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THE VALUE OF VASCULARIZED TENDON TRANSFERS WITH FREE FLAPS IN PERIORAL AND CHEEK RECONSTRUCTIONS

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SUMMARY

The effect of gravity and the process of ageing neccesitate the support of flaps used for the reconstruction of facial tissues. Perioral and cheek reconstructions exemplify this problem.

Support can be obtained using vascularized tendons with free tissue transfers. The radial forearm neurocutaneous flap with the vascularized palmaris longus tendon and the neurocutaneous lateral arm flap with the vascularized triceps tendon are useful options for resurfacing of these defects.

We present 6 cases to highlight the use of the vascularized tendon transfer with free neurocutaneous tissue transfers as composite flaps in perioral and cheek reconstructions.

ZUSAMMENFASSUNG

Die Bedeutung der vaskulisierten Sehnen als Teil der freien Lappen bei der Perioralen- und Wangenrekonstruktion

Cunha-Gomes D., Kavarana N.

Der Einfluß der Schwere (Gravitation) und der Veralterungsprozeß verlangen notwendig die Unterstützung der bei der Rekonstruktion von Gesichtsgewebe angewandten Lappen. Als Beispiel dieses Problems dienen die Perioralen- und Gesichtsrekonstruktionen.

Die unbedingte Unterstürzung kann man durch die Anwendung der vaskulisierten Sehnen als Teil der frei übertragenen Lappen gewinnen. Der neurokutanne Lappen aus der radialen Seite des Unterarms mit der vaskulisierten Sehne m. palmaris longus und der neurokutanne Lappen aus der lateralen Seite des Arms mit der vaskulisierten Sehne des Triceps stellen die geeignete Möglichkeit für die Rekonstruktion dieser Defekte dar.

Die Autoren führen 6 Beispiele ein, die die Anwendung der vaskulisierten Sehne als Teil des freien neurokutannen kombinierten Lappens bei der Perioralen- und Gesichtsrekonstruktionen aufheben.

Key words: vascularized tendon transfer, perioral reconstructions

Areas of the face like the angle of the mouth and the lower eyelid or the infraorbital unit of the cheek, require support against the effect of gravity. Unsupported, the angle of the mouth descends lower than the opposite side marring the symmetry of the face. Also, the loss of the orbicularis oris muscle continuity manifests functionally as drooling from the angle of the mouth.

The cheek unit sans support hangs down, especially in the region of the nasolabial fold. Ectropion of the lower eyelid results in scleral show and epiphora.

In order to prevent this a vascularised tendon transfer with free flaps has been used to provide support for such reconstructions.

REPRESENTATIVE CASES

Case 1: A 28-year-old male, tobacco and betel nut chewer, required excision of a well-differentiated squamous cell carcinoma involving the rt. angle of the mouth, buccal mucosa and cheek skin (Fig. 1).

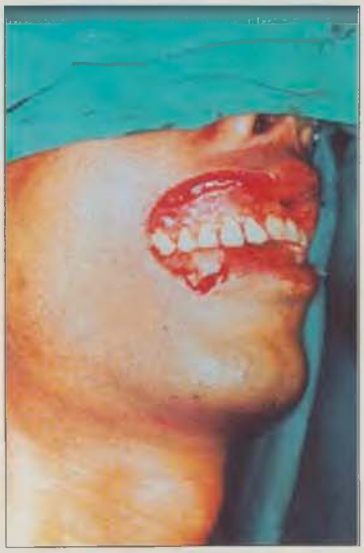

Fig. 1. Case 1: Defect resulting from excision of rt. angle of mouth and buccal mucosa.

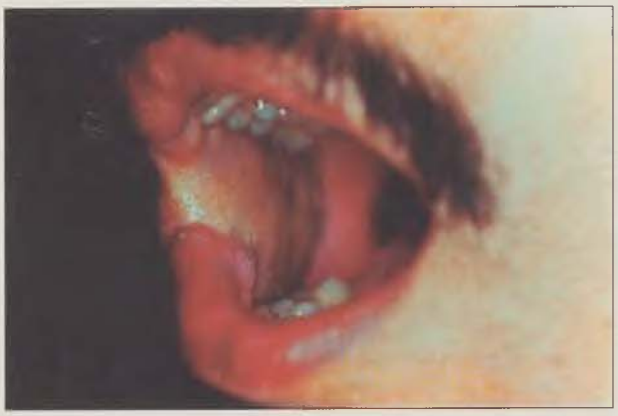

Fig. 2. Case 1: Post-operative intraoral view.

A radial forearm – palmaris longus (PL) tendon composite transfer of 9 x 4.5 cms was harvested. After the vascular anastomosis was completed and the flap revascularized, the lateral cutaneous nerve of the forearm was coapted to the greater auricular nerve in the neck. The palmaris longus tendon was tunnelled through the upper and lower lips and sutured to the cut edges of the orbicularis muscle.

Post-operatively, the flap settled down well, though the PL sling appeared to be slack. This resulted in drooling of fluids from the reconstructed angle of the mouth. 6 weeks after the first surgery, under local anaesthesia the PL tendon was reefed and tightened through small incisions at the junction of the flap and the upper and lower lips. Adequate mouth opening and oral competance was tested.

The patient, now 24 months post-operative, has a well-settled flap with good support and competence of the oral sphincter (Fig. 2).

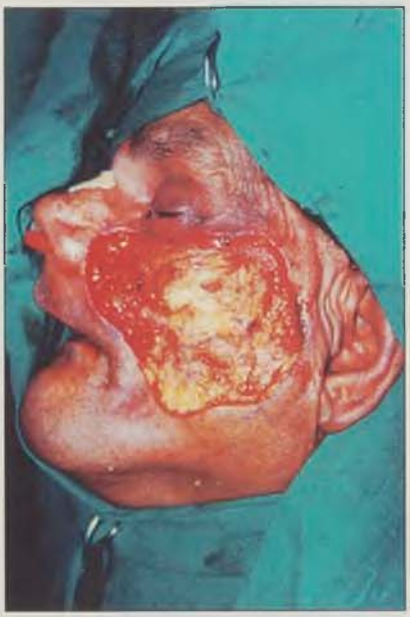

Fig. 3. Case 2: Excision of squamous cell carcinoma over the cheek.

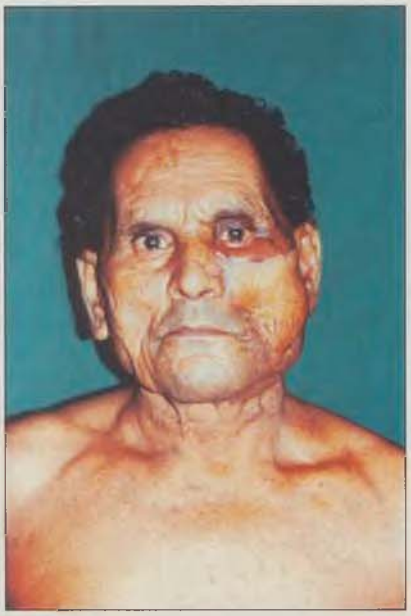

Fig. 4. Case 2: Post-operative – front view.

Case 2: A 64-year-old male presented with a lesion over the lt. cheek, shown on biopsy to be a squamous cell carcinoma (Fig. 3).

A neurocutaneous radial forearm – PL tendon composite transfer of 12 x 10 cms was raised, transferred and vascularized. The flap was planned so that the PL tendon was situated transversely in the cranial part of the flap and was used to pre-

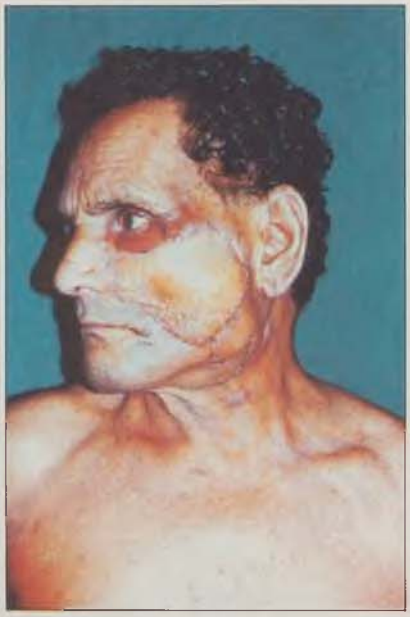

Fig. 5. Case 2: Post-operative – lateral view.

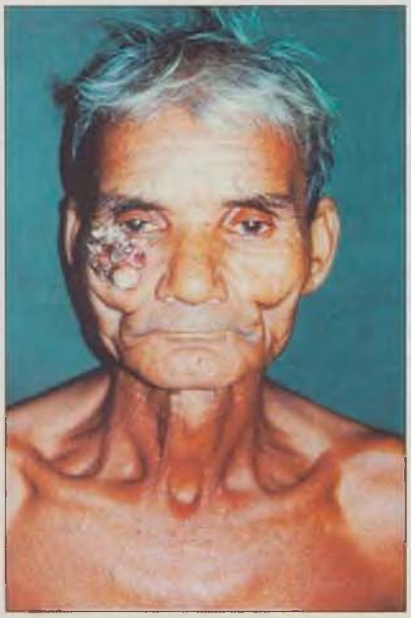

Fig. 6. Case 3: Pre-operative view.

Fig. 7. Case 3: Post-operative view.

vent its sagging, thereby avoiding ectropion of the lower eyelid.

The tendon was tunnelled medially and hitched to the periosteum of the nasal bone. Laterally, it was sutured to the deep temporal fascia. After 14 months the flap and the lower eyelid appear to be well supported (Figs 4, 5).

Case 3: A 63-year-old male patient had a squamous cell carcinoma over the infraorbital region of the lt. cheek (Fig. 6). An excision of the lesion resulted in a 6 x 9 cm defect. The lower eyelid was essentially intact except for the loss of the lower orbital fibres of the orbicularis occuli.

A lateral arm flap with vascularized triceps tendon was used for resurfacing of the defect. The tendon was positioned in the upper edge of the flap in order to support the flap as well as to prevent downward displacement of the lower eyelid.

A thrombus at the venous anastomosis required a re-exploration and revision of the anastomosis. The flap settled down well except for hyperpigmentation (Fig. 7).

DISCUSSION

Perioral and cheek defects essentially require soft, supple and sensate skin and mucosal replacement. Flaps transferred to these defects are adversely affected by the force of gravity as well as the effect of ageing. This results in sagging of the flap as well as distortion of landmarks adjoining the flap. This is especially evident at the angles of the mouth, the base of the nostril and the lower eyelid.

Support for the transferred flap is essential in order to counter sagging of the flap. This can be obtained if the flap is fixed to the underlying bone or to functioning muscle units.

The composite neurocutaneous radial forearm – palmaris longus tendon free flap and the lateral arm – triceps tendon composite free transfer are good options for these defects. They transfer sensate, durable, supple vascularized skin in a single stage (1, 2).

The PL tendon, though present in about 85 % of the population, has been used widely in tendon replacement and other hand surgeries. As a vascularized transfer, with the free radial forearm flap, the viability of the tendomuscular unit is enhanced. It is enclosed within the fascial sheath of the flap. Therefore, its incorporation within the composite flap supports it against the effects of gravity.

In perioral defects, the vascularized tendon is used to reconstruct the orbicularis oris and obtain adequate oral competence. The tension in the neo-sphincter maintains its dynamics. After 6 weeks, a minor surgery under local anaesthesia may be required to reef the tendon in order to balance the forces of mouth opening and competence.

In cheek reconstructions we have found that the tendon supports the flap and prevents a drag on the lower eyelid and the of the ipsilateral nostril. In the future, should the flap sag, the tendon can be readjusted to maintain the flap in the proper position.

The use of the composite radial forearm – PL tendon transfer has been described for lip and chin defects (3–7). Furuta documented angle mouth reconstructions using this flap (7). Others have noted the requirement of support for free flap transfers (6, 8, 9). The brachioradialis tendomuscular unit has also been used in conjunction with the radial forearm free transfer for the same purpose (9).

Another striking advantage with this method is the relatively flatter donor area produced over the distal forearm, which offers a receptive bed for the split thickness graft. When the PL tendon is left behind, graft take in that region is invariably a problem.

Table 1. Patient profile

S.N.	Name/Age/Sex	Defect	Flap	Tendon	Result	Complication	Follow Up
1	S.M./28/M	rt. angle mouth (skin + buccal mucosa)	neurocut. radial forearm	palmaris longus	good	nil	24 months
2	L.D./53/M	lt. angle mouth (skin + buccal mucosa)	neurocut. radial forearm	palmaris longus	good	nil	21 months
3	G.K./67/M	total lower lip + chin + rt. cheek	neurocut. radial forearm	palmaris longus	good	nil	14 months
4	S.S./64/M	lt. cheek	neurocut. radial forearm	palmaris longus	good	nil	14 months
5	A.F./33/M	lt. angle mouth (skin + buccal mucosa)	neurocut. radial forearm	palmaris longus	good	nil	13 months
6	B.M./63/M	rt. infraorbital region	neurocut. lateral arm	triceps tendon	good	venous anast. re-explored	12 months

We offer the option of the free neurocutaneous vascularized tendon composite transfer for perioral and cheek defects as it can achieve a soft, supple, sensate and thin mucosal and skin replacement. It provides normal stomal dimensions and adequate oral competence, prevents sagging of the flap and the angle of the mouth, and effectively supports the lower eyelid.

REFERENCES

1. Song, R., Gao, Y., Song, Y., Yu, Y., Song, Y.: The forearm flap. Clinics in Plastic Surgery, 9: 21, 1982.

2. Soutar, D. S., Scheker, L. R., Tanner, N. S. B., McGregor, I.: The radial forearm flap: A versatile method for intraoral reconstruction. Br. J. plast. Surg., 36: 1, 1983.

3. Sakai, S., Soeda, S., Endo, T., Ishii, M., Uchiumi, E.: A compound radial artery forearm flap for the reconstruction of lip and chin defect. Br. J. plast. Surg., 42: 337-338, 1989.

4. Sadovec, R. C., Luce, E. A., McGrath, P. C.: Reconstruction of the lower lip and chin with the Composite Radial Forearm – Palmaris Longus Free Flap. Plast. reconstr. Surg., 88(2): 209-214, 1991.

5. Serletti, J. M., Tavin, E., Moran, S., Coniglio, J. U.: Total lower lip reconstruction with a sensate composite radial forearm – palmaris longus free flap and a tongue flap. Plast. reconstr. Surg., 99(2): 559-561, 1997.

6. Emiroglu, M.: Utilization of the Palmaris Longus Tendon for maintaining contour in free fasciocutaneous radial forearm flaps. J. reconstr. Microsurg., 12(5): 321-324, 1996.

7. Furuta, S., Sakaguchi, Y., Iwasawa, M., Kurita, H., Minemura, T.: Reconstruction of lips, oral commisure and full thickness cheek with a composite radial forearm palmaris longus free flap. Ann. plast. Surg., 33(5): 544-547, 1994.

8. Kushima, H., Iwasawa, M., Kiyono, M., Ohtsuka, Y., Hataya, Y.: Functional reconstruction of total lip defects with a radial forearm free flap combined with a depressor anguli oris muscle transfer. Ann. plast. Surg., 39(2): 182-185, 1997.

9. Takada, K., Sugata, T., Yoshiga, K., Miyamoto, Y.: Total upper lip reconstruction using a free radial forearm flap incorporating the brachioradialis muscle: report of a case. J. oral maxillofac. Surg., 45(11): 959-962, 1987.

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CAPSULAR CONTRACTURE IN AUGMENTATION MAMMAPLASTY

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SUMMARY

In a group of 331 augmentation mammaplasties performed at the Clinic of Plastic Surgery in Prague from 1994–1998, capsular contracture was recorded in 6 % cosmetic and 12 % of reconstruction operations. The main cause was, in particular, the quality of tissue surrounding the implants along with individual disposition. The incidence of this complication did not differ significantly in relation to the placement of the prosthesis beneath the muscle or gland. Based on histological analysis and the clinical finding, it is obvious that disintegration of the prosthesis does not always lead to severe grades of capsular contracture. It is despite the presence of foreign bodies in the connective tissue.

ZUSAMMENFASSUNG

Die kapsulare Kontrakturation bei der Augmentationsmamaplastik

Dušková M., Sosna B., Kletenský J., Vrtišková J.

In der Gruppe von 331 Augmentationsmamaplastiken, die an der Klinik für plastische Chirurgie in Prag in den Jahren 1994–1998 durchgeführt wurden, erschien die kapsulare Kontraktur bei 6 % der kosmetischen und bei 12 % der Rekonstruktionsoperationen. Als Hauptursache der Entstehung zeigte sich vor allem die Gewebequalität rundherum des Implantats gemeinsam mit der individuellen Disposition. Das Vorkommen dieser Komplikation war statistisch von keiner gravierenden Bedeutung in der Abhängigkeit der Prothesestellung unter dem Muskel oder der Drüse. Aufgrund der histologischen Analyse und des klinischen Befundes ist offenbar, dass die Prothesedesintegration nicht immer zur Entstehung der schwereren Stufen der kapsularen Kontrakturation führt und das auch trotz des Vorkommens der fremden Körper im Zellgewebe.

Key words: augmentation mammaplasty, capsular contracture

Aseptic inflammation and development of a connective tissue layer is the natural response of the organism to an inert foreign body. Thus a fibrous capsule surrounding an implant is also the same reaction. However its excessive formation has a negative effect on the functional and aesthetic effect of augmentation mammaplasty.

The main cause leading to the development of this undesirable complication is above all the insufficiency of soft tissues covering the implant. Also the organisation of haematoma or wound infection or the disintegration of the prosthesis may produce thicker collagen layer (5, 6, 11, 12, 18–21). Last but not least, individual disposition also plays a role (3, 7, 9). Furthermore, the effect of the type of implant is mentioned. Its placement beneath the gland or muscle is very important (1). In the disintegration of prostheses, a more potent connective tissue reaction is assumed. In the clinical picture a hardening of the breast is manifested. A shape change and possible pain can be present.

The classification of Baker, first presented in 1976, is still used (1, 5, 6). It is based on objective findings. They classify capsular contracture into four grades:

I – natural appearance, normal palpation;
II – natural appearance, but more firm palpation;
III – apparent distortion of shape, firm palpation;
IV – unequivocal spheroid deformity with apparent irregularities, firm palpation.

Boswicks's characteristics of practical importance:

1st grade – readily compressible, normal soft breast;
2nd grade – minimal resistance on compression, breast slightly firm, but acceptable;
3rd grade – firm breast, unacceptable, indicated for reoperation.

As a histological correlate, various authors describe the composition of a „physiological" capsule as fibroblasts and fibrous connective tissue. A capsular contracture while contains in addition contractile myofibroblasts. This characteristic implies fibrous tissue immaturity of a capsular contracture. Moreover, in both types, depending on the type of elastomer used, foreign bodies may be present (silicone drops). The thickness of the capsule does not always correspond to the clinical features (2, 16, 21).

