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THE HYPERTELORISM-HYPOSPADIAS (BBB) SYNDROME IN MEMBERS OF TWO FAMILIES

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The association of hypertelorism with hypospadias as a hereditary determined malformation was described for the first time by Opitz et al. (1965). Subsequently Opitz et al. (1969a) reported its occurrence in members of three families and Christian et al. (1969) described this syndrome in another family. Up to the present time less than thirty well documented cases were reported in the literature. The main anomalies described in males consisted of hypospadias and hypertelorism or telecanthus associated sometimes with mental retardation, cleft lip and palate, and with some less severe or uncommon anomalies. In females this malformation is manifested by hypertelorism or by telecanthus, which is mostly present in mothers of involved males. Since though far there is no definite evidence of male-to-male transmission of the anomaly a recessive X-linked hereditary mechanism is assumed. Yet it is not possible to exclude the role of an autosomal dominant inheritance with male sex limitation.

Simultaneously with this syndrome Opitz et al. (1969b) described an identical combination of malformations in four brothers accompanied by laryngo-tracheo-esophageal anomalies. Further studies showed that the extent of these anomalies could markedly differ. In some cases they could not be detected and were manifested only by dysphagia and respiratory distress (stridor) with hoarseness occuring only during childhood. In other cases they consisted of severe clefts incompatible with life. Again these anomalies were recorded mainly in males, while females had predominantly hypertelorism or dysphagia alone. Yet very severe forms of larygo-tracheo-esophageal anomalies might occur in females as well, as illustrated by two lethal cases (Kasner et al., 1974; Arya et al., 1980). At least three documented cases of male-to-male transmission of the anomaly (Funderburk and Stewart, 1978; Farndon and Donnai, 1983; Chemke et al., 1984) were suggestive of an autosomal dominant type of heredity with partial male sex limitation. These observations and other facts (see

below) showed that in practice both syndromes should be regarded as two distinct forms of a similar complex of malformations though a distinct expression of one and the same gen could not be excluded. The authors who described initially these syndromes used in the first case the designation BBB syndrome (the initial capital letters of the names of the first three described families) and in the second case the G syndrome (the initial capital letter of the name of the described family). More illustrative than a designation with an acronyme would be most certainly the use of the hypertelorism-hypospadias syndrome or the hypertelorism-hypospadias-dysphagia syndrome.

The present communication deals with the BBB syndrome in six individuals (three of them were females) in two unrelated families.

Case reports

The occurrence of individual anomalies in affected males is presented together with earlier findings in table 1. The results of measurements in the ocular region and the dermatoglyphic findings in individuals of both sexes

Tab. 1. Manifestations in previous and current cases

Clinical features	Previous cases	Present cases			Total	
		I	II	III	abs.	%
Hypertelorism/telecanthus	19/20	+	+	+	22/23	95.7
Hypospadias	19/21	+	+	+	22/24	91.7
Low total ridge count	6/10	+	—	—	7/13	53.8
Strabismus/exotropia	9/20	—	+	+	11/23	47.8
Mental retardation	11/21	—	—	—	11/24	45.8
High nasal bridge	11/21	—	—	—	11/24	45.8
Distal axial triradius	7/15	+	—	—	8/18	44.4
Widow's peak	10/20	—	—	—	10/23	43.5
Cranial asymmetry	9/21	—	+	—	10/24	41.7
Cryptorchidism	9/21	—	—	—	9/24	37.5
Ears low-set/retroinclined	8/21	+	—	—	9/24	37.5
Postcranial skeletal defects	6/17	+	—	—	7/20	35.0
Cleft lip and palate	7/21	+	—	—	8/24	33.3
Diastasis recti	5/19	—	—	—	5/22	22.7
Cardiovascular anomalies	5/20	—	—	—	5/23	21.7
Low posterior scalp line	3/17	+	—	—	4/20	20.0
Forehead prominence	2/17	+	—	—	3/20	15.0
Neck short/wide	2/17	+	—	—	3/20	15.0
Anus ectopic/imperforate	3/21	—	—	—	3/24	12.5
Epicanthic folds	3/21	—	—	—	3/24	12.5
Inguinal hernia	2/19	—	—	—	2/22	9.1
Prominent metopic suture	2/21	—	—	—	2/24	8.3

Case I = Family I — proband
Case II = Family II = proband
Case III = Family II — brother of the proband

are presented in Table 2 and 3. The diagnosis of telecanthus is based on measurements of the distance between the inner eye canthi and on the calculated values of three indexes, the diagnosis of hypertelorism on the interorbital distance measured on X-ray films and on the values of two indexes (Tab. 2).

Family No. I.

Proband (Fig. 1, 2): A boy, the second child of normal unrelated parents was born at term in vertex presentation after a pregnancy free of any complications. His mother was aged 23 years and his father 26 years. At birth the child was alert and weighed 4100 grams. He had bilateral complete cleft

Fig. 1 Family I, proband: Telecanthus and hypertelorism, bilateral cleft lip and palate repaired in childhood

Fig. 2 Family I, proband: Low-set retroinclined ear lobe, short nose, retrocheilia due to cleft

lip and palate and penile hypospadias without cryptorchism. The proband was subjected to several surgical interventions at our Department of Plastic Surgery, inclusive of a repair of the cleft lip and palate, a prolongation of the columella, repair of the shape of his nose, as well as of the lip with an enlargement of vestibular sulcus and straigtening of the hypospadic penis. According to the opinion of sexuologs a further repair of hypospadias is not required, since it does not interfere with sexual function. The boy showed no signs of mental retardation, he started to walk at the age of one year and co-operated well during subsequent check-up examinations.

Tab. 2. Ocular measurements in three male patients and three female cariers (with differences from the norm expressed in terms of SD)

	Intercanthal distance		IC	ZIC	CIC	Interorbital distance		Interorbital index		Zygo-interorbital index	
Family I											
Propositus	43	+4.1 SD	43.0	31.4	7.6	35	+3.2 SD	31.0	+2.8 SD	25.0	+3.8 SD
Mother	42	+4.9 SD	44.7	29.4	7.7	no X-ray films					
Sister	45	+6.2 SD	46.4	32.4	8.3	no X-ray films					
Family II											
Propositus	46	+5.1 SD	44.2	30.9	7.7	37	+4.0 SD	30.3	+2.5 SD	24.7	+3.6 SD
Brother	39	+2.5 SD	41.9	27.3	6.8	31	+1.6 SD	27.7	+1.4 SD	21.2	+1.6 SD
Mother	40	+4.0 SD	40.0	29.2	7.2	34	+3.0 SD	29.8	+2.3 SD	25.0	+3.8 SD

IC = index intercanthalis (euryopia above 38, hypertelorism above 42)
ZIC = index zygomaticointercanthalis (euryopia above 25.5*, hypertelorism above 29*)
CIC = index circumferenceintercanthalis (euryopia above 6.4*, hypertelorism above 7.2*)

*Šmahel (1978)

An examination at the age of nineteen years showed small stature (body height 166 cm, body weight 56 kg), well developed muscles and a good physical condition. His face was symmetric with frontal prominence, malar hypoplasia, a very short nose and hypertelorism according to all used criteria (Tab. 2). The ear lobes were situated at a low level, retroinclined and slightly deformed (both the tragus and the antitragus were missing). His neck showed a slight pterygia with a low hair line. A spina bifida of the cervical spine was present at the level of C_{4-5}. His trunk and extremities showed no signs of malformations. During the check-up examination at the age of fifteen years the neurologist observed that his trunk could not sustain some movements with a persisting instability of both upper extremities but a satisfactory taxis. There were slight signs of a divergence of the right eye bulbus. ECG showed a partial bundle branch block without any signs of an organic heart disease. The ENT findings were normal. Difficulties associated with food intake during his early childhood were due exclusively to the cleft lip and palate. Routine laboratory studies were within the range of the norm. He had a typical dermatoglyphic pattern with numerous arches, a low TRC (45), and a bilateral distal axial triradius.

The mother of the proband (Fig. 3): The signs of the syndrome consisted only of telecanthus (Tab. 2) and of hypertelorism on palpation. But for a prominent forehead, there were no signs of anomalies on her face, ear

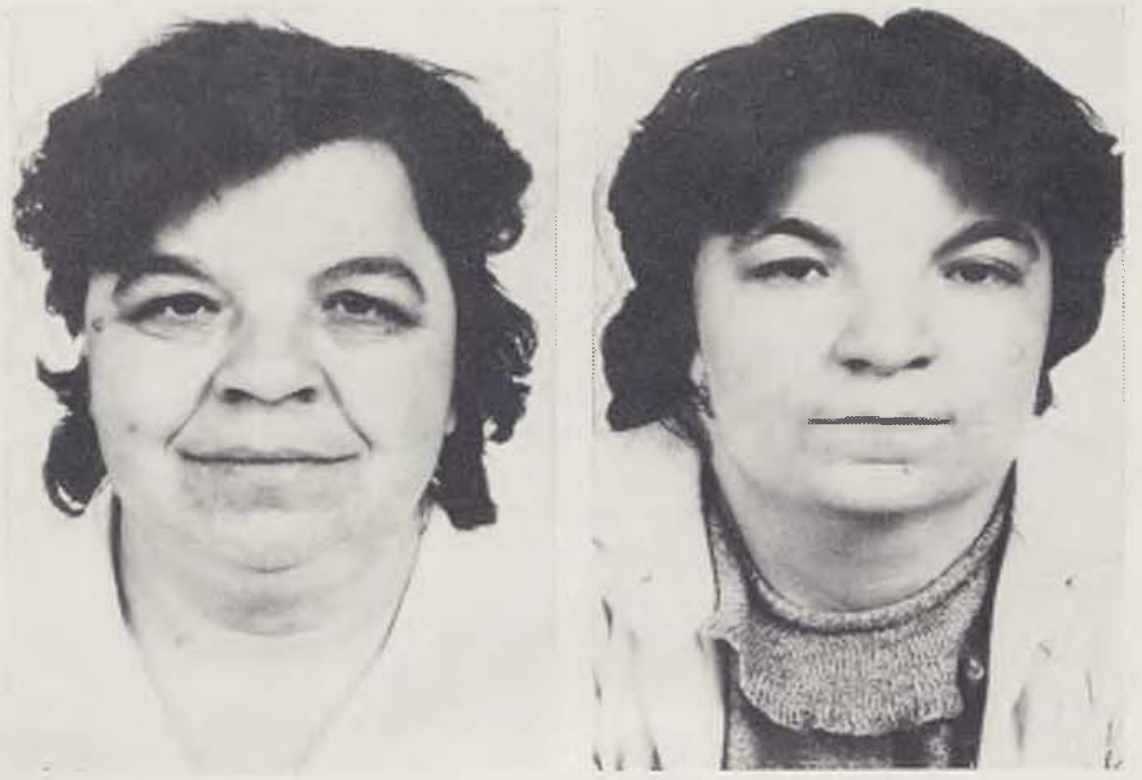

Fig. 3 Family I, mother of the proband: Telecanthus and hypertelorism
Fig. 4 Family I, sister of the proband: Telecanthus and hypertelorism, supraglabellar prominence, short and wide neck

lobes or extremities. She never suffered from dysphagia or respiratory distress. Her medical history showed a normal first pregnancy, while in the course of her second pregnancy there was an exposure to karosen during her full-time work. Her two brothers and one sister and their children were all of them normal. Their photographies and those of her parents showed no signs of telecanthus. Of interest were the dermatoglyphic patterns with bilateral distal axial triradius and two opposite loops on the right hypothenar with triradii t and t'. Finger patterns and the TRC were unchanged (Tab. 3).

Tab. 3. Dermatoglyphic finding

		Finger patterns					TRC	t in %	
Family I									
Propositus	dx	Lu	A	Lu	A	Lu	45	21.9'	
	sin	Lu	A	A	Lu	A		24.3'	
Mother	dx	W	Lu	Lu	Lu	Lu	143+?	11.0	30.0'
	sin	W	W	—	W	Lu		30.0'	
Sister	dx	D	Lr	Lu	Lu	Lu	100	24.2'	
	sin	Lu	Lu	Lu	Lu	Lu		19.6'	
[1]Nephew	dx	W	Lu	Lu	Lu	Lu	148	13.9	
	sin	Lu	Lu	Lu	Lu	Lu		12.5	
Family II									
Propositus	dx	Lu	Lr	Lu	Lu	Lu	178	11.8	
	sin	Lu	Lr	Lr	Lu	Lu		13.9	
Brother	dx	D	W	W	W	W	245	13.4	
	sin	D	W	W	W	W		15.0	
Mother	dx	Lu	W	W	W	W	215	10.0	
	sin	W	D	W	W	W		11.0	
[1]Father	dx	W	W	W	W	W	225	12.1	
	sin	D	W	D	W	D		12.2	22.4'

[1]normal, *larger by more than 1 SD (**2 SD), +smaller by more than 1 SD — missing finger, dis = dissociation

The sister of the proband (Fig. 4): She had a more marked telecanthus than all other members of her family, as well as hypertelorism on palpation (Tab. 2). Her face showed a conspicuous frontal prominence, the ear lobes were retroinclined and the tragus was almost absent. Her neck was wide with slight pterygias and a low hair line, but the X-ray films of the spine failed to disclose any pathologic changes. No further anomalies were

revealed. The dermatoglyphic pattern showed a slight reduction of TRC (100), an increase of papillary lines between the digital triradii, bilateral proximal loop on the thenar and in particular a distal axial triradius on both palms (Tab. 3).

At the age of nineteen years she gave birth to a healthy boy (birth weight — 2900 grams) who subsequently showed an adequate development without any signs of increased interocular distance (Tab. 2). His dermatoglyphic patterns were within the range of the norm (Tab. 3), but for the missing c tri-

a—b	b—c	c—d	Hypoth.	Interdigit.				th.	Main lines			
41	16+	35	Lr	0	0	Ld	0	0	11	9	7	5′
42	23	35	0	0	0	Ld	0	0	11	9	7	5′
37	38*	33	Lr/Lu	0	Ld	0	Ld	0	8	6	5′	3
40	33	28	Lr	0	Ld	0	Ld	dis	7	5″	5″	5′
48*	36*	52*	Lr	0	0	Ld	0	Lp	11	9	7	3
51**	42*	46	Lr	0	0	Ld	Ld	Lp	7	9	5′	3
50**	23	43	0	0	0	Ld	0	0	11	9	7	5′
54**	c missing		Lr	0	0	0	0	0	9	X	5″	3
46*	33	45*	Lr	0	0	Ld	0	0	9	9	5″	3
51**	33	45*	Lr	0	0	Ld	0	0	11	9	7	3
39	31	44	0	0	0	0	0	0	9	7	5″	5′
41	37*	41	0	0	0	0	0	0	9	7	5′	3
37	33	35	0	0	0	Ld	0	0	11	9	7	5′
33	34	33	0	0	0	Ld	0	0	11	9	7	3
42	31	46*	0	0	0	0	0	0	7	5″	5″	5′
35	33	40	Lu	0	0	0	0	0	7	5″	5″	3

radius on the left, and a bilateral increase of the a—b ridge count. Her two further pregnancies were terminated in the 2nd and 4th months by spontaneous abortion. Data on the fetuses are not available. Both pregnancies were neither planned nor controlled. Her husband is an unrelated healthy male.

The father of the proband: Is a normal male. No inborn malformations occurred among his relatives.

Family No. II.

Proband (Fig. 5): The boy was born after the second pregnancy, both parents are normal and unrelated (his mother was 25 and his father 33 years). The pregnancy continued for seven months without complications, but in the 8th month his mother was admitted to a hospital because of threatening abortion. She was delivered in the 34th week of gravidity (vertex presentation) of an alert child weighing 3000 grams. The newborn had scrotal hypospadias, the testes were within the scrotum divided into labial folds encircling the basis of the clitoriform penis. The anus was in its normal localization. The boy is at the present time aged sixteen years, he is healthy, robust (body height 172 cm, body weight 93 kg), and a normal intelligence quotient. He has both a telecanthus and hypertelorism (Tab. 2) with a marked depression of the glabella extending to the root of the nose and slight facial hypoplasia on the left. The ear lobes are slightly protrusive with some deformation of the helix, but are situated at the normal level. During childhood he was treated for stammer and at the age of eight years he was operated upon for strabismus. He suffered never from dysphagia or respiratory distress. No other anomalies were disclosed, the pubic hair was of feminine type. The ECG tracing and the laboratory tests were normal. The dermatoglyphic pattern showed no deviations (Tab. 3).

