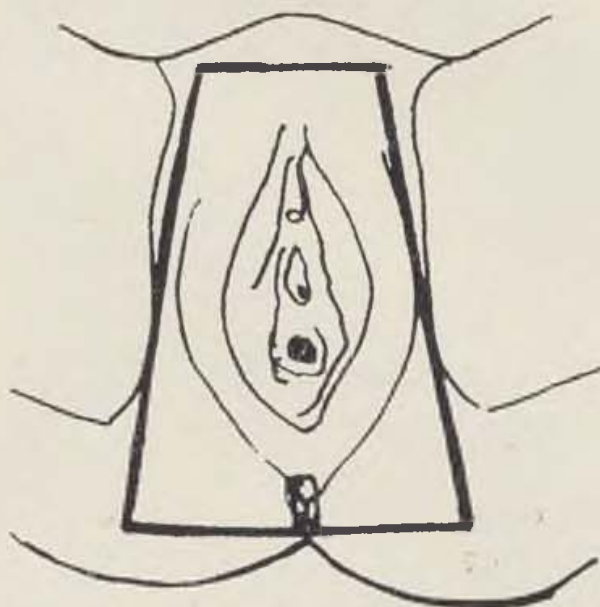
The perineogluteal region with the adjacent portions of the thighs and the sacral region have an oval, elliptical or circular shape (Scheme 2) depending on the pelvic dimensions and on the volume of soft tissues. A line drawn horizontally across the femorogluteal furrow divides the whole region into two parts with the characteristic oval of the lateral and superior profiles.

In this area, at the site of the sacrococcygeal intergluteal furrow we can find the superimposed shape of a triangle with its base on the posterior perineal wall and its apex in proximal position. The symmetrical bend in the median line forms a duplication, the depth of which is individual depending

on the mass of the gluteal muscle and on the degree of approximation of the two walls.

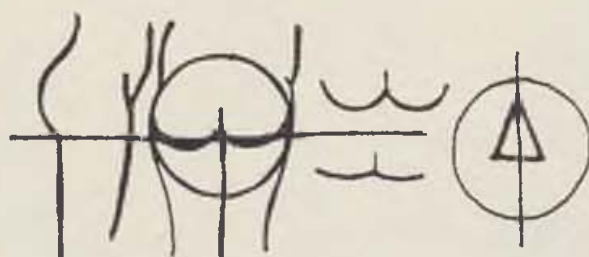
The rest of the details of each of the regions are individual.

Also to be taken into account are the dynamic functional changes in the shape of the whole of the region concerned with the above described furrow permitting maximum use of skin and tissue elasticity in cases of distension or compression.



Scheme 1 — Irregular rectangle of the female perineum in gynaecological position

The mutual contact of the skin surface in connection with the movements, friction, hair and sweating in the region contribute to the development of various skin diseases and other complications [1].



Scheme 2 — Shapes of the gluteo-perineo-femoral region in standing position

The above described specificities are made even more complex in connection with the specific function of the anal muscle — i. e. constriction and release of the ring-like sphincter — related to continence which is sometimes assisted by gluteal muscle contraction.

The above listed facts pertaining to the functional topographic anatomy of the region with its characteristic features account for the strict requirements involved in the planning and execution of plastic-surgery reconstruction of the region.



Fig. 1. — Radical haemorrhoidectomy according to Whithead-Rand with perforated skin flap from the neighbourhood

#### Pre-operative Preparation

This is highly important in view of periodic defecation. Added to this, there is the specific architecture and relief of the region stimulating the accumulation of physiological secretions of the skin and the preservation of bacterial infection [1, 2]. The most important part is the pre-operative preparation of the intestinal tract involving mechanical cleaning and bacterial elimination. Pre-operative treatment should start 4 to 5 days before the planned operation with the identification of the bacterial flora on the thighs, in the gluteal, perianal and perineal regions, and from rectal swabs [3].

At our unit, we prefer the per os administration of antibiotics which are not normally resorbed from the intestinal tract, or sulphonamides in combination with laxatives and a diet containing a minimum of bulk. During the last two pre-operative days we have the patient clysterized twice daily while reducing per os diet.

In patients with protective colostomy, the distal intestinal segment is irrigated with antiseptic solutions.

The skin within the area of the operating field receives two days of pre-operative treatment. Packs with spirit and 3% Rivanol solution, lately also with Hibitane (ICI-UK) solution, are applied to prevent skin irritation or damage resulting from the use of common disinfectants [1].



## Indications for Skin Plastic Operations in the Perianal, Perineal and Surrounding Regions

These can be divided as follows:

1. Skin tissue defects. These are the result of injury involving loss of tissue, burns, ulcerative and destructive processes (decubiti, haemangiomas) or operations requiring tissue and skin removal;



Fig. 2. — Post-partum recto-vaginal fistula — perineum and rectovaginal septum missing

2. Congenital anomalies of the anal, perianal, perineal regions, pilar cysts, spina bifida, etc;
3. Chronic nonspecific inflammatory processes — perianal fistulae, hydroadenitis chronica suppurative Verneuil;
4. Disfiguring scars resulting from injury, burns, operations;
5. Tumours and conditions resulting from tumour treatment.

### Contraindications

The following are contraindications of skin plastic operations in the regions concerned:

1. The presence of active pathogenic bacterial flora.
2. Intestinal or gastric involvement with diarrhoea.
3. Inflammatory process in the operating field zone or in its vicinity. A recent history of inflammation in the same region.
4. Inflammatory involvement of the genitalia.

With the indications and contraindications as well as the results of pre-operative treatment accurately assessed, we can plan the plastic skin oper-

ation proper taking advantage of tissue replacement opportunities in the perianal, perineal and surrounding regions. Should these prove to be inadequate we have to make a decision between a tube flap or a free skin graft taken from a remote area. The surgical technique must be in keeping with the requirements of physiological surgery. Of particular importance is the use of fine instruments, precision and sparingness at work.



Fig. 3—4 — Skin plastic reconstruction of the perineum following radical surgery for carcinoma of the rectum — neovagina, neorectum, neoperineum

Our firm performed the following operations meeting all the above listed conditions:

1. A skin and mucosa plastic operation necessitated by radical haemorrhoidectomy using the Withead-Rand method with a perporated skin flap taken from the neighbourhood — a total of 80 operations.



Fig. 5—6—7 — Extirpation of pilonidal polycystosis — gluteal muscle and skin plastic reconstruction with adjacent tissues shifted in



2. A perianal skin plastic operation in cases of anal sphincter and perianal skin deformation using tissues from the neighbourhood — a total of 9 operations.
3. Perineal skin plastic operations in cases of recto-vaginal traumatic and post-partum deformities (primary and secondary) — a total of 38 operations.



Fig. 8 — Operation according to Scott for perianal and perineal pruritis — strip of full-thickness skin excised according to scheme involving exchange of opposite triangles on the perineum

4. Skin plastic reconstruction following primary extirpation of the rectum in cases of carcinoma — a total of 5 operations.
5. Skin plastic operations in pilondial cysts (using adjacent tissues, pedicled perforated flaps, etc.) — a total of 23 operations.
6. Skin plastic operations for hydroadenitis perianalis chronica suppurativa Verneuil — a total of 6 operations.
7. Skin plastic operations for congenital malformations (hypoplasia of the whole perineal region, spina bifida, haemangioma, etc.) — a total of 15 operations.
8. Skin plastic operations following burns — a total of 5 operations.

A total of 181 operations were performed with their extent, nature, peculiarities and results representing the essence of the problem of skin plastic reconstructions in the perianal, perineal and neighbouring regions.

#### Post-operative Care

The nature of post-operative care differs according to the size (extent) of the operation, the existence of colostomy, the need for post-operative drainage or antibiotic irrigation of the tissues, and according to the size of the operating field.

The most important precautions in patients without colostomy include artificially induced obstipation, diet or just parenteral nutrition.

