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Repair of saddle nose deformity in Wegener's granulomatosis and ectodermal dysplasia*

Wolfgang Pirsig, Sibylle Pentz, Heinrich Lenders

Section for Rhinology and Rhonchopathies, Department of Otorhinolaryngology, University of Ulm, Germany

SUMMARY

Two cases are reported involving surgical treatment of a saddle nose deformity due to Wegener's granulomatosis and ectodermal dysplasia, respectively. The association of ozena with both diseases requires special consideration for any type of transplants because of a high risk of infectious complications. By extranasal incisions the nasal dorsum has been successfully reconstructed by transplanting autogenic conchal cartilage. There has been no resorption or displacement of the transplant after twelve months in the case of ectodermal dysplasia, and after 25 months in the patient with Wegener's granulomatosis despite a severe recurrence of this disease.

Key words: Wegener's granulomatosis, ectodermal dysplasia, saddle nose, autogenic cartilaginous transplant, nasal dorsum reconstruction

INTRODUCTION

The saddle nose is a typical feature of patients with long-term Wegener's granulomatosis and of patients suffering from ectodermal dysplasia. Both diseases display severe symptoms of ozena, which requires special consideration when a surgical treatment of the deformity is planned. Different intranasal surgical attempts to control ozena have been reported in the literature (Loch, 1963; Huizing, 1965, 1969), but we could not find a paper referring to the therapy of saddle nose deformity in both diseases. This communication reports a method for the reconstruction of the nasal dorsum via extranasal incisions and by autogenic transplants from the auricle.

Wegener's granulomatosis, described in 1939, is a systemic disease of unknown origin that is regarded as an immunological disorder. Initially, it affects the mucosal linings of the upper airway, before infiltrating the lungs, kidneys and other organs. Wegener (1939) defined this disease by the following histological findings: necrotizing vasculitis and focal necrotizing multiple granuloma with nephritis. An auto-antibody to a lysosomal protein can be demonstrated by the anti-neutrophil-cytoplasmic antibody test in the serum of the patient. The immunological concept of this disease is also supported by the positive response to pulsed intravenous cyclophosphamide and high-dose oral steroids.

Ectodermal dysplasia combines a rare group of genetic disorders which are transmitted in a recessive way linked to the X-chromosome. It is characterized by absent or diminished numbers of structures derived from the ectoderm. The syndrome consists of hypo- or anhydrosis, anodontia or reduced numbers of deformed teeth, and atrophic rhinitis. Other symptoms may be: sparse hair growth, nail dystrophies, malformation of the external ear, congenital sensorineural hearing loss, hypomastia or amastia, diminished salivary and lacrimal secretions, and mental deficiency. The skin is dry, nasal cavities are greatly expanded and show an extraordinary atrophy of the turbinates (Gil-Carcedo, 1982). According to Stevenson and Kerr (1967), ectodermal dysplasia occurs in an estimated 1 per 100,000 live births and was first described by Thurnman (1848). In a postscript of the same year, Thurnman also refers to a female with ectodermal dysplasia, who had already been mentioned by Williams. The first male affected by ectodermal dysplasia was depicted by Guilford in 1883 (Figure 1), and the first female was photographed by Goeckermann in 1920. Both patients displayed severe saddle nose deformities.

CASE REPORTS

PV1966

A female (1.82 m, 69 kg, age 24) came to our hospital for correction of her saddle nose deformity. Diagnosis of ectodermal dysplasia had been constituted by paediatricians