Fig. 5 Family II, proband: Telecanthus and hypertelorism, slight facial hemihypoplasia, glabellar depression

Fig. 6 Family II, brother of the proband: Slight telecanthus (euryopia), glabellar depression

The brother of the proband (Fig. 6): He was born after the first uncomplicated pregnancy (his mother was 22, his father 30 years) in the 36th week of gravidity, by vertex presentation. He was alert and weighed 3200 grams. He had penile hypospadia, the testes were situated within the well developed scrotum. The boy who is now aged 20 years, is of a mesomorph athletic type, and an active sportsman (body height 171 cm, body weight 69 kg). He has a normal intelligence quotient. Parameters used for the assessment of the orbital region confirmed an enlarged interorbital area classified as euryopia (Tab. 2). The intercanthal distance was characteristic of telecanthus (+2.5 SD). His face was symmetric with a depression of the glabella up to the root of the nose similarly as in his brother. At the age of five years he had surgical repair of strabismus. He complained never of dysphagia or respiratory distress. There were no other anomalies, his pubic hair was of masculine type. The ECG tracing and laboratory tests were within the range of the norm. The dermatoglyphic pattern showed exclusively composite (two triradii) finger patterns with a high TRC (245, Tab. 3). A simian line was present on the left palm.

The mother of the proband (Fig. 7): During her pregnancy she received no drugs, she was not ill, and was not subjected to X-ray studies or vaccination. She suffered never from gynecologic complaints. At the age of 32 years she underwent surgery for a goitre and at the time of examination was treated with thyreoglobulin. Her face showed signs of telecanthus

Fig. 7 Family II, mother of the proband: Telecanthus and hypertelorism, facial hemihypoplasia, glabellar depression

and hypertelorism (Tab. 2), as well as a marked depression of the glabella, similarly as in her two sons. There was also a slight hypoplasia of the left part of the face with an asymmetry and obliquity of the oral slot and a deviation of the nose. The left ear lobe was protrusive with a slightly deformed helix. Other deviations on her face, trunk or extremities were not disclosed. At the age of six years she was operated upon for strabism. She suffered never from dyspnea or dysphagia. With the exception of one finger all other fingers showed composite dermatoglyphic patterns with a high TRC (215). She has one normal brother. Photographies of her brother, his daughter and grandson and of the parents failed to reveal definite signs of a telecanthus in any of these relatives.

The father of the proband: A normal male, with a small interocular distance (31 mm). Of interest was only his dermatoglyphic pattern with bilateral simian lines, composite patterns on all fingers, a high TRC (225) and triradii t and t′ on the left side (Tab. 3). No inborn anomalies were reported in his parents or his other relatives.

DISCUSSION

All three affected males had a moderate or marked hypospadias and definite signs of telecanthus/hypertelorism. Neither of them, similarly as the three female cariers, suffered in early childhood or later from dysphagia or respiratory distress. Their history revealed no aspiration accidents or hoarseness. These symptoms represent pathognomic criteria for the distinction between the G and BBB syndrome. In the family pedigrees of individuals with the first syndrome they occur almost regularly in most affected individuals (Pedersen et al., 1976; Fundenburk and Stewart, 1978), while they are always absent in the second syndrome. As a further distinctive characteristic has been reported a high nasal bridge in the BBB syndrome, contrary to a flat bridge and an anteversion of the nares in the G syndrome (Fundenburk and Stewart, 1978). However in our series of affected individuals we failed to disclose a high nasal bridge and da Silva (1983) on the contrary described a flat nose in two patients with the BBB syndrome (one of them was an adult). Similarly Cordero and Holmes (1978) reported on the basis of their own experience that this characteristic does not represent a regularly occurring distinctive characteristic during childhood. Its drawback consists in the strong subjective influence of its determination which is affected also by racial differences. An anteversion of the nostrils can be again transformed by the presence of a cleft wih typical deviations of the nose and of the nares. Thus the validity of these differentiating criteria is limited. In the differential diagnosis represents a helpful characteristic a cranial asymmetry which occurs frequently in the BBB syndrome (Fig. 5, 7).

The other anomalies occur in both syndromes, inclusive of mental retardation, cleft lip and palate, ear lobe anomalies and dermatoglyphic characteristics with a distal axial triradius and low TRC. In this group of anomalies are of particular interest clefts. Of the eight documented cases in individuals

with the BBB syndrome seven had a bilateral cleft lip and palate, though this type of cleft accounts only for one quarter of individuals with cleft lip and palate and as little as an eighth of all types of cleft lip and/or palate. The relative frequency of individual anomalies associated with this syndrome is presented in Table 1. In this table are included the findings described by Opitz et al. (1969a) in nine patients, by Christian et al. (1969) in four patients, Michaelis and Mortier (1972) in one patient, Gonzales et al. (1977) in one patient, Funderbuk and Stewart (1978) in two patients, Cordero and Holmes (1978) in two patients, da Silva (1983) in two patients, and in three of our own patients. Some insufficiently documented cases were not included. The review of these series of patients provide evidence that occasionally either hypospadias (in two of four cases described by Christian et al., 1969) or hypertelorism (one patient of Opitz et al., 1969a) are missing. Approximately in one half of the patients occur mental retardation, strabismus, widow's peak, high nasal bridge, cranial asymmetries, distal axial triradius, and low TRC. One third of individuals has in addition cleft lip and palate, ear lobe anomalies (usually retroinclination), postcranial skeletal defects and cryptorchismus and one fourth inborn cardiovascular diseases, diastasis recti, low posterior scalp line etc. However, all but one cases of widow's peak were reported in the first two studies, in six patients only later on the basis of photographies (Gonzales et al. 1977). Most probably this abnormal pattern would not occur so frequently as stated. But disturbances in this region were confirmed by the sometimes occurring frontal or metopic prominence, which might be associated with hypertelorism.

Of particular importance for the practice was the diagnosis of hypertelorism (telecanthus), which represents the sole manifestation of this syndrome in heterozygote (hemizygote) females. In our series three affected males and three female cariers had definite evidence of hypertelorism, or telecanthus. Of the index values used for the assessment the highest degree of objectivity was obtained with the zygomatico-intercanthal index (the distance between the inner eye canthi in terms of per cent of facial width), where 29 units represented the marginal value of hypertelorism in our population (Šmahel, 1978). The intercanthal index is affected by simultaneously increasing distance of the inner and outer canthi (similarly as of the orbital margins in the interorbital index) while the circumference-intercanthal index is affected by the intense growth of the neurocranium during the first three years of life and by the neurocranial size. The review of the measurements of the orbital region in the first fourteen patients showed that all but one had definite signs of telecanthus determined on the basis of the intercanthal distance. Telecanthus was demonstrated in seven of nine female cariers, euryopia was present in one (Gonzales et al., 1977). Thus a certain percentage of cariers did not show the main symptom of this syndrome. The common occurrence of strabism was related most probably to the increased interocular distance. Similarly there was a frequent association of cryptorchism with hypospadia, in particular in severe forms of the anomaly.

The weight of our patients at birth was suggestive of a normal prenatal development, and the height-weight and bodily parameters documented an adequate postnatal growth. A smaller stature had only our proband with the cleft, but both his parents were small as well. The circumference of the neurocranium was within the range of norm in all individuals examined.

The dermatoglyphic patterns showed changes which could prove useful in the detection of female cariers. The affected males had in addition to a low TRC and a distal axial triradius often also t asymmetries. The rate of these characteristics was increased equally in heterozygote females. Gonzales et al. (1977) summed up the reports in the literature and found t asymmetry in four of six female cariers, while five had at least one distal triradius. He ascertained in his series an excessively low TRC in two female cariers in contrast to the normal sister of the proband. These findings were in good agreement with our observations in the first family (Tab. 3). Both heterozygote females had a bilateral distal axial triradius, the sister of the proband had a slightly reduced TRC and his mother had on the right another t triradius. The patient had an extremely low TRC and a bilateral t'. The normal son of the female carier showed none of these dermatoglyphic deviations. The reports in the literature revealed that normal sisters of affected males equally had more commonly a distal axial triradius (5/7; Opitz et al., 1969a; Gonzales et al., 1977). This arose the question whether some of them did not represent female cariers. The above mentioned dermatoglyphic deviations were not recorded in the second family. There were, on the contrary, composite finger paterns and a high TRC. They could represent a hereditary familial characteristic, since they were recorded in both parents, especially in the normal father (of a hereditary character was suggestive the presence of a simian line in the father and in his son with a high TRC). The hereditary characteristic was, beyond any doubt, the depression in the region of the glabella and possibly also the occurrence of strabism. However, it is necessary to mention the differences in the occurrence of other anomalies and deviations between the two families, consisting especially in the presence of cleft lip and palate, slight cervical pterygias and the low-set retroinclined ear lobes in the first family. They are suggestive of the differing expression of genes in different families. It is not possible to exclude the existence of distinct variants of this syndrome. In the affected pedigrees a high frequency of twins was recorded (Gonzales et al., 1977).

In prognostic view it is important to underline the differences between the BBB and G syndrome. In the first syndrome it is possible to give preference to the female sex by arteficially induced abortion of male fetuses. This procedure is less reliable in the G syndrome which can result in severe disorders also in females (dysphagia, stridor, lung hypoplasia, aspiration accidents, Little and Opitz, 1971; Biervliet and van Hemel, 1975), as confirmed by lethal outcome in two female patients (Kasner et al., 1974; Arya et al., 1980). In practice it take into account in each newborn with hypospadias and hypertelorism the possibility of an association with a laryngotracheoesophageal malformation and

to subject these patients to a through examination. The studies should be aimed also at the detection of heterozygote females where a precise analysis of facial physiognomy and of the dermatoglyphic patterns represent valuable diagnostic criteria. One case report dealing with a female patient with hypertelorism, mental retardation, and anomalous location of the urethral orifice (an analogy of hypospadias) underlines the necessity of gynecological examination (Reed et al. 1975).

From the two above mentioned syndromes differs the gonosomal recessive Aarskog syndrome which is characterized by small stature and a ventral scrotal fold over the base of the penis. Another reported patient had hypospadias, mental retardation, and bilateral ocular coloboma with a low TRC (Halal and Farsky, 1981). His father had a slight hypospadias, only very slight hypertelorism and an unilateral distal axial triradius. His late brother had an unilateral coloboma of the iris, his mother was normal. Since this proband had a distance between the inner eye canthi well within the range of the norm and his father had only a marginal value of this characteristic which was not confirmed by X-ray study, there was not definite evidence of hypertelorism. The presence of the BBB syndrome seems unlikely, since so far it was never described in association with ocular coloboma. If the BBB syndrome would be present in this patient this finding would represent the first reported male-to-male transmission of this anomaly (one doubtful case was described also by Opitz et al. 1969a). Another case of male-to-male transmission was reported by Stoll et al. (1985). They recorded in the son of their patient in addition a psychomotoric retardation associated with cardiovascular anomalies, unilateral cryptorchism and a distal axial triradius on both palms. The facial physiognomy of his father, however, showed no signs of telecanthus and the reported interorbital distance was larger (39 mm) than the intercanthal distance (38 mm) which appears highly improbable. The authors admitted that the BBB syndrome was not necessarily present in this individual. Thus this single case did not decide which of the two hereditary mechanisms was responsible for the BBB syndrome.

SUMMARY

The occurrence of hypospadias-hypertelorism (BBB) syndrome was described in two unrelated families with three affected males and three female cariers. In the first family the affected proband had penile hypospadias, hypertelorism, and complete bilateral cleft lip and palate. Hypertelorism was present also in his mother and sister. The proband had a prominent forehead, low-set

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retroinclined ear lobes, slight cervical pterygias, with a low hair line, spina bifida C_{4-5}, and a typical dermatoglyphic pattern with a very low TRC and bilateral distal axial triradius. His sister had retroinclined ear lobes and slight cervical ptarygias with a low hair line, both the mother and sister had prominent forehead and a bilateral distal axial triradius on their palms. In the second family two brothers had penile and scrotal hypospadias and hypertelorism, their mother had hypertelorism alone and all three had strabism with a marked depression in the region of the glabella. The dermatoglyphic patterns failed to show any deviations, an increase of the TRC appeared to represent a hereditary familial characteristic. The mother and one of her sons had a slight facial hemihypoplasia. None of the affected individuals in these two families showed mental retardation, cryptorchism or inborn heart disease. These observations were suggestive of the X-linked inheritance or autosomal dominant inheritance with male sex limitation. The physiognomic facial charecteristics, as well as the dermatoglyphic patterns could prove helpful in the identification of female cariers. The prevention consists in the prenatal preference of female fetuses.

RESUME

Syndrome d'hypertélorisme — hypospadias (BBB) dans deux familles

Horák, I., Šmahel, Z.

L'apparition du syndrome d'hypertélorisme — hypospadias (BBB) a été constaté dans deux familles sans parenteé, avec trois hommes atteints et trois femmes-porteuse de la tare. Le sujet de la première famille présentait une hypospadie pénile, un hypertélorisme et une division labiale et palatine bilatérale totale. La mère et la soeur du sujet étaient atteintes par l'hypertélorisme. Chez le sujet, on a en plus constaté un front proéminent, des pavillons d'oreille situés bas, en rétroinclinaison, de légères ptérygies de cou, des limites de la zone chevelue situées en bas, un spina-bifida sur le niveau de C_{4-5} et un dessin dermatoglyphique avec TRC très bas et un triradius axial distal bilatéral. Chez la mère et la soeur, on a trouvé un front proéminent et un triradius axial distal bilatéral, chez la soeur encore des pavillons d'oreille en rétroinclinaison, avec les indices de ptérygies du cou et des limites basses de la zone chevelue. Dans la seconde famille, deux frères étaient atteints par un hypospadias pénil et scrotal et par un hypertélorisme, chez la mère on a constaté un hypertélorisme, tous les trois présentaient de strabisme avec une dépression importante de la région de glabelle. Il n'y avait pas d'anomalies dermatoglyphiques, TRC étant augmenté représentait une caractéristique héréditaire de la famille. Personne des deux familles ne se trouvait en rétardation mentale, ne présentait de cryptorchisme et n'était atteint par aucune lésion cordiale.

Les examens correspondent à une hérédité liée récéssivement au chromosome X, qui comporte également les signes de dominance, autosomatique, restreinte au sex masculin. Les signes de physiognomie du visage, éventuellement les caractéristiques dermatoglyphiques, peuvent contribuer à l'identification des femmes — porteuse de tare. Comme méthode de prévention, on recommende le choix prénatal du sex féminin du foetus.

ZUSAMMENFASSUNG

Ein Syndrom von Hypertelorismus und Hypospadie (BBB) in zwei Familien

Horák, I., Šmahel, Z.

Es wird das Vorkommen eines Syndroms von Hypospadie und Hypertelorismus (BBB) in zwei nicht miteinander verwandten Familien bei drei befallenen Männern und drei übertragenden Frauen beschrieben. In der ersten Familie war der Proband von peniler Hypospadie, Hypertelorismus und vollständiger beiderseitiger Lippen- und Gaumenspaltung betroffen, die Mutter und Schwester von Hypertelorismus. Ferner hatte der Proband eine vorstehende Stirn, niedrigsitzende retroinklinierte Ohrmuscheln, leicht angedeutetes Halspterygium mit niedrigsitzender Haargrenze, spina bifida C_{4-5} und ein typisches dermatoglyphisches Bild mit sehr niedrigem TRC und bilateralem distalem axialem Triradius. Die Mutter und die Schwester hatten eine vorstehende Stirn und einen beiderseitigen distalen axialen Triradius, die Schwester ausserdem retroinklinierte Ohrmuscheln und angedeutetes Halspterygium mit niedrigsitzender Haargrenze. In der zweiten Familie waren zwei Brüder von peniler und skrotaler Hypospadie und Hypertelorismus befallen, die Mutter hatte nur Hypertelorismus, jedoch alle drei Strabismus mit ausgeprägter Depression in der Region der Glabella. Die Dermatoglyphen zeigten keine Abweichungen, ein erhöhter TRC erschien als vererbliche Familiencharakteristik. Die Mutter und ein Sohn hatten leichte Hämihypoplasie des Gesichts. Kein Mitglied der beiden metroffenen Familien war mental retardiert, hatte Kryptorchismus noch einen Herzfehler. Die Befunde entsprechen einer rezessiv an Chromosom X gebundenen Vererblichkeit,aber auch autosomalisch dominierend unter Einschränkung auf das männliche Geschlecht. Die Merkmale in der Physiognomie des Gesichts, evtl. die dermatoglyphische Charakteristik kann zu einer Identifizierung der Frauen als Übertragender beitragen. Die Methode einer Vorbeugung ist die pränatale Wahl des weiblichen Geschlechts der Frucht.

RESUMEN

Hipertelorismo-hipospadía (BBB) síndroma en dos familias

Horák, I., Šmahel, Z.

Está descrita la incidencia de la síndroma de hipospadía-hipertelorismo (BBB) en dos familias no parentezcas con tres hombres afectados y tres mujeres-portadoras. En primera familia el probado fué afectado por la hipospadía de penis, por hipertelorismo y por completa hendedura bilateral labial y paladar; su madre y hermana fueron afectados por hipertelorismo. El probado tenía la frente más prominente, bajamente localizados lóbulos retroinclinados, ligeramente distinguidas pterigias del cuello con baja frontera de pelo, spina bifida C_{4-5}, típico cuadro dermatoglífico con muy bajo TRC y con el distal triradial axial bilateralmente. La madre y la hermana tenían la frente prominente y bilateralmente distal triradial axial, la hermana tenía más los lóbulos retroinclinados y distingidas pterígias de cuello con la frontera baja de pelo. En segunda familia fueron afectados dos hermanos por la hipospadía de penis y por hipertelorismo; la madre tenía solamente hipertelorismo, todos tres luego el estrabismo y la depresión acentuada en la zona de la glabela. Dermatoglifos no mostraron diferencias, elevado TRC parecía por una característica heredera familiar. La madre y su hijo tenían una ligera hemihipoplasia del rostro. Nadie de los afectados de ambas familias no fué mentalmente retardado, no tenía criptorquismo ni una afección cardíaca.