Adequate compression of the tissues used for the reconstruction is another important condition. For satisfactory drainage we lead the drains as far away from the anal opening as possible, trying to remove them at the earliest opportunity. Drainage involves the use of systematic periodic irrigation with antiseptic or antibiotic solutions depending on the sensitivity of the microbial flora present.

J. H.

#### SUMMARY

The authors analyze and generalize on their experience of the special requirements of skin plastic operations in the perianal, perineal and neighbouring tissues as these have so far received very little literary attention. They report on the functional topographic anatomy of the shapes involved, on pre-operative treatment and indications for skin plastic operation in those particular regions, on the surgical techniques and procedures, on wound treatment and subsequent post-operative care. The authors' theoretical and practical conclusions are based on their experience of 181 operations performed in the regions concerned, which they claim justifies a distinction to be made between that particular reconstructive surgery and skin plastic operations in other parts of the body.

#### RESUME

##### **Les spécialités des opérations cutanées plastiques dans la région périanale, périnéale et dans les régions voisines**

Temelkov, T., Trošev, K.

Les auteurs analysent et généralisent leurs expériences avec les spécialités des opérations cutanées plastiques dans la région périanale, périnéale et dans les régions voisines qui sont rarement mentionnées dans la littérature.

L'article présente anatomie fonctionnelle et topographique des formes, préparation avant l'opération, indications des plasties cutanées de ces régions, leurs contre-indications, description de la technique opératoire, soins immédiats et ultérieurs post-opératoires.

Dans leurs conclusions théoriques et pratiques, les auteurs profitent des expériences obtenues au cours de 181 opérations de ces régions ce qui donne le droit de différencier celles-ci des plasties cutanées sur les autres parties du corps.

#### ZUSAMMENFASSUNG

##### **Besonderheiten plastischer Hautoperationen im perianalen, perinealen und umliegenden Gebiet**

Temelkov, T., Trošev, K.

Die Autoren analysieren und verallgemeinern ihre Erfahrungen mit Besonderheiten plastischer Hautoperationen im perianalen, perinealen und umliegenden Gebiet, wovon es bisher in der Literatur sehr wenig Berichte gibt.

Inhalt der Publikation ist die funktionstopographische Anatomie der Formen, die Vorbereitung der Operation, die Indikation plastischer Hautoperationen in diesem Gebiet, ihre Kontraindikationen, die Operationstechnik und der Inhalt der Operationen sowie die unmittelbare und spätere Fürsorge nach der Operation. Die Autoren stützen

sich bei ihren theoretischen und praktischen Schlussfolgerungen auf ihre Erfahrungen, die sie bei 181 ausgeführten Operationen in diesem Gebiet gewonnen haben und die sie dazu berechtigen, solche Operationen von plastischen Hautoperationen an anderen Körperteilen zu unterscheiden.

## RESUMEN

### **Peculiaridades de las operaciones dermoplásticas en las zonas perianal, perineal y aledañas**

Temelkov, T., Troshev, K.

Los autores analizan y generalizan sus experiencias respecto de las operaciones dermoplásticas en las zonas perianal, perineal y las vecinas, tema sobre el cual hasta ahora poco se ha informado.

La publicación se refiere a la anatomía topográfica funcional de las formas, la preparación preoperativa y la indicación de las operaciones dermoplásticas de estas zonas, su contraindicación, la técnica y el contenido de las operaciones, el tratamiento postoperativo inmediato y posterior.

Al hacer sus conclusiones teóricas y prácticas los autores se basan en sus experiencias adquiridas en 181 operaciones aplicadas en dichas zonas, cuya realización los autoriza a hacer diferencia entre estas operaciones y las dermoplásticas efectuadas sobre las demás partes del cuerpo.

## REFERENCES

1. **Vozhkov, M., Vozhkova, K., Yarmova, B., Runkov, R.:** Microflora in Surgical Affections, TNTM Conference, Varna — 1982.
  2. **Dukhtsev, Y. B., Kugayevski, Y. B., Martinova, T. I.:** Pre-operative and Post-operative Treatment of Patients with Anal Sphincter Insufficiency. NII Proceedings, 1978, 32.
  3. **Dukhtsev, Y. B., Korneva, T. K. et al.:** Suppurative Microflora in Patients with Anal Sphincter Insufficiency. Pirogov's Surgery 4; 66, 1982.
  4. **Troshev, K.:** Method for Skin Plastic Operation in Spina Bifida (Author's own observation 28089/March 14, 1980).
  5. **Troshev, K.:** Problems of Skin Plastic Surgery in Haemangiomas. Med. Archives — Sofia, XIX, 3; 65—70, 1981.
  6. **Hartz, R. S., Shields, T. W.:** Healing of the Perineal Wound. Arch. Surg., 115, 4: 471, 1980.
  7. **Hurwitz, D. J.:** Closure of a Large Defect of the Pelvic Cavity by an Extended Compound Myocutaneous Flap Based on the Inferior or Gluteal Artery. Brit. J. plast. Surg., 33: 256, 1980.
  8. **Mc Graw, J. B., Dibbell, D. G., Carraway, J. H.:** Clinical Definition of Independent Myocutaneous Vascular Territories. Plast. reconstr. Surg., 80, 3; 342, 1977.
  9. **Marasco, L., Maviglio, P., Donno, G.:** Sinus (cisti) pilonidalis: trattamento mediante plastica a Z multiple. Riv. ital. Chir. Plastica, 10, 1: 63, 1978.
  10. **Souza, L. J., Shinde, S. R.:** Primary Perineal Closure after Proctectomy in the Female. J. roy. Coll. Surg. Edinb., 25: 129, 1980.
  11. **Woods, J. E., Irons, G. B., Masson, J. K.:** Use of Muscular, Musculocutaneous and Omental Flaps to Reconstruct Difficult Defects. Plast. reconstr. Surg., 69, 2: 191, 1977.
- More literature available at the authors.

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## CONTRIBUTION TO THE CRYOTHERAPY OF MALIGNANT MELANOMA

J. MĚSTÁK, J. FALTÝN, J. ŠTRÍTESKÝ

Local cryodestruction of tissue has been already used as a routine procedure in neurosurgery, proctology, gynaecology, stomatology etc. Local freezing of tissue (up to the temperature of  $-196^{\circ}\text{C}$ ) followed by gradual separation of produced cryonecroses becomes important also in treatment of benign and malignant dermatic tumours.

Recently, cryodestruction has been used even for treatment of malignant melanomas not only for lentigo malignant melanoma, where the cryotreatment is prevalently indicated, but also for nodular malignant melanoma. But prior to the freezing itself, a diagnostic excision of tumour is made for its histological confirmation and the treatment is usually very protractive with regard to long separation of necroses or to necessary repetition of the procedure in cases in which the malignant melanoma has not been totally removed by the cryodestruction. For this reason, we do not regard cryodestruction as a satisfactory treatment especially of malignant melanomas in case of the nodular type in spite of its stimulating effect on immunity response in patients with malignant melanoma. We prefer an extensive excision followed by conservative treatment in indicated cases.

Mortality rate from malignant melanoma still remains considerably high, in spite of unceasing endeavours of surgeons, immunologists, chemotherapeutists and other specialists in bordering fields. It is an alarming fact, that in patients with malignant melanoma anamnesticly in state of "clinical latency" even during long periods this tumour expands quickly after operation in the form of local and even distant skin metastases, the regional lymphatic nodules are attacked, the disease is eventually getting generalised and patients die. A possible reason may be e. g. defective immunity mechanisms of organism which, according to current knowledge, tend to a higher probability of disease dissemination after surgical intervention then in comparison with the immuno-



logically nondefective organisms. Nevertheless, it is possible that a higher migration of tumour cells from the primary tumour is caused by surgical intervention. Surgeons themselves do not deny possible migration of cells during the operation. They recommend not only to begin an extended excision of malignant melanoma from the point situated most closely to the lymphatic drainage in order to block primarily lymphatic paths leading towards the regional lymphatic nodes, but some of them even choose the method of delayed defect transplantation in order to liquidate the tumour definitely by wide drainage.