PURPOSE OF THE STUDY

Evaluation of histological findings was compared with clinical appearance of cases with or without disintegration of silicone gel prosthesis.

MATERIAL AND METHODS

From 1994–1998 at the Department of Plastic Surgery in Prague, a total of 331 augmentation mammaplasties were performed. This group coprised 242 surgeries with cosmetic indication and 89 reconstruction operations. In all cosmetic operations bilateral surgery was performed. In reconstruction the surgery was unilateral in 67 patients and bilateral in 22. The most of patients were 20–35 years old in cosmetic operations and 36–55 years old in reconstruction operations.

The types of implants used and their numbers are shown in Fig. 1. The majority was textured gel round shaped implant type with moderate profile.

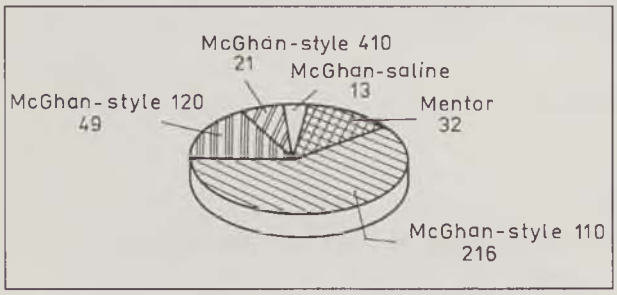

Fig. 1. The types of implants used and their numbers.

In reconstruction the implants were placed beneath the muscle in 100 % of the cases. In cosmetic augmentation the placement of the implant beneath the gland predominated by approximately 6 : 1. The selection of approach is indicated in Fig. 2.

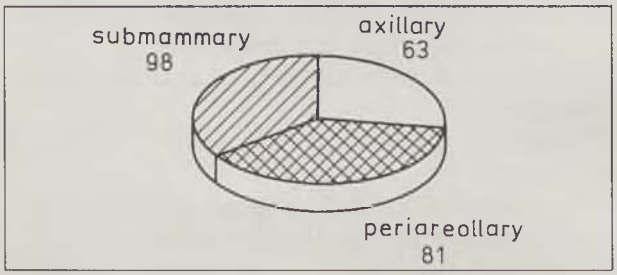

Fig. 2. The selection of the surgical approach.

Redon drainage of the wound was used in 95 % of the cases. Preventive systemic administration of broad-spectrum antibiotics was used in 100 %, at least in prophylactic doses. During re-operations a random sample of the capsule was taken for histological examination. This was done in cases of clinical contracture as well as in cases showing signs of disintegration of the prosthesis or in cases involving a simple change of the implant, normally because of a negatively perceived asymmetry after unilateral reconstruction.

RESULTS

Early complications, such as bleeding, seroma or dehiscence was observed in 5 % of the patients.

Late complications included capsulation, malposition and infection. The number of these complications was slightly above 6 % of cases.

A total of eight patients were re-operated on after cosmetic augmentation. Seven times due contracture when capsulectomy was performed. Reoperation was made in cases of grade III and IV according to Baker. In one case due to infection the implant was removed. Subsequently after healing a reimplantation was performed. Twice a re-operation was made due to a change in the shape and a finding of disintegration of the prosthesis.

As to reconstruction augmentation, three patients with characteristics of Baker IV were reoperated on. However the total incidence of this complication was 12 %. In all instances patients were involved after extensive prophylactic subcutaneous mastectomy or mastectomy with subsequent irradiation. In one instance a patient was re-operated on due to a progressing asymmetry with disintegration of the prosthesis. Here is a conclusion of the histological examination of tissue from the vicinity of the ruptured prosthesis. In all cases a connective tissue capsule surrounding the breast implant was found. That had focally located variations in thickness. The thickness varies from 0.25 to 1.25 mm. The connective tissue of the capsule was practically cell-free. It was formed at some sites by amorphous hyaline tissue (Fig. 3). On the inner side of the fibrous capsule surface was no lining. Focally, cellular epithelia elements in a palisade-like pattern were found. They rarely resembled synovial lining (Fig. 4). Even synovial metaplasia may be considered. In the capsule the presence of focal proliferation of minor capillaries and rare small lymphocyte infiltrates were found. Moreover, there were also areas of connective tissue with small cavities. They were intermittently lined with very flattened cells with filamentous nuclei. At some sites light-refracting greenish masses were observed in the cavities. Exceptionally a giant cell reaction was observed around these masses.

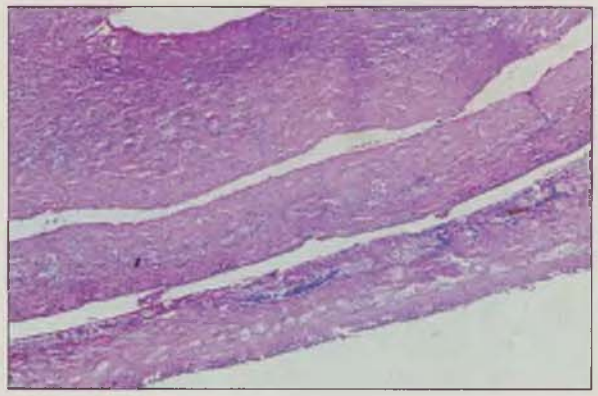

Fig. 3. Connective tissue of capsule, in rare instances with foamy macrophages.

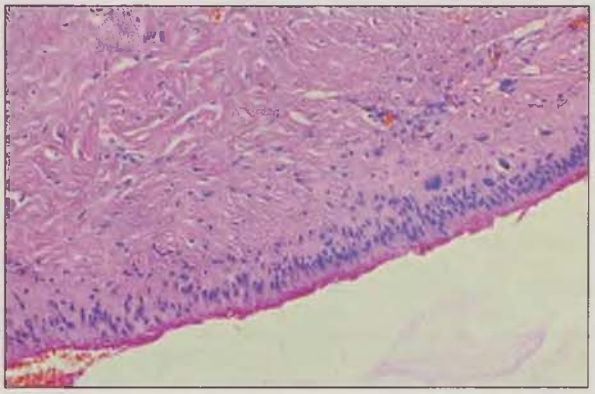

Fig. 4. Cellular elements on inner side of capsule arranged in a palisade-like pattern.

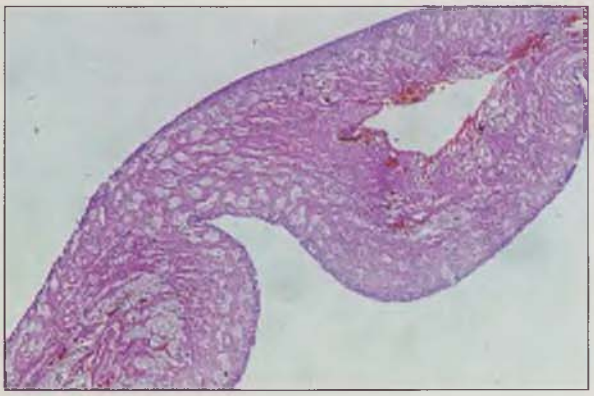

Fig. 5. Connective tissue capsule with numerous foamy macrophages.

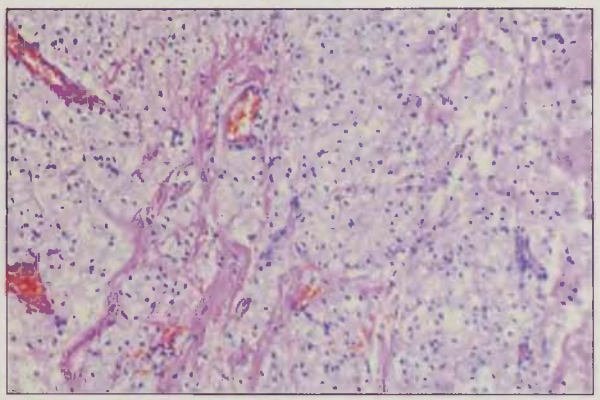

Fig. 6. Accumulation of foamy macrophages in capsular tissue.

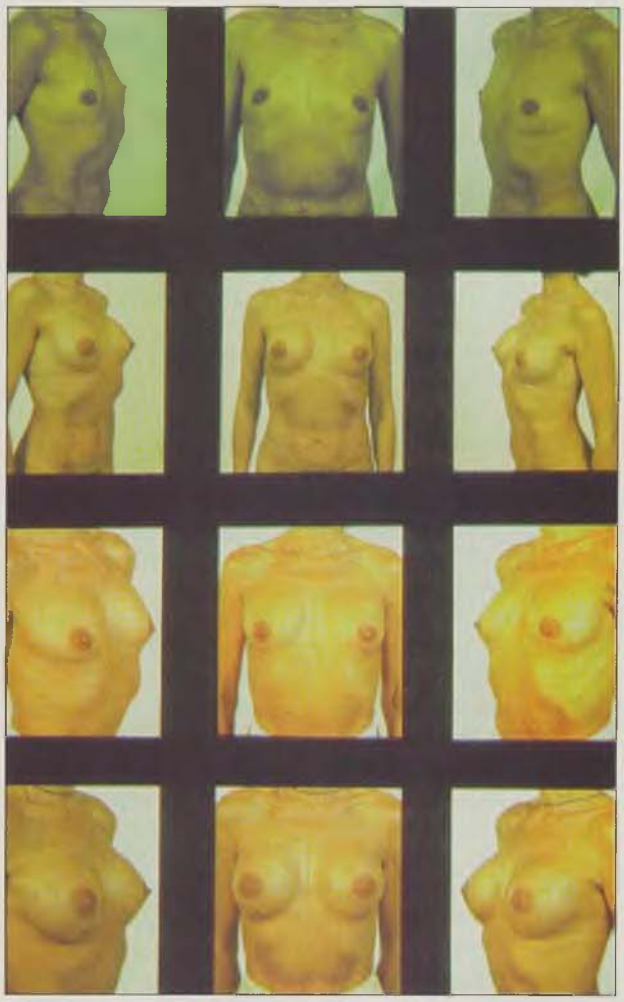

Fig. 7. The change of shape after prosthesis disintegration.

In other cases numerous dispersed foamy macrophage elements were found at some sites. While at others they were more closely grouped (Figs 5, 6).

The phagocytosed material in the macrophage elements was not polarised. A more remote tissue reaction in adjacent lymph nodes was not proved.

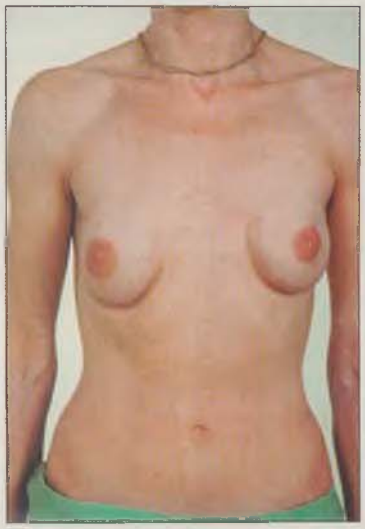

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Fig. 8. Severe capsular contracture in case of immediate breast reconstruction, no implant rupture was found.

On a clinical case the author demonstrates the response of normal tissue to rupture of the prosthesis (Fig. 7) and severe capsular contracture in case of intact implant which was immediately implanted after mastectomy (Fig. 8).

DISCUSSION AND CONCLUSION

Based on health records of patients operated on at the Department of Plastic Surgery in Prague from 1994–1998, the incidence, causes and treatment of capsular contracture were retrospectively analysed. The described number of complications is low (2, 4, 14, 15, 23). But only follow up when the patients are still in the care of the Department is quite objective.

The evaluation of late complications is influenced by the incongruous period of postoperative follow-up. The reason is the patient's varying willingness to check-up examinations, especially they have no complaints. Presumably it is not uncommon for them to ask for help other departments. The significant difference of relation to the implant used was not observed. The same was found in relation to the site of the implant. In all cases a very similar type of elastomer was involved. Prostheses were almost round shaped with a textured silicone shell filled by silicone gel. The ratio of contractures found in cosmetic indications and in reconstruction is logical. It corresponds to the tissue alteration after mastectomy or radiotherapy (7, 8, 10, 13).

Analysing of prostheses ruptures, that condition was manifested by a change in shape. Histologically an enhanced local tissue reaction was found. The thickness of the connective tissue capsule was as much as 1.25 mm with a massive macrophage reaction. However it was not always manifested as toughening of the tissue and pain. In this study the extreme cases of pathological reaction and exulceration were not seen at all (17).

REFERENCES

1. Baker, R. R.: The Management of Breast Cancer with Immediate or Delayed Reconstruction. Advanc. Surg., 25: 51-64, 1992.
2. Bantick, G. L., Taggart, I.: Mammography and Breast Implants. Br. J. plast. Surg., 48-52, 1995.
3. Becker, H.: Quantitative Assessment of Postoperative Breast Massage. Plast. reconstr. Surg., 95: 355-356, 1990.
4. Berger, K., Bostwick III., J.: What Women Want to Know about Breast Implants. QMP, 1995.
5. Boswick, III., J.: Plastic and Reconstructive Breast Surgery. Vol. I, II, St. Louis 1990.
6. Bostwick III., J.: Breast Reconstruction Following Mastectomy. CA Cancer J. Clin., 5: 289-304, 1995.
7. Brody, S. G.: Safety and Effectiveness of the Silicone Breast Implant. Plast. reconstr. Surg., 99: 835-847, 1994.
8. Camara, D. L., Sheridan, J. M., Kammer, B. A.: Rupture and Aging of Silicone Gel Breast Implants. Plast. reconstr. Surg., 98: 828-836, 1993.
9. Charavel, M., Bremond, A., Courtial, I.: Psychosocial profile of women seeking breast reconstruction. Eur. J. Obstet. Gynaec. reprod. Biol., 74(1): 31-35, 1997.
10. Chrristina, Y., et al.: Silicone Implant Rupture Diagnosis Using Computed Tomography: A Case Report and Experience with 22 Surgically Removed Implants. Ann. plast. Surg., 33: 532-546, 1994.
11. Dušková, M.: Complications of Reconstructive Surgery in Breast Carcinoma Therapy. Proceedins. 9th International Congress on Breast Diseases, Houston, April 28–May 2, 1996.
12. Dušková, M., Kaňková, H.: Subcutaneous Mastectomy. Acta Chir. plast., 4: 116-21, 1995.
13. EQUAM: Consensus of meeting 1999.
14. Fee-Fulerson, K., et al.: Factors Contributing to Patient Satisfaction with Breast Reconstruction Using Silicone Gel Implants. Plast. reconstr. Surg., 7: 1420-1426, 1996.
15. Figueira, A. S. S.: Breast Reconstruction with Silicone Prosthesis. 9th International Congress on Brest Diseases, Houston, April 28–May 2, 1996.
16. Grotting, J. C.: Reoperative aesthetic and Reconstructive Surgery. Vol. II, St. Louis 1995.
17. Lebovic, G. S., et al.: Clinical Outcomes Following Removal of Silicone Breast Implants. 9th International Congres on Breast Diseases, Houston, April 28–May 2, 1996.
18. Lectures, Panel Discussions, and Proceedings, 11th Congress of the International Confederation for Plastic, Reconstructive and Aesthetic Surgery, Yokohama, 1995.
19. Lectures, Panel Discussions, and Proceedings, 12th Congress of the International Confederation for Plastic, Reconstructive and Aesthetic Surgery, San Francisco, 1999.
20. Lectures, Panel Discussions, and Proceedings, XVth Congress of the International Society of Aesthetic Plastic Surgery, Tokyo, 2000.
21. Mc Carthy, J. G.: Plastic Surgery, Vol., 6, Philadelphia, 1990.
22. Peters, W., Keystone, E., Smith, D.: Factors Affecting the Rupture of Silicone-Gel Breast Implants. Ann. plast. Surg., 32: 449-451, 1994.
23. Taylor, S. R., Gibbons, D. F.: Effect of Surface Texture on the Soft Tissue Response to Polymer Implants. J. biomed. mat. Res., 17: 205-227, 1983.

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ALTERNATIVE METHOD FOR THE RECONSTRUCTION OF DEFECTS WITH A LOSS OF MORE THAN HALF OF THE UPPER EYELID

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SUMMARY

The authors wish to present an alternative technique for the reconstruction of defects involving the loss of more than half of the upper lid. In a situation where the standard techniques could not be used due to the specific nature of the local findings, the described method seems to be a sound alternative for lid reconstruction.

ZUSAMMENFASSUNG

Die alternative Art der Rekonstruktion der Defekte mit Verlust mehr als einer Hälfte des oberen Augenlides

Kokavec R., Fedeleš J.

Die Autoren präsentieren die alternative Art der Rekonstruktion des oberen Augenlides, wenn mehr als die Hälfte dieses Lides zerstört wurde. In der Situation, wo die Standarttechniken nicht benützt werden können, stellt die beschriebene Methode eine geeignete Methode dar.

Key words: upper eyelid reconstruction, Frické transposition flap, cartilaginous graft

A 35-year-old patient suffered, as a co-driver, damages in a car accident in 1999 (multiple lacerocontused wounds of the face and an opened comminutive dislocated fracture of the nasal bones). The lacerocontused wounds of his left upper and lower eyelids were sutured initially in his district hospital. Six days after the date of the accident he was admitted to our department for eyelid reconstruction due to the left upper eyelid defect. The defect extended over more than one half of the lid, about 2 x 0.5 cm in size, with a total loss of the tarsus and conjunctiva (Fig. 1).

METHODS

Under general anesthesia, during the first part of the operation the absent conjunctiva was replaced through mobilization and moving of the remaining ambient part of the conjunctiva onto the defect. Afterwards, the tarsus reconstruction was performed using a free cartilaginous transplant from the nasal septum cartilage (Figs 2, 3). The skin coverage was made by a transposition flap of Frické. For the next 7 days canthal tarzoraphia limited eyelid movement and increased the chance for better cartilaginous transplant and transposition flap integration (Fig. 4). After three weeks, during the second part of the operation, the flap was detached and the stem was sutured back to the "mother area".

DISCUSSION

There are some techniques recommended for the reconstruction of injuries with a loss of more than one half of the upper eyelid. In our view, in these cases techniques which involve defect reconstruction from the intact contralateral or homolateral eyelid should not be preferred. We are thus able to avoid the risk of needless complications (ectropium, entropium etc.). Also, in the case presented by us, some of these techniques (Mustarde, Cutler-Beard) could not be used due to the local finding of a scar on the lower eyelid.