Datos descubiertos corresponden a la herencia recesivamente ligada sobre el X cromosoma, pero también a la autosomal dominante con restrincción en el sexo masculino. Los signos de la fisionomía del rostro, eventualmente las características dermatoglíficas, pueden contribuir a la identificación de las mujeres-portadoras. El método de la prevención es elección prenatal del sexo femenino del feto.

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I. A STUDY OF THE INCIDENCE, SEX-RATIO, LATERALITY AND CLINICAL SEVERITY IN 3,660 PROBANDS WITH FACIAL CLEFTS IN CZECHOSLOVAKIA

M. TOLAROVÁ

Facial clefts as one of the common congenital malformations have been studied at our genetic unit (founded by one of the world's first plastic surgeons F. Burian) for twenty years from the aspects of heredity, etiology and incidence of this anomaly.

MATERIAL AND METHODS

Our present series consisted of 3,660 probands with cleft lip with or without cleft palate, and probands with isolated cleft palate born in Bohemia (Czechoslovakia) between 1975 and 1982. This sample was used for the estimation of the incidence, sex-ratio, laterality and clinical severity of the anomaly.

Data were collected from two sources. First, from patients coming to our unit, which — based on Burian's tradition — had for many years been in good collaboration with the Department of Plastic Surgery in Prague (the only centre where children with clefts, born in Bohemia, are operated on) and, second, the data were completed by comparison with governmental monitor of birth defects. In this way, practically all cases of cleft were collected and recorded, and the children, usually between 3—8 months of age, were seen at our genetic unit. During the examination, all microforms of clefts as well as associated malformations or syndromes were diagnosed. As a part of the genetic examination, the probands' parents and siblings were also examined for the same reasons.

RESULTS AND DISCUSSION

1. Incidence

Cases of syndromes and orofacial clefts associated with two or more other malformations were excluded from the incidence calculations (Table 1). For non-syndromic and non-multiple cases, the incidence 0.4396 per 1000 live birth

Table 1. Cleft lip with or without cleft palate and isolated cleft palate among live-born in Bohemia in 1964—1982

Type of cleft	Isolated cases			Cleft + 1 ass. anom.		Isolated cases + cleft + 1 ass. nom.		Cleft + 2 or more ass. anom.		Syndromes		All cases	
	No	%	incid.*	No	%	No	incid.*	No	%	No	%	No	%
CL	753	91.38	0.4112	52	6.32	805	0.4396	15	1.82	4	0.48	824	100.00
CLP	1263	83.21	0.6898	144	9.48	1407	0.7684	91	6.00	20	1.31	1518	100.00
CL ± P	2016	86.09	1.0160	196	8.36	2212	1.2081	106	4.52	24	1.03	2342	100.00
CP	963	72.08	0.5259	140	10.62	1103	0.6024	52	3.95	163	12.37	1318	100.00
CL ± P + CP	2979	81.39	1.6270	336	9.19	3315	1.8105	158	4.32	187	5.10	3660	100.00

*total number of live births 1.831,036 (1964—1982 in Bohemia)

(1 : 2275) for cleft lip (CL) only, 0.7684 (1 : 1301) for cleft lip and palate (CLP) and 0.6024 (1 : 1660) for isolated cleft palate (CP) was established.

The incidence of this anomaly is usually rather stable in certain populations, and significant differences are described only in different races. Although the incidence of all kinds of clefts is roughly 1 : 500 live births in Caucasians, one sixths of this value is observed in the black population (Table 2).

Table 2. Incidence of cleft lip and/or palate according to studies by different authors

Author (year)	Country (town)	CL	CLP	CL±P	CP	All types of clefts
Niswander and Adams (1967)	USA, Indians	—	—	1.38	0.59	—
Chung and Myrianthopoulos (1968)	USA, Caucasians	—	—	1.45	0.68	—
	USA, Negroes	—	—	0.73	0.44	—
Klásková (1973)	Czechoslovakia (1964—1971)	—	—	1.30	0.60	1.90
Melnick et al. (1980)	Denmark (1941—1970)	—	—	—	0.47	—
	Denmark (1941—1968)	—	—	1.30	—	—
Czeizel (1980)	Hungary (1970—1976)	0.34	0.57	1.16*	0.48**	—
	Budapest (1962—1967)	—	—	1.03	0.27	—
Chapman (1983)	New Zealand, Caucasians	—	—	1.20	0.64	—
	New Zealand, Maors	—	—	0.40	1.87	—
Tolarová (1984) (present study)	Czechoslovakia (1964—1982)	0.44	0.77	1.21	0.60	1.85

*including 0.25 for CL±P with associated anomaly
**including 0.05 for Pierre Robin syndrome and 0.12 for CP with associated anomaly

As you can see from Table 2, there are some differences in the incidence of isolated cleft palate, which seems to be a much more heterogenous anomaly. As far as we know, the highest incidence for the cleft palate (1.867) was published by Chapman (1983) for Maoris living in Auckland (New Zealand).

2. Sex-ratio

According to the classical multifactorial threshold (MF/T) hypothesis (Carter, 1969), the threshold level is dependent on the sex of the proband. This means that the individual of the sex, which is more rare, needs a higher number of polygenes to express the anomaly. This hypothesis assumes that these individuals have a greater chance to transmit the defect to the next generations.

The values of sex-ratio found in our sample (Table 3) corresponded with other findings (Table 4). The significantly higher proportion of males was found in cleft lip and in cleft lip and palate. The value of 1.85 was found for both types of cleft. The more severe forms had higher values (2.01 of CLP as distinct from 1.61 for CL) in correspondence with findings by other authors (Czeizel, 1980; Fraser, 1980; Melnick et al., 1980), although our value is slightly lower. Fraser (1980b) explained the sex differences in CL and CLP on the bas-

es of his previous experimental studies. Their experimental work shows that palate closure is delayed in embryos with cleft lip because of the large median process or prolabium obstructing the forward movement of the tongue. He assumed the prolabium to be smaller or protruding less in females. This could also explain the higher difference in sex-ratio between CL and CLP in Japanese compared to Caucasians (Fraser, 1980b).

Table 3. Sex-ratio in orofacial clefts

Type of cleft	Male	Female	Total	Sex-ratio
CL	497	308	805	1.61 1 : 0.620
CLP	939	468	1407	2.01 1 : 0.499
CL±P	1436	776	2212	1.85 1 : 0.540
CP	467	636	1103	0.73 1 : 1.362

Table 4. Sex-ratio in orofacial clefts according to studies by different authors

Author (year)	Country (town)	CL	CLP	CL±P	CP
Fogh Andersen (1942)	Denmark	1.90 (138)	2.40 (360)	—	0.75
Knox and Braithwaith (1963)	USA	1.60	2.10	—	0.70
Meskin (1968)	USA	1.50	2.20	—	—
Czeizel (1980)	Budapest (1962—1967)	—	—	1.54	higher
	Hungary (1970—1976)	—	—	1.56	in female
Koguchi (1980)*	Japan (1941—1970)	0.94	2.02	—	0.60
Fraser (1980)	USA and Canada	1.80	2.00	—	—
Melnick et al. (1980)	Denmark	1.73	2.35	2.10	—
Padron et al. (1982)	USSR (Moscow)	—	—	1.33	0.64
Iregbulem (1982)	Nigeria	1.10	1.00	—	1.00
Tolarová (1983)	Czechoslovakia (1964—1982)	1.61	2.01	1.85	0.73

*reviewed data of eight authors

3. Laterality

In the cleft lip and cleft lip and palate cases, the left side is significantly more frequently affected than the right. Also in experimental animals unilateral cleft lip is more common on the left side, whether spontaneous or induced.

In our study the left side was twice as much affected as the right one (Table 5), and this ratio was not influenced by the severity of the affection or by the proband's sex (Tolarová, 1984a).

4. Severity

The proportion of CL to CLP cases in our sample was 0.57 (688/1215); higher in females (398/262 = 0.65) than in males (426/817 = 0.52). This is the subsequent order of CL/CLP ratio in respect to sex and uni- or bilaterality:

1. female proband with unilateral affection 233/286 = 0.81
2. male proband with unilateral affection 398/577 = 0.68
3. female proband with bilateral affection 29/112 = 0.25
4. male proband with bilateral affection 28/240 = 0.11

The proportion of CL to CLP is greater in Orientals than in Caucasians. The racial differences in face shape could be the cause (Fraser, 1980 b). According to Fraser (1980b), the excess of this ratio, which seems to be fairly consistent, could represent an etiologically different type of cleft lip, for which there is good experimental evidence.

Table 5. Laterality of unilateral cases of cleft lip with or without cleft palate

Type of cleft		Male		Female		Total	
		No.	%	No.	%	No.	%
CL	sin	278	69.8	163	70.0	441	69.9
	dx	120	302	70	30.0	190	30.1
	total unilat	398	100.0	233	100.0	631	100.0
CLP	sin	380	65.9	199	69.6	579	67.1
	dx	197	34.1	87	30.4	284	32.9
	total unilat	577	100.0	286	100.0	863	100.0
CL±P	sin	658	67.5	362	69.7	1020	68.3
	dx	317	32.5	157	30.3	474	31.7
	total unilat	975	100.0	519	100.0	1494	100.0

Bilateral cases are rarer than the unilateral ones. This ratio is in relationship with the severity of the affection. While in the isolated cleft lip the bilateral coses occur in 8.28 %, cleft lip and palate cases in 32,75 % are bilateral. These findings are in agreement with the Hungarian (Czeizel, 1980: CL ± P 20.1 % of bilateral cases) and Danish data as well (Melnick et al., 1980: CL 9 % of bilateral cases, CLP 22 % of bilateral cases). It is interesting to note that in the black population a significantly greater number of bilateral cases is found.

When combining the sex of the proband and the severity of the affection, not only a prevalence of the male in both left and right unilateral cases but also a prevalence of the male sex in bilateral CL and CLP were found (Table

6). The highest difference was found in right side unilateral CLP cases — 2.26, and the lowest difference in bilateral CL cases — 0.97.

As our sample is almost a complete selection — all the cases born in a certain region over a certain time period were collected — we supposed we would be able to confirm Melnick's results (Melnick et al., 1980), who found the M/F ratio to be rising with increasing clinical severity. Our sample which is only half as large as the Danish data (1,895 Danish cases) confirms only the

Table 6. Clinical severity of cleft lip with or without cleft palate

Type of cleft		Male		Female		Total	
		No.	%	No.	%	No.	%
CL	unilat	398	93.4	233	88.9	631	91.7
	bilat	28	6.6	29	11.1	57	8.3
Total		426	100.0	262	100.0	688	100.0
CLP	unilat	577	70.6	286	71.9	863	71.0
	bilat	240	29.4	112	28.1	352	29.0
Total		817	100.0	398	100.0	1215	100.0
CL+P	unilat	975	78.4	519	78.6	1494	78.5
	bilat	268	21.6	141	21.4	409	21.5
Total		1243	100.0	660	100.0	1903	100.0

Table 7. Sex-ratio in CL+P by severity of cleft

Type of cleft		Melnick et al. (1980)				Tolarová (1984)			
		male	female	total	M/F ratio	male	female	total	M/F ratio
Unilateral	CL	386	226	612	1.71	398	233	631	1.71
Bilateral	CL	44	22	66	2.00	28	29	57	0.92
Unilateral	CLP	655	294	949	2.23	577	286	863	2.02
Bilateral	CLP	199	69	268	2.88	240	112	352	2.14
Total		1284	611	1895	2.10	1243	660	1903	1.88

difference of sex-ratio between CL (1.61) and CLP (2.01). On the contrary, the continuity of the increase from 1.61 in unilateral CL cases is interrupted by a decreased value 0.97 in bilateral CL cases. It could be suggested that different factors specific for each population were involved. A still larger sample of data is required in order to test this hypothesis.

In respect to the proportion of the subtypes of clefts classified in terms of the proband's sex and clinical severity, eight subgroups could be distinguished in our sample:

1. CLP unilateral male 30.33 %
2. CL unilateral male 20.91 %
3. CLP unilateral female 15.03 %
4. CLP bilateral male 12.61 %
5. CL unilateral female 12.24 %
6. CLP bilateral female 5.89 %
7. CL bilateral female 1.52 %
8. CL bilateral male 1.47 %

As is presented above, some differences in CL and CLP can be found. But it seems to be more reasonable to sum up the CL and CLP cases for this kind of analysis, because the values of empiric risk figures in our data (Table 8) also show the same trend. The use of associated cleft palate as an indicator of severity does not result in such consistent trends (Fraser, 1980b).

Table 8. Cleft lip with or without cleft palate: proportion of the affected individuals in children and in sibship (genealogical analysis)

Probands (No)			Sibs	Children	Total	
			cleft/total	cleft/total	cleft/total	%
Males	unilateral	(304)	12/529	16/444	28/973	2.88±0.54
	bilateral	(77)	6/125	8/93	14/218	6.42±1.72
Female	unilateral	(162)	17/330	10/263	27/593	4.55±0.88
	bilateral	(35)	1/62	7/55	8/117	6.84±2.42
Both sexes	unilateral	(466)	29/859	26/707	55/1566	3.51±0.47
	bilateral	(112)	7/187	15/148	22/335	6.57±1.40
Total		(578)	36/1046	41/855	77/1901	4.05±0.46

The recent reevaluation of Fogh Andersen's data from Denmark, as carried out by Melnick et al. (1977, 1980), discussed the validity of the classical multifactorial threshold model in CL ± P cases. Melnick and coworkers (1977) rejected the MF/T hypothesis in favour of a single major locus (SML). The analysis was carried out on two subgroups of cases — familial and non-familial. It seems a rather complicated division — because there are quite a few cases which became familial after several years, although they were sporadic at the beginning. The present view (Fraser, 1980 a, b) is that the MF/T model should not be abandoned yet and that more studies should be made to try and identify the possible "major" genes for susceptibility, for example in the face shape.

The same as in several other studies our data presented here seem to correspond quite well with the MF/T model of inheritance — the rarer subgroup has a higher risk of recurrence than the more common one, in which a lower risk of found (Tolarová, 1983, 1984a, b; 1985).

The lowest empiric risk figures (2.88 ± 0.54) were found when the proband was male with unilateral CL ± P. The correspondence of the incidence and the empiric risk figures was as follows:

CL ± P unilateral male	2.88 ± 0.54	51.23 %
CL ± P unilateral female	4.55 ± 0.88	27.27 %
CL ± P bilateral male	6.42 ± 1.72	14.08 %
CL ± P bilateral female	6.84 ± 2.42	7.40 %

From this study just as from other similar studies of facial clefts in man, the conclusion can be drawn that there are still some unanswered questions as to the etiology and heredity of this anomaly. A multicentre study of facial clefts could be very useful at the present time.

SUMMARY

A total of 3,660 probands with cleft lip with or without cleft palate, and probands with isolated cleft palate born in Bohemia (Czechoslovakia) between 1964—1982 were examined. As these children represented all cases born in a certain region and during a certain period of time, the precise value of the incidence was calculated. For non-syndromic cases the incidence 0.4396 per 1000 live births (1 : 2275) for cleft lip (CL) only, 0.7684 (1 : 1301) for cleft lip and palate (CLP) and 0.6024 (1 : 1660) for isolated cleft palate (CP) was established. The value of sex-ratio (M : F) 1.85 was found for CL and CLP together. The more severe forms had higher values (2.01 for CLP compared to 1.61 for CL). The same as in other studies, a prevalence of the female sex was found in CP cases (0.73). An evaluation of the clinical severity showed a higher proportion of bilateral cases in CLP (32,7 %) as distinct from CL (8,28 %). As for the unilateral cases, the left side was twice as much affected as the right (L : R = 68.3 : 31.7), and this ratio was not influenced by clinical severity or by sex.

RESUME

Évaluation de fréquence, de sex-ratio, de latéralité et du degré de la division chez 3660 personnes examinées, atteintes d'une division oralofaciale, sur le territoire tchécoslovaque

Tolarová, M.

On a exécuté l'analyse d'un groupe de 3660 personnes atteintes de bec-de-lièvre complet ou incomplet, nées dans les années 1964—1982 en Bohème (Tchécoslovaquie).

