

European Journal of

# Plastic Surgery

Organ of the  
European Association  
of Plastic Surgeons  
and the European  
Burns Association



Springer



0930-343X(200601)28:6;1-X



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## Invasive, aggressive basal cell carcinoma: carcinoma basocellulare terebrans—ulcus terebrans

Received: 28 July 2005 / Accepted: 8 December 2004 / Published online: 29 October 2005  
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**Abstract** The clinical picture, treatment and course of invasive, aggressive basal cell terebrans carcinoma of the face, scalp and neck in eight patients is presented in detail. Based on the analysis of these patients, as well as our experience of a total of 93 patients with aggressive, invasive basal cell carcinoma, treated so far, the conclusion is reached that basal cell terebrans carcinoma develops mainly on recurrences, that is on residual tumors, after inadequate surgical and radiation therapy. It was observed that the most aggressive tumors are those, which develop in residual tumors after radiation therapy. Due to extraordinary infiltrational growth, especially into deep structures (muscles, bones, cartilages, dura, and brain), even with extensive, mutilating operations one can never be sure that the tumor is radically removed. That is why in such cases, after extensive, mutilating operations, radiation therapy is recommended, if possible.

**Keywords** Skin tumors · Carcinoma · Basal cell carcinoma · Basal cell terebrans carcinoma

### Introduction

Basal cell carcinoma (carcinoma basocellulare, epithelioma basocellulare, basalioma) is an epithelial tumor of skin characterised by local infiltrative growth, sometimes even destructive, as with any carcinoma. Even though it is locally aggressive it very rarely metastasizes [2, 6], and thus it can be regarded as a semimalignant tumor. This tumor makes up 70% of all skin tumors, two-thirds of which occur in the upper face. Basal cell carcinoma tu-

mor cells originate from pluripotent primordial cells of the basal layer of the epidermis, outer area of the hair follicle, and other skin annexa [1, 3, 5]. The etiopathogenesis of the tumor is not fully understood. It probably develops by sincarcinogenous action of different exogenous and endogenous factors [6]. This tumor is clinically manifested in many forms. The most malignant one is that of carcinoma basocellulare terebrans—Ulcus terebrans. This begins as a rodent ulcer (the late phase of noduloulcerative form), and then infiltrates and destroys subcutis, fascia, muscle, cartilage, bone, meninges, and brain. In this way extensive mutilation occurs with accompanying deformity followed by bleeding from the tumor and secondary infection. The tumor occurs most frequently in the middle part of the face and the scalp. If it penetrates the orbit, destruction of orbital contents and loss of vision follows. If it penetrates the brain, the situation is usually lethal. In spite of the extensiveness of the destructive process there is only slight pain [4]. The most effective therapy is surgical [6].

The aim of this paper is to present, in more detail, some patients with the typical extensive, aggressive basal cell terebrans carcinoma, a different surgical treatment for the same patients, the results of this treatment, and also to share some observations on the characteristics and treatment of basal cell terebrans carcinoma. In addition to this, we wish to emphasize this entity and its behavior, which has been unjustifiably ignored in the literature for decades.

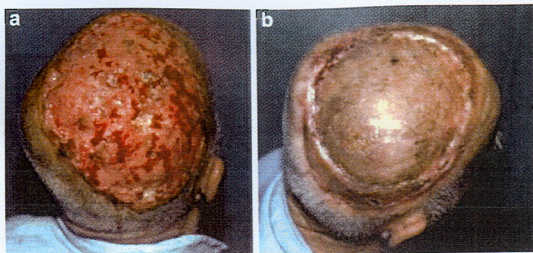
### Patients and methods

A retrospective analysis of 93 patients with aggressive terebrans basal cell carcinoma treated surgically was performed. In 15 patients (16%), the tumor developed primarily, in 30 patients (32%) after surgical therapy, and in 49 patients (52%) after radiation therapy. In all patients radical surgical resection was performed. In 54 of the patients, wide, extensive, mutilating excisions were performed. In 30 of these 54 patients, postoperative radiation

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Fig. 1



therapy was given, and fatal recurrence occurred in nine cases (30%). In 24 patients postoperative radiation therapy was not given, and fatal recurrence occurred in 10 cases (41%). After excision of the tumor in all patients, various reconstructive procedures were performed (split-thickness skin grafts, large local flaps, vascularized musculocutaneous flaps, musculocutaneous flaps transferred by microsurgical technique), depending on the site and size of the tumor, the age and general condition of the patients. Only 8 out of 93 cases will be presented in detail.