Undoubtedly, when the mobilization and moving of the remaining ambient part of the conjunctiva to the defect is impossible, a technique of a "composite graft" from the nasal septum carti-

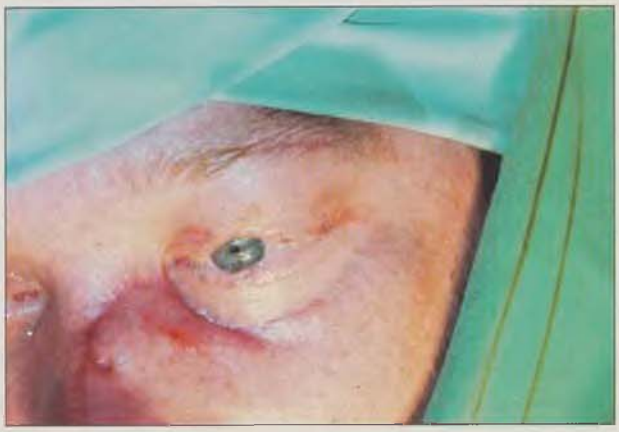


Fig. 1. The defect extended over more than one half of the lid, about 2 x 0.5 cm in size, with a total loss of the tarsus and conjunctiva.

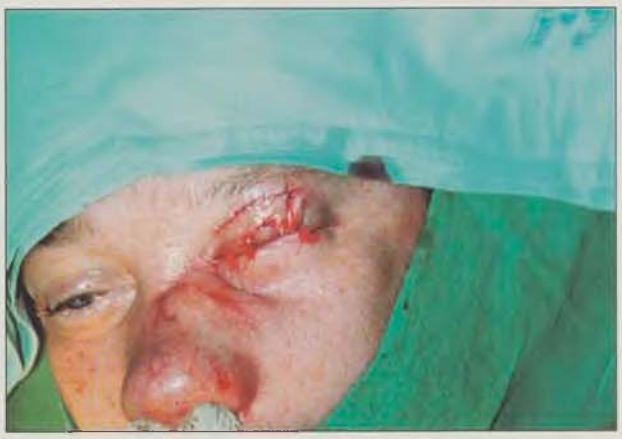


Fig. 4. The transposition flap of Frické sutured into the defect with the canthal tarzoraphia.

Fig. 2. Free cartilaginous transplant from the nasal septum.

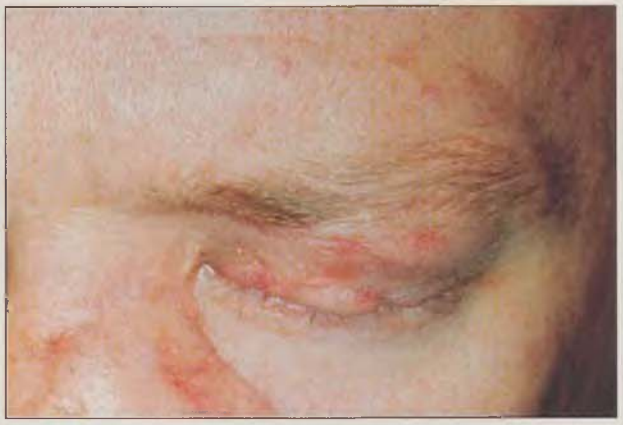
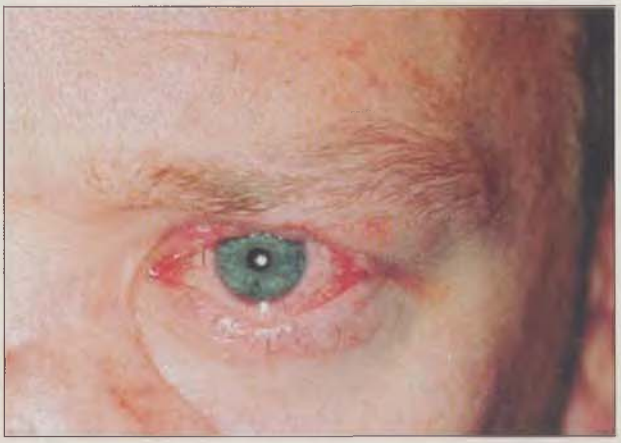


Fig. 5.

Fig. 3. The tarsal reconstruction using a free cartilaginous transplant from the nasal septum cartilage.



Figs 5, 6. Post-operative status shows an adequate functional and aesthetic result.

lage for conjunctivae and tarsa reconstruction is recommended.

The technique of eyelid reconstruction described above gives an adequate functional and aesthetic result (Figs 5, 6), and in our opinion represents a reliable surgical alternative to the standard methods for the reconstruction of defects involving the loss of more than one half of the upper eyelid.

REFERENCES

1. Cutler, N. L., Beard, C.: A method for partial and total upper lid reconstruction. Am. J. Ophthal., 39: 1, 1955.
2. Callahan, A.: Free composite lid graft. Arch. Ophtal., 45: 539, 1951.
3. Cole, J. G.: Reconstruction of large defects of the upper eyelid. Am. J. Ophthal., 64: 376, 1967.
4. Černý, J., et al.: Speciálna chirurgia. Chirurgia krku a hlavy. In: Fedeleš, J. (Ed.): Osveta, Martin, p. 492, 1995.

5. Holt, J. E., Holt, G. R., van Kirk, M.: Use of temporalis fascia in eyelid reconstruction. Ophthalmology, 91: 89, 1984.
6. Mustardé, J. C.: Reconstruction of the eyelids. Ann. plast. Surg., 11: 149, 1983.
7. Mustardé, J. C.: Major reconstruction of the eyelid, the functional and aesthetic considerations. Clin. plast. Surg., 8: 227, 1981.
8. Rogers, B. O.: History of oculoplastic surgery: the contributions of plastic surgery. Aesth. plast. Surg., 12: 129, 1988.
9. Smith, B., English, T. P.: Techniques available in reconstructive surgery of the eyelid. Br. J. Ophthal., 54: 450, 1970.
10. Tenzel, R. R.: Reconstruction of central one-half of an eyelid. Arch. Ophthal., 93: 125, 1975.
11. Youens, W. F., et al.: Full thickness lower lid transplant. Arch. Ophthal., 77: 226, 1967.

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HOURLY DIURESIS IN PATIENTS WITH EXTENSIVE BURNS

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SUMMARY

The values of hourly diuresis in 40 patients with extensive burns were investigated and expressed graphically during the stage of burn shock. By analysis of the values and the patient's condition, a marked effect of the mental state on the course of burn shock and further treatment at the intensive care unit of the Prague Burns Centre was found. On a preliminary basis the effects of some drugs on the values of hourly diuresis were investigated.

ZUSAMMENFASSUNG

Die einstündige Diurese bei weit verbrannten Patienten

Bláha J.

Die Werte der einstündigen Diurese wurden bei 40 weit verbrannten Patienten verfolgt und sie wurden in der Zeit des akuten Brandwundeschocks graphisch ausgedrückt. Anhand der Werteanalyse und des aktuellen Zustandes des Patienten wurde festgestellt ein beduetender Einfluß der Psychik auf den Verlauf des akuten Brandwundeschocks und auf die weitere Behandlung an der Intensivstation der Brandwundeklinik in Prag. Orientierungsweise verfolgte man die Auswirkungen einiger Medikamente auf die Werte der einstündigen Diurese.

Key words: burn shock, hour diuresis, psychic state

Every patient with extensive burns suffers from shock due to the loss of fluids associated with generalized oedema, caused by the release of multiple mediators influencing cell membranes.

One of the crucial factors used to follow-up the development of burn shock is diuresis. Although the hourly urine output and its specific weight are very sensitive to changes in the gen-

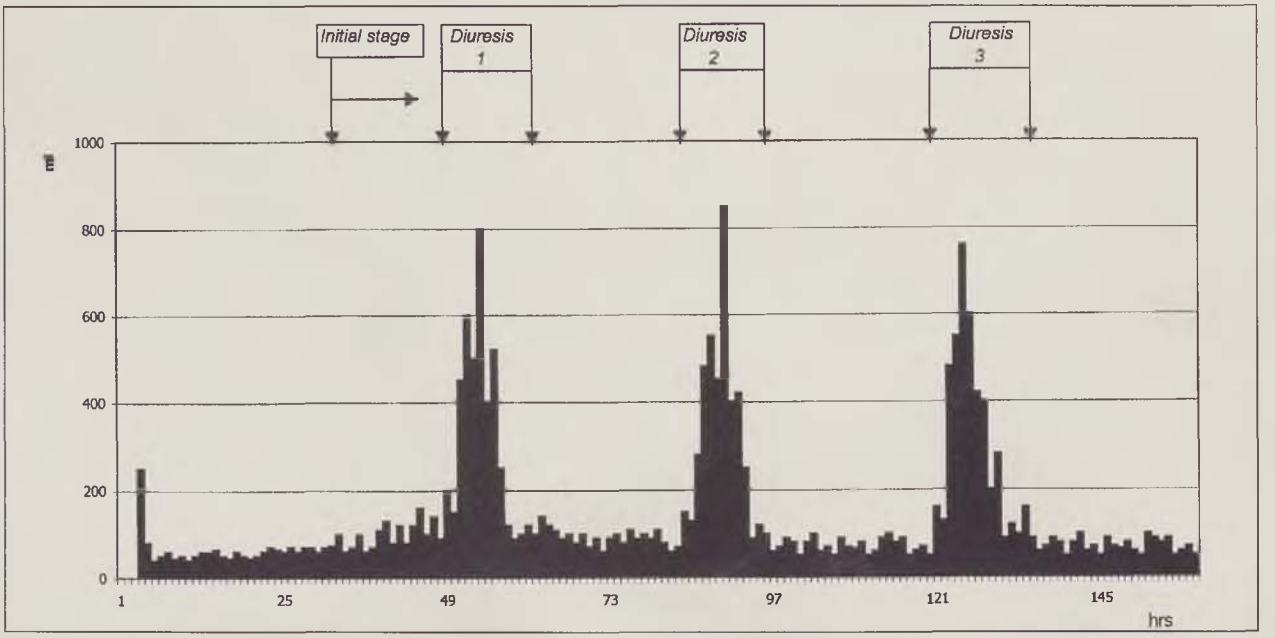

Fig. 1. Model graph to demonstrate the hourly diuresis of a patient with burns on 30–40 % of the body surface. In the initial stage the diuresis is limital, the values are almost stable. Then starts the preparatory stage and shock is terminated by potent diuresis. Later periods with high and optimal diuresis alternate throughout the period of treatment at the intensive care unit.

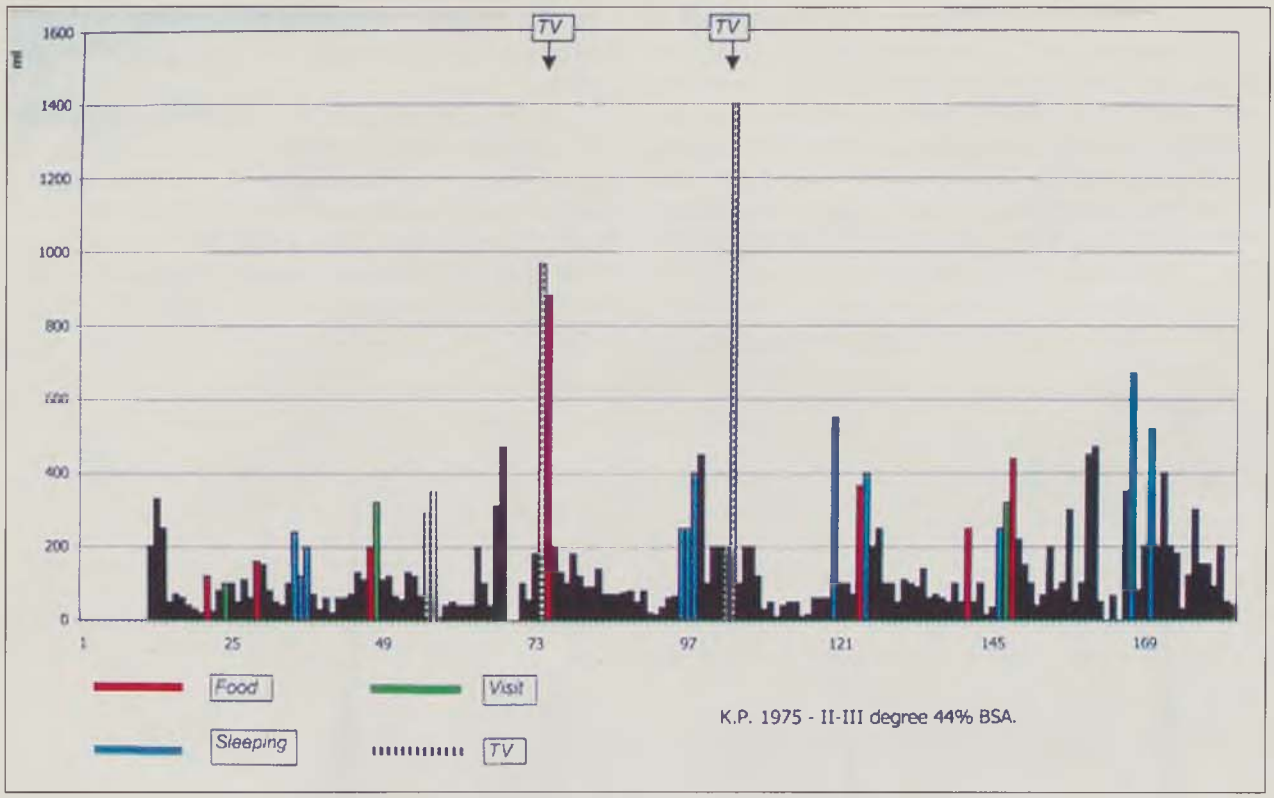

Fig. 2. Example of the effect of food intake, visits, TV and sleep on hourly diuresis.

eral condition of the patient, they cannot be evaluated separately in isolation from other important parameters such as the haematocrit, mineralogram, blood pressure, pulse, respiration rate and quality of respirations, the state of the patient s consciousness etc.

The hourly diuresis is monitored carefully in all trauma intensive care units. It is well known, however, that in patients with extensive burns changes in diuresis are extremely marked and from these changes various important facts can be derived that so far, might have escaped attention.

The basic postulate in this respect was the rule that a simple picture has a much greater informative value than a group of assembled numerical data. The first indication of this was the experience of recent years, when at the Burn Centre, the effect of Seropram on the course of burn shock was investigated and the value of

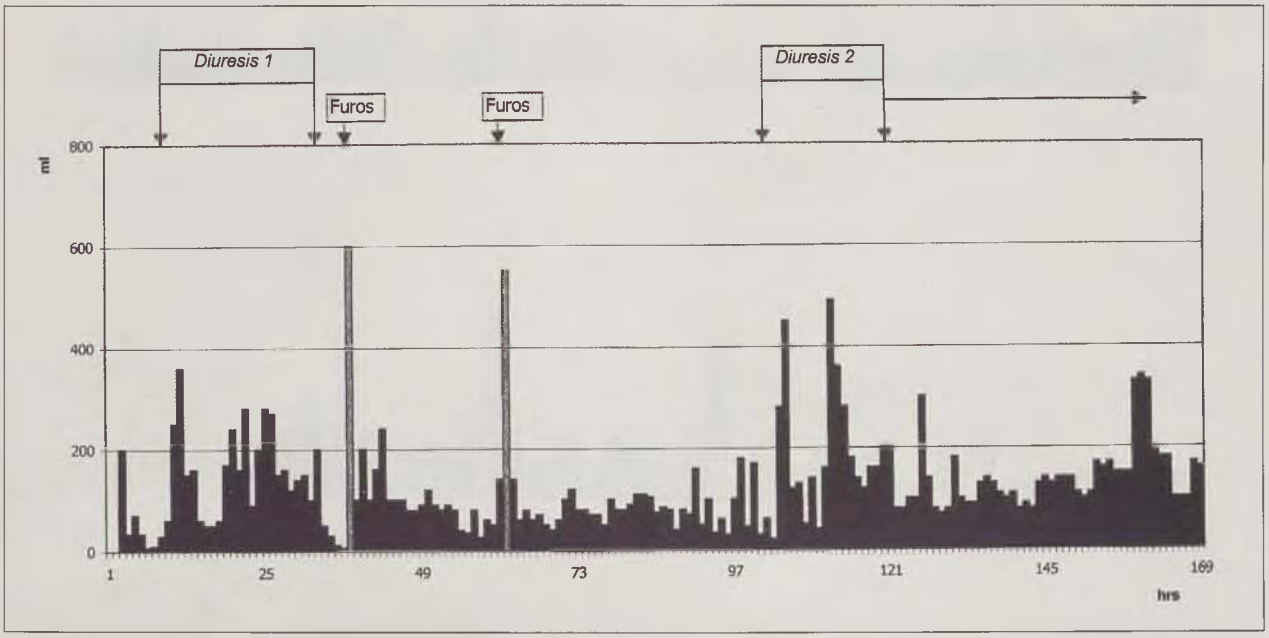

Fig. 3

graphic expression of a numerical series, recorded by the nursing staff in the intensive care unit was noted. The study of the obtained graphs, revealed some order. Only one basic factor was lacking, i.e. finding a unifying denominator common to all patients, not only for patients with burns but also for other patients with severe injuries or diseases. Nothing of that sort was found in the literature. The complicated character of different injuries and the marked individual differences of the affected subjects prevented the finding of a uniform factor.