Comme il s'agissait d'un dépistage complet, de tous les enfants atteints de bec-de-lièvre sur un territoire précis et lors d'une période longue, il était possible de désigner les valeurs précises de la fréquence d'atteinte. Après l'élimination de syndromes et de malformations multiples, on a établi les valeurs suivantes: pour la division labiale 0,4396 à 1000 nouveaux-nés vifs (1 : 2275), pour la division labiale et palatine 0,7684 (1 : 1301) et pour les divisions palatines isolées 0,6024 (1 : 1660). La valeur de sex-ratio a été 1,85 pour les divisions labales avec ou sans division palatine. Les formes plus graves du point de vue clinique ont eu les valeurs plus élevées (2,01 pour la division labiale et palatine contre 1,61 pour la division labiale). Pareillement à d'autres études, la prévalence de femmes (0,73) a été constatée chez la division palatine isolée. L'évaluation de l'étendue clinique nous a amené à la constatation d'un haut pourcentage des types bilatérales de bec-de-lièvre (32,75 %), en comparaison de divisions palatines (8,28 %). L'atteinte de la partie gauche a été 2 fois plus fréquente que celle de la partie droite (68,3 % : 31,7 %). Cette relation n'était pas influencée ni par l'étendue. de division, ni par le sex des personnes examinées.

ZUSAMMENFASSUNG

Die Einschätzung der Häufigkeit, der Betroffenheit der Geschlechter, der Lateralität und des Umfangs der Spaltbildungen bei 3660 Probanden mit orofacialer Spalte aus der Tschechoslowakei

Tolarová, M.

Es wurde eine Analyse einer Gruppe von 3660 Probanden mit Lippenspalten mit oder ohne Gaumenspalten vorgenommen, die in den Jahren 1964—1982 in Böhmen (Tschechoslowakei) geboren wurden. Da es sich um das vollständige Auffangen aller Kinder mit Spaltenbildungen in einem bestimmten Gebiet und einer verhältnismässig langen Zeitspanne handelte, war es möglich, genaue Werte der Häufigkeit des Vorkommens zu berechnen. Nach Ausschliessung von Syndromen und vielfachen Missbildungen wurden folgende Werte festgestellt: Lippenspaltung bei 0,4396 von 1000 lebend geborenen Kindern (1 : 2275), Lippen- und Gaumenspaltung bei 0,7684 (1 : 1301) und isolierte Gaumenspaltung bei 0,6024 (1:1660). Die Betroffenheit der Geschlechter ergab für Lippenspaltungen mit oder ohne gleichzeitiger Gaumenspaltung den Wert von 1,85. Klinisch schwerwiegendere Formen wiesen höhere Werte auf (2,01 für Lippen- und Gaumenspalten und 1,61 für Lippenspalten). Ebenso wie bei weiteren Studien wurde für die isolierte Gaumenspaltung ein Übergewicht der Frauen festgestellt (0,73). Die Einschätzung des klinischen Umfangs führte zur Feststellung eines hohen Prozentsatzes der beiderseitigen Typen bei Lippen- und Gaumenspaltungen (32,7 %) gegenüber einer blossen Lippenspaltung (7,02 %). Die linke Seite war doppelt so häufig betroffen als die rechte Seite (68,3 % : 31,7 %). Dieses Verhältnis wurde weder vom Umfang der Spaltung noch vom Geschlecht der Probanden beeinflusst.

RESUMEN

Valoración de la incidencia de hendiduras buco-faciales distribuídas según sexo, lateralidad y extensión, en 3660 casos en Checoslovaquia

Tolarová, M.

Se analizó un grupo de 3660 casos con dendidura labial, con o sin acompañamiento de hendidura del paladar, nacidos en el período comprendido entre 1964—1982 en la región de Chequia (Checoslovaquia). Debido a que la muestra abarcaba toda la población afectada por hendiduras buco-faciales en una región y un cierto período de tiempo, los resultados obtenidos pueden representar estadísticamente las cifras correspondientes a otras poblaciones. Al eliminar los casos que no correspondían debido a síndromes o diferentes malformaciones, fueron obtenidos los siguientes valores: para hendiduras del labio 0,4396 para 1000 nacidos vivos (1:2275), para hendiduras de labio y paladar 0,7684 (1 : 1301), y para hendiduras aisladas del paladar 0,6024 (1 : 1660). Agrupados por sexo, el valor obtenido en casos de hendiduras del labio acompañadas o no por hendiduras del paladar fué de 1,85. Las formas clinicamente más graves presentaron valores superiores (2,01 para hendidura del labio y del paladar en comparación con 1,61 para casos con hendidura del labio solamente). Se comprobó la prevalencia en mujeres de hendiduras aisladas del paladar (0,73) lo que fué comprobado en investigaciones ulteriores. La valoración según la extensión cliníca condujo a la comprobación de un alto porcentaje de tipos bilaterales con hendiduras de labio y paladar (32,7 %) en comparación con los que presentaban hendiduras del labio solamente (8,28 %). La sesión en el lado izquierdo se presentó con el doble de frecuencia que la del lado derecho (68,3 % : 31,7 %), no influyendo en esta proporción la extensión de la hendidura ni el sexo delos casos muestreados.

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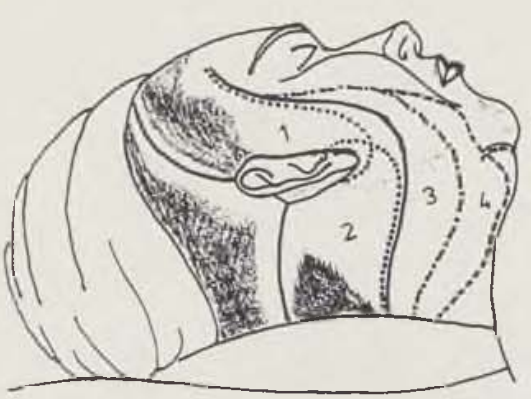
SKIN LIFTING TECHNIQUES IN RHYTIDECTOMY

K. FAHOUN

Skin lifting for rhytidectomy requires adequate, strictly individual, skin mobilization. Depending on the extent of mobilization, rhytidectomy can be classified into regional — temporal and cervical, standard, and radical cervico-facial rhytidectomy. In the mastoidal region, the skin is always thin with no subcutaneuos fat, which makes mobilization there a rather difficult process. The point is then to make sure that the mobilized skin is sufficiently thick in order to prevent the danger of necrosis and, on the other hand, to avoid mobilizing too deep for fear of damaging the auricular nerve with its branches. This can be avoided if we keep strictly to the fascia. On the nape of the neck, the incision and mobilization can go variously far, if need be as far as the posterior median line. Mobilization in the temporal region should be sufficiently deep to avoid damage to the hair follicles. On the cheeks and on the nape of the neck, mobilization should proceed fairly superficially under the skin keeping the blunt tips of the mobilization scissors under constant visual control. This is to avoid any damage to the facial nerve noted for its greatly variable ramifications. The extent of mobilization is again individual, possibly as far as the outer edge of the orbit and the nasolabial sulcus. The general rule is to take a more conservative approach in the case of thin, profusely bleeding types of skin, and a more radical strategy in elderly patients with major prolapse. On the neck, mobilization can go as far as the anterior median line, particularly where we plan to remove submental and submandibular fat.

Skin lifting is effected by traction of the preauricular and retroauricular mobilized cornerpieces. The force of traction depends on the quality of the skin and the degree of the required correction. The thing to be borne in mind throughout the process is the need to preserve the natural expression of the face. The general rule is that the preauricular end ought to be pulled backward and upward roughly in the direction of the connecting line between the tragus and Darwin's prominence on the helix of the pinna. Traction at the retroauricular mobilized skin flap is effected by means of three Kocher's forcepses situated behind the pinna, in the middle portion of the incision and

at its end. As a rule, this is a fanwise type of traction. The forceps behind the pinna pulls upward and towards the auricle, the one in the middle straight upward, and the posterior one upward and backward towards the nape of the neck.



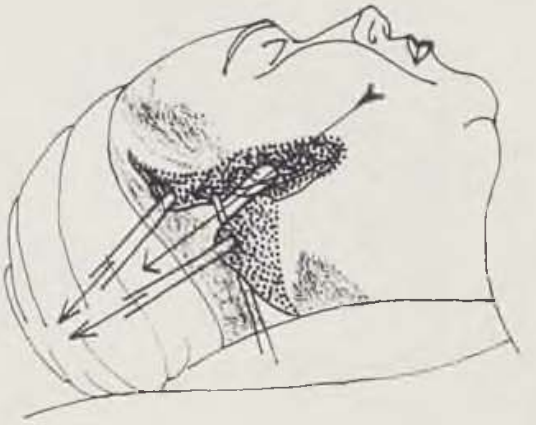


Fig. 1 Extent of mobilization: 1. Temporomalar rhytidectomy — regional, 2. Cervical rhytidectomy — regional, 3. Standard rhytidectomy, 4. Cervicofacial rhytidectomy — radical type

Fig. 2 Preauricular and retroauricular skin flap traction

Bald patches are often seen in the hair-covered part of the temporal region plication as they can thwart the whole surgical effort and, sometimes, add to the dissatisfaction of the patient about to be operated on for reasons of ptosis.

Bald patches are often seen in the hair-covered part of the temporal region in connection with a surgical scar. The mode of healing (such as, e. g., suture fistulae, stitches cutting through, necrosis) can exercise a major influence on the appearance of bald patches. Of equal importance, however, is the right degree of the lifting traction depending on the quality of skin and hair. The surgeon himself can give rise to bald patches during the operation by advancing the preauricular hairline too far or by its wrong reconstruction behind the auricle on the nape of the neck. Bald patches are always a source of the patient's dissatisfaction and often likely to make the surgeon venture more operations which can be extremely complicated and not always exactly successful. A mere excision of the bald patch in the hair-covered part of the temporal region may sometimes bring remedy to what is an unpleasant complication. However, this need not always be the case as the skin excision will inevitably add to the traction in the respective zone. This is certainly not the way to deal with a highly elevated preauricular hairline. In such cases, local flap transfer of hair-covered skin is the only, highly complex, solution. This danger arises mainly in repeated rhytidectomy. Complications of this kind should be prevented already at drawing up the individual plan of operation, by planning the surgical incision required, and, at the time of surgery, by making the direction of lifting traction a strictly individual affair. We certainly cannot

perform the lifting operation in only one well-practised schematic way. Fig. 4 shows a technique of fan-like traction in preauricular skin lifting designed to prevent an excessive elevation of the hairline.

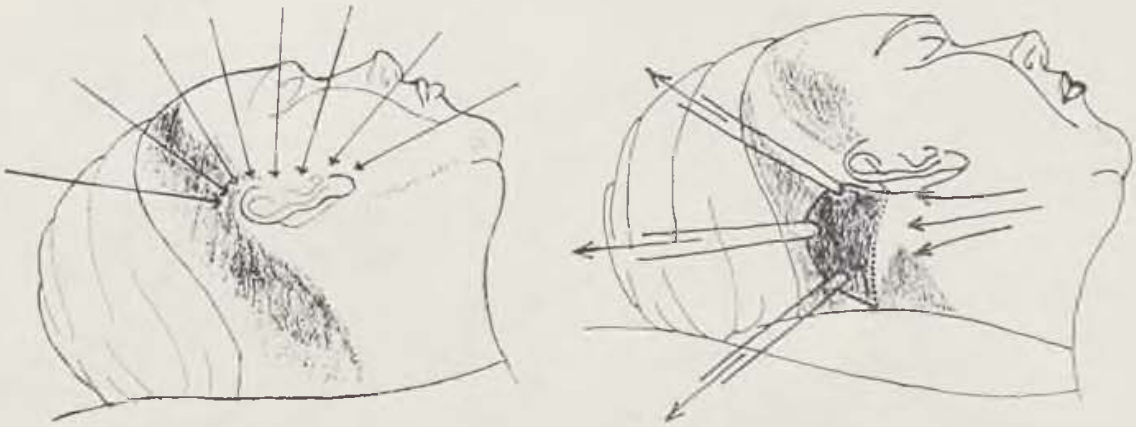


Fig. 3 Directions of retroauricular skin flap traction
Fig. 4 A technique for fan-like traction in preauricular skin lift

Fig. 5 Direction of skin incision in hair-covered part of the nape of the neck

The retroauricular incision should be made fairly high up in the hair, roughly along the continuation of the line connecting the outer corner of the eye with the upper margin of the tragus.

Misreconstructed hairline rather than bald patches constitutes the main potential complication in that region. The choice of the operation incision as required is greatly variable, and ought to be individualized from case to case. Fig. 6 to 9 show different extent of excision and advancement for the correct reconstruction of the hairline on the nape of the neck.

However, led by an exaggerated effort at hairline reconstruction we are not at complete liberty to rotate the mobilized skin corner towards the auricle.

Excessive rotation might result in too much skin being accumulated at the site of the ear lobule. A correct location and suture of the lobule are of great aesthetic importance. Hence why an optimum compromise is necessary. The skin suture in the hairless part of the skin behind the auricle should always be achieved by intradermal continuous suture using an atraumatic needle. This is the way to make the scar the least conspicuous with no need for covering up with hair.

Fig. 6

Fig. 7

Fig. 8

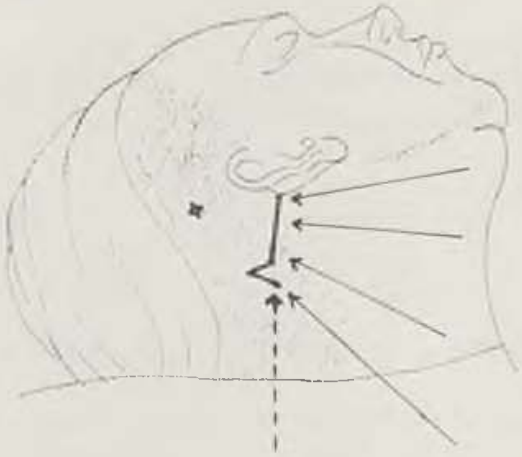


Fig. 9

SUMMARY

There is no stereotype for sugical incisions and skin lifting in rhytidectomy. A case-to-case individual approach is called for there. The author presents diverse variations and techniques for achieving an aesthetically acceptable post-operative scar with minimum damage to the hair-covered part and to the hairline.

RESUME

Méthodes de «lifting» cutané au cours d'une rhitidectomie

Fahoun, K.

Au cours d'une rhitidectomie, les incisions et le «lifting» cutané ne peuvent pas être exécutés par routine. Il faut suivre un plan individuel, cas sur cas. L'auteur allegue plusieurs variations et plusieurs possibilités des méthodes aboutissantes à une bonne cicatrisation postopératoire du point de vue esthétique, sans que les parties chevelue et leurs contoures soient gravement endommagées.

ZUSAMMENFASSUNG

Die Art und Weise eines Liftings der Haut bei einer Rhytidektomie

Fahoun, K.

Operative Schnitte und Lifting der Haut bei einer Rhytidektomie kann man nicht schablonenmassig ausführen, sondern man muss individuell von Fall zu Fall verschiedenartig vorgehen. Der Autor führt verschiedene Varianten und die Möglichkeit an, wie ästhetische und qualitative Narben nach der Operation unter minimaler Beschädigung behaarter Teile zu erzielen sind sowie ihre Grenzen.

RESUMEN

Modos de lifting cutáneo en la rhytidectomía

Fahoun, K.

A los cortes de operaciones y al lifting cutáneo en la rhytidectomía no se les puede realizar rutinariamente. Hay que proceder individualmente — de caso en caso. El autor presenta diferentes variaciones y posibilidades, por los cuales se puede lograr a una cicatriz posoperatoria estéticamente de buena calidad con el daño mínimo de la parte peluda y de su frontera.

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A NEW METHOD OF PRE-OPERATIVE PLANNING OF EAR-TO-SCULL DISTANCE IN PROTRUDING EARS

M. M. MADZHAROV

The importance of pre-operative planning of ear-to-scull distance in protruding ears has been noted by a number of plastic surgeons. Thus, for inst., McDowell (1968) estimated that during operation the top of the helix should reach 10—12 mm from the lateral surface of the scull, the tail of the helix (i. e. at the level of the lower border of the antitragus) — 20—22 mm and half-way between the top and the tail of the helix — 16—18 mm. Tardy et al. (1969) carried out a measurement of the mastoid-helix distance before and during operation, which helped them to achieve symmetrical ears. Spira et al. (1969) pointed out the benefit of using pre-operative and post-operative models of the pinna in analysing the deformities and in evaluation of the results of auriculoplastics, while Wright (1970) considered that the operator should have a mental image of his goal and the post-operative configuration of the auricles. A basic shortcoming of existing methods of pre-operative planning of ear-to-scull distance, both in this country and abroad, is that no pre-operative calculation of this distance is made. During the process of operative intervention plastic surgeons continue being guided by conventional norms and/or by the accuracy of their estimation by eye. The practice, however, shows that norms used by different authors vary considerably. For some mastoid-helix distance could be 1 cm while for others — 1 inch. Besides that existing norms are being applied as a rule to all patients. But as is well-known such norms are strictly individual and should be in direct dependance both between themselves as well as on the width of auricles and heads. On the other hand, closing protruding ears by eye is a subjective evaluation and could lead to errors. Guided by the above considerations, we developed a mathematical method of pre-operative planning of the ear-to-scull distance in dealing with protruding ears.

