■ CASE STUDIES

# Nasal Closure as Definitive Treatment for Epistaxis in Rendu-Osler-Weber Disease

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**Objective:** To show the result of this surgical technique for the treatment of nasal bleeding in Rendu-Osler-Weber disease.
**Method:** After great efforts to obtain informed consent for the procedure, considered as a final therapeutic attempt, we present here the surgical technique used in a severe long-standing case with frequent symptoms that required a large number of admissions to our hospital and caused our patient considerable social, family and work-related psychological problems.
**Results:** The surgical procedure was performed 14 months ago, with no further epistaxis since then, thus bringing to an end the emergency admissions, rated at 7 per year, and reducing consultations at the digestive and psychological clinics. The laboratory studies (haematocrit, complete blood count, bleeding time) have improved greatly.
**Conclusions:** This is a safe technique, well tolerated by the patient, that avoids additional pathologies and reduces costs.

**Key words:** Rendu-Osler-Weber disease. Epistaxis. Nasal closure.

**Cierre nasal como tratamiento definitivo de las epistaxis en la enfermedad de Rendu-Osler-Weber**
**Objetivo:** Conocer las posibilidades terapéuticas que supone esta técnica quirúrgica en el tratamiento de las epistaxis en la enfermedad de Rendu-Osler-Weber.
**Método:** No siendo muy fácil conseguir el consentimiento para su realización y quedando dentro del arsenal terapéutico como medida final, exponemos su desarrollo técnico en un caso de larga evolución, con muchos y frecuentes síntomas que, por obligar a multitud de ingresos hospitalarios, motivaba problemas sociales, familiares, laborales y psicológicos a la paciente.
**Resultados:** Tras 14 meses de practicada la intervención, la paciente no ha vuelto a padecer epistaxis. Antes, la paciente ingresaba en el hospital por este motivo una media de siete veces al año, de modo que la intervención ha producido una notable disminución de las consultas digestivas y psicológicas. Los parámetros sanguíneos evaluados han mejorado ostensiblemente.
**Conclusiones:** Se trata de una técnica eficaz y bien tolerada, que evita otros trastornos a los pacientes y supone eliminación de gastos.

**Palabras clave:** Enfermedad de Rendu-Osler-Weber. Epistaxis. Cierre nasal.

## INTRODUCTION

Rendu-Osler-Weber disease is characterized by telangiectasias, spontaneous bleeding, predominantly epistaxes, and arteriovenous malformations in internal organs. It is an inheritable disorder. It is also known as hereditary haemorrhagic telangiectasia (HHT) with various clinical types, depending on the chromosome affected. It is an angiopathic haemorrhagic diathesis,[1,2] with bleeding due to failures in the vessels themselves and normal plasma coagulation and platelet factors. It has a prevalence rate of 1 case/5000-12 000 people.[3,4]

## MATERIAL AND METHOD

Fifty-one-year-old female, monitored clinically for 10 years, with a family history of the same disorder (her mother, her sister, and a daughter), who has required a great many consultations and hospital admissions (49 in the last 7 years) for epistaxis, analytical alterations, and dysthymic symptoms.

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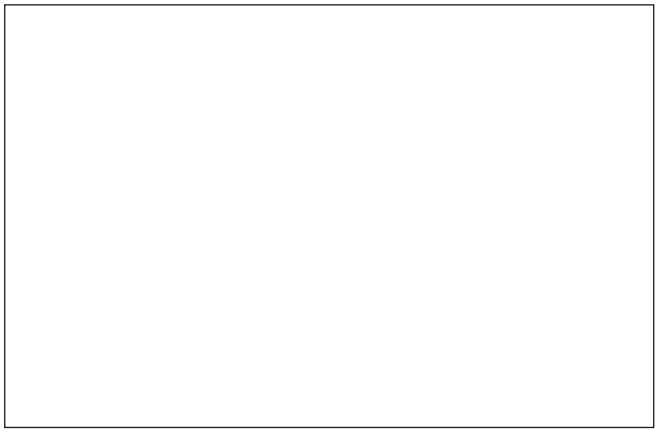

**Figure 1.** Creation of the double skin and mucosal flap in the right nasal fossa (arrows pointing to both flaps). Merogel in the fossa.

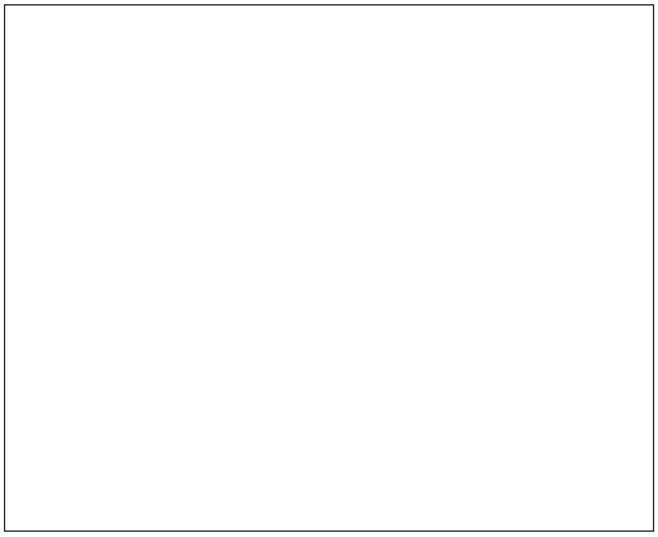

**Figure 2.** Closure of the mucosal flap in the right nasal fossa.

Due to frequent epistaxes, more in the left nostril initially, the patient was seen for the first time in June, 1997. In 1998, the bleeding became more profuse and bilateral, when alterations were seen in her RBC, haemoglobin and haematocrit and she developed chronic anaemia, dizziness, and headaches.