A method preventing the above mentioned migration of tumour cells during the operation is cryofixation of the primary tumour prior to its radical removal. This method aims at a fixation of tumour cells by freezing apparatus in order to prevent a migration of these cells during the operation. With respect to this, an experimental microsurgical proof of a cryosurgical occlusion of mesenterial arterial, venal and lymphatical vessels during the resection of large intestinal carcinoma preventing peroperational expansion of tumour cells during the manipulation with tumour accompanying its radical removal is of importance.



Fig. 1 — Malignant melanoma before surgery

We applied cryofixation of primary tumour in three patients with clinically evident form of malignant melanoma and with regional lymphonodes proved by examination not be hypertrophic. In two cases the tumour was localised

in the region of right humerus and in one case of frontal side of the body (Fig. 1). For freezing of malignant melanoma, we used a cryosurgical system KCH 3A with freezing capacity 60 W developed in State Research Institute of Electrical Engineering Works, Prague-Běchovice (Fig. 2). We used the contact



Fig. 2 — Cryosurgical system KCH — 3 A

freezing method, producing the freezing effect immediately after fine contact and switching-on (Fig. 3). Frozen zone spreads homogenously. The result is a regional hemostasis preventing the expansion of tumour cells into healthy tissue during the intervention. In this case we used a cryosurgical system with 2 cm dia cap and after 2 minutes exposition we froze a dermatic zone of 37 cm dia and 8 mm deep (Fig. 4). Around the frozen zone, we excised extensively the whole block of tissues including underlying subcutaneous tissue and muscular fascia and covered the resulting defect with dermoepidermal graft fixed with sutures knotted over a compressive bolus. Taking into consideration the immunosuppressive effect of general anaesthetics, we try not to prolong the surgery without reason and with respect to that we must stress that cryofixation does not effect duration of the operation. The transplanted skin always took small cases (Fig. 5).

The frozen substances were put into Dewar's flask with liquid nitrogen and stored there for several days. Under room temperature but without "melting" of the substance, we took and prepared small tissue blocks in order to cut 10 microne cryostate slides under temperature of  $-30^{\circ}\text{C}$  for HE stained biopsies and for monophenol-monooxygenase reaction. A malignant melanoma growing into stratum reticulare has been histologically proved in all patients

concerned, in two cases with traces of intradermal expansion (Fig. 6). Therefore in cryofixation the diagnostic excision of tumour prior to its radical removal is not necessary while it is imperative in the case of cryodestruction.

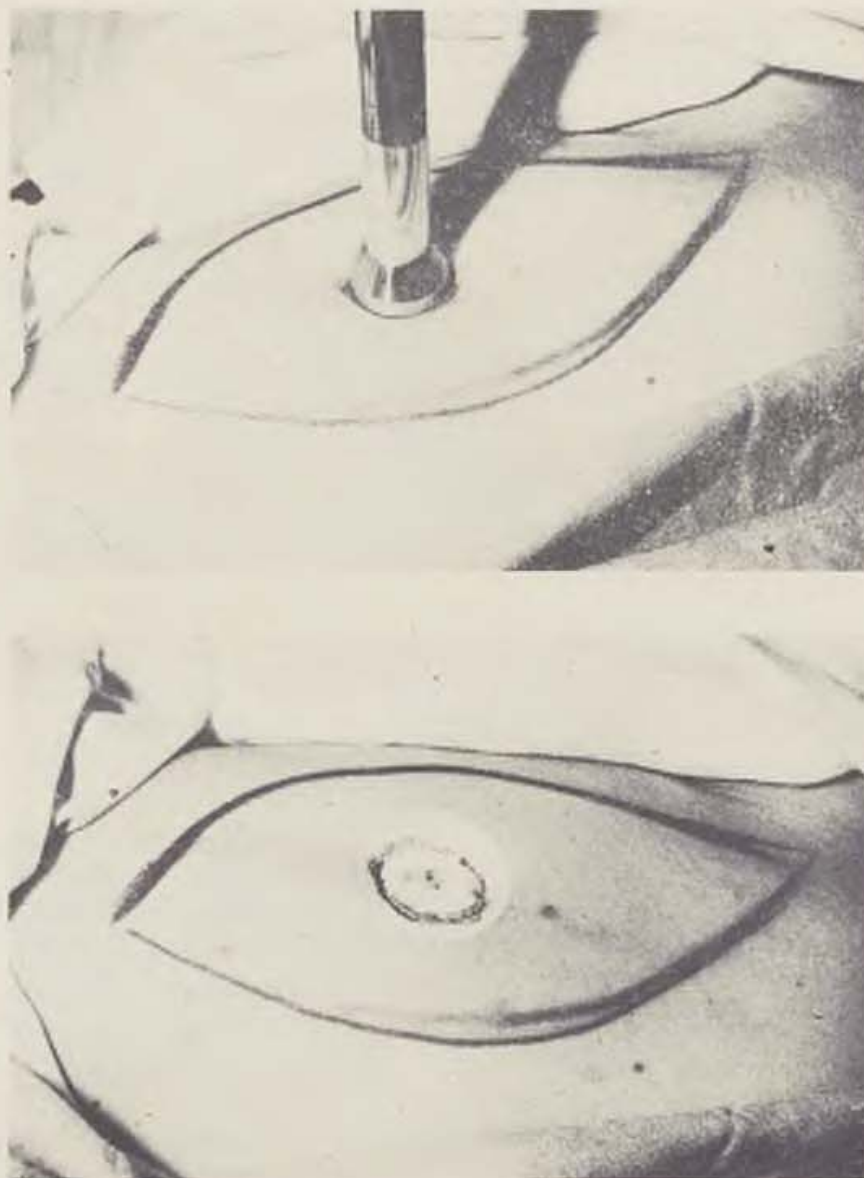


Fig. 3 — Application of cryoprobe to the tumour — Fig. 4 — Frozen zone — details

In order to get a documentary evidence concerning the importance of cryofixation for malignant melanoma treatment, it is necessary to follow the patients for a long period. It may be presumed that opinions concerning the practice of this method will naturally vary in the same way as those concerning the radical treatment, especially the extent and depth of excision or the immediate and delayed transplantation of the defect after the excision. This problem does not concern the surgical intervention only, but it applies also to the conservative treatment of malignant melanoma. We presume that cryofixation will prove to be a satisfying reliable procedure in the prevention of possible per-

operational dissemination of tumour cells. The method would find its place as a complementary method in the complex treatment of malignant metastatic damage to regional lymphnodes in spite of high infiltration of the tumour into the dermis.

The research continues with long-term follow up the patients, who have been treated as described, further we are going on with experiments in animals.



Fig. 5 — Healed in graft

#### SUMMARY

Surgical treatment represents an important component of malignant melanoma complex medical treatment. The authors devoted this study to the problem of local and distant metastases following surgery and to the possibility of their prevention. Besides the well defined radical surgical procedures the authors stress the value of tumour cell cryofixation carried out immediately before surgery. Cryofixation represents a method, that may prevent peroperative dissemination of tumour cells.

#### RESUME

##### **La contribution à la cryothérapie du mélanome malin**

Měšťák, J., Faltýn, J., Strítěský, J.

Le traitement chirurgical représente une partie importante, mais pas seule, du traitement complexe du mélanome malin. Dans leur travail, les auteurs s'occupent du



problème des métastases locales et des métastases éloignées pendant la période post-opératoire, ils cherchent des possibilités de la prévention. A côté de la méthode détaillée qui consiste dans l'intervention radicale, les auteurs soulignent l'importance de la cryofixation des cellules tumorales avant de prélever le mélanome malin. Cette méthode représente une des possibilités d'arrêter l'expansion tumorale de la place d'excision, pendant l'opération.

#### ZUSAMMENFASSUNG

##### Beitrag zur Kryotherapie eines malignen Melanoms

Měšťák, J., Faltýn, J., Stříteský, J.