The essence of the method consists in calculating the mastoid-helix distance, the protruding at the top of the ears and at the earlobe.

1. Calculation of mastoid-helix distance

Mastoid-helix distance is defined on the basis of a trigonometric dependence, according to the formula $h_1 = a \cdot \sin \alpha$, where h_1 is the mastoid-helix

distance $(pal—rm)^2$, a — the width of the auricle $(pra—pal)^2$ and α — the cephalic auricular angle. The triangle where the trigonometric dependence is can be seen on Fig. 1. This formula helped us to draw a table (Table 1), ac-

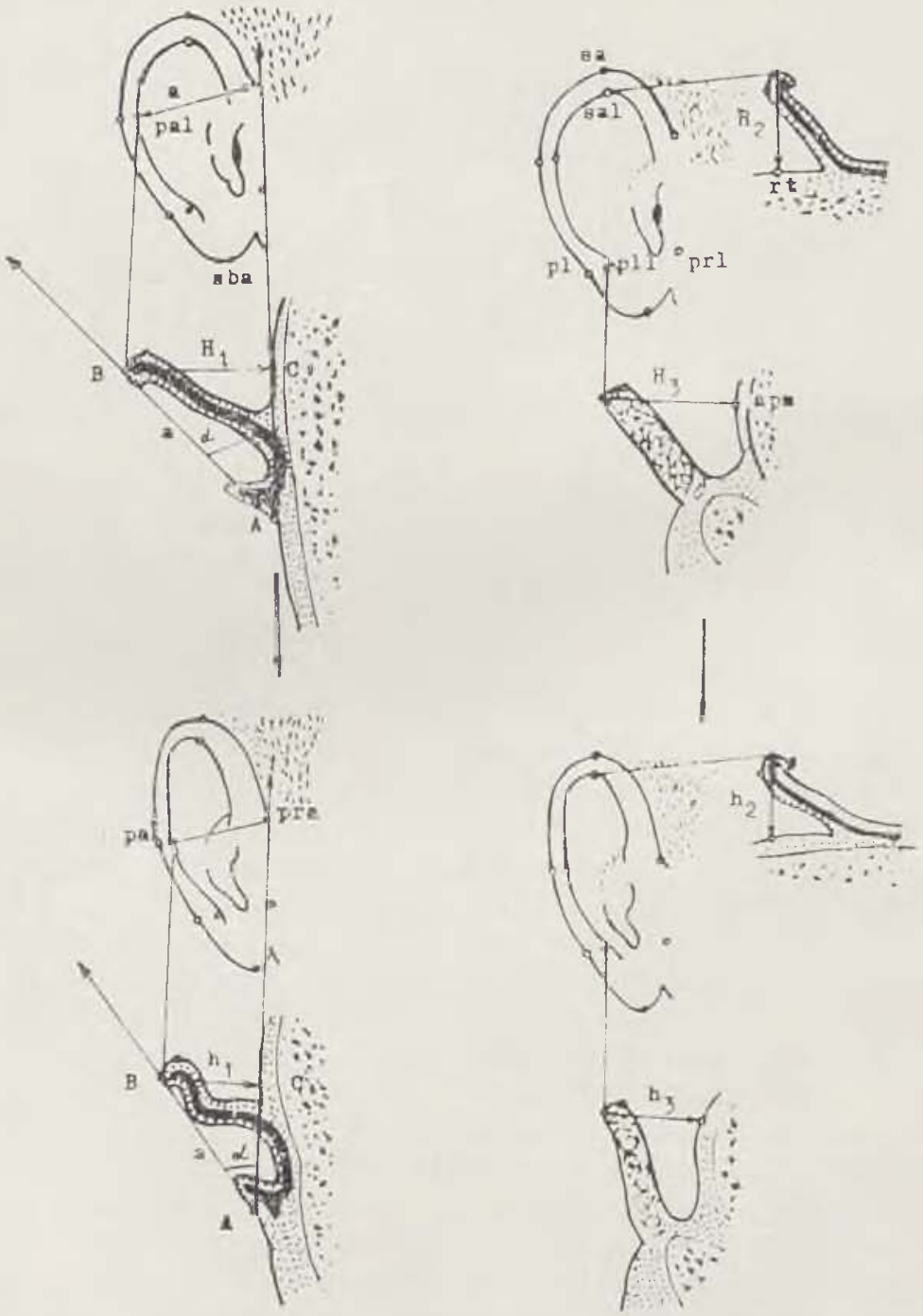

Fig. 1. The triangle ABC where one can see the trigonometric dependence for determining the value of mastoid-helix pre-operative distance (H_1) and the post-operative (h_1) and through it the distance between the top of the ear and the ear lobe on one hand and the scull on the other, (H_2, h_2 and H_3, h_3) respectively.

More important anthropometric points of the ear: sa — superaurale; sal — superaurale laterale; pa — postaurale; pal — postaurale laterale; pra — praeaurale; pl — postlobulare; pll — postlobulare laterale; prl — praelobulare; sba — subaurale; rt — regio temporalis; rm — regio mastoidea; apm — apex processus mastoidei

Table 1. Values of the mastoid-helix distance, used for diagnosis and planning in auriculoplsty of protruding ears

Width of auricle to helix (pra-pal) (mm)	Cephalic-auricular angle		
	14° Major drawing (mm)	24° Moderate drawing (mm)	34° Minor drawing (mm)
21	5.1	8.5	11.7
22	5.3	8.9	12.3
23	5.6	9.4	12.9
24	5.8	9.8	13.4
25	6.0	10.2	14.0
26	6.3	10.6	14.5
27	6.5	11.0	15.1
28	6.8	11.4	15.7
29	7.0	11.8	16.2
30	7.3	12.2	16.8
31	7.5	12.6	17.3
32	7.7	13.0	17.9
33	8.0	13.4	18.5
34	8.2	13.8	19.0
35	8.5	14.2	19.6
36	8.7	14.6	20.1
37	9.0	15.0	20.7
38	9.2	15.5	21.2
39	9.4	15.9	21.8
40	9.7	16.3	22.4
41	9.9	16.7	22.9
42	10.2	17.1	23.5
43	10.4	17.5	24.0
44	10.6	17.9	24.6
45	10.9	18.3	25.2
46	11.1	18.7	25.7
47	11.4	19.1	26.3

cording to which, depending on the width of his or her auricles and the degree of protruding required, the exact mastoid-helix distance can be determined for each individual patient. Defining the maximum and minimum normal values of the cephalic auricular angle has been based on the established by us for the Bulgarians from 0 to 100 years of age the maximum and minimum values of the width of the auricle to the helix (21—47 mm) and the minimum value of the mastoid-helix distance (5.1 mm)[2], and the maximum value of the mastoid-helix distance (26.3 mm)[1], as indicated in the literature.

Manipulation with the table

In auriculoplasty the table is used to differentiate between protruding ears and normal cases and for pre-operative planning of the required by the patient post-operative mastoid-helix distance.

Qualifying ears as protruding is done easily by comparing the figure in the last column of the table, corresponding to the width of the ear under observation, with the value of the mastoid-helix distance. If the value is greater than the figure of the table it means that we have a case of protruding ears.

A basic principle in pre-operative planning of the mastoid-helix distance is that its value should not be higher than the above-mentioned maximum tolerances in the last column of the table. Corrective auriculoplasty in protruding ears is, however, a cosmetic operation which is supposed to meet the aesthetic requirements of the patient and his or her relatives. The plastic surgeon is bound to comply with these requirements. That is why pre-operatively he has to specify the degree of drawing nearer of the auricles as requested by the patient — minor, moderate or major. A drawing nearer is considered as major when the cephalic auricular angle is 14°—20° (practically 14°), moderate — 21°—27° (practically 24°) and minor — 28°—34° (practically 34 °). Based on the patient's request and the value of the width of his ears the exact mastoid-helix distance can be defined by using the table. The difference between this distance and the value of that of the patient indicates how many milimetres the protruding auricles have to be drawn closer. For instance, if a patient with 32 mm protruding auricles and width 39 mm requires a moderate drawing closer, respectively of 24° cephalic auricular angle, the ears have to be drawn to the scull by 16 mm (32 — 16 = 16).

2. Calculation of protrusion of the ear tops

This is done by using the formula $h_2 = K_1 . h_1$, where h_2 is the distance we want to establish between the top of the ear and the temporal region $(sal—rt)^2$, h_1 — the planned mastoid-helix distance $(pal—rm)^2$ and K_1 — a coefficient. The latter is a decimal figure, being the result of dividing the mean value of the distance between the top of the ear and the temporal region by the mean value of the mastoid-helix distance of mature individuals. According to our data for the Bulgarian nationality its value for females is 0.74 and for males — 0.77.

3. Calculation of protrusion of the ear lobes

Here the formula $h_3 = K_2 . h_1$ is used, where h_3 is the looked for distance between the ear lobe and the mastoid process $(pll—apm)^2$, h_1 — the planned mastoid-helix distance (pal—rm) and K_2 — a coefficient. The latter being a decimal figure, the result of dividing the mean value of the distance between the ear lobe and the mastoid process by the mean value of the mastoid-helix distance in mature individuals. For the Bulgarian nationality we have established that its value for females is 0.94 and for males — 0.89.

CONCLUSION

We think the proposed method of planning corrective auriculoplasty in protruding ears represents a qualitatively new type of planning. It is substantiated mainly by mathematical pre-operative determination of mastoid-helix

distance, the distance between the ear lobe and the top of the ear on one hand and the scull on the other, this being done in a strictly individual way depending on the width of the auricle and the aesthetic requirements of the patient. In this way planning of corrective auriculoplasty has been placed on mathematical principles and a better adaptation of the degree of correction to the size of the auricles and the psychic adjustment of the operated patient is achieved, at the same time eliminating any possibilities for subjective errors. Besides, the method deals a blow both to empirical application of the recognized in the literature "norms" of planning and to the theory of the "born plastic surgeon with an accurate eye", who does his planning on the operation table.

SUMMARY

A table for pre-operative calculation of the mastoid-helix distance, and through it of the distances between the top of the ear and the scull and the ear lobe and the scull, has been composed by means of a trigonometric dependance between the width of the auricle and the cephalic auricular angle. The advantage of the method is that planning of auriculoplasty in protruding ears has been based on mathematical principles and all possibilities for subjective errors are eliminated.

RESUME

Nouvelle méthode pour fixer avant l'opération la distance entre le pavillon d'oreille et le crâne, dans le cas des oreilles décollées

Madzharov, M. M.

On a élaboré un tableau de calcul préopératoire pour que l'on puisse désigner la distance entre l'apophyse mastoide et le hélix, ainsi que la distance entre le lobule et la tête. Le compte est fondé sur le rapport trigonométrique de la largeur du pavillon et de l'angle céphalo-auriculaire. L'avantage de la méthode consiste au fait que le plan opératoire de l'auriculoplastie en cas des oreilles décollées s'effectue sur les fondements mathématiques, éliminant ainsi tous les risques d'erreurs subjectives.

ZUSAMMENFASSUNG

Eine neue Methode voroperativer Planung der Entfernung zwischen dem Ohrläppchen und dem Kopf im Fall abstehender Ohren

Madzharow, M. M.

Es wurde eine Tabelle ausgearbeitet zur voroperativen Berechnung der Entfernung zwischen processus mastoideus und helix und mit ihrer Hilfe auch der Entfernung zwischen dem Ohrläppchen und dem Kopf, ebenso wie zwischen der Ohrmuschel und dem Kopf, und zwar auf Grund der trigonometrischen Abhängigkeit der Breite des Ohrs und dem zephalo-aurikularen Winkel. Die Vorteile dieser Methode bestehen darin, dass sie die Planung der Aurikuloplastik bei abstehenden Ohren auf eine mathematische Grundlage stellt und jedwedes Risiko subjektiver Fehler ausschaltet.

RESUMEN

Nuevo método de planeamiento preoperatorio de distancia entre pabellón de la oreja y la cabeza en los casos de las orejas resaltadas

Madzharov, M. M.

Fué elaborada la tábula para el cálculo preoperatorio de la distancia de processus mastoideus y helix, y con su ayuda también de la distancia entre el pico del pabellón de la oreja y la cabeza, lo mismo como entre la perilla de la oreja y la cabeza, a saber — a la base de la dependencia trigonométrica entre la anchura de la oreja y el ángulo cefaloauricular. Las ventajas de éste método residen en que el planeamiento de la auriculoplástica de casos de las orejas resaltadas están puestas en base matemática que elimina cualquier riesgo de equivocaciones subjetivas.

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CLOSURE OF THE LARGE BILATERAL SOFT-TISSUE DEFECTS ON THE ANTERIOR KNEES SURFACES WITH THE USE OF AN ISLAND MUSCLE FLAPS FROM THE GASTROCNEMIUS CASE REPORT

J. PODLEWSKI, L. JANKIEWICZ, M. OPOLSKI, K. MICHALOVSKI

The medial and lateral heads of the gastrocnemius muscles are two of the most useful muscle units in the treatment of soft tissue defects in the lower extremity. During the past several years there were many reports of their usefulness, both as muscle and musculocutaneous flaps (1, 2, 3, 4, 5). The purpose of this report is to present the successful management of the large bilateral soft tissue defects on the anterior knees surfaces resulting from a frost-bite.

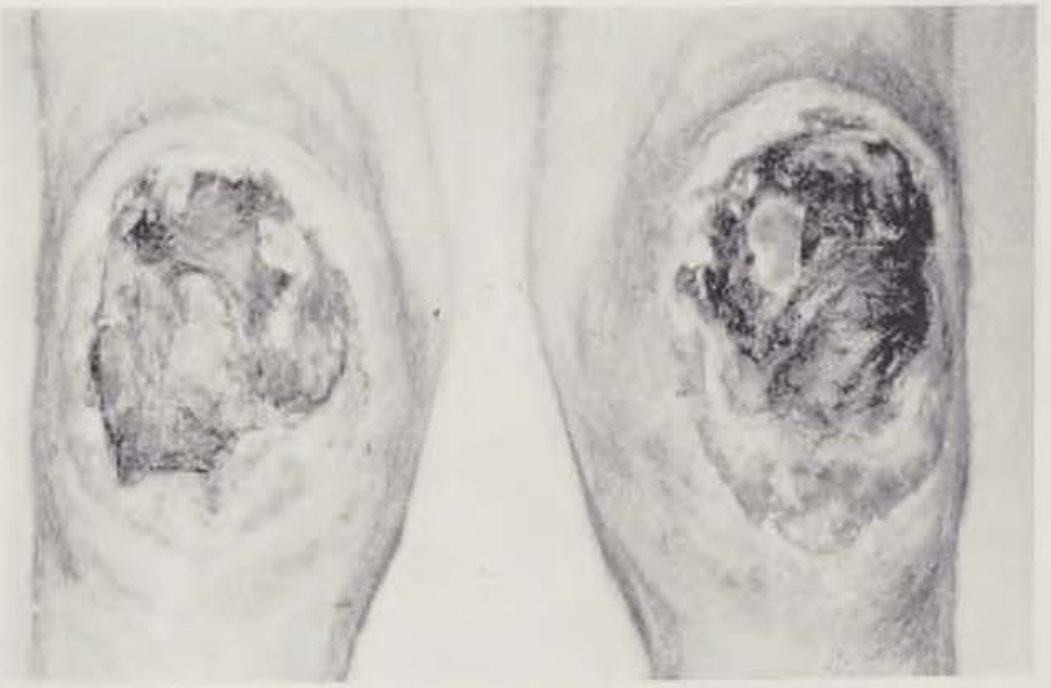

Fig. 1 Frost-bite ulcers of the knees, showing a large amount of necrotic material

Case report

A 53-year-old man had sustained 5 months earlier a frost-bite of both knees. At the time of admission his knees were covered with necrotic tissue

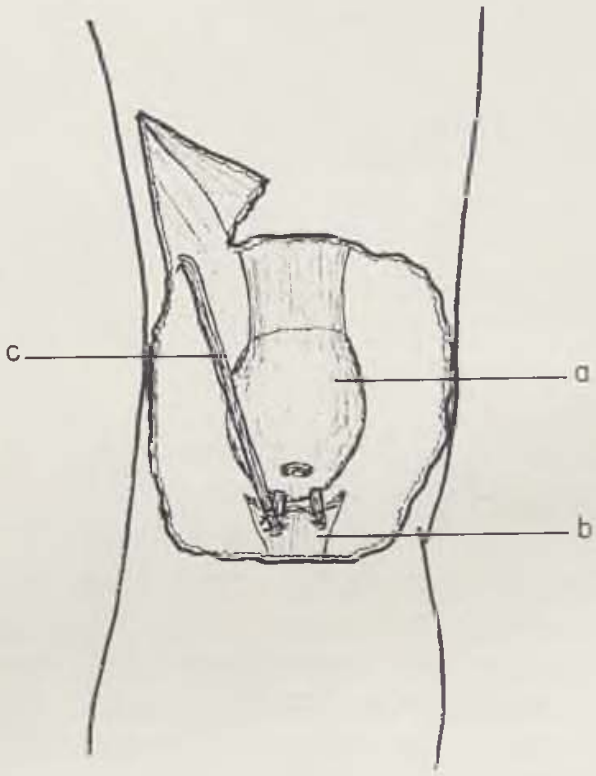

Fig. 2 The infrapatellar tendons were reconstructed with the fascia strips from the fascia lata a — patella, b — infrapatellar tendon, c — strip of fascia lata