#### Case 1

Over 2 years almost the entire scalp had been infiltrated with the ulcerative tumor (Figure 1a). Skull X-ray showed that the tumor had infiltrated the outer cortex and diploe in several sites. It was decided that the tumor and the full-thickness of the skull should be removed, and the defect covered with a latissimus dorsi musculocutaneous flap transferred by microsurgical technique. When the skull was removed from the dura there was a cardiac arrest, which was dealt with successfully. Because of this and his previous cardiac problems, the dura was covered with a split-thickness skin graft (Figure 1b). It was recommended that he wear a firm protector on the head, but he refused and he has lived 10 years without any further treatment.

#### Case 2

Following radiation treatment, this patient's lesions involved the scalp, skull and dura (Figure 2a). The resection involved removal of the skull and the involved dura. The latter was reconstructed with fascia lata (Figure 2b), and the external cover was with a free latissimus dorsi musculocutaneous flap (Figure 2c). He was given postoperative radiation and so far he has survived for 1 year without recurrence.

#### Case 3

A basal cell terebrans carcinoma of the infraorbital region and lower eyelid recurred after seven surgeries and

radiation therapy. It spread to the orbit, maxilla, mandibular ramus, zygomatic, frontal, and temporal bone, as well as the anterior skull base and underlying dura (Figure 3a). The plan was to remove the tumor radically, and to cover the large defect with a latissimus dorsi musculocutaneous free flap. The recipient neck vessels were prepared followed by an en bloc resection of the tumor (Figure 3b). As the specimen was being removed, cardiac arrest occurred. Even though the heart rhythm was restored, the arrest had been for several minutes. The operation was terminated, and she died 3 days later without recovering consciousness.

#### Case 4

This basal cell terebrans carcinoma occurred after radiation and 11 operations. It had started in the supraorbital region (Figure 4a). The first therapy was radiation, but the tumor eventually destroyed the orbit, infiltrated the frontal bone and frontal lobe. The tumor was removed together with the orbit, most of the frontal bone, and part of the underlying frontal lobe. The defect was covered with a free latissimus dorsi musculocutaneous flap (Figure 4b). In 3 months there was a recurrence in the brain and in the musculocutaneous flap, and the patient died soon after this.

#### Case 5

This basal cell terebrans carcinoma of the anterior side of the neck, clavicle and sternal region presented after irradiation therapy and three operations (Figure 5a). The tumor had started on the neck. The initial therapy was radiation. The tumor now infiltrated muscles, and cartilages of the upper ribs bilaterally. The head was held in flexion, the lower jaw was fixed down towards the clavicle, and he could not extend the neck. In order to perform endotracheal anesthesia, a tracheotomy was performed under local anesthesia. The tumor was excised together with removal of all tissue structures down to the clavicle and sternum bilaterally. The cartilages of the first, second and third rib were also removed bilaterally (Figure 5b). The defect was covered with split thickness skin grafts, and the tracheostome was closed with Gore-Tex