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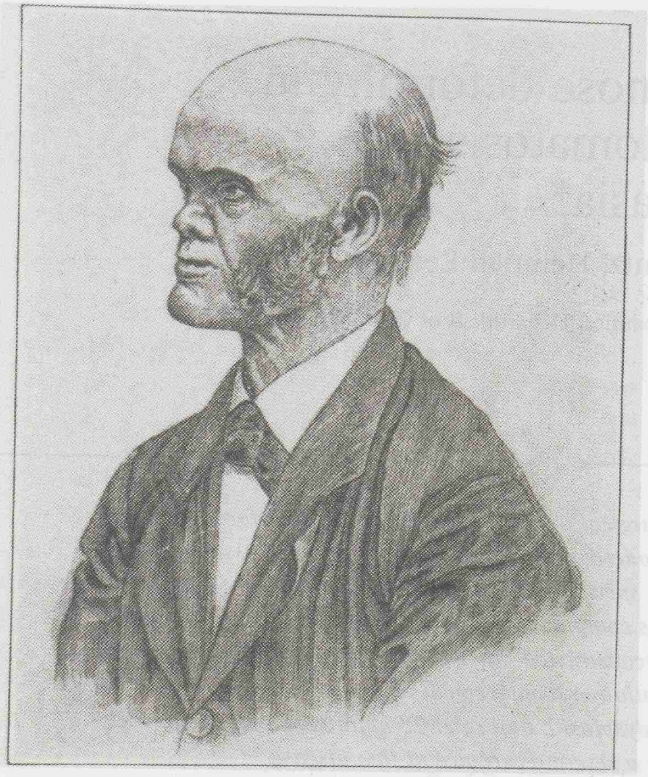

Figure 1. Male with ectodermal dysplasia depicted by Guilford in 1883.

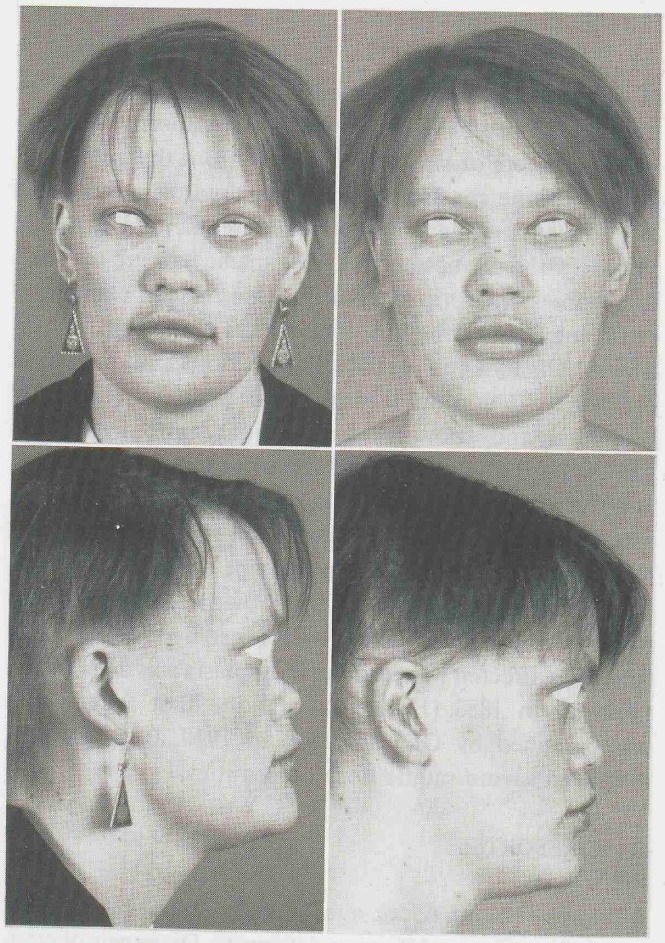

Figure 2. PV1966 with ectodermal dysplasia pre-operatively (left) and eight months after nasal surgery (right).

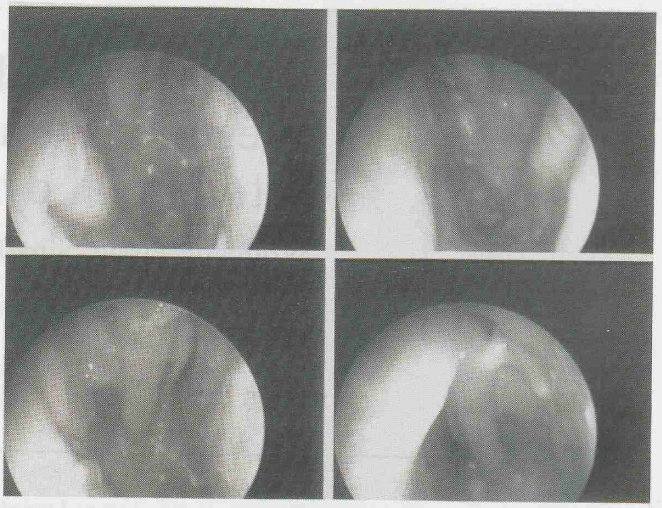

Figure 3. PV1966: video-endoscopic prints of both nasal cavities. Top: right and left inferior turbinates with choanae. Bottom: right and left middle turbinates.