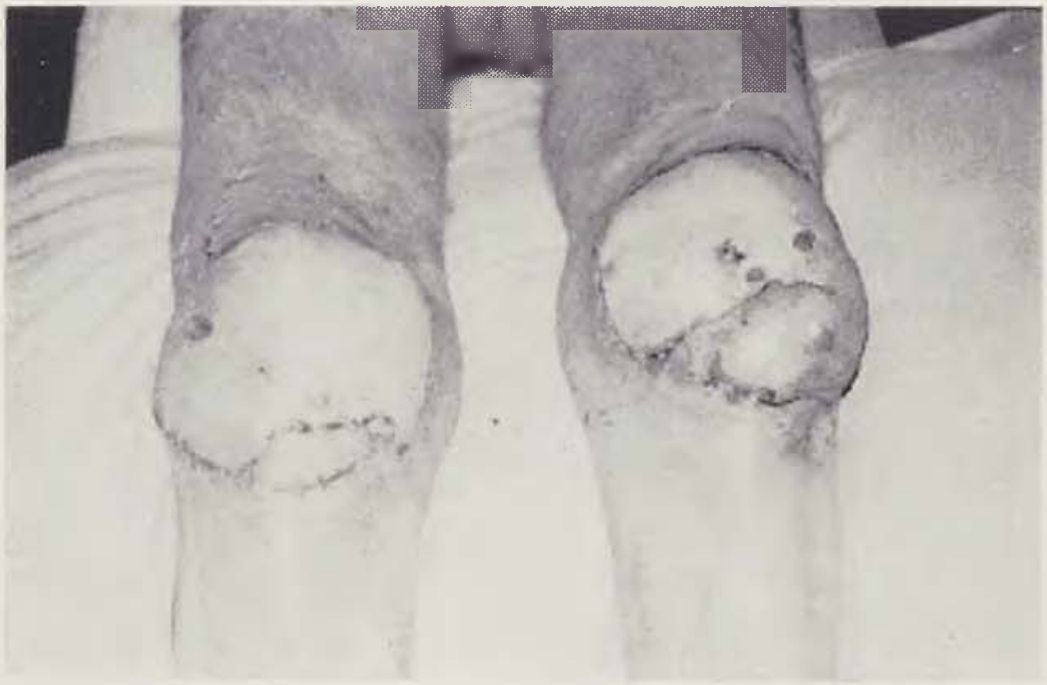

Fig. 3 Early result of treatment

(Fig. 1). The patient had the stable knees, except the avulsion of infrapatellar tendons. After debridement, the infrapatellar tendons were reconstructed with the fascia strips from the fascia lata. The fascia strips were rotated 180° and sutured with steel wire to the distal part of the infrapatellar tendons (Fig. 2). Each of the knee joints were covered with the medial and lateral island muscle flaps from the gastrocnemius muscles. At the same time the muscle flaps were covered with the split skin grafts. After operation the knee joints were immobilized in an open cast for 4 weeks. The flaps healed well, except a small loss of the split skin grafts (Fig. 3). The final result after six months was satisfactory (Fig. 4).

Fig. 4 Late functional result of treatment

DISCUSSION

Gastrocnemiusplasty is the best method for secondary closure of compound injuries of the knee. When the injured areas have been sufficiently debrided, the well vascularized gastrocnemius flaps reconstruction allows a repair of the deep structures in the knee during the one and the same operation. The muscle belly of gastrocnemius will become atrophic later, forming the tissue pad over the knee joint.

SUMMARY

We present a patient with the large bilateral soft-tissue defects on the anterior knees surfaces resulting from a frost-bite. The surgical treatment included: radical debridement of necrotic soft-tissue and bone, reconstruction of the infrapatellar tendons, covering the resulting defects with the medial and lateral island muscles flaps and split-skin graft. The final result was satisfactory.

RESUME

Fermeture d'un vaste défaut bilatéral des tissus moux sur la face antérieure du genou par un lobe libre pris des muscles jumeaux

Podlewski, J., Jankiewicz, L., Opolski, M., Michalowski, K.

On présente un malade avec un vaste défaut bilatérale des tissus moux sur la face antérieure du genou, conséquant d'une engelure. Le traitement chirurgical a comporté l'élimination radicale des nécroses de tissus moux et d'os, la reconstruction du tendon rotulien, le recouvrement du défaut par des lobes médiaux et latéraux prélevés aux jumeaux et la plastie des arrachements cutanés. Le réculta: définitif est satisfaisant.

ZUSAMMENFASSUNG

Das Schliessen eines beiderseitigen ausgedehnten Defekts des weichen Gewebes an der vorderen Oberfläche des Knies durch einen freien Lappen des m. gastrocnemius. Beschreibung eines Falles

Podlewski, J., Jankiewicz, L., Opolski, M., Michalowski, K.

Es wird ein Patient mit einem ausgedehnten bilateralen Defekt der weichen Gewebe an der vorderen Oberflache des Knies als Folge einer Erfrierung vorgestellt. Die chirurgische Behandlung bestand in der radikalen Beseitigung der Reste erfrorener weicher Gewebe und Knochen, in der Rekonstruktion der Sehne unter dem Knie, in der Abdeckung des entstandenen Defekts mit medialen und lateralen Lappen des m. gastrocnemius und in der Transplantation der gespaltenen Haut. Das Endergebnis ist zufriedenstellend.

RESUMEN

Clausura de defecto bilateral extenso de tejidos blandos sobre la superficie anterior de la rodilla por lóbulo libre de m. gastrocnemius. Descripción de un caso

Podlewski, J., Jankiewicz, L., Opolski, M., Michalowski, K.

Está presentado un enfermo con el defecto bilateral extenso de tejidos blandos sobre la superficie anterior de la rodilla en consecuencia del sabañon. El tratamiento quirúrgico concluía la eliminación radical de restos de necrótizados tejidos blandos de huesos, la reconstrucción del tendon de la corva, la clausura del defecto ocasionado por medio de lóbulos mediales y laterales de m. gastrocnemius con transplante de la cutis quebrada. El resultado final es satisfactorio.

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Review

Septo and Rhinoplasty by F. Jeppesen, Viborg, Denmark MUNKSGAARD, International Publishers Ltd, 35, Nörre Sögade, P. O. box 2148, DK-1016 Copenhagen K

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NEW PRINCIPLES OF SURGICAL TREATMENT OF TRAUMATIC TONGUE PARALYSIS

†F. M. KHITROV, A. A. PODKOLZIN, V. M. BEZRUKOV, A. N. LEGOSHIN, V. I. DIDENKO

Issue of surgical treatment of tongue traumatic paralysis is quite current (F. M. Khitrov, 1971 et al.). Disorders of tongue innervation incidence are the subsequence of traumas, neck injuries, post-operative complications, being accompanied by disruption of regional lingual nerves, in particular by sublingual nerves damage (V. I. Grebenyuk and Y. V. Chuprina, 1964).

Clinical picture of given disturbance is quite typical. Tongue mobility loss is followed by chewing and swallowing disturbances, alalia, increased salivation and so on. To swallow the food the patient is obliged to lean his head backwards and to push it into pharynx with finger. Liquid food is swallowed by secondary torturing swallowing motions. In case of unilateral lesion of sublingual nerve the assymetry of tongue is progressing. On the side of lesion it appeares dry. Paralyzed lingual muscles are loosing their bulk in atrophy process, they are getting shorter, as the tongue is deviating to the side of lesion when put forth. In case of bilateral sublingual nerves damage symmetric atrophy is observed, the degree of which is determined by the time expired from the lesion (F. M. Khitrov, 1971; V. V. Kaverina, 1975). Therapeutic treatment of patients of this sort is practically without prospect. The choice of methods of surgical intervention to restore motion function and trophy of paralyzed tongue is conditioned by multiple factors in cases like that. We can choose among them following: general state of patient and the state of denerved tissues of paralyzed tongue to the taken moment, preservation of the respective nerve trunks etc. One of the most substantial factors, determining surgeon's strategy is damage area localization of the respective left or right sublingual nerve.

Mechanical sublingual nerve entirety damages are most frequently identified along its part from the exit of scul cavity up to Pirogoff triangle. It is easier to reveal and separate middle and peripheral end of damaged sublingual nerve for the suture, when the damage is identified before Pirogoff triangle,

as sublingual nerve devides into multiple branches behind it (V. V. Kaverina, 1975; Y. R. Sinelnikov, 1961; Wozniak and Young, 1969). It is not easy to carry out operative interventions anyhow, because of connective tissue growing topographic-anatomic ratio distortion. It makes reinnervation of the respective tissues and organs complicated (E. N. Meshalkin et al., 1964; V. I. Didenko, 1970).

Traditional neurosurgical operations are complicated and sometimes impossible under above stated conditions, as they demand to find out the middle and peripheral end of damaged nerve, to consolidate and fix them to facilitate nerve fibres growth in the initial "casing". When the sublingual nerve damages occure on the distal side of Pirogoff's triangle then it is especially hard.

Viewing all that we propose essentially new modes of disturbed innervation of tongue treatment in these patients: suturing middle end of damaged sublingual nerve into lumen epinervi of lingual nerve in end-to-side fashion. We can advocate the above principle of surgical correction of tongue traumatic paralysis as follows. As anastomosis of sublingual nerve with lingual one gets along quite good in human and mammals, connection of the common sublingual nerve trunk to the lingual one is not antinatural. We can presume, that sublingual nerve will enhance previous anastomosis function and facilitate motion function and trophy of paralyzed tongue restoring, as the lingual nerve is reaching as far as two thirds of tongue. This assumption was corroborated by our subsequent observations.

The second mode is more promissing. Identified middle part of damaged sublingual nerve is sutured to the lumen of the respective lingual artery. Growth of newly formed nerve fibres in arterial lumen under the condition of sufficient oxygenation was revealed to go faster, being orientated free towards the denerved organ. Nerve impulses transmission is getting on from the middle end of sublingual nerve to paralyzed tongue tissues along the wall of "biological casing" in artery and through blood (F. M. Khitrov and A. P. Legoshin, 1977). Given anticipations were corroborated by the following experimental and clinical observations.

Clinical testing was preceeded by experimental investigations in animals. We have studied the outcomes of 20 observations in dogs with unilateral tongue paralysis, performed by section of sublingual nerve in submaxillary area. The second operation was accomplished 3 months later — implantation of before cut sublingual nerve proximal end to respective regional lingual artery. Animals were slottered to carry out pathomorphological research about 3—4 months after the second operation in good state and restored function of tongue.

To carry out pathohistological investigation the specimens of various tongue areas tissues were excised. We were above all interested in the specimen of nerve implantation area into artery. Classical methods of material fixation and colouring were utilized. Special neurohistological modes of sections impregnation with silver were applied besides well tested methods.

Interesting and valuable morphological data were acquired, when investigating "total" sections of tongue root, impregnated with silver nitrite and gold plated in the fashion of Gross-Bilshovski. Preservation of functional, connective,

muscle, epithelium and nerve cells was observed in operated tongue side during the examination (Fig. 1, 2).

Morphological facts found out, are the proof of certain rates of muscle and epithelium tissue cells atrophy and of reducing nerve fibres amount in nerve trunk of operated tongue side. The later is apparently subsequence of the first operation, e. g. section of sublingual nerve and concomitant distal parts of cut nerves Wallerian degeneration.

Fig. 1 Tissues of the right paralysed half of dog tongue, 3 months after operation of the right sublingual nerve section. Expressed signs of atrophy of functional tissue cells of tongue. Colouring with hematoxylin-eosin. Microfotogramm

Our observations are also the evidence of an important fact, that the nerve elements of intraorgan nerve system (neurones I and II of doggel types including their processes and endings in tongue tissues) have not been impaired. Expressed proliferation of glial nerve cells of the proximal end of the cut nerve is detected on nerve implantation into artery site on the background of sclerous changes, forming significant accumulations, and the growth of newly formed nerve fibres, representing prolongation of the cut nerves. Their fine (0,8—1,0 micron in complience with artery orientated symmetrical cylinders, well accepting silver, are identified in intima and subintima layer of artery. Later observations revealed growing nerve fibres also in peripheral parts of operated tongue side, at 2,5—3 cm distally from the nerve implantation into artery.

Thus the outcomes of pathomorphological examinations and their comparison with the data acquired of living testing animals observation are the

evidence of: tissue cells of functional structures and intraorgan nerve system have been preserved when denerved after the first operation and paralyzed half of the tongue of the testing dogs was reinnerved 2 months after the second operation.

Fig. 2 Tissues of the right reinnerved half of dog tongue 3 months after implantation of the middle end of damaged right sublingual nerve into the right lingual artery. Preservation of epithelium, muscle tissue structure of the tongue is identified. Colouring with hematoxylin-eosin. Microfotogramm

Still clinical operation is quite different from the experimental one. This is caused by varied as to level and degree damages of sublingual nerve, furthermore by the certain topographic and anatomic features, as described in essential works of P. K. Anokhin (1935), N. A. Bogoraze (1940), D. Luzha (1973), D. N. Lubotski (1953), A. N. Maksimenko (1963), V. V. Ognev (1960), F. Burian (1967), Hovelacque (1927). Anastomosis of sublingual nerve with the lingual one is purposeful to accomplish on one side and on the other implantation of separated middle end of damaged sublingual nerve (or its fragment) into sublingual artery lumen in case of bilateral sublingual nerve entirely impairment.

The techniques of the above operative intervention are as follows. End of

the found middle part of sublingual nerve, usually with neuroma, is excised with razor in the area of sound tissues. The sign of section haemorrhage can be adopted for the sufficiency index of nerve excise. If there is none, performed nerve cut is insufficient and we have to cut another fragment of nerve. Then the end of nerve is mobilized, neural epithelium is removed cuff like at 3—5 mm proximally from the section level. Submaxillary salinary gland is replaced downwards. We find lingual nerve under the margin of mandibulla. 5 mm of epithelium is dissected longitudinally. Margins of dissected neural epithelium are taken in clamps. Stripped nerve fibres of sublingual nerve are introduced into the formed fissure of neural epithelium, along the lingual nerve. The margins of the later are sutured then to the edges of neural epithelium of sublingual nerve with interrupted sutures in end-to-side fashion (Fig. 3).

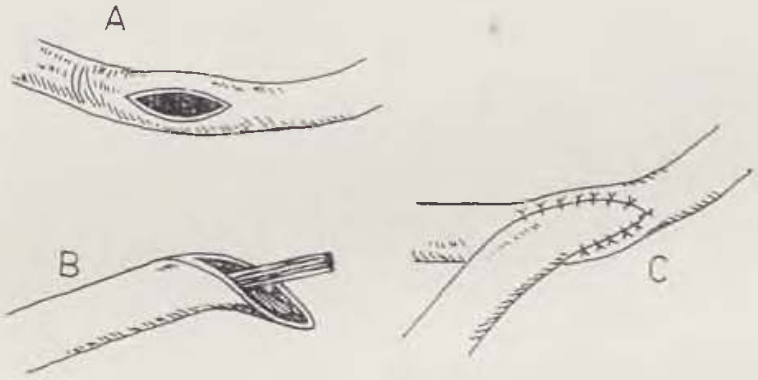

Fig. 3 Scheme. Terminated anastomosis (C) of sublingual nerve end (B) into lumen of neural epithelium of lingual nerve (A) in end-to-side fashion

The end of sublingual nerve implantation into lumen of lingual artery is carried out in stages too. Initially the separated lingual artery is clamped by vascular forcepses placed 1—2 cm from each other. 4—5 mm of artery wall is dissected longitudinally with thin scalpel on this site. The end of sublingual nerve is introduced into the lumen of artery, worked out as described above. Furthermore the margins of neural epithelium are sutured with dissected edges of artery by an intestinal needle applying interrupted sutures in end-to-side fashion. The subsequence is shown in the Figure 4. If the suture is done carefully there is no bleeding when we release clamps. If it may happen the lumen of artery be narrower than the nerve end, we dissect neural epithelium longitudinally at the end of nerve trunk, subdivide it into fibres and introduce merely a part of nerve into dissected vessel to prevent obturation of artery. Then we suture the edges of neural epithelium to the wall of vessel. Remained nerve fibres are placed on adventicius of vascular wall, covered with neural epithelium, common in arteries in this area. In the end the neural epithelium edges are sutured with vessel wall (Fig. 5).