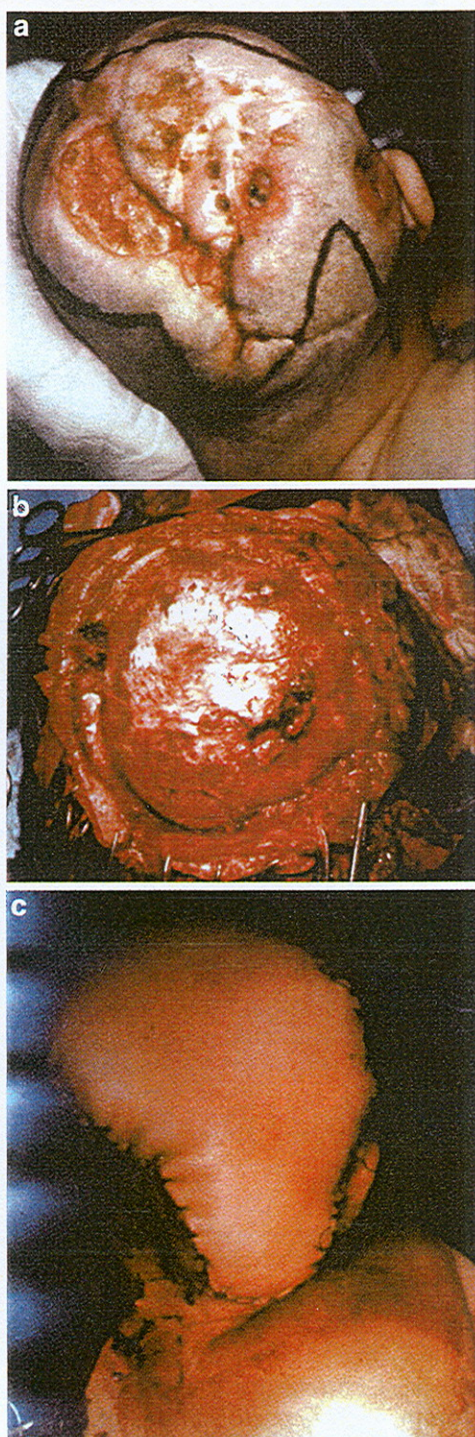


Fig. 2

(Figure 5c). Postoperatively, radiation therapy was given. There was a recurrence 9 months after operation, and this rapidly spread to the remaining muscles, ribs and sternum. The thoracic surgeon felt that the condition was inoperable. The patient died 6 months later.

#### Case 6

Basal cell terebrans carcinoma infiltrating the temporal, zygomatic and orbital regions following previous