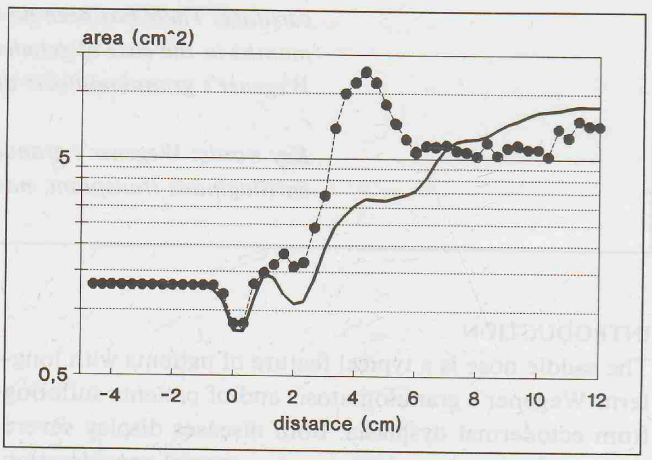

Figure 4. PV1966: acoustic rhinometric curve of the left nasal cavity (closed circles) showing the enlarged cross-sectional areas as a function of distance from the nostril. For comparison's sake, the solid curve represents the cross-sectional areas of a normal, decongested nasal cavity.

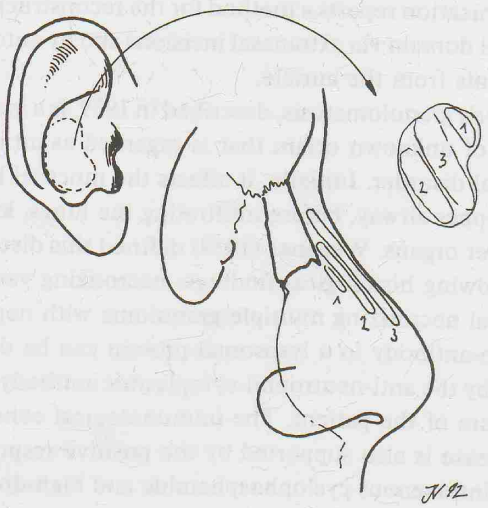

Figure 5. PV1966: sketch of the nasal dorsum reconstruction by auricular cartilage.

when she was aged 15. In her family, one elder brother is known to carry a complete syndrome, and another relative carried a minor form. The patient displayed sparse hair, neither sweat glands nor mammary glands, and had developed nine permanent teeth. From early infancy she had been troubled as well by a dry mouth, nasal crusting, obstructed nasal breathing and intense nasal fetor.

In Figure 2 (left) we see that her midfacial region is changed by a marked saddle nose deformity involving the bony and cartilaginous dorsum. The auricles are caused to protrude slightly by an overgrowth of the conchal cartilage. Nasal endoscopy revealed widely expanded nasal cavities partitioned by a thin septum. The inferior turbinates were almost completely atrophic and the middle turbinates were also reduced in size (Figure 3). Acoustic rhinometry (Figure 4) revealed the enlargment of the cross-sectional areas behind the first notch of the isthmus nasi due to the mucosal atrophy. There was no difference in the acoustic rhinometric curves for the untreated and decongested state.

The nasal dorsum was approached via an incision through the extraordinarily thin skin at the nasal root. The transplant (Figure 5) was obtained from the right auricle by an incision made just inside the anti-helical border, leaving perichondrium attached. The kidney-shaped cartilage was divided and inserted under the skin of the nasal dorsum. External fixation was obtained by Adaptic® and tape. Three mega-propicillin per day were given for one week. Twelve months following surgery the transplant was found without resorption in the correct position (Figure 2, right).