During the essential intravenous administration of fluids it is important to take into consideration the state of the cardiovascular system and the danger of acute cerebral and pulmonary oedema. Nevertheless, it is important to treat hypovolaemia, and the fluid intake is carefully moni-

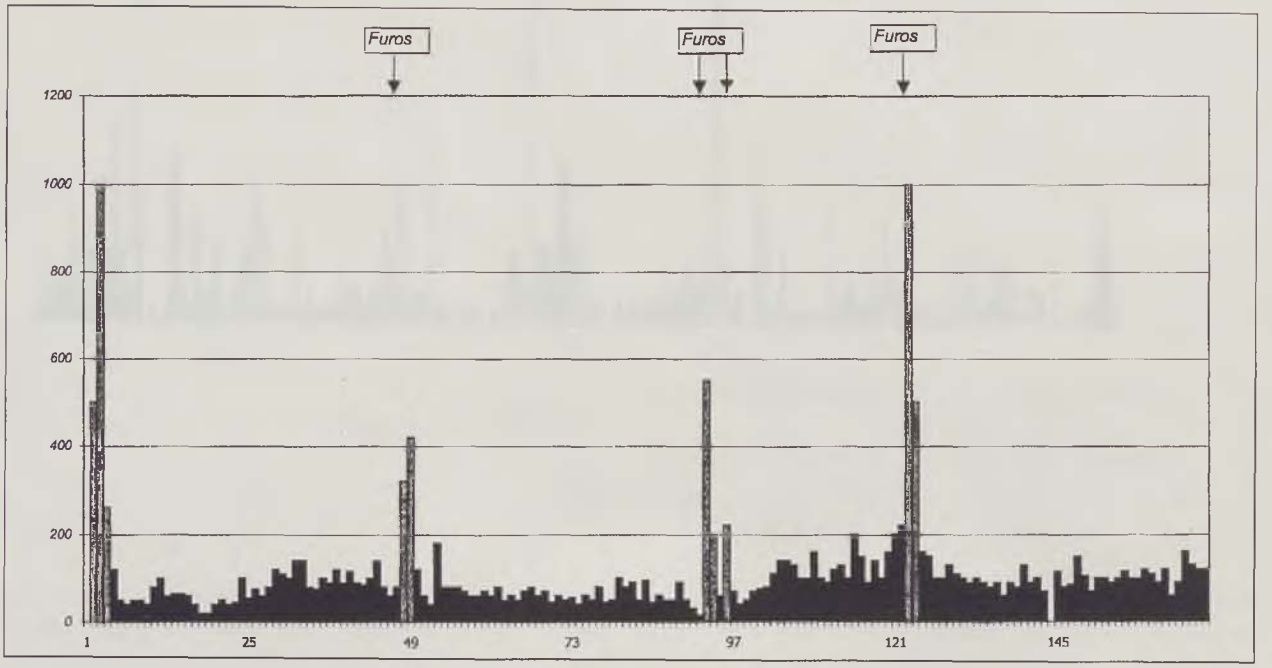

Fig. 4a

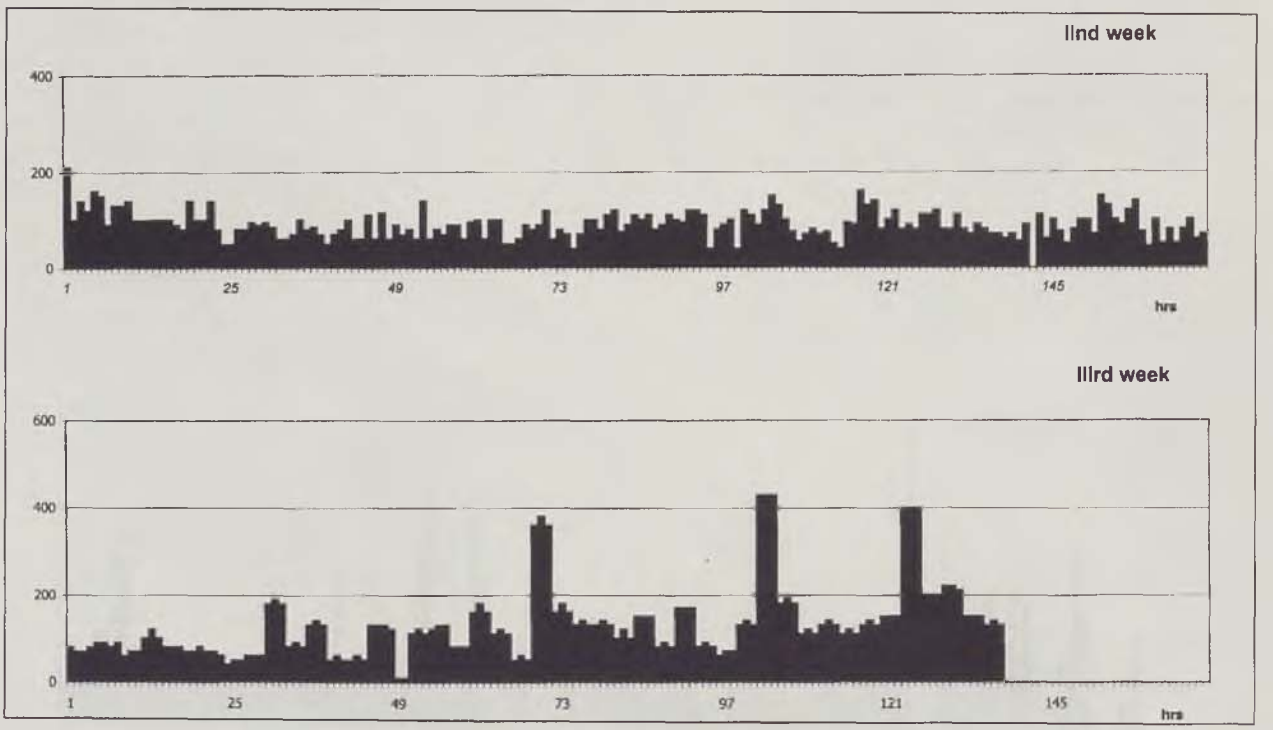

Fig. 4b

Figs 3, 4. Examples of hourly diuresis in two suicides. The first case inflicted himself with burns as a demonstration without the true intention of killing himself. The second case committed the same act in depression and with a serious intention to terminate his life.

tored with regard to the patient's individual condition. The resuscitation with crystalloids and colloids should keep the hourly diuresis within the range of 0.5 and 1.0 ml urine per kg body weight per hour. For an adult this is 35–100 ml/hour. The hourly diuresis is one of the crucial indicators for orientation as to how the resuscitation is progressing.

From a graphic presentation of hourly diuresis, it is apparent that after a certain period of reduced urinary output if the patient is properly resuscitated with fluid the so-called diuretic phase develops, signaling the termination of the burn shock. Also possible changes in the urinary excretion of minerals, i.e. increased potassium and reduced sodium excretion convert. These findings are, however, rare and thus lack standard validity. After termination of the first diuretic stage, further periods of exceptionally high hourly diuresis develop, alternating with lower diuresis varying around 1 ml/kg body weight or slightly more.

If we elaborate a graph sufficiently sensitive to items that are at first close, we reveal a very interesting fact. Before the onset of the diuretic stage proper, very frequently a certain prelude develops and low values alternate with higher ones that are approximately double (Fig. 1). This condition of unsteady diuresis is of varying duration, but in a well-conducted course of shock treatment it lasts for about 24 hours. There are, however, individual differences, and it is necessary to take into consideration the concurrently administered medication. (The investigation is biased most by frequently administered diuretics, although even these provide some interesting information). If the course of treatment is not disturbed by other traumatizing events, potent diuresis develops exceeding by many times the original values. Its scope depends on the amount of administered fluids during the first days after the burns and their superficial losses. Nevertheless, the author observed repeatedly an up-to-tenfold increase of urinary output, i.e. if before the onset of the diuretic stage the diuresis was cca 70–100 ml/hour, after its onset the values were some 700–1000 ml/hour.

MATERIAL

In a group of 40 varied patients hourly diuresis was expressed graphically. By analysis of the graphs and a comparison with the records on the condition and activities of the patients, some unexpected facts were revealed. When investigating the causes of the sudden increase of urinary output of patients with burns (during the period before as well as during the diuretic stage proper) the following facts became evident (Fig. 2).

Immediately before the increased urinary output the patient had a meal, had a visit of close relatives or good friends, was given an analgetic, watched an interesting TV programme etc. It may thus be considered that he felt a certain relief from his complaints, was relieved of certain anxieties, he enjoyed the food which produced pleasant sensations. Diuresis also increases markedly during periods of unrest and disorientation of alcoholics and drug addicts in delirium and during the period of withdrawal symptoms.

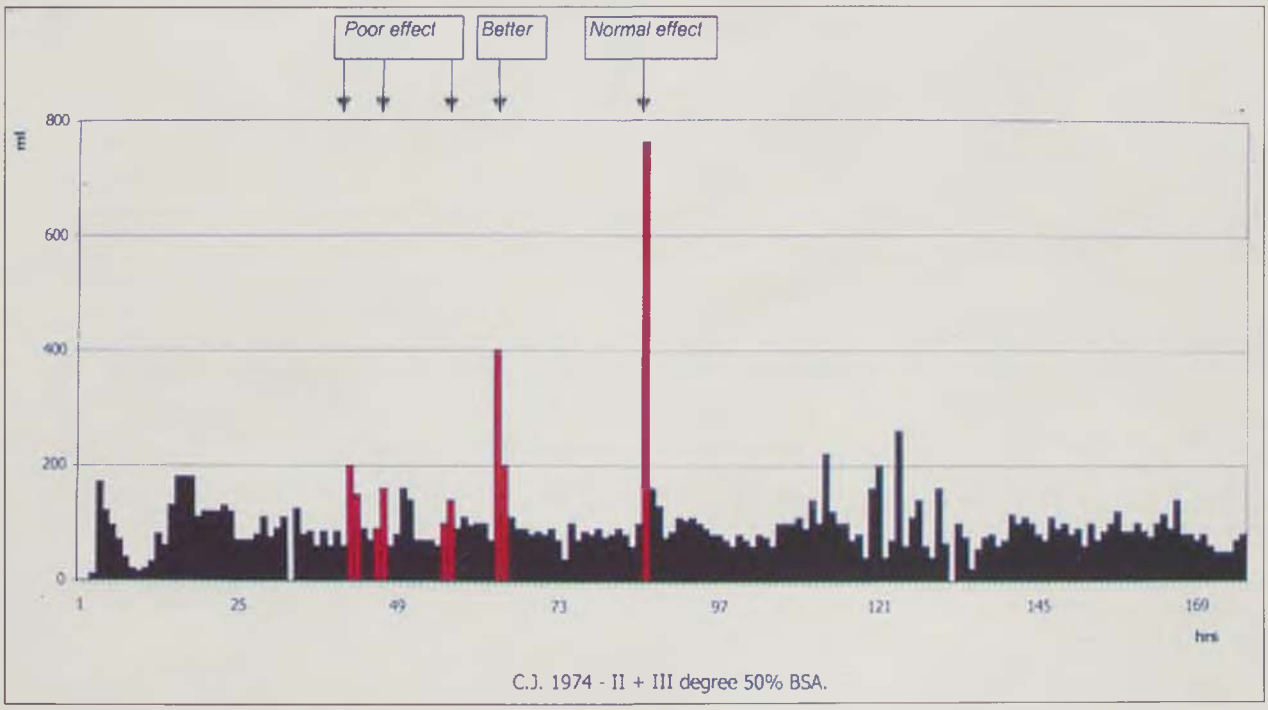

Fig. 5. Effect of diuretic (Furosemide) during the slow administration of fluids in a patient with burns of the airways and a risk of acute pulmonary oedema. Unless the patient is adequately saturated with fluids, the diuretic has little effect. Later the effect increases greatly but does not influence the diuretic stage terminating the shock period, but rather conceals it.

Similar factors were involved closely before the onset of the potent diuresis which terminated the burn shock. There was proposed a marked influence of the mental state of the patient.

This is even more apparent in the treatment of burns of suicidal subjects (Figs 3, 4). These patients can be divided into two groups. If prodromes and the subsequent diuretic stage develop soon after the accident (with regard to the extent of the injury), attempted suicide was intended as a demonstration of a certain objective, which was achieved. If the suicide was meant seriously, however, the patient is angry with the attending staff who have spoiled his intention, the patient is permanently negativistic, aggressive, does not cooperate in treatment and the diuretic stage do not occur or is delayed. And while in the first case the shock is over in 48 hours, in the second case diuresis develops sometimes only after 14–17 days. Again, a marked influence of the mental state on the course of the shock period can be seen.

DISCUSSION

The increase of diuresis may be a sign that the patient is fully aware of his burned body, he becomes reconciled to the given situation and accepts the perspective offered by the medical team as regards recovery. To put it briefly, he finds that the injury can be survived and decides to cooperate. This implies marked mental relief and terminates the stress condition. Additional similar periods continue as therapy proceeds successfully and the patient feels that the staff meets his ideas.

The evaluation of figures is difficult as every patient receives a different medication, and there were very few patients to whome only basic drugs were administered. The graphs of hourly diuresis in alcohol and drug addicts with burns were instructive as they did not differ from each other. This is probably due to the fact that the patient was influenced to such an extent by alcohol or the drug that he was not aware of the burn and therefore extreme amounts of catecholamines which otherwise dominate the stress condition were not released into the circulation.

The effect of different drugs used in burn patients is rather complicated and it must be analyzed on a larger number of patients. The effect of diuretics depends on the successfull resuscitation with fluids (Fig. 5). If during this stage furosemide is used to increase the diuresis, it causes practically no response and the optimal effect is achieved only after the adequate fluid replacement. It does not affect the onset of the diuretic stage proper. However, administration of steroids may have an effect on the course of shock period and can reduce its duration. Steroids, however, have their limitations and a number of well-known disadvantages.

The interactions of concurrently administered drugs are remarkable, and a detailed analysis will follow in the future study.

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MARJOLIN'S ULCER

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SUMMARY

In 1828 John Nicolas Marjolini characterized ulcer with malignant degeneration which developed in scars after burns, but it occurs under varying clinical conditions. Typical feature is the latent period (on average 30 years). It is encountered in 2 forms: a shallow ulcer or exophytic tumour, most frequently on the lower extremities. At the Prague Burn Centre 11 patients were treated since 1978 till 1998. A unique case was a man suffering from congenital form of epidermolysis bullosa who developed Marjolin's ulcer on his foot (histological examination confirmed well differentiated squamous cell carcinoma). After 4 years he died with extreme cachexia and metastatic spread of the tumour, because he refused repeatedly amputation and lymph nodes dissection. To prevent Marjolin's ulcer several recommendations are presented.

ZUSAMMENFASSUNG

Ulcus Marjolin

Königová R., Rychterová V.

Im Jahre 1828 Jean Nicolas Marjolin zwar charakterisierte das Geschwür mit der malignen Degeneration in Narben nach der Brandwundeverletzung, aber in den späteren Publikationen wurde es in Narben verschiedener Herkunft beschrieben. Als typisch gilt eine lange latente Zeitperiode seit der Verletzung bis zur Entstehung der malignen Wendung (im Durchschnitt 30 Jahre). Die Pathogenesion bleibt bis jetzt unklar aber weist auf sgn. "double insult" hin.

Dies zeigt sich klinisch als ein flaches Geschwür oder ein exofythische Tumor, meistens lokalisiert an den unteren Extremitäten.

In den Jahren 1978–1998 wurden 11 Patienten an der Klinik für Verbrennungen der 3. medizinischen Fakultät der Karlsuniversität in Prag hospitalisiert. Es handelte sich um radikal gelöste Exzisionen mit darauf folgenden Autotransplantation. Bei 3 Patienten folgte nachher die Lappenplastik und bei 2 war notwending die Amputation und die Exenteration der inguinalen Knoten. Ein besonderer Fall stellte ein Mann mit der kongenitalen Form Epidermolysis bullosa dar. Bei diesem Patienten wurde mit 32 Jahren histologisch ein geflieste Karzinom an der unteren Extremität erwiesen und in 4 Jahren entstand Exitus letalis in der extremen Kachexion, weil er die Amputation der unteren Extremität wiederholt abgelehnt hatte.

Aus der Sicht der Vorbeugung gilt folgendes zu empfehlen:

– ein frühzeitige Verschluß der Defekte beugt der Infektion und der exzessiven Vernarbung vor;

– die vernarbten Kontrakturen rechzeitig anhand der Rekonstruktionsoperationen lösen und deren Zerfall verhindern;

– die nicht heilende Defekte bioptisch untersuchen

– nach der radikalen Exzision des gefliesten Karzinom nicht das Lappenmaterial anwenden, sondern allmälich oder eventuell über Ettapen der provisorischen biologischen Deckungen decken, um den lokalen Befund verfolgen zu können

Key words: Marjolin's ulcer, burn scar carcinoma, non-healing wounds

From the historical aspect it is important to remind of the work of Cornelius Celsus who mentions cancerous ulcerations in scars already at the time of Christ's birth. In 1828 Jean Nicolas Marjolin published in the Dictionnaire de Medicine (1) a chapter called „Ulcere" characterizing an ulcer with malignant degeneration which developed in scars after burns. Stedman's Medical Dictionary published in Baltimore in 1995 describes Marjolin's ulcer as „...a well-differentiated but aggressive squamous cell carcinoma occurring in cicatricial tissue at the epidermal edge of a sinus draining underlying osteomyelitis..."

This definition, however, does not mention that Marjolin's ulcer can develop under varying clinical conditions: in scars after frostbite, after suprapubic cystostomy, after hidradenitis suppurativa, after lupus erythematodes, after vaccination, in chronic ulcerations in venostasis and in chronic decubiti.

A typical feature is the latent period between injury and the malignant reversal, which is on average 30 years, but may be only one year or conversely as long as 70 years. The younger the patient at the time of injury, the longer the latent period.

The pathogenesis was the subject of several papers: Treves and Pack (1930) considered the release of toxins from damaged tissues as the cause, which along with hypoxia and nutritional deficiency causes cellular mutations in the tissues. According to research in recent years mediators of the inflammatory response are involved (TNF, I1-1). In 1968 Castell and Goldsmith found that immunodeficiency is the predisposing factor for malignity. In 1973 Futrel and Myers tried to prove that obliteration of the lymphatic system reduces the control of tissues on cell mutations. During the last decade Hahn, Kim and Jeon drew attention to the fact that injury per se is not cancerogenic but makes the tissue sensitive to other cancerogens such as irradiation or ultraviolet radiation („double insult").

In clinical practice Marjolin's ulcer is encountered in two forms: 1. a shallow well defined ulcer with nodular elevations at the periphery, 2. an exophytic tumour with papillary granulations which is very aggressive during growth and relapses in as many as 50 %. Both types are as a rule infected and metastatize on average in 30 %. With the rate of metastases rises also the mortality rate (4, 5).

The most frequent site is on the lower extremities (38 %) from where it metastatizes twice as frequently as from other sites. On the upper extremities it was described in 22 %, on the head (2, 3) and neck (mostly in men) it is found in 30 %, on the trunk only in 10 %. These data are derived from thirty references concerned with 200 patients with Marjolin's ulcer. At the Burn Centre (Third Medical Faculty Charles University) during the last two decades (1978–1998) 11 patients aged 23–69 years (8 men and 3 women) with Marjolin's ulcer were treated.

Malignant degeneration occurred in this group once before 2 years and the longest interval after injury was recorded in the oldest patient, after 66 years. The patient had suffered the injury as a child at the age of 3 years.

The ulcer was located on the lower extremities 8 times, on the upper extremity once and in the face twice. All affections were treated radically by excision, seven times with autotransplantation, three times with a plastic operation using a flap and twice it was necessary to make an amputation after a relapse and secondaries in the groin combined with dissections of the inguinal lymph nodes.