Die chirurgische Behandlung ist ein wichtiger, wenn auch nicht der einzige, Bestandteil der komplexen Behandlungsmethode eines malignen Melanoms. Die Autoren befassen sich in ihrer Arbeit mit dem Problem der lokalen und entfernten Metastasen im postoperativen Verlauf sowie mit den Möglichkeiten ihrer Vorbeugung. Ausser der bereits erprobten Methode einer radikalen chirurgischen Intervention betonen sie die Bedeutung der Kryofixierung der Tumorzellen vor der eigentlichen Beseitigung eines malignen Melanoms als eine der Möglichkeiten einer Begrenzung der weiteren Ausbreitung des Tumors während der Operation von der Stelle der Exzision aus.

#### RESUMEN

##### Contribución a la crioterapia de un melanoma maligno

Měšťák, J., Faltýn, J., Stříteský, J.

El tratamiento es parte importante, aunque no única, del complejo sistema de tratamiento del melanoma maligno. En su trabajo, los autores describen el problema de las metástasis locales y distantes en el período postoperativo y la posibilidad de su prevención. Aparte del conocido método de una intervención quirúrgica radical subrayan la importancia de la criofijación de las células tumorosas antes de proceder a eliminar el melanoma maligno, como una de las posibilidades de como impedir la propagación del tumor, durante la operación, desde el lugar de la excisión.

#### REFERENCES

1. Ansell, B. H.: Use of Liquid Nitrogen for Treatment of Lentigo Maligna as Effective. *Dermatology News*, 9, 1, 1976.
2. Faltýn, J.: Pokroky v kryochirurgické léčbě ve FN Praha 10. Sborník „Kryochirurgie jako aplikace nízkých teplot“, Benešov, 1981.
3. Graham, G. F., Stewart, R.: Cryosurgery for Unusual Cutaneous Neoplasms. *J. Dermatol. Surg. Oncol.*, 3:4, 1977.
4. Grana, L., Airan, M., Gordon, M., Johnson, R.: Cryogenic Technique for Resection of Carcinoms of the Colon. *Int. J. Surg.*, 63, 5:53, 1978.
5. Kolektiv autorů: Maligní melanom, Avicenum, Novinky v medicíně, 1980.
6. Lojda, Z., Gossrau, R., Schiebler, T. H.: Enzym-histochemische Methoden. Springer-Verlag, Berlin, Heidelberg, New York, 1976.
7. Love, D.: Cryotherapy Role in Benign Pigmented Lesions. *Dermatology News*, 13, 8, 1980.
8. Málek, Z., Zobač, L., Soukup, F., Krýsl, I., Hora, O., Jelínek, J., Ryska, A., Šafrata, S.: Autonomní kryochirurgický systém s chladicím výkonem 60 W při  $-195^{\circ}\text{C}$ . *Čs. čas. pro fyziku (sekce A)*, 30:485, 1980.
9. Matthaues, W., Scholz, A., Kremlička, L.: Výsledky kryoterapie nádorů v oblasti hlavy. *Čs. Othol.*, 36, 4, 1980.

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## CHONDROECTODERMAL DYSPLASIA (ELLIS – van CREVELD SYNDROME) REPORT OF A CASE

I. HORÁK, Z. ŠMAHEL

Ellis and van Creveld described in 1940 under the name of chondroectodermal dysplasia a new syndrome. This disorder belongs into the group of chondrodysplasias with polydactylia and autosomal recessive mode of inheritance. Main features consist of chondrodystrophia (disproportional dwarfism with shortening of extremities), postaxial polydactylia and ectodermal dysplasia, mostly associated with incomplete median cleft upper lip (or some other lip stigmata), a narrow chest and approximately in one half of affected individuals with congenital heart disease. From the total number of about 150 cases reported in the literature 52 individuals were detected by Mc Kusick et al. (1964) in an inbred population of an Old Order Amish in Pennsylvania, in which subsequently additional cases were disclosed and da Silva et al. (1980) found fifteen affected individuals in a pedigree of a Brazilian family.

The observations reported so far provided a list of changes and individual deviations characterizing this syndrome. Small body height was due exclusively to the shortening of extremities, which was mostly symmetric and progressive distalwards (mesomelic and acromelic brachymelia). The forearms and legs were shorter than the arms and thighs, contrary to classic achondroplasia. There was mostly a more marked shortening of the ulna as compared to the radius and of the fibula as compared to the tibia. This could lead to a proximal dislocation of the radial head, cubitus valgus, or to limitation of the movement range of the elbow. The proximal part of the ulna and the distal part of the radius were usually larger, while on the contrary the opposite ends of both bones were smaller as compared to their normal size. A characteristic feature represented the configuration of the proximal tibial metaphysis which was enlarged with medial displacement of the epiphysis due to the steeper slope of the epiphysial surface of the metaphysis on its medial side which was shorter than on the lateral side. This resulted in a frequent development of genu

valgum. The femora and humeri were curved, there was an occurrence of coxa valga, flattened acetabular roof, acetabular hypoplasia and exostoses on pelvic bones and on the tibiae. The hands showed a definite trend towards synmetacarpia of supernumerary digits and fused carpal bones (mostly os hamatum and os capitatum). The shortening affected to a larger degree medial and distal phalanges as compared to proximal and metacarpal bones. Because of these changes the fingers could not be fully clenched into a fist. There was a frequent occurrence of ossification disorders and changes in phalangeal configuration (e. g. clinodactylia), with occasional reports on pseudoepiphyses and supernumerary carpal bones. Similar anomalies occurred on the lower extremities as well.

Polydactylia is present always on upper extremities, bilaterally and the supernumerary finger on the ulnar side of the hand is usually well developed (type A). Occasionally the numbers of fingers are even larger with a possible syndactylia. Supernumerary metacarpal bones can develop separately, show varying degrees of fusion or are completely missing. On the feet polydactylia occurred only in 27 per cent of individuals involved (Kunze 1980), with a possible syndactylia, in particular of the 2nd and 3rd toes.

A third constant feature represents ectodermal dysplasia reported by Kunze (1980) in 93 per cent of reported cases. The nails are hypoplastic, dystrophic, friable and sometimes even missing (especially on supernumerary fingers). Deciduous and permanent teeth are microdontic with tremata and varying numbers of teeth can be missing. Enamel hypoplasia is common and can result in atypical shape of the teeth. There is a frequent malocclusion and premature eruption, sometimes even in the prenatal period. The hair is occasionally fine, while body hair, the skin and sweating are normal.

A regular feature is a malformation of the upper lip. It consists of a medial colobom within the vermillion, or leads only to a shorter lip attached to the gingiva of the alveolar process by multiple frenula or a hypertrophic frenulum obliterating the labiogingival sulcus. Cranial X-ray films fail to reveal any anomalies (contrary to achondroplasia).

Another characteristic feature is a long frequently malformed chest (pectus carinatum). The ribs are short, hypoplastic. An exception represents an associated anomaly of the lungs and of the bronchial tree. X-ray films of the vertebral column fail to disclose any typical changes.

Cardiac anomalies occur in as many as 60 per cent (Kunze 1980) and consist of a defect of the atrial or ventricular septum. Other described anomalies include cor triloculare or biloculare, transpositions of large vessels, coarctation of the aorta, valvular defects, patent ductus arteriosus etc.

About 22 per cent of involved individuals (Kunze 1980) have genital anomalies (hypospadias, epispadias, hypoplastic penis, cryptorchism and vulvar atresia). Occasionally the malformations include cleft palate, talipes, arthrogryposis, hepatosplenomegaly a. o. The intellect is normal, the sex ratio 1:1. The prognosis quad vitam depends on the presence and severity of congenital cardiac disease and on the deformation of the chest. Approximately in one



half of affected individuals death during early infancy is due to cardiac and respiratory insufficiency, while the other involved individuals have a favourable prognosis.

#### CASE REPORT

A boy M. B., born on May 1, 1966, was treated at the Department for Plastic Surgery, Charles University, Prague, since 1967. The diagnosis of the Ellis-van Creveld syndrome was established.