EG1942

This female felt healthy up to age 46 (Figure 6, inset), at which time she began to suffer from chronic inflammation of the nasal sinuses. In addition to nasal obstruction by severe crusting and a developing saddle nose deformity, she complained of progressive hearing loss in the left ear. Biopsies from the nasal mucosa and radiography led to the diagnosis of Wegener's granulomatosis, and treatment with cyclophosphamide and prednison was begun in February 1988. In a period of remission in June 1989, we found a severe collapse of her nasal bridge, due to loss of the triangular cartilages, parts of the septal structures, and depression of the nasal bones (Figure 6, left). There was a septal perforation in area III, 5 cm in diameter, and the nasal cavities were filled with serosanguineous and malodorous crusts. Until January 1990 the nasal septum completely disappeared except for the membraneous columella. In addition to the cartilaginous saddle, the skin of the nasal dorsum was retracted under the caudal ends of the nasal bones, resulting in a disfiguring furrow along the piriform aperture.

In a first stage a built-up of the nasal dorsum was undertaken by an extranasal incision in the deepest part of the furrow described above. The skin over the resorbed triangular cartilages was elevated. A butterfly-shaped transplant (Figure 7), harvested from the right conchal cartilage, was inserted in order to replace the triangular cartilages. The area over the nasal bones was left untouched. A dressing with Adaptic® protected the nose for one week. During this period the patient received four mega-propicillin each day. A coronal CT-scan of the sinuses in June 1991 (Figure 8) revealed no infiltrations of the nasal cavities and sinuses despite a recurrence of the disease at that time. Figure 8 shows that no structures of the internal nose are left. This recurrence prevented the second stage of nasal

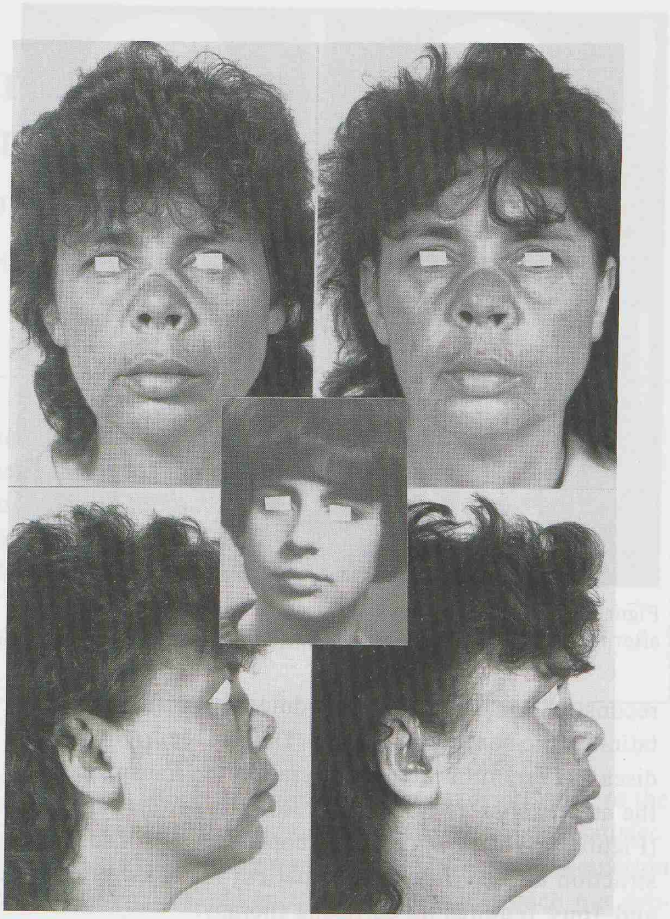

Figure 6. EG1942 with Wegener's granulomatosis pre-operatively (left) and 21 months after nasal surgery (right). Inset: patient's appearance six years before onset of the disease.

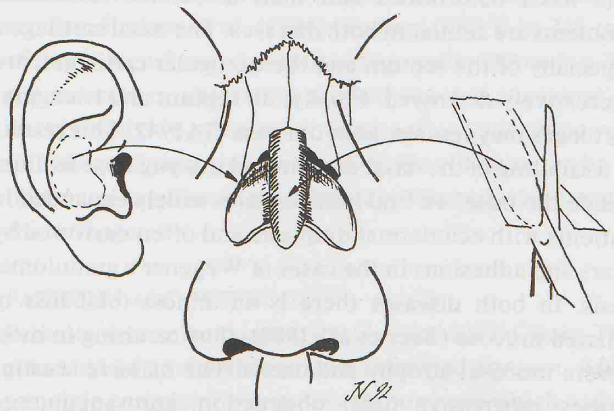

Figure 7. EG1942: sketch of the nasal dorsum reconstruction by auricular cartilage and lyophilized dura.