A unique case was a young man from Litvínov who suffered from the congenital form of epidermolysis bullosa. In 1980 he was, at the age of 23 years, referred to our Burn Centre with total contracture of the stumps of the fingers on both hands, the remaining phalanges being „buried" in the palm, incl. the thumb, and on the feet the distal phalanges were missing. The skin cover was formed by a fragile thin epidermis constantly desquamating and the exposed corium with penetrating granulations bled repeatedly and the bare

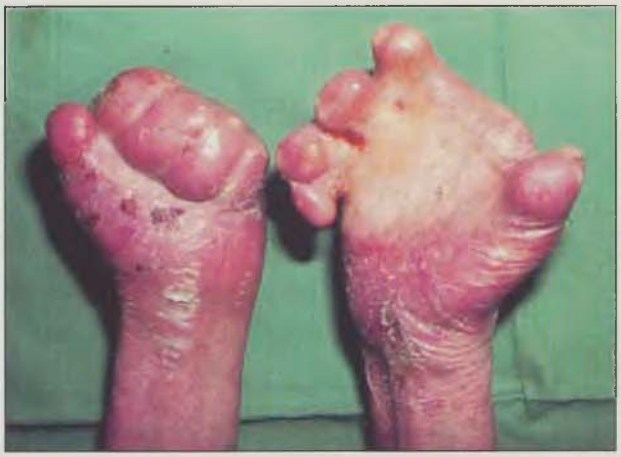


Fig. 1. Flexion contracture of stumps of the fingers: on the right hand still „emburied" in the palm; on the left hand contracture released by means of a full-thickness skin graft.

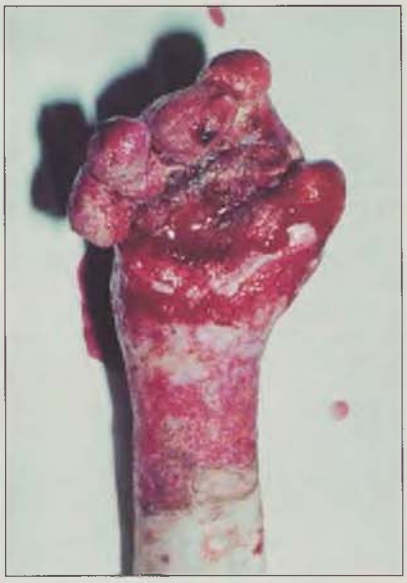


Fig. 2. Under modelling dressing desquamation of epidermis reappeared in the operated area and in the neighbourhood.

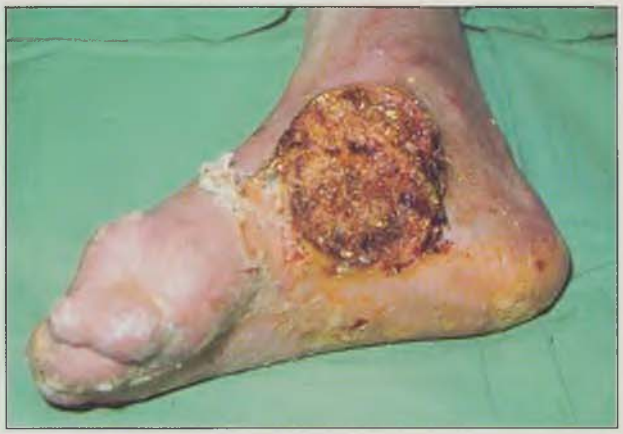


Fig. 3. 10 years later the patient was admitted for exophytic mass that developed in the perimalleolar region on his right foot in site of chronic irritation and inflammation.

areas were permananently infected. During the same year (1980) the right hand was unfolded with insertion of a full thickness skin graft, the skin was taken from the right hypochondrium (Fig. 1). It healed without complications but under the compressive bandage a desquamation of the epidermis developed on the hand and stumps of the fingers (Fig. 2). After xenotransplantation spontaneous epithelization took place (6).

In 1989 during a check-up examination at the out-patient department of the Prague Burn Centre a cauliflower-shaped exulcerated tumour on the instep of the right foot was found (Figs 3, 4) which was radically removed and histological examination confirmed squamous cell carcinoma (Figs 5 a,b). After temporary xenotransplantation the defect was closed by autotransplantation using a dermoepidermal graft. Four months later (November 1989) the patient was again admitted to the Burn Centre with a relapse (10 x 10 cm) which developed at the periphery of the healed autograft. The condition was again treated by radical excision and transplantation.

In October 1991 the patient came again with a disintegrated extremely infected tumour, but repeatedly refused amputation. During 1992 radical excisions were made three times and in 1993 twice (in March and June). In December 1993 he was admitted in extreme cachexia (Figs 6, 7) and extensive disintegration of the area of the instep, heel and perimalleolar region (Figs 8, 9). After ten days in hospital the patient died (Figs 10 a,b).

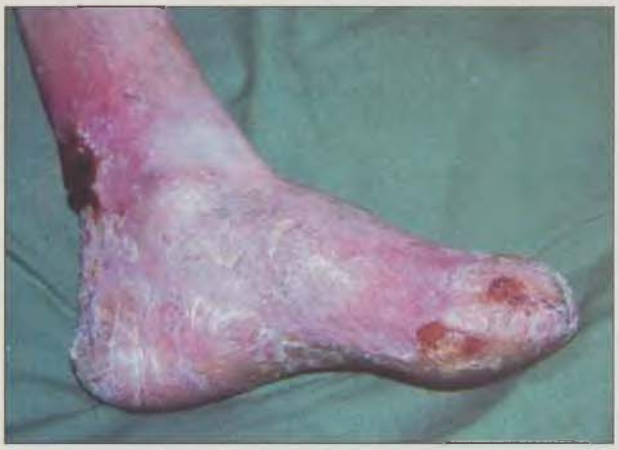
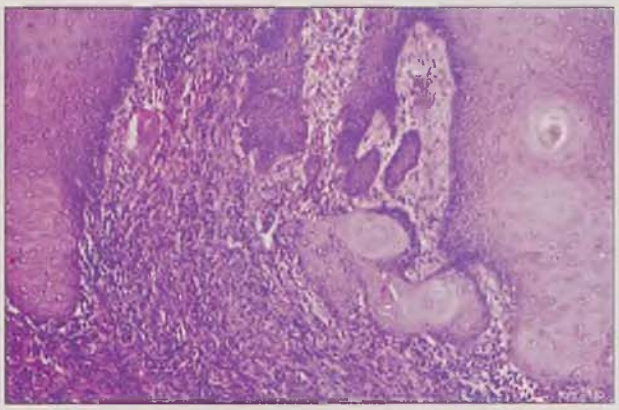
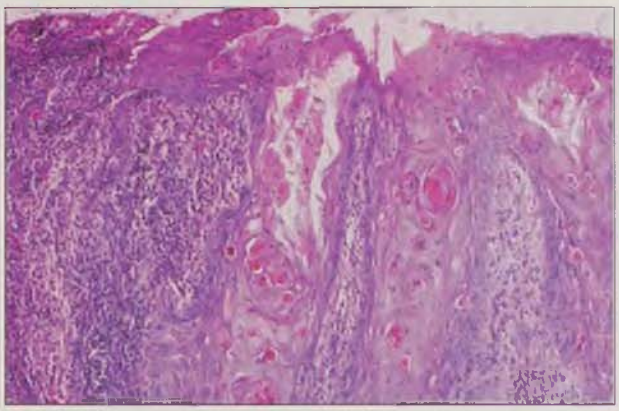


Fig. 4. His left foot with non-healing defects but still without malignancy.



a



b

Fig. 5. Excision of the margin of the Marjolin's ulcer shows at the sites pseudoepitheliomatous hyperplasia of epidermis and in the middle cancerization with invasive squamous cancer, in corium pronounced mixed inflammatody cellulization (a); the surface of the tumour is covered with a crust, the corium is infiltrated by sheets of well differentiated squamous carcinoma with keratinization (b).

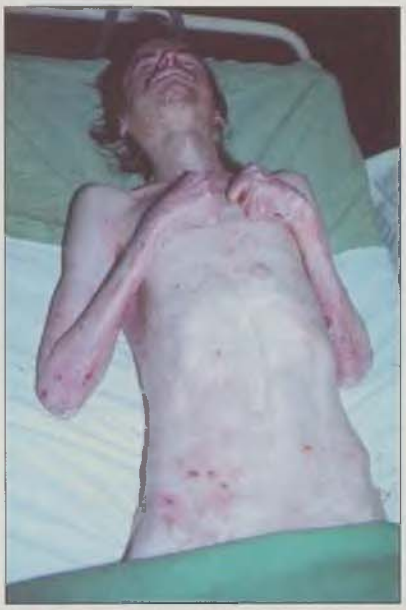


Fig. 6. 2 years later the patient was admitted in extreme cachexia.

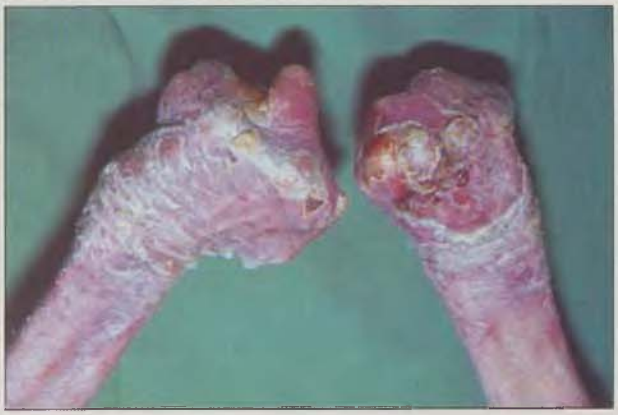


Fig. 7. Recurrence of flexion contracture on both hands.

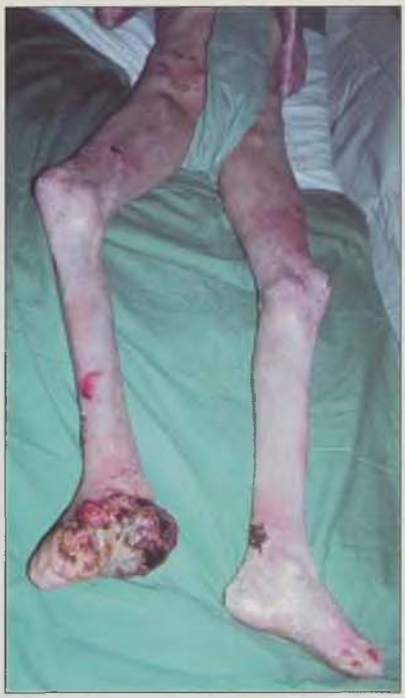


Fig. 8. Extreme cachexia with the right foot covered with an exophytic cancer.

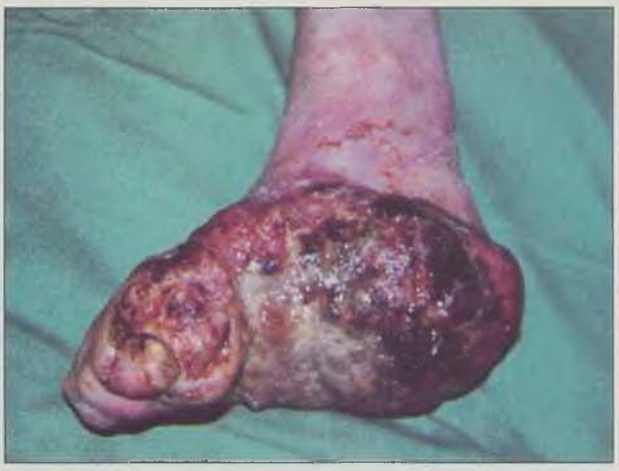
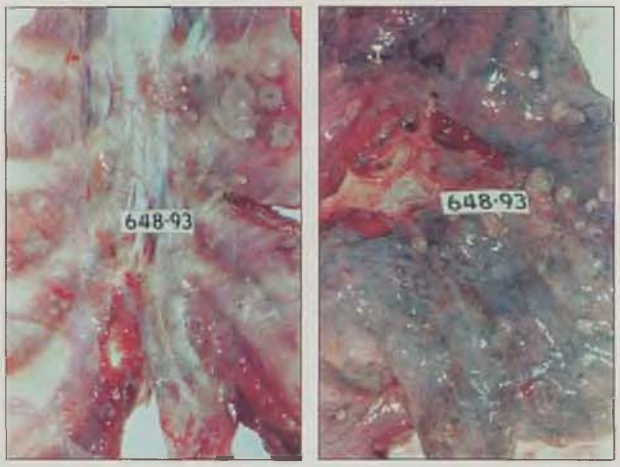


Fig. 9. Detail of the right foot.



a b

Fig. 10. On necropsy metastatic spread was discovered: multiple metastases in the wall of thorax (a); multiple nodular metastases in the parenchyma of the lung (b).

DISCUSSION

Based on analysis of the mentioned cases and studies implemented abroad, in Marjolin's ulcers aggressive treatment is essential, i.e. treatment which involves dissections of the lymph nodes and/or amputation of the affected extremity.

From the aspect of prevention of Marjolin's ulcer the following recommendations are presented:

1. during the primary treatment of wounds (not only after a thermal injury) close the defects as soon as possible and prevent excessive scarring which is promoted also by local infection which is one of the factors of degeneration;

2. treat scarred contractures by proper rehabilitation (compression bandages, elastic dressings, massages, splints) and use reconstructive operations in case of disintegration of scars;

3. medical staff and patients must be aware that long-term life-long follow-up of scars after thermal injuries is necessary and that any local changes in non-healing defects must be examined by biopsy and, depending on the histological evaluation, appropriate radical treatment must be started. The diagnosis is clinical and the pathologist confirms it;

4. it is a general principle that for covering the defect after excision of a squamous cell carcinoma flap material should not be used immediately, to make better observation of the local finding possible. There are however certain situations calling for flap surgery. Exposed vital structures must be covered immediately, while functionally important structures (bones, joints, neuro-vascular bundles) must be closed successively, after a provisional stage of temporary biological covers.

REFERENCES

1. Marjolin, J. N.: Dictionnaire de Medicine. Paris, Bechet, 1828.

2. Clements, B., Lewis, H., et al.: A late, fatal complication of a high energy thermal injury to the scalp. Ann. plast. Surg., 35, 6: 650-653, 1995.

3. Uzunismail Adnan: Marjolin's ulcer of the scalp after 45 years. Plast. reconstr. Surg., 1: 198-199, 1995.

4. Flemming, M. D., Hunt, J. L., et al.: Marjolin's ulcer: a review and reevaluation. J. burn Care Rehab., 11: 160-169, 1990.

5. Bernstein, S. C., Lim, K. K., et al.: The many faces of squamous cell carcinoma. Derm. Surg., 22: 243-254, 1996.

6. Türegün, M., Nisanci, M., Güler, M.: Burn scar carcinoma with longer lag period arising in previously grafted area. Burns, 23, 6: 496-497, 1997.

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POLAND-MÖBIUS SYNDROME AND DISRUPTION SPECTRUM AFFECTING THE FACE AND EXTREMITIES: A REVIEW PAPER AND PRESENTATION OF FIVE CASES

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SUMMARY

The author summarizes hitherto assembled experience with the clinical and genetic characteristics of Poland's and Möbius syndrome. Five selected case-records with this disease and the sequence of the Poland-Möbius syndrome are presented. Another case-record is devoted to an allied syndrome, hypoglossia-hypodactyly, found in a spontaneously aborted fetus. For establishment of a more accurate symptomatology, an irreplaceable place is held by anthropometric examination; for objectifying the asymmetry of the chest the so-called cyrtogram, the chest circumference recorded by means of a wire, is valuable. From the aspect of genetic counseling, preconception care is always provided to mothers from families with reproductive intentions, as well as ultrasonographic examination of the fetus in areas of assumed acral symptomatology (signaling phenotype). In two families ultrasonography was used for prenatal diagnosis. Invasive prenatal diagnosis by amniocentesis was employed in a family with Möbius syndrome. In these families dermatoglyphs have certain common characteristics, such a tendency towards simple patterns. In the wider family of one of our patients we detected in a cousin Parkes-Weber-Klippel-Trenaunay's syndrome, which may indicate common vascular predisposing factors.

ZUSAMMENFASSUNG

Das Poland-Moebius Syndrom. Das Syndrom und das Disruptionsspektrum, das das Gesicht und die Gliedmaßen betrifft

Kuklík M.

Der Autor faßt die gegenwärtigen Erfahrungen mit klinischen und genetischen Charakteristiken des Poland und Moebius Syndroms zusammen. Es werden präsentiert 5 ausgewählte Kasuistiken mit dieser Krankheit, respektiv die Sekventionsfolge vom Poland-Moebius Syndrom. Für die Präzisierung der Symptomatologie ist unentbehrlich die anthropometrische Untersuchung, zur Objektivisierung der Brutkorbassymetrie sgn. Cyrtogram (mithilfe eines Drahts festgestellter Brustkorbumfang). Aus der Sicht der genetischen Beratung wird immer die präkonzeptielle Behandlung bei Müttern mit Reproduktionsabsichten appliziert, genauso wie eine ausführliche Ultraschalluntersuchung der Frucht in Gebieten der voraussetzlichen Gliedmaßensymptomatologie (der signale Fenotyp). In zwei Familien wurde die Ultrasonographie zur pränatalen Diagnostik angewandt. Die invasive pränatele Diagnostik mithilfe der Amniozenthese wurde darüber hinaus in einer Familie mit dem Moebiussyndrom durchgeführt. Die Deramtoglyphen sollen bei diesen Familien gemeinsame Charakteristiken als Tendenzen zu einfachen Mustern feststellen. In breiterer Familie stellten wir bei einem unserer Patienten bei seinem Cousin das Vorkommen vom Parkes-Weber-Klippel-Trenaunaysyndrom, was auf die gemeinsamen dispositionelen vaskularen Faktoren zeigen kann.

Key words: Poland's syndrome, Möbius syndrome, vascular disruption sequence, genetics, dermatoglyphs

Poland-Möbius syndrome is an overlapping disruption spectrum of inborn defects affecting the face and extremities. The majority of authors are inclined to think that Poland's anomaly is part of the Möbius spectrum. There is, however, a number of other disruption syndromes affecting the face, e.g. the syndrome of hypoglossia – hypodactyly, which greatly resembles Möbius syndrome. The common denominator of all these syndromes and sequences is a disruption sequence on a genetic or non-genetic basis. Depending on the site of the vascular disruption anomaly, there are differences in symptomatology, and the definition of different syndromes is also associated with this problem.

Möbius syndrome was described as early as 1888 by the German neurologist P. J. Möbius. At present, just under 300 cases have been described with varying degrees of affliction of the craniofacial area and extremities. Möbius syndrome is a rare condition with a frequency of 1 : 500 000.

Poland's syndrome (sequence) was described in 1841 as the unilateral absence of the pectoralis major muscle and ipsilateral dermal syndactyly

of the hand. The reported frequency is 1 : 20 000 – 1 : 30 000. It is maintained that 10% of all patients with syndactyly have Poland's sequence.

Möbius syndrome has a number of synonyms – such as paralysis oculofacialis, agenesis nuclearis, akinesia algera, and diplegia facialis congenita – which more or less characterize the disease (17, 40). A typical feature is paralysis of the facial muscles and paralysis of lateral eye movements to a varying degree, which can be unilateral or bilateral.