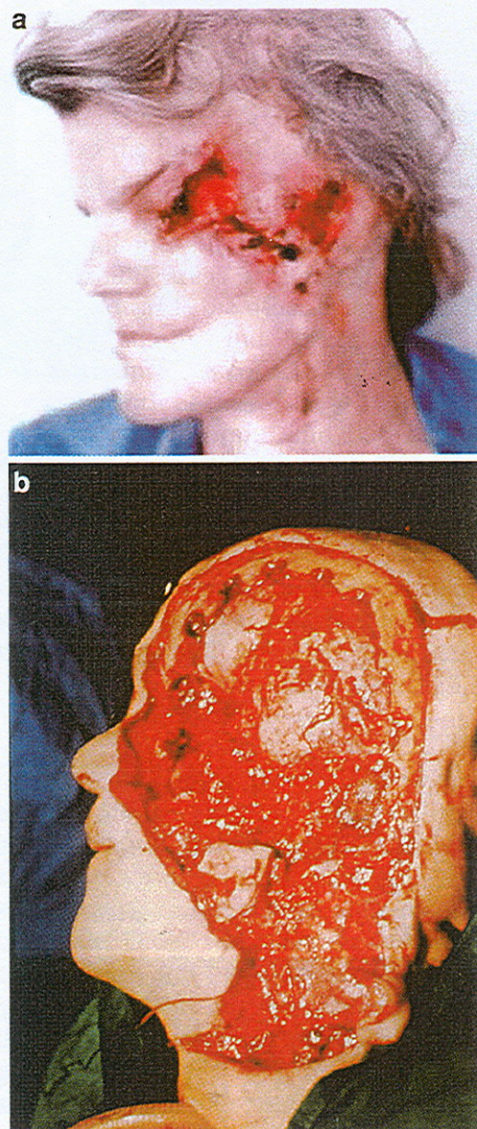


Fig. 3

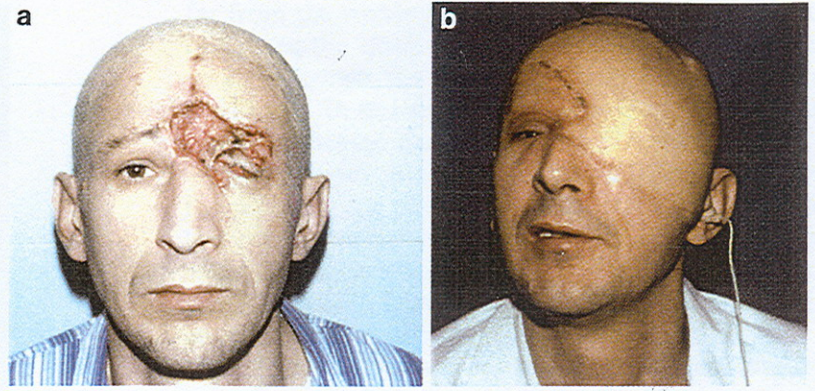
surgery (Figure 6a). The tumor had begun in the temporal region. It was removed en bloc with the remainder of the temporal muscle, lateral orbital wall, part of zygoma, the lateral half of the eyebrow, and the lateral half of both eyelids (Figure 6b). The facial nerve was preserved except for the frontal branch. The defect was covered with large cervicothoracic flap together with reconstruction of eyelids (Figure 6b, c). Postoperatively, radiation therapy was given. He has survived 3 years and 4 months without recurrence.

#### Case 7

A multicentric basal cell terebrans carcinoma of the scalp involving the ear and temporal bone; he had had several previous operations (Figure 7a). The tumor was removed en bloc with the external, middle and internal ear, part of the temporal bone, mastoid and skull base from the mastoid to jugular foramen. The trunk of facial nerve was preserved (Figure 7b). Tumors of the scalp



Fig. 4



were also removed. After the excision there was insufficient scalp to cover the defect in the temporal and auricular region. The defect was covered with temporal and sternocleidomastoid muscles, and a split-thickness skin graft was applied (Figure 7c). He was irradiated postoperatively, and there has been no recurrence for 2 years and 4 months.

#### Case 8

Basal cell terebrans carcinoma of the eyelids, orbit, zygomatic bone and upper part of maxilla, this had recurred after radiation therapy (Figure 8a). The tumor was removed together with exenteration of the orbit and resection of the zygomatic bone and upper maxilla. Due to his serious heart condition and frequent episodes of bradycardia during the operation the idea of reconstruction was abandoned and the wound was covered with a split thickness skin graft (Figure 8b). The patient died 3 years later of heart failure, but without recurrence.

#### Discussion

Based on the analysis of these patients it was noted that basal cell terebrans carcinoma develops mostly in residual tumors, after inadequate surgical and radiation therapy. The latter situation results in the most aggressive lesion. The lesions are more malignant than the primary tumor. The best prevention of recurrence and the best aesthetic results are achieved by radical primary surgical intervention. Due to inadequate excision of the tumor on the face, nose and eyelids, in order to preserve aesthetics, many patients are mutilated later because of the recurrence, i.e. of the residual tumor and subsequent surgical treatment. Inadequate primary therapy of basal cell carcinoma is often the cause of death. We strongly believe that primary radical surgical intervention gives much better results, in every way, than primary radiation therapy, and furthermore radiation therapy is not indicated in cases of basal cell carcinoma. This should be reserved for those in which it is impossible to perform surgical therapy and in cases of invasive terebrans basal cell carcinoma after extensive, mutilating operations.