**Family history:** The boy was a desired child by his normal unrelated parents. At the time of delivery his mother was aged 25 years and his father 29 years. As far as could be detected the boy was the only member of the kinship with an inborn anomaly. The sister of his mother and the sister of his father are both normal and the latter has normal children. During her first pregnancy the mother of the boy had some bleeding in the second month of pregnancy which subsequently continued without further complications up to term. No infections, or exposure to drugs or radiation were reported. The delivery at term was spontaneous with parietal presentation. The child weighed 3,350 g and measured 49 cm. He started to cry immediately, no resuscitation was required. His psychomotoric development was slightly retarded during early infancy. After the age of one year he was able to walk with support, his first tooth erupted at the age of five months and the obliteration of the major fontanella was recorded at 18 months of life. At the age of nine months the boy developed a severe bilateral pneumonia associated with seizures and followed by bilateral otitis media. Within his 21st month of life he suffered again from severe bronchopneumonia. Subsequently no severe infection developed. During his hospital treatment numerous studies were performed inclusive of the following results obtained: karyotype 46 XY; a normal ECG pattern, as well as ocular and auditive findings, while neurologic studies disclosed a marked hypotony and increased excursion of joints due to chondrodysplasia; a negative complement fixation response to toxoplasma antigens both in the child and his mother; laboratory tests were within the range of normal; chest X-ray disclosed an atelectasis of the right upper lung lobe (at the age of nine months). The boy was characterized as a small lively baby. Blood groups: in the boy B, Rh<sup>+</sup>, N, in his father O, Rh<sup>+</sup>, MN, in his mother B, Rh<sup>+</sup>, MN.

In 1977 his mother had a second pregnancy and on January 31, 1978 was delivered of a normal boy (3,400 g, 49 cm). Up to the present time his development is satisfactory. At the age of 3 years and 9 months his basic body parameters corresponded exactly to the norm of the Czech population (body height 102 cm, body weight 16 kg, sitting height 59.5 cm etc.).

**Global findings:** During a check-up examination at the age of sixteen years the patient had a short stature (160.5 cm, i. e. -1.6 SD) with appropriate body weight (51 kg, -1.2 SD). His physical condition was relatively good (Fig. 1). The smaller body height was due exclusively to the shortening of lower extremities, his sitting height corresponded exactly to the norm (89 cm, 0.0 SD). The arm span (141.5 cm) was by 19 cm smaller than his body height



and reflected the shortening of upper extremities. He had a conspicuously narrow chest. If not mentioned otherwise all deviations described were determined during the check-up at the age of sixteen years, but for the X-ray films made at the age of fifteen years. Anthropometric data of body height and weight were compared with the norms obtained during a nation-wide study



Fig. 1. Boy aged 16 years with a narrow chest and crosswise asymmetric shortening of extremities, especially of the left forearm.

[Prokopec et al. 1973], sitting height and dimensions of upper extremities according to Škvařilová (1975), those of the lower extremities according to Krejčovský (Thesis) and trunk characteristics according to norms reported by Bláha et al. (1982).

Chondrodysplasia was associated with a disproportioned shortening of extremities: the upper extremities by 4.3 SD on the right and by 5.9 SD on the left (difference 5 cm), the lower extremities by 3.1 SD on the right and by 2.7 SD on the left (diff. = 2 cm). Thus the pelvis was slightly inclined towards the right side (Fig. 1). The shortening was mesomelic and acromelic

[the arm bilaterally by 2.0 SD, the forearm by 4.8 SD on the right and by 7.9 SD on the left, the hand by 5.4 and 6.4 SD resp., the thigh by 2.9 and 2.5 resp., the leg by 3.5 and 3.2 SD resp., and the foot by 2.2 bilaterally]. The values showed a crosswise asymmetry. A difference between both sides occurred in both segments of the lower extremities, while in the upper extremities it was limited entirely to the forearms. The ulna was shortened more markedly (to 78 % on the right and 64 % on the left of its normal length) than the radius (82 % on the right and 78 % on the left; according to Maresh's norm, from Lusted and Keats, 1978). Thus the radius was curved on both sides and on the left his head showed a marked luxation proximalwards (Fig. 2). A slight proximal dislocation was recorded equally on the right (Fig. 3) with a similar dislocation of ulnar head in the left wrist (Fig. 2). Thus the shortening of the forearm (to 75 % on the right and 60 % on the left) was due mainly to the shortening of the ulna, while that of the leg was caused, without any doubt by the shortened tibia. Shortening of individual phalanges of the hand are presented on Tab. 1. In metacarpal bones it amounted on the average to -4.2 SD, in proximal phalanges to -4.1 SD, in medial phalanges to -8.5 SD and in distal phalanges to -6.5 SD. The slightest shortening of fingers was present in thumbs and the most marked shortening in the three middle fingers (Tab. 1). For these differences were responsible middle and basal phalanges. Terminal phalanges were shortened uniformly (but for the thumb).

X-ray studies revealed global hypoplasia of the forearm bones, with particularly marked hypoplasia of the ulnar head on the right (Fig. 4) and of the radial head on the left (Fig. 2). The head of the right radius was malformed as well (Fig. 3). The humeri were curved and hypoplastic and the apophyses of the lateral epicondyls were missing (Fig. 2, 3). The proximal epiphysis of the left ulna, inclusive of the proc. coronoideus was larger (Fig. 2), while on the right no conspicuous deviations were evident, similarly as in the terminal distal parts of the radii. Chondrodystrophic changes were observed equally on the epiphyses and within their growth zones. Distal epiphyseal spaces of both radii and ulnae were irregular with invaginations into the metaphyses (Fig. 4). On the ulnar side the radial epiphysis was very narrow and the cartilage showed a zone of provisional calcifications (Fig. 4, 5). No normal concave contour of the radiocarpal joint was recorded (Fig. 4, 5). Epiphyses of most proximal phalanges and metacarpal bones are cone-shaped with invaginations into metaphyses (Fig. 5). The epiphyses on middle and terminal phalanges frequently were ill defined. Their absence was confirmed in most cases by X-ray films of the left hand at the age of 8.5 years with clear evidence of a pseudoepiphysis of the metacarpus and of the proximal thumb phalanx. Neither metacarpal width (Tab. 1) nor those of phalanges were reduced, but for some exceptions (the 3rd finger and the 2nd and 3rd metacarpal bones on the left). The carpal angle was obtuse (161° on the right and 150° on the left; the norm = 134°).

With the exception of larger and wider great toes, digital hypoplasia was present on the feet as well, with bilateral supernumerary os tibiale externum

[1X1.5 cm). Bilaterally shortened were predominantly the 2nd and 3rd metatarsal bones with malformations in basal regions. No pathologic changes were demonstrated within the region of knee joints and thus the position of both lower extremities was parallel. The feet were flat and slightly valgus. During hospital treatment at the age of nine months X-ray studies disclosed foci of chondromatosis in iliac, ischiac and pubic bones, as well as in the distal metaphysis of the right fibula and left ulna. No further changes were described.



Fig. 6. Both hands prior to surgery (with simian lines).

Digital anomalies were associated with limited joint mobility. All DIP joints showed a marked limitation of motion, the 3rd PIP on the left was immobile. These changes represented a further reason why the patient could not clench a fist. Both elbow joints, on the contrary, were capable of hyperextension. No other obvious deviations of joint motion from normal were found.

**Polydactyly:** Our patient had postaxial polydactyly of type A with an involvement of all four extremities. Six digits were present originally on his right hand with slightly webbed interdigital spaces (Fig. 6). The extra little finger was smaller and thinner and, with a rudimentary metacarpus. The left hand had eight digits with doubled third, fourth and fifth fingers. The fingers were thinner and shorter than on the right and a syndactyly with fingers grown together mostly up to the nail basis was present (Fig. 6). The duplicated third finger had a single common metacarpus, the fourth metacarpus was partially divided (bifurcation), the pattern of the little finger was not documented (there was probably a single metacarpus). Carpal bones were separated without synostosis, the os hamatum was enlarged and deformed, the hamulus osis hamati was not visualized. Both feet had six toes, the duplicated little toes had a com-

mon metatarsal bone (Fig. 7). There was a slight syndactyly between the great and the second toes.