The anatomical correlate of these changes is the congenital absence of the nuclei of cranial nerves, which can be verified by dissection (41). This may be due to a disruption sequence of the Möbius type or to primary agenesis of the nuclei of the cranial nerves (2, 27).

Facultative, inconstant manifestations result from haemorrhagic lesions due to a disruption sequence caused by a vascular anomaly that is a predisposing factor (ibidem).

Poland's syndrome is, compared with Möbius syndrome, in this sense a more frequent vascular anomaly located in the distal portions of the central nervous system, extremities and tongue. Poland's anomaly (sequence) is considered a subset of the manifestations of Möbius syndrome and is concurrent with the latter in 15 % of patients. It occurs, however, more frequently separately and can be considered a microform of Möbius syndrome (25).

Möbius syndrome

Möbius syndrome affects the VIth and VIIth cranial nerves with manifestations of bilateral central paresis, less frequently with unilateral manifestations (29). Pareses of the IIIrd, Vth, IXth and XIIth cranial nerves are less frequent. The first manifestations at a neonatal age are manifested by a mask-like face due to congenital bilateral damage of the lower motor neuron.

Just as Möbius syndrome and Poland's anomaly overlap, the same may happen in the syndrome of hypoglossia-hypodactyly. The common denominator of the two nosological units can be unilateral hypoplasia of the tongue. For Poland's syndrome, on the other hand, athelia and polythelia are typical (34).

For Möbius syndrome the orofacial symptomatology is typical with fasciculations of the tongue, poor speech and pronunciation, and medium or mild hypoplasia of the mandible that calls for orthodontic treatment (6). For Möbius syndrome, a defect of the elastic cartilage of the ear is typical and retraction of the middle portion of the face. Infants with this affliction have difficulties with ingesting food, suffer from hypersalivation, have a small mouth and the function of the soft palate is inadequate.

The orthopedic symptomatology of Möbius syndrome involves in 50 % of cases defects of the extremities, incl. hypoplasia and syndactyly in 20 % of cases and deformities of the extremities in 30 %. Less frequent manifestations are clinodactyly, brachydactyly, polydactyly, contractures, dysplasias of the coxae and fibrous ankylosis of the joints, e.g. of the temporomandibular joint (15, 20).

Less frequent manifestations of Möbius syndrome are organ defects such as congenital heart disease, genitourinary abnormalities and hypogenitalism.

Ophthalmologic defects include nystagmus, ptosis of the eyelids, strabism, epicanthus and corneal ulcerations. Only in 10–15 % of cases are mental disorders due to mental retardation observed. The patients of school age are usually sociable.

Neurological manifestations with disorders of motor functions are manifested as a mild spastic diplegia. Supranuclear deafness was shown by the method of evoked potentials. The absence of the pectoralis major and minor muscle may by associated with a defect of the m. trapezius, m. quadriceps femoris, m. serratus and m. semimembranosus and other muscle anomalies. Neurological manifestations are one of the dominant symptomatologies. Paresis or plegia of the oculomotor nerves is found. The disorder of the XIIth cranial nerve is manifested by an immobile palate. Impaired deglutition is potentiated by a micromandible. In some children the facial motility improves with advancing age (23).

The heredity can be autosomal dominant with a variable expression, at least in some cases, in relatives microsymptoms in the orofacial area without a reducing acral defect are found. Despite the fact that autosomal dominant transmission or polyfactorial determination is involved, the majority of patients have mostly sporadic fresh mutations with a restricted reproductive possibility of the carriers – reduced reproductive fitness (2, 9).

The gene location of the syndrome is not clear so far; however, on the basis of studies of reciprocal translocations at the site of breakage 1p23 and 13q13 in a family with Möbius syndrome, it seems to be related with chromosome 1 and 13 (42).

Poland's syndrome

Poland's syndrome (35) is, compared with Möbius syndrome, a better known, more frequently diagnosed syndrome. In 1980 there were some 300 published cases while in 1983 already some 500.

The minimal diagnostic criteria of Poland's syndrome are unilateral aplasia of the m. pectoralis major and an ipsilateral anomaly or defect of the hand. Facultative manifestations include absence of the nipple of the mammary gland and, in women, inadequate development of the mammary gland.

It has been classified into three sub-groups depending on 3 defomities of the hand (26), described as A, B, C.

The most frequent (A) is symbrachydactyly associated with brachymesophalangia without oligodactyly – i.e. 25 % of these patients have shorter fingers II to IV with thicker medium phalanges.

In group B the distal phalanges of the IInd to IVth fingers are absent.

Group C suffers from ectrodactyly (cleft) of the hand with an absence of other defects of the carpal bones. This group is rare.

Main symptoms: The syndrome includes (see above) aplasia of the m. pectoralis major and minor. At the same time there is hypoplasia or aplasia of the homolateral thoracic wall. Frequently, flattening of the ipsilateral defect of the chest is observed and rarely an osseous defect of the chest or the absence of fingers, ankylosis of fingers, or facultative hypoplasia of the forearm and arm.

The biomechanical interpretation of the symptomatology of the chest is that it is deformed due to aplasia of the pectoralis major muscle, and externally the m. serratus anterior is apparent.

Organ disorders are not typical – facultatively, supernumerous mammary glands are present as part of so-called polythelia. Disorders of the major vessels are fairly frequent such as coarctation of the aorta and dextrocardia (7). Individual hernias of the pulmonary tissue into intercostal spaces are observed as well as inguinal and umbilical hernias (26). Goldberg and Mazzei (13) describe microcephaly and occipital encephalocele, Rattan et al. (37) also a cleft in the area of the lumbosacral spine. Hemivertebrae were also described.

The laterality of findings and the sex ratio are remarkable. A predilection towards laterality is described by various authors. In Poland's syndrome mostly boys are affected; according to Lazjuk the ratio is 3 : 1, while in Möbius syndrome the ratio is 1 : 1. The results reported by various authors can differ (26). The majority (45 %) reported in the literature are dextrolateral. The remainder are partly bilateral or with a crossed symptomatology.

The etiopathogenesis is in many respects still obscure (26). Mostly sporadic cases are involved. A monogenic autosomal dominant transmission has also been described. There exist several syndromological descriptions in siblings and in the offspring of patients, not only in the latter, but also in remote relatives. The etiopathogenesis was studied, e.g., by Goldberg and Mazzei (13), Bouvet et al., (5), David (10) and Lazjuk (26).

In general a heterogeneous etiopathogenesis is assumed, as a rule local disorders of morphogenesis, while a minority are caused by multifactorial genetically determined stenosis of the subclavian artery (36).

Genetic counseling and consultations, therapy and management: According to experience, there is no substantially increased empirical risk of re-occurrence of the defect in a subsequent pregnancy in families with one affected child, if neither of the parents is affected. Within the framework of genetic consultation in sporadic cases, a detailed examination of the parents of sick children is essential, focused on stigmas such as anomalies of dermatoglyphics and minor shortening of the palms. There may also be micromanifestations of the above symptoms (if they are absent, the prognosis is favorable; if there are minimal manifestations, a risk up to 50 % in the offspring must be foreseen).

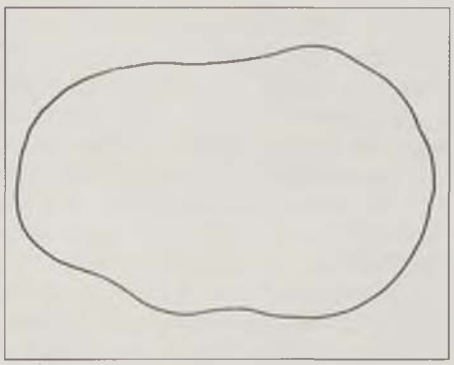
Therapy and management: Patients with Poland's syndrome frequently need plastic surgery of the phenotype (33). Also, more severe deformities of the thoracic wall call for an operation by a thoracic surgeon (28, 12, 18, 19, 14, 21, 16). An integral part of therapy is genetic counseling, governed by the general principles outlined above, incl. counseling such as psychotherapeutic consultations. At a later age, treatment of rheumatoid arthritic manifestations is necessary.

Differential diagnostic considerations: The latter must also include other syndromes of the orofacial disruption spectrum such as glossopalatinal ankylosis, Hanhart's syndrome and the hypoglossia-hypodactyly syndrome.

CASE-RECORDS

Case 1

Patient born 13. 6. 1962, child of healthy non-related parents, born of first pregnancy (Fig. 1a,b).



a | b

Fig. 1. Case 1: Phenotype of patient with Poland's syndrome – typical unilateral deformity of the chest on the right side caused by aplasia of the m. pectoralis major (a); cyrtogram at age of 26 years and 3 months, chest circumference recorded by soft wire preserving the structure (b).

In the phenotype he has hypoplasia of the right scapula, clavicle, and the second to fifth rib on the right side and aplasia and deformity of the right pectoralis muscle. No other developmental defects were revealed. The patient surffers from hypotension, but congenital heart disease was ruled out. Syndactyly is not present.

The phenotypic picture shows a patient of above average height, 183 (+ 0.84 SD) and a body weight of 86.6 kg (+ 0.86 SD). The length of the anterior wall of the trunk and lower extremities is normal. As to circumferential measures in relation to height, only the circumference of the abdomen is above average (1.38 SD). Skinfolds are of average value with the exception of an above-average skinfold above the triceps (+ 2.4 SD) and on the abdomen (+ 2 SD). There were no significant deviations on cephalometry; the patient is a mesocephalic and mesoprosopic type. The length of the upper extremity is below average (index in relation to height -2.2 SD).

The width dimensions confirm extremely narrow shoulders, the biacromial index to the body height being -4.4 SD. The distance of the right acromiale from the median plane is reduced due to the deformity of the chest. The deformity of the chest was confirmed by a cyrtogram. The pelvis is relatively broad; the acromiocristal index is significant increased by 4.3 SD. The sagittal diameter of the chest is reduced in relation to the transversal diameter by 1.2 SD, and the thoracic index is reduced by 1.2 SD; the index of the chest circumference in relation to body height is normal.

Dermatoglyphs: On the fingers there is a predominance of ulnar loops; there are whorls only on the thumb of the upper left extremity and on the index finger of the right hand. The pathways of the main papillary lines have a normal physiological pattern. On the right hypothenar is a radial loop; in both IIIrd interdigital spaces are distal loops. The atd angles are in normal positions, and there is nothing remarkable about the flexion ridges.

The total ridge count is 167, i.e. above average.

The right hand is shortened. The IInd, IIIrd and Vth middle phalanges of the fingers of the right upper extremity are shorter: II. 2 cm on the right as compared with 2.5 cm on the left, III. 2.4 cm on the right as compated with 3.5 cm on the left, IV. exception – no shortening: 2.5 cm is the length of the middle phalanx on both hands, V. 1.5 cm on the right, on the left no shortening – 2 cm.

The patient has a healthy sister; their father suffers from nephrolithiasis, while the mother is in good health. The father's sister suffers from a gastric ulcer and primary hypertension; her son and daughter are in good health.

The grandparents with the exception of the mother's mother are dead; the cause of death of the father's mother was bronchogenic carcinoma at the age of 61 years.

No marriage among relatives was found back to the third generation.

Genetic counseling: The empirical risk of a re-occurrence of the defect is low, although a new dominant mutation cannot be ruled out. Microsymptoms were not detected in either the parents or the sister. During a future pregnancy of the spouse, ultrasonographic checks of fetal development were recommended, possibly foetoscopy. Preconception care is also indicated for the wife.

Case 2

The genetic ambulance was visited on the recommendation of a gynecologist by a pregnant mother of a child with Poland's syndrome. The patients was born on 13. 8. 1953. She had menarche at the age of 13 years, regular cycle 28/7 days, painful. As regards occupational exposure to potential teratogens and mutagens, she worked with benzene for cca 6 months before her first pregnancy, and again before the second pregnancy. She had not suffered from any serious diseases, only the usual exanthemic diseases of childhood incl. German measles.

The patient's husband has a history of rheumatic fever, but no other serious diseases.

Genealogical examination: Three siblings of the proband are in good health; one niece suffers from sideropenic anaemia. The patient's mother is one of monozygotic twins; her sister died at the age of 20 years from leukemia. The brother of the proband's mother died as a child. The proband's father had repeated myocardial infarctions and suffers from crural ulcers. The husband's sister has two healthy children and has no problems herself. The husband's parents are in good health, further antecedents are without problems, and no marriage between relatives was found back to the third generation.

History of pregnancy: The proband delivered a son on 17. 9. 1982 after her first pregnancy, complicated by inflammation of varicose veins. The son suffered during delivery a fracture of the left clavicle and only after three days syndactyly of the IInd to Vth finger on the right side, while the absence of the pectoralis major muscle was manifested only later during growth at the age of 2 months.

At the time of consultation the second pregnancy was under way, (XIIIth week of pregnancy) without complications, only slight nausea. Ultrasonographic examination confirmed the vitality and integrity of the pregnancy; a further check-up was made two months later. The cardiac activity was regular, the fetal movements adequate, no gross deviations in the morphology of the fetus were found, and the size of the fetus corresponded to the stage of pregnancy. Delivery of a healthy son occurred during the 40th week, 3480 g/50 cm, spontaneous normal delivery.

At the age of 3 years, the height of the patient is within medium range and the body weight corresponds to about the 50th percentile (Figs 2–4). The head and chest circumference are within nor-

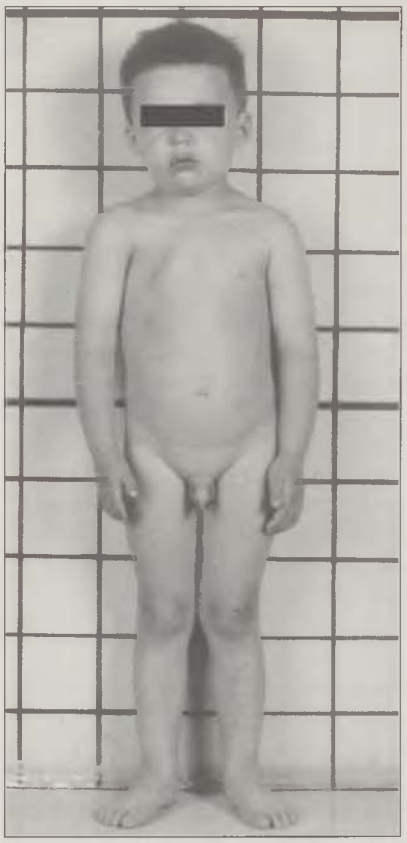


Fig. 2. Case 2: Phenotype of patient with Poland's syndrome – unilateral deformity of the chest on the right caused by aplasia of the m. pectoralis major, on the same side syndactyly of the hand.

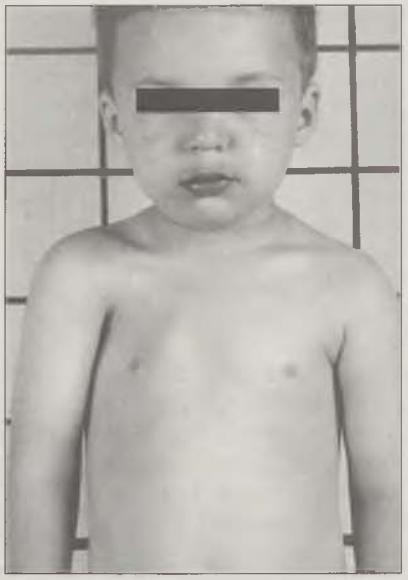


Fig. 3. Case 2: Photograph of the chest shows in detail a dextrolateral deformity with an asymmetric pattern of the clavicles at the age of 3 years.

mal range. The lower portion of the body is prolongated, and the difference in relation to the body height is +1.5 SD. The width measurements and the circumferential measures are within the normal range. As to cranial dimensions, the reduced width of the face is striking. Marked asymmetry affects the upper extremities. The length of the arms on both sides are the same, the forearm displays a dextro-sinistral difference of 3 mm, the length of the hand is 15 mm, and the width of the hand 6 mm. The asymmetry also affects soft tissues as ensues also from the values of the circumferential measures, where the lateral deviations of the arm circumference are 6 mm.

Fig. 4. Case 2: Detail of patient, posterior view. Note the higher position of left scapula and conversely hypoplasia and the lower position of the right scapula.

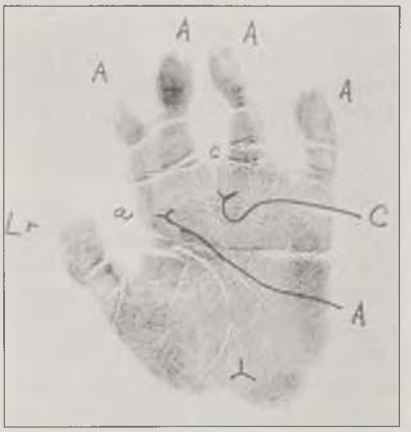


Fig. 5. Case 2: Dermatoglyphs of the right hand – reduction of number of triradii, consistent with syndactyly.

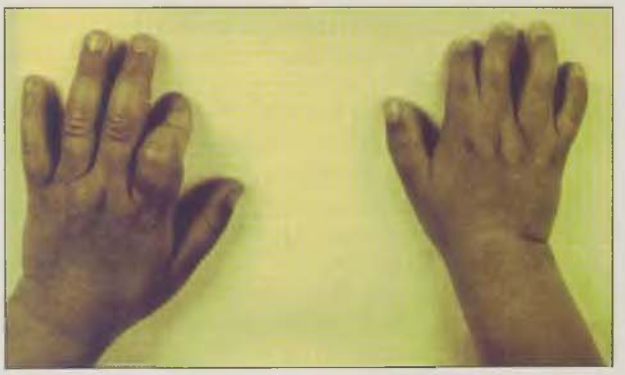


Fig. 6. Case 2: Detail of the hands.

The psychomotor development is adequate and the patient prefers the left hand.

Dermatoglyphic examination (Figs 5, 6): On the right side on the extremity afflicted with

syndactyly of the 2nd and 3rd finger, a classical four-finger ridge and a quite atypical pattern of subdigital triradii. Only the a and c radii are present and 2 unusual ulnar loops; on the right thumb a radial loop. The axial triradius is in the basic position t; the angle cannot be evaluated due to the absence of the triradius. On the other fingers of the right hand only arches are found. On the left, unaffected hand there are no remarkable dermatoglyphic findings.

The patient's mother has the main papillary lines without any remarkable features; the axial triradii are in the basal positions t. The ab distances are somewhat smaller – on both hands always 36. On the right hand a transient four-finger crease. On the fingers a predominance of ulnar loops, TRC = 144; there is a relatively higher frequency of secondary flexional creases. On the right hand there are lower values of the ab ridge count 31, while on the left side 32; bc ridge count 23 and 27, respectively. Distal loops were found the IVth interdigital spaces on both sides.