The panic attacks and depression the patient suffered due to her bleeding and other social and employment reasons became more severe, with broad treatment, self-motivated overdoses obligating psychiatric consultation.

Several treatments were initiated, cauterizations, local sealing, embolizations, etc, but in the light of the scant positive outcomes with respect to her bleeding, it was agreed with the patient, after clearly having the technique and its consequences explained to her, that a bilateral nasal closure would be performed. The surgery took place in September, 2006 by means of a circular incision in both nostrils, with 2 flaps (a skin flap and a mucosal flap) in each fossa (Figure 1), which are sutured separately (Figure 2), creating a false chamber between both closures to form an intermediate fibrous area and definitive closure. The technique ceases to be straightforward, however, when bleeding occurs and it may be impossible in the absence of continuous aspiration. Our lack of experience led us to place small Merogel tampons in both nostrils as a precaution prior to closure, as well as gauze tampons fixed in the cavum with silk sutures to be removed 48 hours later, leaving the Merogel tampons in place until they had spontaneously dissolved. Whether due to the tampons or to the nasal closure, there was a foul smell that lasted 12 months. The new functional status of the nose, without bleeding, the patient's psychological well-being improved from the very beginning and has persisted until the present time. She is calmer, euthymic and has no problems of having to breathe through her mouth.

An imaging study was carried out a month and a half post-operatively (Figure 3) and normal function was seen in both sinonasal edges, although the inflammatory response

**Figure 3.** Magnetic resonance image a month and a half after surgery. The inflammatory reaction is still visible in the right fossa and sinuses. The left side has been normalized.

**Figure 4.** Three months after surgery.

Diagnostic Criteria

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| 1 Epistaxis: spontaneous and recurrent |
| 2 Multiple cutaneous mucosal telangiectasias in specific places |
| 3 Dominant autosomal heritance |
| 4 Arteriovenous-type fistulae in internal organs |
| Three of the 4 criteria are needed to establish a diagnosis of the disease. |

is still visible in the thickening of the sinonasal mucosa (Figure 4) on the right side. Fourteen months following surgery, we no longer fear relapse of bleeding.

## DISCUSSION

This autosomal dominant transmission disease presents three different genetic types, I, II, and III, which cannot be distinguished clinically, although type I is associated with pulmonary arteriovenous malformations; type II, with hepatic manifestations (as in our case), and type III, with transformations in tumour growth beta factor.[5-7]

Bleeding is especially common in the nasal mucosa and epistaxis is the only initial symptom in more than 90%-93% of patients.[8] They begin at 6-10 years of age and become worse starting at age 35. They are located in the anterior part of the fossa, the turbinate, the floor of the nostril and the valve area, and if there are septal perforations, along their edges or in more posterior regions of the nasal fossa.[9] Telangiectasias, in addition to the ones located in the nose, may be present in the mouth and skin of the face, on the hands, ears and thorax.

The pulmonary arteriovenous malformations are located in the inferior lobes and cause dyspnoea, cough, pulmonary and pleural bleeding and hypoxaemia. They are treated by means of embolization through a catheter in single- or multi-stage procedures.[10-13]

Hepatic arteriovenous manifestations are characterized by intrahepatic shunts, intraparenchymal telangiectasias or, as in our case, increased number and thickness of the hepatic vessels.

In the brain, telangiectasias, angiomas or aneurysms may require neurosurgical techniques.

The disease is diagnosed on the basis of clinical manifestations, examination and at least three of the four criteria stipulated in 2000 for this disease must be met (Table).

Complications are directly related to spontaneous bleeding, due to failure in the collagen and elastin in the vascular wall, severe anaemia that is well-tolerated by the patients, aesthetic problems, physical, emotional and social limitations[14] and psychological issues related to the duration of the illness.

Several different types of treatments have been used to treat this disorder, although many have merely served to alleviate it and have tended to fail to resolve the problem: local lavage, cauterizations, fibrin glue (sealant or in aerosol), which has at times been deemed to be highly useful,[15] Ethibloc or other sclerosing agents for peripheral percutaneous embolizations in the area of bleeding,[16,17] arterial ligature of the anterior ethmoid, embolizations of the maxillary artery,[18] dermoplasties, which also entail a protracted foul smell post-operatively and fail to achieve good outcomes, albeit sparing the airways and sense of smell. When none of these achieves a positive result, it is possible to resort to nasal closure, which prevents epistaxis, but forces the patient to breathe through the mouth permanently. After seeing the course of illness in this patient for several years during which time she underwent many of the previously-cited treatments, we resorted to surgical nasal closure; in our opinion, successfully and satisfactorily.

Therefore, we believe that nasal closure can represent an efficacious treatment well-tolerated by these patients. As a secondary outcome, we have observed an improvement in our patient's mental health, eliminating hospital admissions and emergency treatment.

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