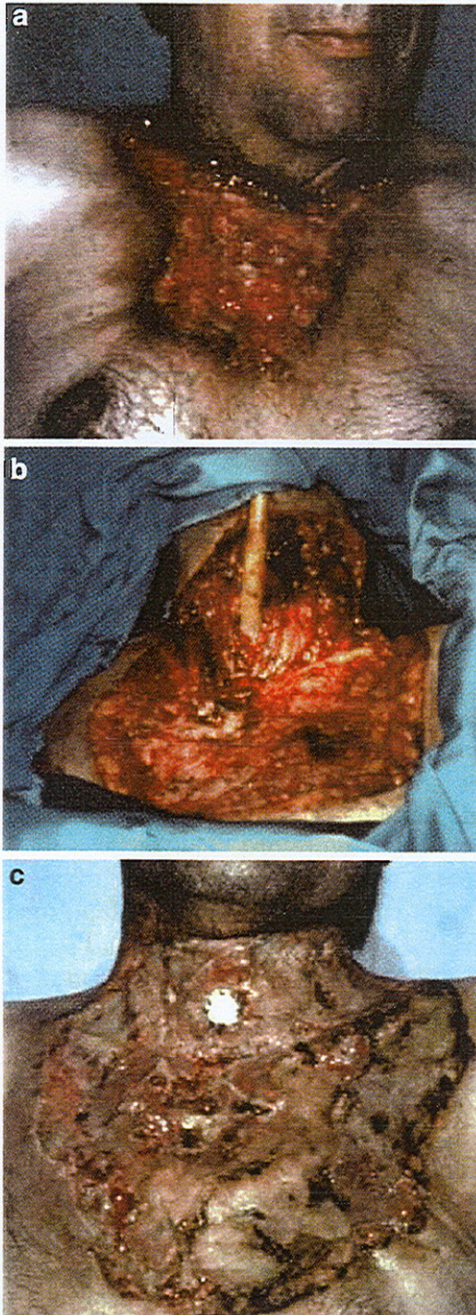


Fig. 5



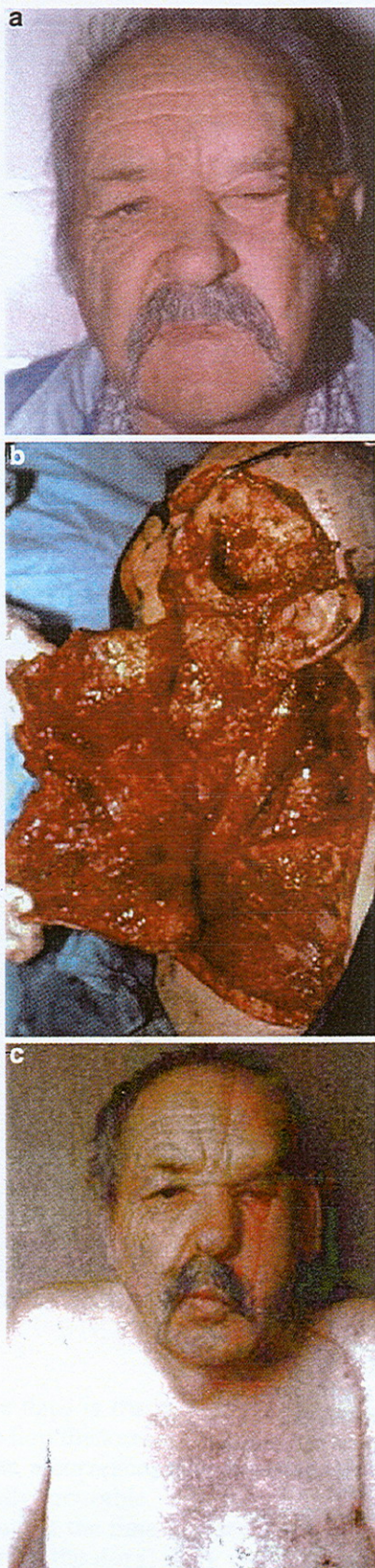


Fig. 6

If basal cell terebrans carcinoma infiltrates deeper structures (muscles, bones, cartilages, and dura), it is impossible to determine the limits of the tumor because it often infiltrates, in bands, into the tissues, which are

far from the visible periphery of the tumor. If this tumor infiltrates the skull diploe, the bands of the tumor can spread through the diploe far from the visible limit of the tumor. In our series there was one case in which the tumor infiltrated the forehead part of frontal bone, with the intact inner table, but the tumor spread far through the diploe of the parietal bone, to the top of the head. If the tumor spread into the orbits it infiltrates quickly towards paraorbital areas i.e., the paranasal sinuses, sometimes even towards the other orbit. In two patients, after radiation therapy and several operations and recurrences, the tumor destroyed both orbits resulting in loss of vision. In both of these cases the tumor extended from the orbit into the brain, and both patients died. These cases were not included because of lack of photo documentation.

If the tumor infiltrates the outer table of the skull and into the diploe, regardless of the fact that the inner table looks intact on X-ray, CT, and MRI, there is a distinct possibility that the tumor has infiltrated by narrow bands into the inner table and even the dura. In these cases, during the surgical resection of the tumor, the full-thickness skull should be removed. Sometimes, even when the tumor macroscopically and on X-ray infiltrates only the outer table of the skull, it penetrates, by invisible bands, through the diploe and the inner table. The dura is a significant barrier against penetration of the tumor into the brain. If the tumor infiltrates the dura the prognosis is much more unfavorable. By removing of the dura we are never sure that the tumor has been excised radically enough. In these cases it is possible that the tumor has penetrated the brain by narrow invisible bands. An additional major surgical problem is that if the tumor infiltrates the dura it is in the walls of the venous sinuses.