The patient's father has on both hands a pattern of papillary lines without any remarkable features. The axial triradii are in the basic positions t. Atypical features were found in the IVth interdigital spaces on both sides; on the left thenar is a carpal loop and indicated four-finger ridge. On the fingers is a predominance of ulnar loops with TRC 106. On the right hand there are lower values of the ab ridge count – 32, while on the left side 41; bc ridge count on the right 21 and on the left 26; cd ridge count on the right is 15 and on the left 22.

Case 3

The patient, a 35-year-old man, was followed-up on a long-term basis due to Poland's syndrome and asymmetry of the chest. In the phenotype: ptosis, strabism, asymmetry of the chest, and hypoplasia of the pectoralis minor muscle on the left.

He is the child from a second pregnancy, preceded by a spontaneous abortion complicated by maternal hypertension and influenza infection with imminent abortion. The delivery was at term with section due to a rupture of the uterus, with a birth weight of 2600 g and length of 47 cm. Dysplasia of the hips became evident at the age of 3 months and strabism at the age of 6 months.

The first manifestations of the syndrome were apparent during the 6th to 9th months, first scapula alta, with asymmetry of the chest only appearing later. The phenotype at the age of four years was characterized by the left scapula in a more cranial and, at the same time, in a discrete medial position. The spine deviates slightly to the left in the Th region. Aplasia of both pectoralis muscles was found on the left side. Both anterior costal ends are invereted parasternally. A considerable degree of valgosity of the knees was observed. At the age of 6 years the patients still suffered from enuresis, which persists intermittently.

Signs of Möbius sequence were also found at the age of 6 years: ptosis of the eyelids, asymmetry of mimics, hypofunction of the facial nerve, convergent strabism, left deviation of the tongue, hypofunction of n. XII sin., n. III, IV and VI on the right side.

Evaluated as the hyperactive form of mild cerebral dysfunction.

Dermatoglyphs were obtained by the mirror technique using color and foils. There is a predominance of simple patterns of ulnar loops. Simple flexional (palmar) ridges (ape) were not found; radial loops are present on the hypothenars, and there are also atypical features in the region of the c, d triradii.

He has 46,XY normal karyotype.

The above changes were expressed objectively by anthropological methods.

The mother was treated unsuccessfully for infertility (postinflammatory changes).

Case 4

The patient comes from a family with an isolated incidence of Möbius syndrome, age 12 years at the time of the first examination, from the 3rd pregnancy.

During her first pregnancy, his mother had a spontaneous abortion during the 10th week and concomitant myomatosis of the uterus; from the second pregnancy a healthy sister of the patient was born. After termination of the pregnancy, the mother underwent a hysterectomy. During pregnancy with the proband the mother had excessive weight increments (21 kg during the 40th week of pregnancy). The birth weight of the patient was 3200 g, the length 50 cm; the parents' ages at the time of delivery were 29 (mother) and 26 years (father). The immediate post-partum adaptation was satisfactory, the psychomotor development normal.

The defect was assessed after delivery as paresis of the VII cerebral nerve along with the presence of acral defects, syndactyly of the upper right extremity and shortening of the hand, with cleft of the 2nd and 3rd finger (Fig. 7). The left lower extremity is characterized by symbrachydactyly.

From birth his face has the typical appearance of a mask with dropping anguli oris and ptosis of the eyelids; he cannot close the lips completely, and the mouth is extremely small (Figs 8, 9). The patient is unable to close the eyelids completely. Occasional spontaneous movements of the bulb are also observed.

Important anthropometric parameters at the age of 12 years (Figs 10a, b): right hand shortened by 3 standard deviations, left planta by -1.8 SD, circumference of right forearm by -2.4 SD. The neurocranium was narrow (-2 SD), similarly as the cranial base (-3.4 SD) and the bizygomatic distance (-3 SD). The left lower extremity was reduced by -1.5 cm.

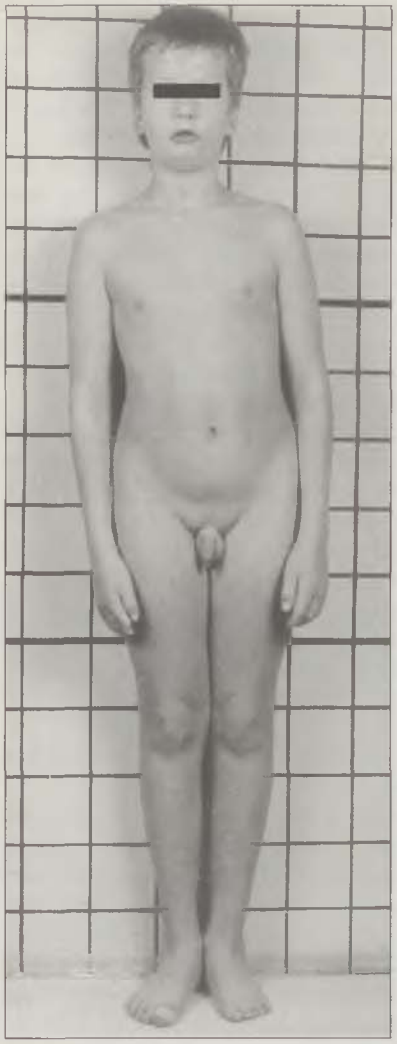


Fig. 7. Case 4: Möbius syndrome, whole figure at age of 12 years – scoliotic posture, low position of the right nipple, symbrachydactyly of the left foot.

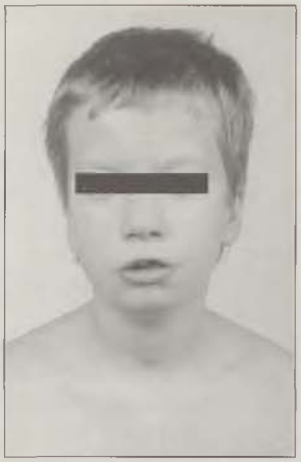


Fig. 8. Case 4: Möbius syndrome, detail of the face – slight paresis of facial muscles, masklike face, eversion of the lower lip (for detailed characteristics, see text).

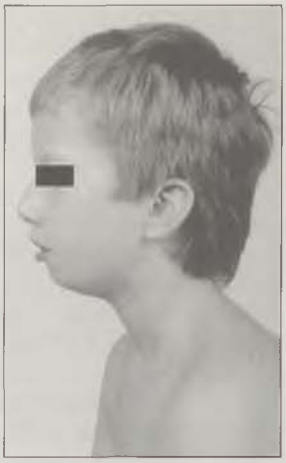
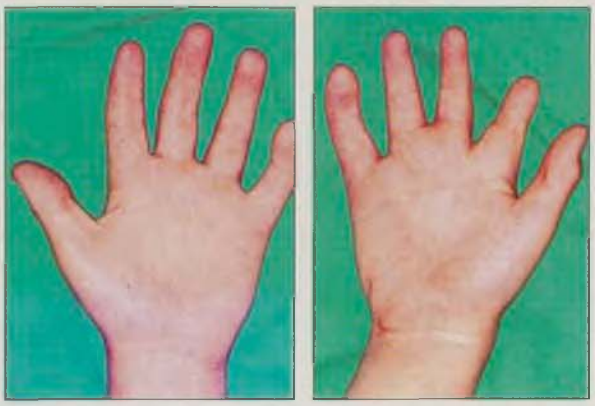


Fig. 9. Case 4: Profile of head and face.



a b

Figs 10 a,b. Case 4: Palms – cleft of the right palm between the 2nd and 3rd fingers.

The development of the chest can be evaluated as asymmetric, hypoplasia to aplasia of the pectoral muscles on the right side and associated sinistroconvex scoliosis of the thoracic spine. The nipple on the right side is low.

On intraoral examination at the age of 12 years, a marked cariosity of the teeth was revealed and crossed occlusion in the frontal area. The permanent molars were extracted in all quadrants due to destruction by caries. For preservative reasons the frontal teeth are provided with protective crowns made from precious metals. The eruption of teeth corresponds to the patient's age. The tongue is in a central position, and hypoplasia of the tongue was not found. The palatine arches are symmetrical; the motility of the soft palate is restricted. Due to microstomia, opening of the mouth is restricted; the interincisal distance is 24 mm. The area of the TM joints is not painful on palpation, when opening and closing the mouth without pathological phenomena. The patient's speech is faint and mumbling. While rinsing his mouth during dental examinations, he supports his lower lip to prevent water escaping from his mouth. The patient attends elementary school with average progress.

The described typical appearance of the face is due to the central paresis of the VIth and VIIth cranial nerves, and as a result to impaired innervation of the m. rectus bulbi temporalis and the mimic muscles.

The impaired motility of the soft palate, which is the main cause of the impaired speech, also indicates an affliction of the IXth cranial nerve, the ramus styloglossus of which innervates the muscle bearing the same name, as well as the m. glossopalatinus and levator veli palatini. The impaired speech is due to the weakened mimic muscles. The increased cariosity of teeth is explained by poor hygiene of the oral cavity. The small oral cavity makes perfect cleaning of the teeth difficult, while the poor mobility of the deformed hands contributes to the inadequate brushing of the teeth. The weakened mimic muscles have a reduced role in self-cleansing.

Anthropometric examination of the proband's sister does not reveal marked deviations that would indicate any minimal symptomatology. In the proband's sister, prenatal diagnosis was made by ultrasonography and examination of the amnotic fluid. The results revealed a normal male karyotype, 46,XY. She gave birth to a healthy boy; the intrauterine and postnatal development of the child is quite normal.

Case 5

The patient is an 29-year-old woman. She was the child of the second pregnancy of healthy, un-

related parents. The first pregnancy of her mother was terminated by induced abortion. The client was born at full term with a low birth weight of 2 700 g. In the pedigree a marriage between relatives was ruled out back to the 3rd generation.

A congenital defect such as Poland-Möbius syndrome was assessed immediately after delivery with a dextrolateral localization, suffering from syndactyly of the fingers of the right hand, a smaller palm, absence of the right pectoralis major muscle and left-sided paresis of the facial nerve. Her mental and psychomotor development were normal; later she developed thoracolumbar scoliosis of the spine with shortening of the right lower extremity by 1 cm. The right nipple is hypoplastic; after puberty hypoplasia of the right mammary gland was found. She also suffers from myopia and astigmatism and from partial bilateral paresis of the VIth cranial nerve. Compression of the teeth in both jaws.

Height 163 cm, body weight 64 kg, cardiorespiratory system compensated. Anthropometric examination revealed hypoplasia of the right upper extremity (-2.7 SD incl. shortening of the forearm by -2.4 SD).

Had long-term preconception care, and from her third pregnancy she gave birth to a healthy daughter. Ultrasound examinations made did not reveal congenital defects of the extremities as a signalizing phenotype.

The dermatoglyphic findings are quite atypical with low frequencies of papillary lines on the fingers with reduced TRC (30) and atypical features of the palmar triradii, where their reduction is associated with syndactyly on the affected right hand. Simple digital patterns such as arches are amply represented. There are also atypical flexional ridges on the palm. The non-afflicted extremity on the left side serves as an intraindividual control.

The history of the proband's pregnancy is also remarkable. The first pregnancy at the age of 26 years ended by a spontaneous abortion after 3 months. The second pregnancy at the age of 27 years also failed – a spontaneous abortion was induced during the 20th week of gestation by means of prostaglandins because congenital heart disease was diagnosted – hypoplasia of the left heart, female fetus.

Genetic counseling focused on a possible elucidation of repeated defects in the offspring, consedered by the authors as an association of Poland's and Möbius syndrome. Autosomal, dominant heredity with a 50% risk of re-occurrence in the offspring of the affected woman could not be ruled out.

DISCUSSION

The combination of Möbius and Poland's syndrome is described in the literature rather rarely. We found this association in one of our female patients. At least in some of the cases a disruption variability is involved, as mentioned, e.g., by Larrandaburu et al. (25). Also the contralateral involvement of the extremities in this syndrome is of interest (39). Minguella and Cebrera (31) presented a report on a large number (38) or patients from Spain, 28 boys and 10 girls. There was a slight predominance of leftsided afflictions, 20/18. This is a relatively new finding as compared with the formerly reported lateral predilection.

The authors paid attention to anomalies of the hand and divided the patients, based on clinical and radiological findings, into five groups – without syndactyly, with syndactyly and brachymesophalangy (hypoplasia or aplasia of the middle phalanx), type 3 with syndactyly incl. the thumb, type 4 – longitudinal deficiency of some fingers or the radius and type 5 – with a transversal absence of the skeleton of the hand.

The most frequent anomaly is aplasia of the middle phalanx of the fingers (15 cases) or its hypoplasia (another 15 cases). We also observed this finding in one of our case-records. A cleft hand was observed in two patients with manifestations of Möbius syndrome but not in isolated Poland's syndrome.

Anesthesiologists draw attention to possible complications of anesthesia in Poland's syndrome caused by paradoxical breathing due to the deformity of the chest wall (38). In this syndrome emphysematous bullae were also described (21).

There are also reports that syndactyly in Poland's syndrome is usually more serious from a functional aspect than other types of syndactyly, even when surgery was successful (22).

Dysontogenetic changes predispose in general also to oncogenic lability of the organism. Evidence of this effect was provided by the work of Athale and Warrier (1) describing Wilms tumor in a patient with Poland's syndrome. The position is also complicated by the fact that patients with Poland's syndrome are more liable to develop leukemia, and this must be taken into account during radiation treatment and chemotherapy; therapeutic modifications are necessary. As to other tumors in Poland's syndrome, neuroblastomas have been described (8).

Poland's syndrome also holds an important place within the framework of morphological anomalies of the chest and mammary gland in girls (14). It is thus an important field for plastic surgery. Reconstructive surgery is focused on the thoracic wall, incl. implants (18) and mammary glands making use of silicone (12, 28).

As a disorder of mesodermal tissues is involved, the finding of hemocoagulation disorders is not surprising (11). The vascular etiology is suggested by the joint finding of a defect of the atrial septum and Poland-Möbius syndrome described by Matsui et al. (30). Bouvet (1995) mentions atypical features of the blood supply of the afflicted extremity by plethysmography. Hy-

poplasia of the subclavian artery is assumed – on the same side at the affect extremity. Obviously, this is not so in all patients but, on the other hand, a common disruption sequence „subclavian artery supply disruption sequence" (SASDS) is assumed, which explains the common pathogenesis not only of Poland's and Möbius syndrome but also of Klippel-Feil's syndrome (sequence), isolated terminal acral defects and Sprengel's anomaly. The critical period of this abnormal pathogenesis (dysontogenesis) is the sixth week of gestation. Explanation for the vascular theory obviously does not apply to cases with a contralateral symptomatology (36).

REFERENCES

1. Athale, U. H., Warrier, R.: Poland's syndrome and Wilms tumor: an unusual association. Med. Pediat. Oncol., 30: 67-68, 1998.
2. Baraitser, M.: Genetics of Meobius syndrome. J. med. Genet., 14: 415-417, 1977.
3. Beer, G. M., Komopatscher, P., Hergan, K.: Poland's syndrome and vascular malformations. Br. J. plast. Surg., 49: 482-484, 1996.
4. Bouvet, J. P., Maroteaux, P., Briard-Guillemot, M.: Poland's syndrome: clinical and genetic studies – physiopatology. Nouv. Press med., 5: 185, 1976.
5. Bouvet, J. P., Leveque, D., Bernetieres, F., Gras, J. J.: Vascular origin of Poland syndrome? Eur. J. Pediat., 128: 17-26, 1978.
6. Braye, F., Souchere, B., Franc, C., Freiderl, M.: Mobius syndrome: therapeutic proposal from 2 cases. Rev. Stomat. Chir. Maxillofac., 97: 332-337, 1996.
7. Burkhardt, H., Buss, J.: Dextrocardia and Poland syndrome in a 59 year old patient. Z. Kardiol., 6: 639-43, 1997.
8. Caksen, H., Patiroglu, T., Ozdemir, M. A., Patiroglu, T. E., Poyzaroglu, M. H., Tercan, M.: Neuroblastoma and Poland syndrome in a 15-year-old boy. Acta paediat. jap., 39: 701-704, 1997.
9. Collins, D. L., Schimke, R. N.: Moebius syndrome in a child and extremity defect in her father. Clin. Genet., 22: 312-314, 1982.
10. David, T. J.: Nature and etiology of the Poland anomaly. New Engl. J. Med., 287: 487-489, 1972.
11. Fedorov, J. J., Rozanova, L. S.: Disorders in the hemostatic system in the Poland syndrome. Vestn. Khir., 156: 78-79, 1997.
12. Gatti, J. E.: Poland's deformity reconstructions with a customized, extrasoft silicone prosthesis. Ann. plast. Surg., 39: 122-30, 1997.
13. Goldberg, M. J., Mazzei, R. J.: Poland syndrome: a concept of pathogenesis based on limb bud embryology. Birth Defects: Orig. Art Ser., 13: 103-115, 1977.
14. Grolleau, J. L., Pienkowski, C., Chavoin, J. P., Costagliola, M., Rochiccioli, P.: Morphological anomalies of breasts in adolescent girls and their surgical correction. Arch. Pediatr., 4: 1182-1191, 1997.
15. Hanson, P. A. et al.: Moebius syndrome and facioscapulohumeral muscular dystrophy. Arch. Neurol., 24: 31, 1971.
16. Harpf, C., Schwabegger, A., Hussl, H.: Carpal median nerve entrapment in a child associated with Poland's syndrome. Ann. plast. Surg., 42: 458-459, 1999.
17. Henderson, J. L.: The congenital facial diplegia syndrome: clinical features, pathology and aetiology. A review of sixty one cases. Brain, 62: 81-403, 1939.
18. Hodginson, D. J.: Chest wall implants: their use for pectus excavatum, pectoralis muscle tears, Poland's syndrome, and muscular insufficiency. Aesthetic. Plast. Surg., 21: 7-15, 1997.
19. Jasonni, V., Lelli-Chiesa, P. L., Repetto, P., Torre, M., Nobili, F., Mazzolas, C., Martucciello, G.: Congenital deformities of the chest wall. Surgical treatment. Minerva pediat., 49: 407-413, 1997.
20. Jorgenson, R. J.: Moebius syndrome, ectrodactyly, hypoplasia of the tongue and pectoralis muscles. Birth Defects, 7: 283-284, 1971.
21. Kondo, N., Koshima, R., Sakurada, T., Kikuchi, Y., Kusajima, K.: A case of Poland's syndrome with giant bulla resected by using autologos fascia lata. Koybu-Geka, 51: 343-345, 1998.
22. Kramer, R. C., Hildreth, D. H., Brinker, M. R., Bennett, J. B., Thompson, L., Lumsden, R. M., Cain, T. E.: A comparison of patients with different types of syndactyly. J. Pediat. Orthop., 18: 233-238, 1998.
23. Krueger, K. E., Friedrich, D.: Familiare kongenitale Motilitaetstoerungen der Augen. Klin. Mbl. Augenheilk., 142: 101-117, 1963.
24. Kupper, H. J.: Anesthesia in Poland syndrome (letter). Can. J. Anaesth., 46: 513-514, 1999.
25. Larrandaburu, M., Schuler, L., Ehlers, J. A., Reis, A. M., Silveira, E. L.: The occurence of Poland and Poland-Moebius syndromes in the same family: further evidence of their genetic component. Clin. Dysmorphol., 8: 93-99, 1999.
26. Lazjuk, G. I., Lurie, I. V., Čerstvoj, E. D.: Nasledstvennyje sindromy množestvennych vroždennych porokov rozvitija. Medicina, Moskva 1983.
27. Legum, C., Godel, V., Nemet, P.: Heterogeneity and pleiotropism in the Moebius Syndrome. Clin. Genet., 20: 254-259, 1981.
28. Longaker, M. T., Glat, P. M., Colen, L. B., Siebert, J. W.: Reconstruction of breast asymmetry in Poland's chest wall deformity using microvascular free flaps. Plast. reconstr. Surg., 99: 429-436, 1997.
29. Masaki, S.: Congenital bilateral facial paralysis. Arch. Otolaryng., 94: 260-263, 1971.
30. Matsui, A., Nakagawa, M., Okuno, M.: Association of atrial septal efect with Poland-Moebius syndrome: vascular disruption can be a common etiologic factor. A case report. Angiology, 48: 269-271, 1997.
31. Minguela, S. J., Cabrera, G. M.: Poland's syndrome. A report of 38 cases. An. esp. Pediat., 48: 143-147, 1998.
32. Moebius, P. J.: Ueber angeborenen doppelseitige Abducensfacialis Laehmung. Munch. Med. Wochenschr., 35: 91-108, 1988.
33. Paulhe, P., Magalon, G.: Expansion in breast surgery. Ann. Chir. plast. esthet., 41: 467-480, 1996.
34. Perez-Aznar, J. M., Urbano, J., Garcia-Laborda, E.: Breast and pectoralis muscle hypoplasia. A mild degree of Poland's syndrome. Acta Radiol., 37: 759-762, 1996.
35. Poland, A.: Deficiency of the pectoral muscles. Guys Hosp. Rep., 6: 191, 1841.
36. Powell, C. V., Coombs, R. C., David, T. J.: Poland anomaly with contralateral ulnar ray defect. J. med. Genet., 30: 423-424, 1993.
37. Rattan, K. N., Budhiraja, S., Pandit, S. K.: Poland's syndrome with meningomyelocele. Indian J. Chest Dis., 38: 259-261, 1996.
38. Sethuraman, R., Kannan, S., Bala, I., Sharma, R. K.: Anaesthesia in Poland syndrome. Can. J. Anaesth., 45: 277-279, 1998.
39. Silengo, M., Leroner, M., Seri, M., Boffi, P.: Lower extremity counterpart of the Poland syndrome. Clin. Genet., 55: 41-43, 1999.
40. Sprofkin, B. E., Hillman, J. W.: Moebius Syndrome: Congenital oculofacial paralysis. Neurology (Minneap.), 6: 50-54, 1956.
41. Stabile, M., Cavaliere, M. L., Scarano, G., Fels, A., Valiani, R., Ventruto, V.: Abnormal B. A. E. P. in a family with Moebius syndrome: evidence for supranuclear lesion. Clin. Genet., 25: 458-463, 1984.
42. Ziter, F. A., Wiser, W. C., Robinson, A.: Three-generation pedigree of a Moebius syndrome variant with chromosome translocation. Arch. Neurol., 34: 437-442, 1977.