We have observed 5 patients with tongue paralysis of traumatic etiology in ward department of the Central Research Institute of Stomatology. They have

undergone invasive operations of cancer on lower lip, larynx, angioma of the root of tongue, furthermore after gunshot injury and serious trauma of neck. The age of patients fluctuated from 17 up to 65 years (see Table 1). As we can see in the table the revision of sublingual nerves was carried out in two patients (in one with tongue paralysis after gunshot and in the patient, who has undergone angioma radix linguae excision). The full interruption of sublingual nerves was identified in both of them in the area before Pirogoff triangle. Excision of nerves was assumed to carry out, suture of their ends on one side and neurolysis of the opposite nerve trunk. The end of middle sub-

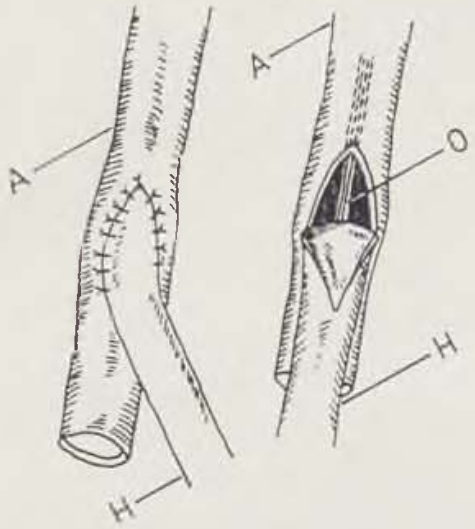

Fig. 4 Scheme of terminated nerve-vascular anastomosis in end-to-side fashion introducing sublingual nerve into the lumen of lingual artery
A — artery, H — nerve, O — axial cylinders of sublingual nerve

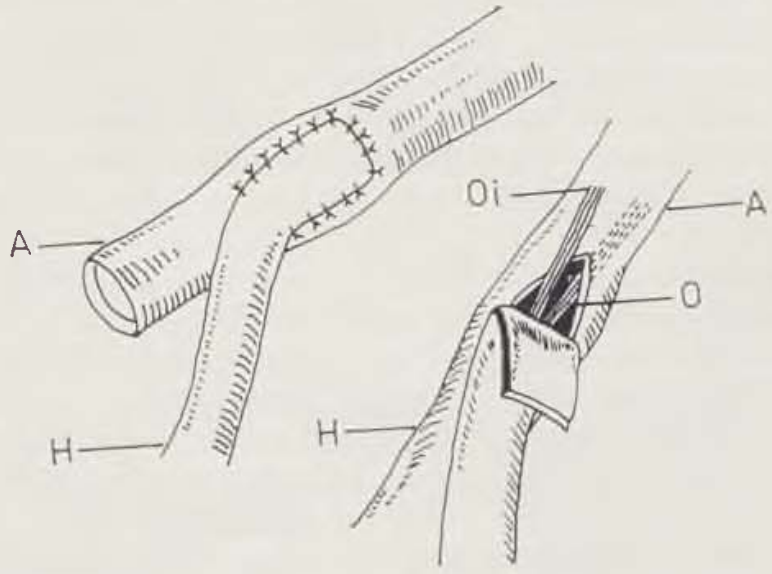

Fig. 5 Scheme of terminated neuro-vascular anastomosis in end-to-side fashion
A — artery, H — nerve, O — part of sublingual nerve, introduced into artery, 01 — part of sublingual nerve, fixed to the adventicius of artery

Table 1. Data of clinical observations

Patient No.	Age	Causes of tongue paralysis	Time in years	Mode of operative intervention	Results of treatment (since one year after operation)
1	42	Consequence of gun shot neck injury	1/2	Excision of nerves, suture of one sublingual nerve ends and neurolysis of the opposite one	Partial improvement
2	17	After excision of angioma of radix linguae	1 1/2	Excision of nerve ends and their suture	Partial improvement
3	53	After invasive operation of lower lip cancer in Vanakh fashion	3	Implating middle nerve end of damaged left sublingual nerve into the sublingual left artery, anastomosis of the right sublingual nerve with the right lingual nerve	Significant improvement
4	34	Consequence of neck trauma	1	The same	Significant improvement
5	65	Radical operation of larynx cancer	4	The same	Significant improvement

Note. All the patients were males, with bilateral tongue paralysis.

lingual nerve fragment on one side was implanted in lingual artery lumen, and on the opposite side the middle fragment end of sublingual nerve was connected with lingual nerve in accord with above described methodology.

We present one case report of the above observations.

Patient E., 53 years old was examined in maxillary-facial surgery department of the Central Research Institute of Stomatology from February 2 until April 4, 1974. He suffered from traumatic tongue paralysis. Condition after operation.

Anamnesis. Radical operation of lower lip cancer was carried out, with concomitant bilateral tongue paralysis on January 4, 1969. Operation of tongue reinnervation was performed on December 1, 1973. The middle end of the left sublingual nerve was implanted into the left lingual artery, and the middle end of the right sublingual nerve was anastomosed with the right lingual nerve during the later operative intervention under the general intratracheal anasthesia.

Inspecting the patient after a year no complains occured, talking ability was fully restored. Tongue motions without pathological signs. Atrophical tongue changes were visually insuspicious. Electric-miographical examination revealed, the positive outcome of surgical treatment occured mostly due to

reinnervation of the half of tongue, where middle sublingual nerve fragment was implanted onto respective artery trunk of the left lingual artery (Fig. 6 and 7). Due to topographical features of blood supply we have observed the growth of regenerated axons of sublingual nerve also in the right half of a

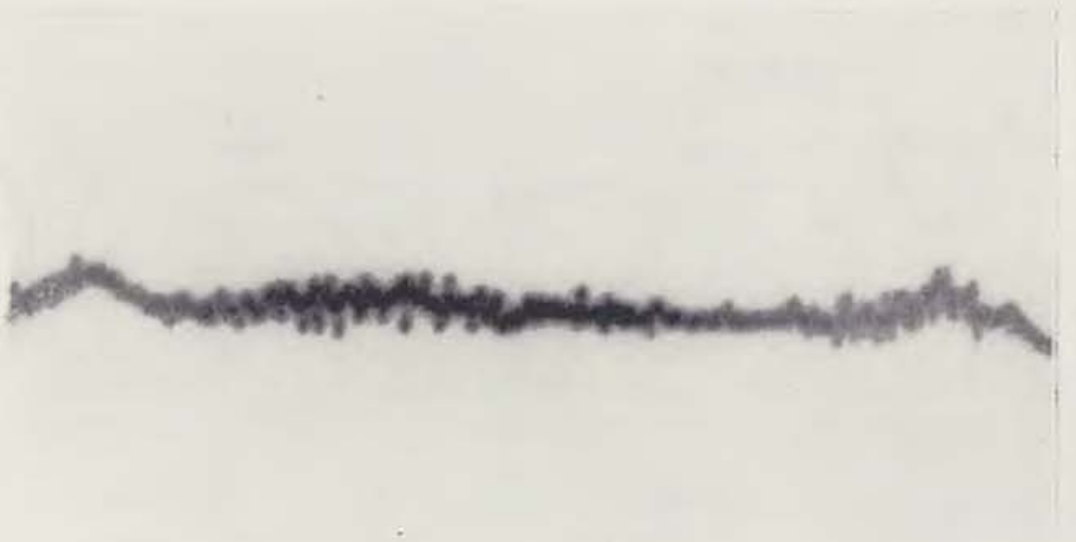

Fig. 6 EMG of the right half of the tongue of the patient E. before operation. We observe entire missing of bioelectrical potentials, sign of tongue muscles paralysis

Fig. 7 EMG of the left half of the tongue of the same patient one year after operation of tongue in middle end of damaged sublingual nerve implanting into respective lingual artery fashion. Group biopotentials are generated, it is the sign of bioelectrical tongue activity recovery

tongue. Biopotentials EMG arising simultaneously in the right half of the tongue with those in the left one were evidence of it.

The sensitivity reoccurence, caumesthesia and also cloning twitching were observed in the given patient on the second day after operation.

Our clinical experience enables to suppose the proposed method of implanting middle end of damaged sublingual nerve into respective lingual artery lumen to be optimal in the repertoire of methods to correct traumatic paralysis of tongue.

Experimental data analysis and the first successful outcomes of clinical observations give us the chance to take the proposed new treatment principles of traumatic tongue paralysis caused by sublingual nerves damages to be purposeful. In particular the strategy of middle end of damaged nerve implanting into respective sublingual artery. They enable to solve effective the issue of motion function and trophy of paralysed tongue restoration.

Viewing the successful clinical implementation of the new methodology of tongue paralysis treatment, we recommend to apply it generally in the practice of reconstructive surgery and transplantology.

SUMMARY

The new principles of surgical correction of traumatic tongue paralysis are presented. One of them represents anastomosing the middle end of damaged sublingual nerve and the respective lingual artery. The second one — anastomosing middle end of damaged sublingual nerve with the respective lingual nerve.

The application of the above fairly new method of treatment instead of the old ones, the first mode in particular, guarantees manding of function and trophy of paralysed tongue.

Conclusions are based on objective data of 20 experimental and 5 clinical observations.

The new approach to the tissue of tongue reinnervation was successfully implemented in clinical practice. The experience acquired enables to predict the possibility of its wide application in the practice of reconstructive surgery and organ transplantation.

RESUME

Justification de nouvelles méthodes du traitement chirurgical de la paralysie de langue posttraumatique

Khitrov, F. M., Podkolzin, A. A., Bezrukov, V. M., Legochin, A. N., Didenko, V. I.

On apporte une justification de l'utilisation de nouvelles méthodes des corrections chirurgicales de la paralysie posttraumatique de langue. L'une des méthodes consiste en la création de l'anastomose entre le bout médian du nerf hypoglosse endommagé avec l'artère de langue correspondante, l'autre est fondée sur l'anastomose du bout médian du nerf hypoglosse avec le nerf de langue correspondant.

Ces nouvelles méthodes de traitement se distinguent par une nouvelle qualité, particulièrement en comparaison avec les méthodes traditionnelles. Les nouvelles méthodes, surtout la première, rendent possible un optimal renouvellement des fonctions de la langue paralysée.

La conclusion est basée sur les données objectifs, obtenus au cours de 20 observations expérimentales et de 5 observations cliniques.

Les expériences avec l'introduction heureuse des nouvelles méthodes dans la pratique clinique, qui représentent une nouvelle solution du problème de renouvellement du système nerveux de langue, permettent prévoir les possibilités ouvertes aux nouvelles méthodes dans la pratique de la chirurgie réparatrice.

ZUSAMMENFASSUNG

Begründung der neuen Methoden chirurgischer Behandlung einer Zungenparalyse nach einem Unfall

Khitrow, F. M., Podkolzin, A. A., Bezrukow, V. M., Legoschin, A. N., Didenko, V. I.

Es wird die Anwendung neuer Methoden der chirurgischen Korrektion einer Zungenparalyse nach einem Unfall begründet. Eine dieser Methoden ist die Bildung einer Anastomose zwischen dem mittleren Ende des beschädigten Nervs unter der Zunge und der entsprechenden Zungenarterie. Eine andere besteht in Anastomosieren des mittleren Endes des beschädigten Nervs unter der Zunge mit dem entsprechenden Zungennerv.

Die Anwendung dieser qualitativ neuen Methoden der Behandlung anstatt der traditionellen Methoden, insbesonders die Anwendung der erstgenannten Methode, gestatten die optimale Erneuerung der Funktion der paralysierten Zunge.

Diese Schlussfolgerungen gründen sich auf die objektiven Angaben aus 20 experimentellen und 5 klinischen Beobachtungen.

Die Erfahrungen mit der erfolgreichen Einführung der neuen Methode bei der Lösung des Problems einer Erneuerung des Nervensystems der Zunge in die klinische Praxis gestatten es, die Möglichkeit ihrer Verbreitung in der Praxis der rekonstruktiven Chirurgie und Organtransplantation vorauszusehen.

RESUMEN

Motivación de nuevos métodos de tratamiento quirúrgico de parálisis post-traumática de lengua

Khitrov, F. M., Podcolzin, A. A., Bezrukov, V. M., Legochin, A. N., Didenko, V. I.

Se presenta la motivación para el empleo de nuevos métodos en la corrección de parálisis post-traumática de la lengua. Uno de ellos consiste en la formación de anastomósis entre el extremo central del nervio lingual inferior afectado y la arteria lingual correspondiente; el otro consiste en la anastomósis del extremo central del nervio lingual inferior afectado con el correspondiente nervio lingual.

El empleo de estos métodos de tratamiento, cualitativamente nuevos, en lugar de los tradicionales, posibilita, especialmente con el primer método, lograr una renovación de la función de la lengua paralizada.

Las conclusiones están basadas en datos objetivos obtenidos del estudio de 20 casos experimentales y 5 clínicos.

Las experiencias logradas con la aplicación exitosa del nuevo método en la solución del problema de renovar el sistema nervioso de la lengua en la práctica clínica, permite pronosticar que existen posibilidades de que se amplie su uso en la práctica de la cirugía reconstructiva y en el transplante de órganos.

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THE USE OF LASER IN SURGICAL SPECIALTIES

V. SEKAN, M. BROZMAN, J. FEDELEŠ, L. KOŠŤÁL, Š. ZBOJA

Once the properties of laser had become known a multitude of experimental studies appeared to show the scope of its potential uses in technology and medicine. There was hardly a single area where laser had not been tested. Today it is obvious that it is well under way on its triumphant course.

Ophthalmology became the first branch of medicine where laser techniques proved their worth. On passing through the eye, light energy changes into thermal energy and, unless the value of 60 °C is exceeded, coagulation necrosis develops on the retina. Direct laser radiation can be used to destroy tumours (haemangiomas, naevi, retinoblastomas), foci of inflammation on the retina and the choroid. Laser is used for iridotomy, trabeculotomy, for the treatment of glaucoma, in cases of detached retina.

As a result of the good experience of laser work in ophthalmology, laser scalpels — CO_2 laser — are coming into use. This amounts to the exploitation of the known properties of laser radiation. Let us give the characteristics of some of the advantages of laser scalpel employment:

1. Sharply delineated incision of fine edges with no need to bring the cutting instrument into direct contact with the tissue surface. The layers of cells destroyed by thermocoagulation necrosis are extremely narrow and, with a few exceptions, remain confined to the target focus of the laser beam.

2. Capillary bleeding is totally suppressed as a result of microthrombotization in the small vessels, while arterial bleeding, if present, can be arrested by repeated coagulation using focussed or even unfocussed beams of radiation.

3. The depth of incision can be regulated, depending on the speed of cutting, on laser output changeability, on focus shifting.

4. The wound heal very quickly and well with no major fibrosis developing in the neighbourhood; the results are comparable to the conventional scalpel with reasonable resistance to the usual types of infection.

5. Laser permits micro-operations unmanageable with conventional instruments, a fact of importance mainly for microsurgical operations.

6. Absolute sterility of incision is another major advantage.

7. Wound resistance to secondary infection is lower only in cases of massive inoculation of the fresh wound; laser radiation has a sterilizing effect on infected wounds.

Surgery

Relatively successful experiments with CO_2 laser in primates (Goldman, 1967) and dogs (Náprstek, 1971; Hall, 1971) prompted the idea of using the laser scalpel for liver resection. Many authors envisage its future use mainly in oncological conditions of the liver with the patient being free of the danger of intraabdominal haemorrhage, with a strong enough fibrous scar developing in the wound, and with the liver enzymes activity in the vicinity of the incision steadily growing normal.

Skobelkin (1979) reported the laser scalpel potential uses in alimentary tract surgery (resection of the stomach, hemicolectomy, coagulation of bleeding gastric ulcer).

In the years 1979—1980, Tomšů was able to avail himself of the first Czechoslovak device of the CO_2 type. His comparative studies corroborated literary reports on non-anatomical resections of the hepar (reduced blood loss, shorter time of operation). He made incisions of the skin, subcutis, muscle, cartilage and vessels, non-anatomical resections of the liver, lungs, stomach, the small and large intestines without prior preparation of the intestinal tract, using single-layer atraumatic suture. There was not a single case of anastomosis dehiscence or skin infection. On the basis of those favourable experimental results he ventured mastectomy in 1981.

According to Tomšů, CO_2 laser is indicated in the following situations:

1. wherever major blood loss is feared
2. in oncology — to prevent tomour cells spreading to the deeper parts or penetrating into the lymphatic and venous circulation
3. the need to excise or evaporate necrotic tissue.

Another type of surgical laser, the Neodym — YAG apparatus, used in the surgery of the digestive tract and the urinary organs has great potential uses ahead of it since, using fibrooptical means, its beam can be introduced into the body cavities.

Cochrane (1980) made large-intestine anastomosis in 75 rats following intestinal discission with the conventional scalpel, the laser scalpel, and electrocautery. On the 7th post-operative day he examined the anastomosis for strength and patency. As for the former, there were no significant differences between the three types of intestine discission. The classical scalpel and laser produced about the same patency, only the electrocautery resulted in lumen stenotization and reduced thickness of the intestinal wall.