Because of these facts it is impossible to determine if, in cases of terebrans basal cell carcinoma, and in spite of an extensive mutilating surgical procedure, the whole tumor has been radically removed. The analysis of our patients showed that in cases where radiation therapy was given, after the extensive, mutilating operations, there were fewer fatal recurrences. That is why we advise radiation therapy to be used postoperatively in such cases. In practice, it is the last possible therapy for such patients.

Because of the uncertainty after radical tumor removal when there has been infiltration of the skull, primary reconstruction of calvarial bone defects is never performed. In the reconstruction of large defects of scalp and skull bone, after removal of the tumor, the best results are achieved by use of the latissimus dorsi musculocutaneous free flap. This flap gives a relatively good protection for the brain. If such a defect is located in the occipital region then it is possible to successfully reconstruct the defect using a vascularized musculocutaneous vertical trapezius flap. If the defect is large, this flap can be raised bilaterally. For successful reconstruction of postoperative face defects, the vascularized musculocutaneous pectoralis flap is advised. The disad-



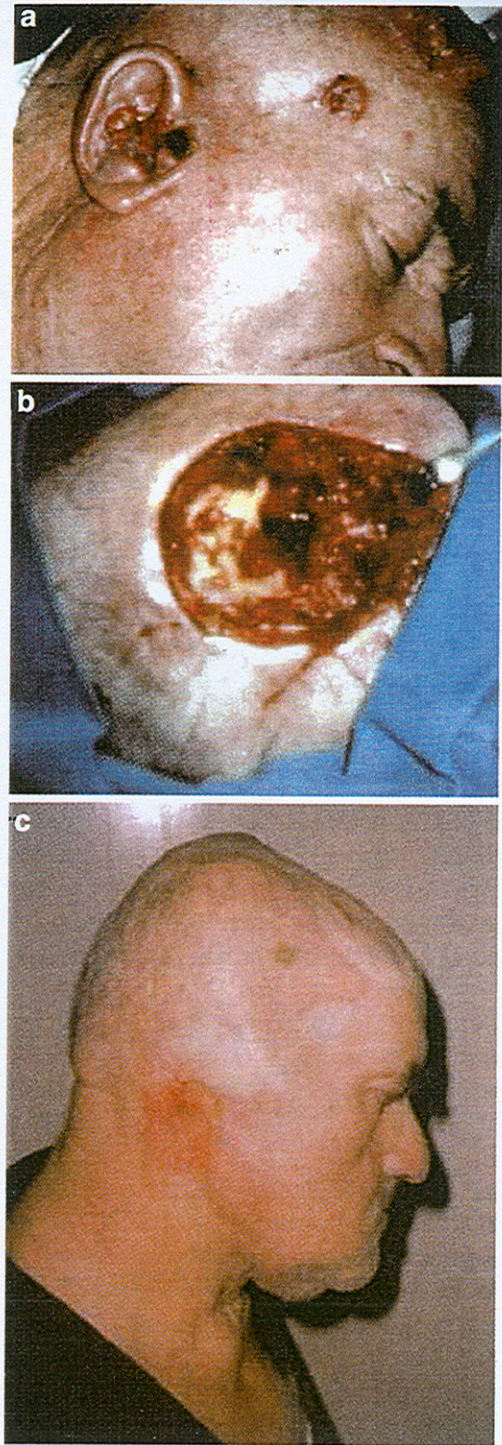


Fig. 7

vantage of these flaps is the inability to diagnose recurrence due to the flap thickness. This, in effect, is not a big problem because recurrence, after such extensive operations, is virtually incurable.

In two patients at the time of resection of a large part of basal skull from the dura, there was a cardiac arrest. Apart from several reasons for this complication it is possible that irritation of the dura, over a large surface area, initiates a meningocardiac reflex resulting in cardiac arrest.



Fig. 8

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