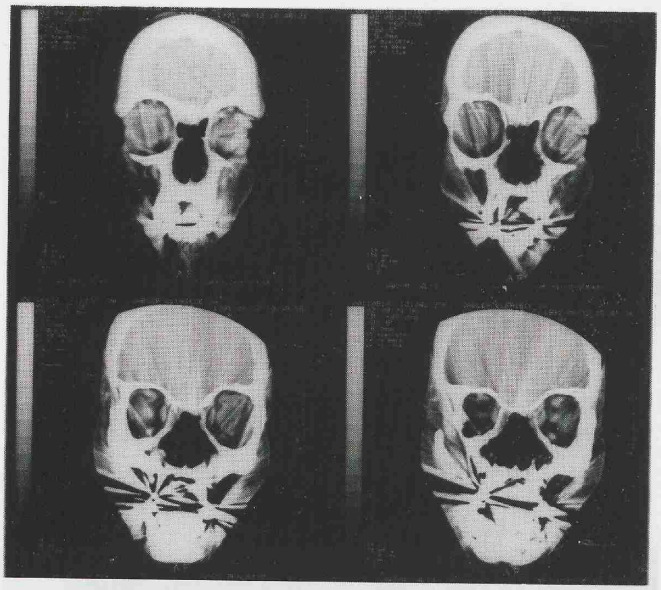

Figure 8. EG1942: coronal CT-scan of the sinuses 15 months after nasal surgery.

reconstruction, which was scheduled for further augmentation of the bony nasal dorsum. Despite recurrence of the disease, 25 months after the operation the transplant from the auricle showed no sign of resorption or displacement (Figure 6, right). We plan the second stage of dorsal reconstruction at a point when the patient resumes remission and, thus, requires no cortisone therapy.

HOW TO RECONSTRUCT THE SADDLE NOSE DEFORMITY

The nasal deformities and their associated functional problems are similar in both diseases. The nasal cartilages, especially of the septum and the triangular cartilages, are successively destroyed. Finally, no septum and triangular cartilages may remain, as in our case GA1942. This results in a saddling of the nasal dorsum and in a pug nose feature. Inside the nose, we find nasal cavities widely expanded in patients with ectodermal dysplasia and often narrowed by scars and adhesions in the cases of Wegener's granulomatosis. In both diseases there is an almost total loss of ciliated mucosa (Baer et al., 1988), thus resulting in most severe mucosal atrophy and ozena. The massive crusting causes progressive nasal obstruction, serosanguineous discharge and, due to secondary infection, a fetor.

These nasal pathologies have to be considered when surgical repair of the saddle deformity is planned. Ten days before nasal surgery we treat ozena by installing glucose powder into the nasal cavities, several times per day. In addition, lavages with saline solution help to remove the crusts. This treatment reduces all types of intranasal bacteria and eliminates the fetor for approximately ten days, and is continued after surgery. Reconstruction of the cartilaginous dorsum by septal built-up is not possible because of atrophy or loss of septal structures. Because there are always pathogenic bacteria in the atrophic nasal cavities, approaches to the nasal dorsum by intranasal incisions bear the risk of infection of the dorsal transplant. Therefore, we prefer to reconstruct the nasal dorsum via external incisions. These incisions are placed in the nasal root or in furrows of the skin due to the altered anatomical findings.

We prefer a cartilaginous transplant obtained from the cavum conchae of the auricle because it constitutes a nearly ideal transplant, especially for those two diseases. The autogenic conchal cartilage is not involved in either disease and has a very high rate to survive without resorption after being transplanted into other regions (Tardy, 1985). The shape of this transplant ideally adapts to the shape of the nasal dorsum and little sculpting is required. It can easily be inserted by the extranasal incision with a minimum of elevation of the dorsal skin. It is important not to overdo the dorsal reconstruction by inserting too much cartilage in one stage, thus preserving a sufficient blood supply for the dorsal skin. Usually a fixation of the transplant is not necessary. The nose is packed using a gauze impregnated with an ointment containing polymyxin-B, neomycine, bacitracin, and hydrocortisone. These internal nasal dressings are left in place for three days.

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Prof. Dr. Wolfgang Pirsig
ENT-Department
University of Ulm
Prittwitzstraße 43
D-7900 Ulm
Germany