Fingers of the left hand were gradually separated and their number was reduced (the extra 3rd, 6th and 8th digits were removed) with the resection



Fig. 7. Both feet prior to surgery. Shortening of 2nd and 3rd toes, slight syndactyly between the great and the 2nd toes, nail dystrophy.

of the duplicated metacarpus. On the right hand the interdigital space between the thumb and the second finger was enlarged and the 6th digit resected. Extra little toes on the feet were removed as well. The width of the repaired hands was  $+1.4$  SD on the right and  $-1.3$  SD on the left; the corresponding figures on the feet were on both sides  $-0.5$  SD. There was an ulnar subluxation of the 3rd finger on the left (after the removal of its duplicate on the radial side of the metacarpus — Fig. 5).



Fig. 8. Teeth at the age of 15 years. Agenesis of lower lateral incisors, enamel hypoplasia deforming the teeth shape (e. g. conical premolars).



Ectodermal dysplasia: The teeth were irregularly spaced and the patient has been subjected to orthodontic treatment since his childhood (Fig. 8). Our patient had a marked hypoplasia and abrasion of enamel resulting in teeth deformation. Dental caries was recorded already during his hospital



Fig. 9. The boy at the age of 9 months. Narrow long trunk, deformed chest with lateral depressions and protruding sternum, medial coloboma of the upper lip.

treatment at the age of 21 months. Panoramic roentgenogram confirmed the agenesis of lower lateral incisors. The third molars consisted of a single unerupted tooth on the upper right side. The nails were dystrophic and especially on the feet fragile (they were present on all fingers and toes). The hair and the skin were normal.

Cleft upper lip: The cleft or a coloboma within the median plane of the vermillion (Fig. 9) proceeded across the lip upwards at first in the form of a deeper and then of a shallow groove up to the columella. A broad middle frenulum was inserted rather deeply between the great incisors; beyond the latter proceeded in a relatively broad alveolar process a groove which passed into a gothic palate. The uvula was uniform. On the left approximately 0.5 cm beyond the tip of the tongue a solid lingual cyst of the size of a pea was present. Extirpation of the cyst, lip suture and removal of the frenulum were carried out at the age of eighteen months.

**Chest deformations:** On presentation the boy had a marked chest deformation with bilateral lateral depressions and a keeled breast (pectus carinatum, Fig. 9). Thus the trunk was narrow and apparently long (chest circumference corresponding to the age of 21 months amounted to 40 cm, i. e.



Fig. 10. Boy aged 16 years. Scoliosis with rightward deflected curvature and pelvic slope, in association with a more marked shortening of right lower extremity.

—5.2 SD). X-ray study revealed at the age of 21 months irregular thin ribs with enlarged terminal parts, yielding the pattern of a rachitic rosary. Congenital dysplasia of some lung segments and the low level of the diaphragm were diagnosed as well. The chest deformation showed a substantial improvement after the age of eleven years, yet there was still a narrow pigeon breast (Fig. 1) with short hypoplastic ribs and enlarged osseous ends of the 2nd up to the 7th ribs. Chest circumference at the age of 16 years was smaller by —2.8 SD, its width by —4.4 SD and its depth by —1.7 SD. While the narrow trunk was characterized by a smaller width of the shoulders (by —2.5 SD) and abdominal circumference (by —1.5 SD), the pelvic width corresponded to the

Tab. i. Differences of metacarpal, phalangeal and finger lengths and of metacarpal widths in a patient, expressed in terms of standard deviation (X-ray measurements)

	Metacarpus <sup>1</sup>		Proximal <sup>1</sup> phalanx		Middle <sup>1</sup> phalanx		Distal <sup>1</sup> phalanx		Finger <sup>2</sup>		Metacarpal <sup>3</sup> width	
	dx	sin	dx	sin	dx	sin	dx	sin	dx	sin	dx	sin
I.	-2.9	-3.3	-2.7	-2.7	-	-	-7.2	-3.6	-4.8	-2.7	-1.0	-1.1
II.	-5.0	-4.8	-5.4	-5.8	-8.2	-9.5	-6.2	-6.2	-6.0	-6.7	-2.3	-4.0
III.	-4.0	-3.8	-3.8	-5.8	-10.8	-8.8	-6.4	-6.4	-7.0	-7.3	-1.9	-3.5
IV.	-6.2	-3.5	-3.3	-4.1	-9.8	-7.8	-6.9	-7.6	-6.4	-7.3	+0.7	-1.7
V.	-4.6	-4.0	-2.8	-4.4	-6.4	-6.4	-6.9	-7.8	-4.6	-6.2	-1.3	-1.6
$\bar{x}$	-4.2		-4.1		-8.5		-6.5		-		-	

<sup>1</sup>norms according to Garn et al. (Lusted L. B. and Keats T. E., 1978: Atlas of Roentgenographic Measurement)

<sup>2</sup>norms according to Blåha for males aged 19 (Doklād M., ed., 1976: Human Growth and Physical Development)

<sup>3</sup>norms according to Parish for adults (Lusted L. B. and Keats T. E., 1978: Atlas of Roentgenographic Measurement, Year Book Medical Publishers, 4th ed., Chicago)

note: <sup>2,3</sup> do not change substantially after the age of 15

norm ( $-0.3$  SD). The spinal column was scoliotic, the curvature was deflected rightwards (Fig. 10) and thus compensated the more marked shortening of the right lower extremity with hemivertebra L1 and L2. The anteroposterior curvature of the spinal column was less pronounced and the thoracic kyphosis was missing.

**Other findings:** No congenital heart disease was demonstrated on ECG, the patient was free of any symptoms and therefore no further methods of cardiac examinations were used. During childhood he had phimosis and a small penis, but at the age of fifteen years the sexuologic examination yielded normal results. No other anomalies were disclosed. The dimensions of the neurocranium and of the face were within the range of  $-0.5$  to  $-1.5$  SD (but for a few exceptions). The interocular distance was unchanged, the intercanthal index was 35.6. He had an average intelligence. Longitudinal studies of his growth and dermatoglyphic patterns will be described in another paper (Šmahel et al. in preparation).

#### DISCUSSION

The diagnosis of the Ellis-van Creveld syndrome should be based on the presence of three cardinal features consisting of evidence of chondrodystrophia and ectodermal dysplasia, as well as postaxial polydactylia. These anomalies are regularly associated with upper lip malformation, while a further major feature, i. e. a narrow and deformed chest is related to chondrodystrophia. The presence of a congenital heart disease is of auxiliary importance only. Our patient had all five basic features of this syndrome, only the congenital heart disease, which is not regularly present, was missing.

X-ray studies play a major role in the diagnosis of this syndrome. They disclose not only changes in the shape of individual structures, but also chondrodystrophic changes within the growth zones of bones which, conceivably, are not specific and are less conspicuous or even missing in adults. This could be the reason why no obvious deviations from normal are visualized on X-ray films of knee joints in our patient (since the growth zones are no more clearly defined). Conceivably the changes are not necessarily evident in all regions. The variability of expression of the involvement of other systems are documented in patients with teeth anomalies, yet without any nail dystrophy (da Silva et al. 1980). On the contrary in other individuals an involvement of teeth is missing (Goor et al. 1965). Postaxial polydactylia is not necessarily present since da Silva et al. (1980) also described in one patient from a pedigree of fifteen affected individuals a combination of postaxial and preaxial polydactylia, and Dayer (by Ellis and Andrew 1962) mentioned one patient with an extra digit on the radial side.