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SOUHRNY

Význam vaskularizovaných šlach jako součásti volných laloků při periorální a tvářové rekonstrukci

Cunha-Gomes D., Kavarana N.

Účinek gravitace a proces stárnutí nutně vyžaduje závěs laloků užívaných při rekonstrukci obličejových tkání. Příkladem tohoto problému jsou periorální a tvářové rekonstrukce.

Nezbytný závěs lze získat použitím vaskularizovaných šlach jako součásti volně přenášených laloků. Neurokutánní lalok z radiální strany předloktí s vaskularizovanou šlachou m. palmaris longus a neurokutánní lalok z laterální strany paže s vaskularizovanou šlachou tricepsu jsou vhodnou možností pro rekonstrukci těchto defektů.

Autoři uvádějí 6 případů vyzdvihujících užití vaskularizované šlachy jako součástí volného neurokutánního kombinovaného laloku při perorálních a tvářových rekonstrukcích.

Kapsulární kontraktura u augmentační mammaplastiky

Dušková M., Sosna B., Kletenský J., Vrtišková J.

V souboru 331 augmentačních mamaplastik, provedených na Klinice plastické chirurgie v Praze v letech 1994–1998, se vyskytla kapsulární kontraktura v 6 % kosmetických a 12 % rekonstrukčních operací. Jako hlavní příčina vzniku se ukázala především kvalita tkáně okolo implantátu společně s individuální dispozicí. Výskyt této komplikace neměl statisticky významný rozdíl v závislosti na uložení protézy pod svalem či žlázou. Na podkladě histologického rozboru a klinického nálezu je zřejmé, že desintegrace protézy vždy nevede ke vzniku těžších stupňů kapsulární kontraktury, a to i přes přítomnost cizích těles ve vazivu.

Alternativní metoda rekonstrukce defektu po ztrátě více než poloviny horního víčka

Kokavec R., Fedeleš J.

Autoři popisují alternativní metodu rekonstrukce defektu při ztrátě více než poloviny horního víčka. V situaci, kdy standardní techniky nemohou být použity v důsledku specifických okolností a místního nálezu, popsaná metoda se jeví vhodnou alternativou pro rekonstrukci víčka.

Hodinová diuréza u rozsáhle popálených

Bláha J.

Hodnoty hodinové diurézy u 40 rozsáhle popálených pacientů byly sledovány a graficky vyjádřeny v období akutního popáleninového šoku. Rozborem hodnot a aktuálního stavu pacienta byl zjištěn významný vliv psychiky na průběh akutního popáleninového šoku a další léčby na JIP Kliniky popáleninové medicíny v Praze. Orientačně byly sledovány účinky některých léčiv na hodnoty hodinové diurézy.

Ulcus Marjolin

Königová R., Rychterová V.

Rakovinové ulcerace v jizvách byly popsány již Corneliem Celsem. V roce 1828 charakterizoval Jean Nicolas Marjolin vřed s maligní degenerací v jizvách po popáleninovém úrazu, ale v pozdějších publikacích byl popisován v jizvách nejrozmanitějšího původu. Typické je dlouhé latentní období od úrazu do vzniku maligního zvratu (průměrně 30 let). Patogeneze je dosud nejasná, ale upozorňuje se na tzv. double insult.

Klinicky se projeví jako mělký vřed nebo exofytický tumor, s nejčastější lokalizací na dolních končetinách.

Od roku 1978 do 1998 bylo hospitalizováno na Klinice popáleninové medicíny 3. LF UK v Praze 11 pacientů řešených radikální excizí s následnou autotransplantací. U 3 nemocných následovala poté laloková plastika a u 2 byla nezbytná amputace a exenterace ingvinálních uzlin. Zvláštním případem byl muž s kongenitální formou epidermolysis bullosa, u něhož byl ve věku 32 let histologicky prokázán dlaždicový karcinom na dolní končetině.

Za 4 roky v extrémní kachexii nastal exitus letalis, protože amputaci dolní končetiny opakovaně odmítal.

Z hlediska prevence je nutné doporučit:

– včasným uzávěrem defektů zabránit infekci a excesivnímu jizvení;

– jizevnaté kontraktury včasně řešit rekonstrukčními operacemi a zabránit jejich rozpadu;

– nehojící se defekty vyšetřovat biopticky;

– po radikální excizi dlaždicového karcinomu nepoužívat lalokový materiál, ale krýt postupně, ev. přes etapu provizorních biologických krytů, aby bylo možno sledovat místní nález.

Polandův-Moebiusův syndrom a disrupční spektrum postihující obličej a končetiny: přehledová studie a popis pěti pacientů

Kuklík M.

Autor shrnuje současné zkušenosti s klinickými a genetickými charakteristikami Polandova a Moebiusova syndromu. Je prezentováno 5 vybraných kazuistik s touto chorobou, respektive sekvenčním sledem Polandova-Moebiusova syndromu. Pro upřesnění symptomatologie má nezastupitelné místo antropometrické vyšetření, k objektivizaci asymetrie hrudníku tzv. kyrtogram, drátem zachycený obvod hrudníku. Z hlediska genetického poradenství je vždy aplikována prekoncepční péče u matek v rodinách s reprodukčním záměrem, stejně tak podrobné ultrazvukové vyšetření plodu v oblastech předpokládané končetinové symptomatologie (signální fenotyp). Ve dvou rodinách bylo využito ultrasonografie k prenatální diagnostice. Invazivní prenatální diagnostika pomocí amniocentézy byla navíc provedena v rodině s Moebiusovým syndromem. Dermatoglyfy mají u těchto rodin určité společné charakteristiky, jako tendence k jednoduchým vzorům. V širší rodině jsme u jednoho z našich pacientů zastihli u bratrance výskyt syndromu Parkes-Weber-Klippel-Trenaunay, což může ukazovat na společné dispoziční vaskulární faktory.

BOOK REVIEW

Mark May, Barry M. Schaitkin: **The Facial Nerve (May's Second Edition).**
Thieme, New York, Stuttgart, 2000.

In the preface the facial nerve is described as the second most burdened nerve in the human body after the vagus nerve. This is, however, only one of many reasons why so much attention is paid to it. The entire monograph devoted to the problem of the facial nerve contains 877 pages and is divided into six sections and a total of 40 chapters. Seven appendices follow. The whole work is conceived to provide complete clinical sources for all physicians who participate in the diagnosis and treatment of diseases of the facial nerve. It is based above all on the 35-year experience of the first author, who has treated more than 3500 patients suffering from these diseases. Moreover, 47 authors from all parts of the world participated in its preparation. The first section is devoted to applied basic scientific and clinical findings. Embryonic development, clinical anatomy, neurobiology, degeneration, regeneration and other basic processes with an impact on the biology of the nerve are discussed. Rather unusual is the chapter on the facial nerve in animals written by two veterinary surgeons. The chapter should facilitate possible model operations in animals.

In the second section methods suitable for general examination of the patient with paresis of the facial nerve are summarized. This is followed by a detailed section concerned with the treatment of patients with paresis of the facial nerve. Causes of the development of pareses – from injury to infectious diseases and others are discussed as well as the incidence of pareses in children and their causes etc. In the fourth section the authors analyze the causes and treatment of facial hyperkinesia, comprising treatment with botulotoxin, selective myectomy and microvascular decompression. For the surgeon, sections 5 and 6 are of greatest interest. They are devoted to surgery proper of the facial nerve and in particular to possible reanimation of the paretic face. The basic principles are defined and the principal techniques are explained step-by-step, supplemented by information on the selection of patients, indications, contraindications and the development of paresis. For example, after impairment of the trunk of the facial nerve, the best results of surgical treatment are achieved within 30 days after development of the paresis. When the facial nerve or another of the cranial nerves is used, the upper limit is cca 2 years after development of the paresis. As to basic techniques, the authors mention restoration of the nerve by a nervous graft, cross face grafts, anastomosis n. XII–VII, XII–VII jump graft and others. The chapter on transposition discusses in great detail the possibilities for using the m. temporalis, m. masseter and digastric muscle.

Somewhat in the background is the chapter on free muscle grafts, which are in general quite frequently used and preferred in some departments. In my opinion this chapter could be more extensive. On the other hand the chapter devoted to sequelae of facial nerve paresis for the function of the eye is very well elaborated. The authors mention all possibilities – from implantation of golden weights, palpebral springs, implantation of cartilage, tarsography, excision and others. For completeness sake, static methods are also mentioned, which include drop of the commissure and restored symmetry of the face as well as combinations of different methods at different time intervals to achieve a final result optimal for the patient. In the chapter on "Cosmetic Camouflage" the reader is informed how it is possible to improve the cosmetic effect by make-up, hairstyle, clothes and cosmetics.

Completely novel are the appendices that summarize the personal experience of patients with different types of damage of the facial nerve and its subsequent treatment and repair.

The whole publication is a unique summary of extensive information about an important cranial nerve. It will be useful for physicians engaged in theoretical disciplines, but in particular ENT specialists, ophthalmologists, surgeons, and plastic and maxillo-facial surgeons for whom the publication is a very instructive handbook of special surgial procedures.

Jiří Kozák, M. D.

AWARD OF THE G. WHITAKER INTERNATIONAL BURNS PRIZE FOR 2000 PALERMO, ITALY

At meeting held on March 25th, 2000, at the seat of the G. Whitaker Foundation, Palermo, after examining the scientific activity in the fields of research, teaching, clinical organization, prevention and cooperation presented by various candidates and in consideration of the high level of the candidates, the Adjudicating Committee unanimously decided to award the prize for 2000 to BASIL PRUITT, JR., M. D., Commander and former Director, US Army Institute of Surgical Research – Texas. The prize is awarded with the following motivation:

"Having graduated from a most prestigious University, Harvard, in 1957, Dr. Pruitt was drafted in the Army almost immediately, during the Vietnam war, and began a career in trauma and burns care in war and peace, which continues until today, over 43 years: for 27 years directed and commanded the US Army Burn Centre, now called the Institute of Surgical Research.

On retirement he continued his teaching and operating activity as Professor of Surgery at the University of Texas Health Sciences Center at San Antonio.

At all times he has operated, clinically treated, directed, guided research and administered burn care at the highest level, making his Centre the large internationally recognized Centre of burn excellence in the world.

An analysis of his many publications as primary author (among his 415 papers) reveals that he has shown deep interest and has brought scientific contributions to all the aspects of burns disease. It must be emphasized that burns disease concerns all the parameters of human physiology and pathology; these render Dr. Pruitt's work all the more capital. This global view is also evident in the work of his collaborators, colleagues, and researchers whom he has inspired."

G. WHITAKER INTERNATIONAL BURNS PRIZE – PALERMO (Italy)

Under the patronage of the Authorities of the Sicilian Region for 2001

By law n. 57 of June 14th 1983 the Sicilian Regional Assembly authorized the President of the Region to grant the "Guiseppe Whitaker Foundation", a non profit-making organisation under the patronage of the Accademia dei Lincei with seat in Palermo, an annual contribution for the establishment of a "G. Whitaker International Burns Prize" aimed at recognising the activity of the most qualified experts from all countries in the filed of burns pathology and treatment.

The amount of the prize is fixed at twenty million Italian Lire. The prize will be awarded every year by the month of June in Palermo at the seat of the G. Whitaker Foundation.

The Adjudicating Committee is composed of the President of the Foundation, the President of the Sicilian Region, the Representative of the Accademia dei Lincei within the G. Whitaker Foundation, the Dean of the Faculty of Medicine and Surgery of Palermo University, the President of the Italian Society of Plastic Surgery, three experts in the field of prevention, pathology, therapy and functional recovery of burns, the winner of the prize awarded in the previous year and a legal expert nominated in agreement with the President of the Region as a guarantee of the respect for the scientific purpose which the legislators intended to achieve when establishing the prize.

Anyone who considers himself to be qualified to compete for the award may send by January 31st 2001 his detailed curriculum vitae to: Michele Masellis M. D., Secretary-Member of the Scientific Committee G. Whitaker Foundation, Via Dante 167, 90141 Palermo, Italy.

ANNALS OF BURNS AND FIRE DISASTERS

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- varia

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NABÍDKA KNIH Z NAKLADATELSTVÍ MAXDORF

HEMODIALÝZA

Doc. MUDr. Sylvie Sulková, CSc. a kol.

Precizně a přehledně napsaná monografie, základní dílo oboru. Kapitoly pokrývají téma od historických poznámek a demografických dat, přes technické aspekty, zajištění cévního přístupu, vlastní provedení dialýzy, sledování pacienta až po přehled možných komplikací (akutních i chronických) a charakteristiku specifických situací (nutrice, dialýza u dětí, u diabetiků, psychosociální problematika, vztah dialýzy a transplantace). Zahrnují i alternativní hemoeliminační postupy včetně užití při intoxikacích a jsou doplněny základy farmakokinetiky při selhání ledvin. ***Kniha je určena:*** nefrologům, internistům, diabetologům, kardiologům.

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PRAKTICKÁ DIABETOLOGIE, 2. rozšířené vydání

Prof. MUDr. Vladimír Bartoš, DrSc., Doc. MUDr. Tereza Pelikánová, DrSc. a kol.

Druhé rozšířené a aktualizované vydání úspěšného knižního titulu. Autorský kolektiv předních českých diabetologů vytvořil příručku, která je bez nadsázky nepostradatelná pro každého lékaře. Diabetes mellitus se stal jednou z nejzávažnějších nemocí současnosti, a to jak z hlediska rozšíření, tak z hlediska následků a komplikací. Počet diabetiků v ČR je přibližně 500 000. ***Kniha je určena:*** každému lékaři bez rozdílu oboru.

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MUDr. Miroslava Navrátilová, Prof. MUDr. Eva Češková, DrSc., Doc. MUDr. Luboš Sobotka, CSc.

Kniha je podrobným průvodcem všemi stavy poruch příjmu potravy, se kterými se v psychiatrii lze setkat. Kniha obsahuje podrobné návody jak postupovat při stanovení diagnózy, kvantifikaci malnutrice a především jak dosáhnout úspěšné realimentace pacientů. Prevalence mentální anorexie (MA) u dospívajících dívek a mladých žen je 1%. Pacientky s MA se dožijí průměrného věku 33 let, každý 5. případ nemoci končí letálně. ***Kniha je určena:*** především psychiatrům, dále pak ostatním pracovníkům, kteří pečují o psychiatrické nemocné. Podrobné a praktické zpracování nutriční problematiky činí knihu vhodnou pro všechny lékaře, kteří se chtějí seznámit se základy klinické výživy jako takové

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Monografie je výsledkem spolupráce autorů převážně z Ústřední vojenské nemocnice a 3. LF UK FN Královské Vinohrady v Praze – chirurgů (otorinolaryngologa, neurochirurga, stomatochirurga a oftalmologa) s radiology. Většina publikovaných vyobrazení byla klinicky i operačně ověřena. Doplněna jsou schémata od jednoho ze zakladatelů české otorinolaryngologie prof. MUDr. Ervína Černého, DrSc. Publikace vznikla z naléhavé potřeby seznámit širokou lékařskou obec, rentgenové laboranty i střední zdravotnické pracovníky s novými zobrazovacími metodami hlavy, o kterých nebylo u nás od konce šedesátých let souborně publikováno.

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