Urology

Khromov (1969) drew attention to the fact that following laser incision of the kidney the healing process does not produce full-value cicatrical tissue,

that post-operative bleeding is minimal, and that the adjacent parenchyma is intact. Meiraz (1977) made experimental partial nephrectomy in the cat with very good effects. The results were checked by angiographical and histological means. Mulwaney and Beck (1968) studied laser beam effects on urinary concrements in the bladder and kidneys. Exposed to laser radiation the concrements developed cavities inside subsequently to be broken up. Ultimately, they became disintegrated and evaporated. Hofstetter (1979) employed a laser urethrocystoscope in neoplastic diseases of the urinary bladder.

In other words, urology makes use of laser for kidney pole resection, for purposes of urological oncology, for urethral strictures, for dealing with urinary concrements in hollow organs.

E. N. T.

Of late, some authors have studied the potential uses of laser in otorhinolaryngology. Laser beams are likewise channelled endoscopically. This method is used in combination with microsurgical techniques. For upper respiratory tract work, medical engineers have developed special endotracheal cannules while anaesthesia is effected solely through the intravenous channels (explosive gases).

Strong (1972) used continual CO_2 laser for laryngeal surgery to remove nodes from the vocal cords, cysts, keratoses, carcinomas and papillomas. The results were remarkable in all the cases. Laforet (1976) published a report on laser resection of the trachea for carcinoma. Laser bronchoscopy in combination with intrabronchial surgery represents yet another potential use of laser radiation.

No doubt, malignant tumours will remain the chief area of indication for laser, though the technique has also been used for the treatment of haemorrhagic teleangiectasis of the nasal mucosa. There are some new prospective uses of laser for the treatment of congenital subglottal haemangioma, for the preservation of hearing by means of cochlear electrode implantation, for microneural surgery in hemifacial spasm, and for reinnervation of the paralyzed larynx (Tomšů, 1982).

B. Healy (1979) reports on a large group of diseased children where he used CO_2 laser in operations on the respiratory passages for such diagnoses as: choanal atresia, papilloma of the mouth and nose, teleangiectasis, lingual tonsillectomy, tonsillectomy, laryngeal papilloma, neurofibroma, subglottal haemangioma, subglottal stenosis including the congenital variety, laryngeal lymphoedema, vocal cords granuloma and nodules, tracheal papilloma, stenosis and granuloma, in other words, a really broad field of indications.

Gynaecology

Gynaecology, too, has included laser techniques in its instrumentation. Demonstration of spermatozoal motility in depolarized light, cervical lesions, neoplasms, carcinoma of the vulva and vagina, perineal condylomatosis, as well as

oncosurgical operations from the laparotomical approach — all these open up new possibilities for laser surgery. As a recent addition, studies have appeared on laparoscopic operations on the Fallopian tubes using laser techniques (Tomšů, 1982).

Neurosurgery

In neurosurgical practice, laser treatment is used for epilepsy, for selective hypophysectomy and for nervous pathway transsection for excruciating pain alleviation. There are good prospects for the treatment of intracranial tumours, especially for the absence of haemorrhage and for the possibility of breaking up the tumour (stereotaxy).

Oncology

Tissue destruction is the basic principle of the favourable effect of laser energy in oncology. Very often, a single flash of adequate laser energy will do to destroy the tumour. The assumption is that each tumour requires "its own" wavelength and radiation energy. In what is an advantage over X-rays, ionizing radiation, cytostatics and chemotherapeutics, the laser bean can be focused precisely on the area of indication. The neighbouring tissues sustain only minimal damage or none at all. The depth of laser beam action depends on tissue thickness and on the scope of the surgeon's ability to keep the operating field under visual control, so that the uses of laser are limited to superficial tumours or to tumours inside cavities or lumina. Because of their high pigment content, malignant melanoma cells exhibit the greatest degree of affinity to laser radiation. The explosive power of a laser radiation impulse can drive isolated tumour cells as deep as 1.5 cm into the neighbouring tissues. Dissemination into the air can go as far as 2.5 metres (Ketcham, 1970). Hence why it is essential to use high-energy radiation to effect progressive necrosis of the whole of the tumour mass spreading along with coagulation necrosis from the site of the original impact. Where high-energy radiation is unavailable (1000 J/cm^2) the tumour can be destroyed by repeated impacts of lower energy.

The oncolytic effect of laser radiation can be enhanced by the simultaneous application of more therapeutical methods. Chemotherapy and ionizing radiation therapy are well suited for the purpose. Many studies have been published recently on the favourable effect of laser beams on malignant melanoma, on malignant melanoma metastases, on pigmented as well as non-pigmented basalioma, on squamocellular carcinoma, epithelioma, on breast cancer, on sarcoma as well as lymphoma. Vascular tumours — malignant as well as benign ones — also represent a vast field of indications.

SUMMARY

After the properties of laser became known, a number of experimental and clinical studies appeared to explore the scope for its uses in medicine. The first to make use of laser techniques was eye surgery soon to be

followed by other surgical disciplines — general surgery, urology, E.N.T. surgery, gynaecology, plastic surgery, oncology, neurosurgery. The use of the laser scalpel in all the surgical specialties has a common denominator in its undisputed advantages: well-defined incision with no direct contact between the tool and the surface of the tissue; capillary and arterial bleeding can be laser-handled unless large vessels are involved; the wounds heal soon and well; there is sterility of incision; wound resistance to secondary infection is worse only in case of massive inoculation of the fresh wound.

RESUME

L'utilisation du laser dans les spécialités chirurgicales de la médecine

Sekan, V., Brozman, M., Fedeleš, J., Košťál, L., Zboja, Š.

Quand on a connu le fonctionnement du laser, un grand nombre de travaux expérimentaux et cliniques ont surgi qui avaient pour but de découvrir les possibilités pour s'en servir en médecine.

La première spécialité médicale, où on a adopté la technique de laser, a été l'ophtalmologie et, désormais, d'autres disciplines chirurgicales ont suivi: chirurgie générale, urologie, O.R.L., gynécologie, chirurgie plastique, cancérologie, neurochirurgie.

Le dénominateur commun d'application du laser dans les disciplines chirurgicales sont ses avantages incontestables: l'incision aux bords fermes, l'absence de contact du bistouri avec la superficie du tissu. L'hémorragie capillaire ou artérielle peut être maitriseée par la technique de laser, s'il ne s'agit pas d'artères à grand lumen. La guérison des plaies est rapide et bonne, l'incision stérile. L'endurence des plaies envers l'infection secondaire est moins forte uniquement au cas où une massive inoculation de la plaie fraîche soit présente.

ZUSAMMENFASSUNG

Die Anwendung von Laser im chirurgischen Fachgebiet der Medizin

Sekan, V., Brozman, M., Fedeleš, J., Košťál, L., Zboja, Š.

Als die Eigenschaften der Laser bekannt wurden, tauchte eine Menge experimentaler und klinischer Arbeiten auf, die sich mit den Möglichkeiten seiner Ausnutzung in der Medizin befassten.

Das erste medizinische Fachgebiet, in dem die Laser-Technik angewendet wurde, war die Ophthalmologie, dann kamen hinzu die chirurgischen Disziplinen — die allgemeine Chirurgie, die Urologie, die Ophthorhinolaryngologie, die Gynäkologie, die plastische Chirurgie, die Onkologie, die Neurochirurgie. Der gemeinsame Nenner der Anwendung des Laser-Skalpells in den chirurgischen Disziplinen sind seine unzweifelhaften Vorteile und zwar: scharf begrenzte Inzisionen, kein direkter Kontakt des Schneideinstruments mit der Oberfläche des Gewebes, Bewältigung von Gefäss- und Schlagaderblutungen durch den Laser, sofern es sich nicht um Gefässe von grösserem Lumen handelt, raschere und bessere Verheilung von Wunden, sterile Inzisionen, Widerstandsfähigkeit der Wunden gegen sekundäre Infektionen, nur dann schlimmer, wenn es sich um eine massive Inokkulation eines frischen Wunde handelt.

RESUMEN

El empleo del laser en los ramos de cirugía de medicina

Sekan, V., Brozman, M., Fedeleš, J., Košťál, L., Zboja, Š.

Al reconocer las propiedades de laser aparecieron numerosos trabajos experimentales y clínicas, consagrados a su uso en la medicina.

Por primer ramo de medicina, en el cual emplearon la técnica de laser, fué la medicina oftálmica, después se añadieron las disciplinas de cirugía — la cirugía general, urología, otorrinolaringología, ginecología, la cirugía plástica, oncología, neurocirugía. Por un denominador mútuo del uso de escalpelo de laser en las disciplinas de cirugía son sus ventajas indiscutibles, a saber: una incisión agudamente limitada, sin contacto directo del instrumento cortante con la superficie del tejido, hemorragia capilar y arterial está dominada por laser, si no se trata de vasos de más grande lumen, las heridas se curan más rápidamente y bien, la incisión es estéril, la resistencia de las heridas hacia la infección secundaria es más peor solamente en casos de la inoculación masiva de la herida fresca.

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IN MEMORY OF LENIN PRIZE LAUREATE PROFESSOR FEDOR MIKHAILOVICH KHITROV

On June 1, 1986, the Soviet medical science suffered a great loss. After a serious and protracted disease, Professor Fedor Mikhailovich Khitrov, Doctor of Medical Sciences, member of the Communist Party of the Soviet Union since 1955, Lenin Prize Laureate, merited scientist of the Russian Soviet Federal Socialist Republic, died aged 84.

He devoted almost 60 years of his life to the development of one of the most important branches of medical science — reconstructive and plastic surgery. Death interrupted the many-sided activity of the scientist and surgeon introducing into practice modern achievements of science and technology.

Fedor Mikhailovich Khitrov was born in 1903. In 1927 he graduated from the Medical Faculty of the North-Caucasian University and entered general practice in Grozny. He specialized in surgery under Professor N. I. Napalkov. Starting from 1932, he worked in the Central Institute of T. O. under the guidance of the famous surgeon, Prof. A. E. Rauer, the founder of plastic surgery.

In 1946, F. M. Khitrov headed the clinical department of jaw and face surgery of CITO which in 1962 was included in the Central Research Institute of Stomatology, Ministry of Health of the USSR.

F. M. Khitrov is the author of more than 120 scientific works on the pro-

blems of surgery, primarily the surgery of the head and the neck (processing and plastic surgery in skull injuries, free transplantation of the skin, transplantation of nerves and muscles). Of special importance in his activities were his studies on the problems of reconstructive surgery of the face and the neck. In 1949, F. M. Khitrov defended his doctoral thesis devoted to plastic surgery of the nose after gunshot injuries using Filatov's graft.

Fedor Mikhailovich and his disciples elaborated a considerable number of new surgical methods of plastic operations on congenital and acquired defects of the face using adjacent tissues and Filatov's graft (pedicle). His works are the original studies concerned with rhinoplasty, formation of the pharyngeal and faucial entrance using a T-shaped pedicle, creation of the pharyngeal skeleton from homocartilage, formation of faucial and esophageal stoma by means of a "skin tube". He was the first to accomplish reconstruction of the alimentary and respiratory passages in cases of complete separation of the larynx, pharynx and esophagus, to improve the methods of surgical treatment of patients with congenital cleft lip and palate; he grounded the indications to fragmentary osteotomy in combination with corticotomy during the correction of serious deformities of the upper jaw. F. M. Khitrov was one of the first to start elaboration and the use in practice of the method of treating patients with paralysis of the tongue focused on the restoration of its mobility by transferring to another place the central end of the sublingual nerve.

F. M. Khitrov mastered perfectly the most complicated methods of reconstructive treatment based on an original system of surgical approaches, on precise knowledge of surgical and topographic anatomy, on the know-how of analysing defects and deformities, on the selection of purposeful succession, the proportions and the terms of the operations carried out.

For his monograph "Plastic replacement of defects of the face and the neck using Filatov's pedicle", Fedor Mikhailovich was awarded the S. I. Spasokukotskii prize in 1955. F. M. Khitrov's most significant achievement are his studies collected in the monograph "Defects and cicatricial overgrowings of the fauces, the cervical portion of the esophagus, the larynx and the trachea, and the methods of their elimination" for which the author was awarded the Lenin prize.

In 1984, the "Atlas of plastic surgery of face and neck" was edited by F. M. Khitrov and published.

Fedor Mikhailovich Khitrov took an active part in the work of the editorial board of the journal "Stomatologiya", was editor of the journal "Acta Chirurgiae plasticae" and editor of the section "Stomatologiya" in BME.

His country greatly appreciated the merits of F. M. Khitrov by awarding him two orders and medals.

Fedor Mikhailovich devoted all his talent to the education of young medical doctors, to the training of scientific workers. His unresting energy has been and will continue to be an example for the young.

The good memory of Fedor Mikhailovich Khitrov, a man of great mental strength, unusual diligence and energy, will forever survive in the hearts of his pupils, companions and colleagues.

NEWS

7th Joint Congress of the Asian and Pacific Federations of the International College of Surgeons

will be held in Taipei, Taiwan, from November 3—7, 1987.

Main themes of symposia are:

— anesthesiology
— cardiovascular surgery
— general surgery
— gynecology and obstetrics
— neurosurgery
— ophthalmology
— orthopedics
— otolaryngology
— pediatric surgery
— plastic surgery
— thoracic surgery
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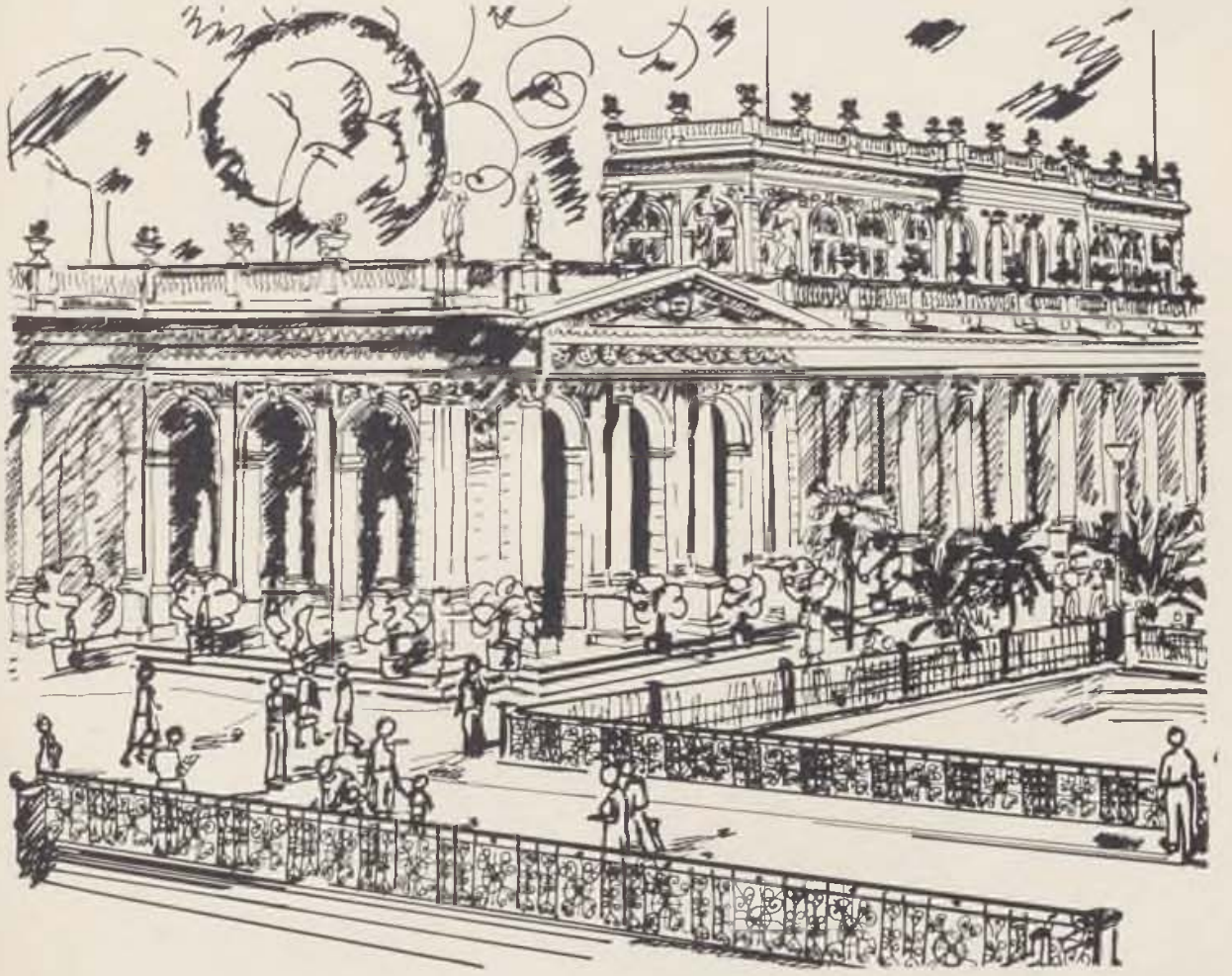
CONTENTS

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 THING 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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