Our patient represented an example of asymmetric involvement of extremities, with a particularly marked difference in the length of his upper extremities. The shorter extremity had on presentation eight fingers and did not represent an exclusively postaxial type of polydactylia (there was also a duplication of the 3rd and 4th fingers). Further there were all degrees of duplications of metacarpal bones, i. e. supernumerary, bifurcated and common



metacarpal bone for two fingers. In our patient the lower extremities were less shortened than the upper extremities. This situation occurs probably more frequently since the length of forearms is determined predominantly by the ulna, which uses to be more markedly shortened than radius, while the length of the leg is determined by the tibia which uses to be less involved than the fibula. This is in agreement with the report by Ellis and Andrew (1962) who described the shortening of long bones in one of their patients as follows: humerus to 67 per cent, radius to 59 per cent, ulna to 48 per cent, femur to 77 per cent, tibia to 61 per cent and fibula to 50 per cent of their normal length. The values reported by Ferrero et al. (1961) in another patient, however, are suggestive of individual differences (humerus 82 %, radius 66 %, ulna 68 %, femur 75 % and fibula 57.5 %, the length of the tibia was not stated).

Laboratory and chromosomal studies fail to reveal any deviations from normal.

The differential diagnosis requires the elimination of other syndromes within the group of chondrodystrophies with polydactylia, inclusive of the Majewski syndrome (with epiglottic anomalies), the Saldino-Noonan syndrome (fetal hydrops without nail dystrophy and cleft lip) and Jeune syndrome (equally without nail dystrophy and cleft lip and often without polydactylia) which has all of them a lethal outcome due to congenital heart disease and in the latter also due to nephritis. It is further necessary to exclude the Mohr syndrome or OFD II which, however, does not belong among chondrodystrophies and is not associated with shortened ribs, X-ray changes of long bones and nail anomalies. The cartilage-hair hypoplasia with fine hair and without an involvement of the ribs and of the pelvis should be taken into consideration as well. Postaxial polydactyly occurs also in some other syndromes as e. g. Meckel, Smith-Lemli-Opitz, Laurence-Biedl-Bardet-Moon a. o.

#### SUMMARY

Ellis-van Creveld syndrome is reported in a boy followed-up throughout a long period of time. He had all five main features of the syndrome, i. e. chondrodystrophia, ectodermal dysplasia, postaxial polydactylia, incomplete median cleft upper lip and a narrow deformed chest; the inconstantly occurring congenital heart disease was not demonstrated. Chondrodystrophia was manifested by small body height due to shortening of extremities of mesomelic and acromelic type. The shortening was crosswise asymmetric. Bone changes on extremities were documented by X-ray studies and changes within growth zones were underlined as well. Malformations of the chest were caused, equally, by chondrodystrophy, however during childhood congenital dysgenesis of some lung segments was also diagnosed. The extent of all deviations from normal was expressed in metrical terms. Polydactylia was present on all four extremities, which had six fingers or toes, but for the left hand where eight fingers grown together (syndactylia) were present. The interdigital spaces were slightly webbed also on all other extremities. The nails were dystrophic, the teeth irregular with enamel hypoplasia and oligodontia. The upper lip with

an incomplete cleft in the midline was attached by a broad frenulum and there was a solid lingual cyst. The family history provided no significant information. Laboratory and chromosome studies yielded normal results. Longitudinal follow-up of his growth and dermatoglyphic patterns were described in a separate paper.

**A c k n o w l e d g e m e n t :**

We wish to express our thanks to Prof. M. Fára, M. D., DrSc., for his kind permission to follow-up his patient and to M. Brejcha, M. D., CSc., for his valuable aid in the interpretation of the X-ray films.

**R E S U M E**

**La dysplasie chondroectodermique (syndrome Ellis - van Creveld) d'un garçon**

Horák, I., Šmahel, Z.

On a décrit un cas du syndrome Ellis - van Creveld d'un garçon soigné longtemps. Tous les cinq signes principaux se sont manifestés, c'est-à-dire manifestation de la chondrodystrophie, dysplasie chondroectodermique, polydactylie postaxiale, division incomplète médiale du lèvres supérieur, thorax étroit déformé, seulement le défaut cardial inconstant n'était pas prouvé. La chondrodystrophie se manifeste par une taille diminuée, conséquence de l'abrégement des membres, type brachymélie mésomélique et acromélique. L'abrégement est asymétrique d'une manière croisée. Des changements sur les os des membres sont documentés par radiographie. Les auteurs constatent aussi des déviations dans les zones de croissance des os. La déformation du thorax est également conséquence de la chondrodystrophie. En enfance on a diagnostiqué la dysgenèse congénitale de quelques segments du poumon. La grandeur de toutes les déviations est exprimée métriquement. La polydactylie atteint tous les quatre membres qui avaient six doigts à l'exception de la main gauche à huit doigts joints par les syndactylies (l'élévation interdigitale se trouve aussi sur les autres membres). Les ongles sont dystrophiques, la denture irrégulière avec l'hypoplasie d'émail et avec l'oligodontie. Le lèvres supérieur avec la division incomplète médiale est joint par une large bride, sur la langue une kyste dure. L'anamnèse familiale n'est pas remarquable, les examens laboratoires et chromosomiques sans les signes pathologiques. Les résultats d'observation longitudinale et d'observation dermatoglyphique seront traités dans un autre travail.

**Z U S A M M E N F A S S U N G**

**Chondroektodermale Dyslasie (Ellis - van Creveld Syndrom)  
bei einem Knaben**

Horák, I., Šmahel, Z.

Es wird ein Fall des Ellis- van Creveld Syndrom bei einem langfristig beobachteten Knaben beschreiben. Vorhanden waren alle fünf Hauptsymptome des Syndroms, d. h. Äusserungen von Chondrodystrophie, Ektodermaldysplasie, postaxialer Polydactylie, eine unvollständige mediale Spaltung der Oberlippe und ein schmaler deformierter Brustkorb, nur ein nichtkonstanter Herzfehler wurde nicht nachgewiesen. Die Äusserung der Chondrodystrophie bestand in geringem Körpergewicht infolge Gliederverkürzung vom Typ einer mesomelischen und akromelischen Brachymelie. Diese Verkürzung war kreuzweise asymmetrisch. Die Veränderungen an den Knochen der Gliedmassen wurden roentgenographisch dokumentiert und auch auf die Abweichungen in den Wachstumszonen der Knochen hingewiesen. Auch die Deformierung des Brustkorbs ist eine Folge der Chondrodystrophie; in der Kindheit wurde noch eine ange-

borene Dysgenese einiger Lungensegmente festgestellt. Die Grösse aller Abweichungen wird metrisch ausgedrückt. Die Polydaktylie befällt alle vier Gliedmassen, die sechs Finger bzw. Zehen haben, mit Ausnahme der linken Hand mit acht Fingern und zusammengewachsenen Syndaktylien (Erhöhungen zwischen den Fingern bzw. Zehen befinden sich auch an den anderen Gliedmassen). Die Nägel sind dystrophisch, das Gebiss unregelmässig unter Hypoplasie des Zahnschmelzes und Oligodontie. Die Oberlippe mit unvollständiger Mittelspalte durch breiten Zaum zusammengefügt, auf der Zunge eine harte Zyste. Die Familienanamnese ist nicht bemerkenswert, die Laboratoriums- und chromosomalen Untersuchungen ohne pathologischen Befund. Die longitudinalen Beobachtungen des Wachstums und der dermatoglyphische Befund werden in einer anderen Studie veröffentlicht.

## RESUMEN

### Displasia condroectodermal (síndrome Ellis - van Creveld) en el muchacho

Horák, I., Šmahel, Z.

Se describe el caso del síndrome Ellis - van Creveld en un muchacho examinado durante largo tiempo. Se manifiestan las cinco características propias del síndrome, es decir condrodistrofia, displasia ectodermal, polidactilia postaxial, escisión medial incompleta del labio superior y un tórax estrecho y deformado, no se ha comprobado, sin embargo, la afección cardíaca inconstante.

La condrodistrofia se caracteriza por baja estatura producto del acortamiento de las extremidades tipo traquimelia acromélica y mesomélica. El acortamiento es asimétrico en cruz. Alteraciones en los huesos de las extremidades se documentan por radiografía observándose igualmente las anomalías en las zonas de crecimiento de los huesos. La deformación del tórax es asimismo producto de la condrodistrofia; en la edad infantil se había diagnosticado además la disgénesis congénita de algunos segmentos pulmonares. El tamaño de estas anomalías se da en escala métrica. La polidactilia afecta las cuatro extremidades, tres de las cuales tenían seis dedos y la mano izquierda ocho, pegados por sindactilias (se observan crecidas sindactilias también en otras extremidades). Las uñas son distróficas, la dentadura irregular con hipoplasia del esmalte, y oligodoncia. El labio superior presenta una fisura medial incompleta y está ligado mediante un ancho frenillo, en la lengua hay un quiste correoso. La anamnesis familiar no presenta nada especial, exámenes de laboratorio y cromosomales sin diagnóstico patológico. Los resultados de la observación longitudinal del crecimiento así como el diagnóstico dermatoglífico serán publicados en otro análisis.

## REFERENCES

1. Bláha, P., et al.: Antropometrie československé populace od 6 do 35 let. Praha, OZZ VS, 1982.
2. Ellis, R. W. B., Andrew, J. D.: Chondroectodermal Dysplasia. J. Bone Jt. Surg., 44B : 626, 1962.
3. Ferrero, N. A., Pozo, O. O., Morresi, E. S.: Chondro-Ectodermal Dysplasia (Ellis-van Creveld Syndrome). J. Bone Jt. Surg., 43A : 1230, 1961.
4. Goor, D., Rotem, Y., Friedman, A., Neufeld, H. N.: Ellis-van Creveld Syndrome in Identical Twins. Brit. Heart J., 27 : 797, 1965.
5. Krejčovský, L.: Thesis under preparation.
6. Kunze, P.: Ellis-van Creveld Syndrome. Kinderärztl. Praxis (Leipzig), 48 : 193, 1980.
7. McKusick, V. A., Egeland, J. A., Eldridge, R., Krusen, D. E.: Dwarfism in the Amish. I. The Ellis-van Creveld Syndrome. Bull. John Hopkins Hosp., 115 : 306, 1964.



8. Prokopec, M., Suchý, J., Titlbachová, S.: Age Children. Anthropologie, 13 : 101, 1975.  
Výsledky třetího celostátního výzkumu mládeže 1971. Čs. Pediat., 28 : 341, 1973.

9. Škvařilová, B.: The Growth Patterns Report of 15 Cases in an Inbred kindred. in the Upper Extremities of Prague School J. Med. Genet., 17 : 349, 1980.

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## NEWS

### Problems and Failures in the Theory, Experiment and Practice of Plastic Surgery. Symposium with International Participation. Brno, October 13—15, 1982.

Department of Plastic Surgery, J. E. Purkyně University, Brno (Czechoslovakia)  
Head Prof. V. Kubáček, M. D., DrSc.

The Symposium took place at the International hotel with more than 100 Czechoslovak and 15 foreign plastic surgeons from a number of European countries including one from the United States taking part.

The meeting went on under the auspices of leading officials of the region of South Moravia as a follow-up to the celebrations of the 65th birthday of Prof. V. Kubáček, M. D., DrSc., head of the Department and, at the same time, Rector of the J. E. Purkyně University. The gala opening took place in the late afternoon hours of October 13 at the Knights' Hall of the New Town Hall and was followed by a get-together cocktail party in the Cross Corridor.

The meeting proper proceeded in 5 sections with Czech, Russian and English as the working languages. A total of 32 papers were presented and 10 posters discussed. Each participant was given the main points of each report. The papers are scheduled to be published at the turn of 1983—1984 in English and in Russian with extensive summaries to appear in Scripta Medica, a journal published by the Brno Medical Faculty.

To introduce the Symposium proper in the morning of October 14, Prof. Kubáček gave a brief substantiation of the subject of the meeting. This was followed by a brief opening section on organizational problems facing plastic surgery. Evidence was presented (Michek, Brno) to show that in the next few years it would be imperative to expand substantially therapeutical facilities of plastic surgery in the South Moravian region to meet the population's requirements.

Another section then gave a summary of the latest knowledge concerning wound healing and the relevant disorders (Pospíšilová, Brno), the effects on healing of ultrasound (Dyson, London), and, eventually, problems of experimental blood supply to the skin using direct skin vessels and musculocutaneous vessels. Both systems of complementary, which is of great importance for practical work (Kartik, Gulyás, Budapest).



Next followed a larger group of papers discussing various general state disorders in severe burns affecting adults and children (staff members and co-workers of the Prague Department of Plastic Surgery Burns Unit, Sauer, Netherlands, Lukášová, Brno). Much attention was devoted to immunological problems, to transfusion and infusion therapy including transfusion of ramified amino acids. A great deal of discussion was devoted to problems of euthanasia in the very severely burned. There were also posters dealing mainly with problems of local treatment for burns.

The third, afternoon maxillofacial section discussed problems involved in primary reconstruction of deformities of the nose in clefts (Anderl, Innsbruck), complications in surgical operations for some of the rarer cases of clefts, and surgical reconstruction of the face in various facial syndromes (Kobus, Polanica, Zdrój), palliative surgery in such cases of severe facial deformities (Bilder, Brno), the use of pectoral flaps (Kozák, Prague), and corrections for the consequences of severed facial nerve. The problem here was how to suppress involuntary asymmetrical facial movements, a case finely illustrated by a documentary film (Clodius, Zurich).

In the evening, participants in the Symposium were taken for a trip to South Moravia to taste samples of wine and to be entertained at a dulcimer music party.

The morning of October 15, the last Symposium day, saw first a brief section devoted to the surgery of the hand and to traumatology. The introductory paper demonstrated the possible replacement of necrotic os lunatum with the wound up tendon of the m. palmaris longus (Rott, Bozděch, Brno). Then there was a report on a case of upper extremity replantation associated with a series of severe general complications (Kubačák, Vyškov, Černý, Brno). A number of posters were displayed (particularly by staff members of the Department of Plastic Surgery, Brno), dealing with early rehabilitation after deliveries of finger flexors, corection of ulnar deviation of fingers in rheumatics, microsurgery in severe hand injuries, treatment for primary severance of flexor muscles (Chváta, Brno) and other injuries.

The last section on different themes included first 3 papers on lymphoedema dealing with such problems as prospects for the reconstruction of impaired lymph drainage (Mandl, Vienna, Clodius, Zurich, Bardychev, Obninsk) or radical operations (Bardychev). These subjects elicited once again a lively discussion. Other problems dealt with included conservative treatment for urethral stenosis using new information about connective tissue (Hájek, Samohýl, Brno), disputes concerning superradical excision in melanoblastomas (Hasman, Plzeň), and the causes of complications and failures in the treatment of decubital ulcers (Riebelová, Brno). There were two papers on cosmetic surgery, one concerning the rate of psychological hazard, to which patients are exposed (Kipikaša, Košice), the other — repair surgery following inadequate and unsuccessful cosmetic operations on the breasts (Bažinka, Brno). The last paper examined the possible uses of laser in surgery (Hubáček, Olomouc).

The Symposium was wound up by two assessments, one presented by Prof. H. Pešková, M. D., DrSc., on behalf of the Czechoslovak participants, the other by Prof. L. Clodius on behalf of foreign guests. Both expressed their satisfaction recommending similar meetings to be organized repeatedly at certain intervals of time.

Before leaving for their homes, a number of the participants were able to inspect the newly reconstructed and expanded Brno Department of Plastic Surgery.

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# STOP FOR A MOMENT AND CONSIDER YOUR HEALTH



DAY AFTER DAY AND YEAR AFTER YEAR YOU ARE CONSTANTLY CHASING SOME AIM OR ANOTHER, YOU STRETCH THE MAINSPRING OF YOUR HEALTH TO THE VERY MAXIMUM. AND HOW LONG DO YOU THINK YOU CAN CONTINUE TO DO SO? REMEMBER THAT YOU HAVE ONLY ONE HEALTH AND FINALLY MAKE UP YOUR MIND TO GRANT IT, AT A VERY REASONABLE PRICE, WHAT IT DESERVES: COMPLEX TREATMENT AT ONE OF THE OLDEST AND THE MOST WIDELY RECOGNIZED SPAS IN EUROPE.

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