

## CASE REPORT

# Woakes' syndrome: Case report

**Lucian Lapusneanu<sup>1</sup>, Cristina Andrei<sup>2</sup>, Daniela Mihalache<sup>3</sup>, Luciana Lapusneanu<sup>4</sup>**

<sup>1</sup>ENT Department, Braila County Emergency Hospital, Braila, Romania

<sup>2</sup>Radiology Department, Braila County Emergency Hospital, Braila, Romania

<sup>3</sup>Anatomopathology Department, Braila County Emergency Hospital, Braila, Romania

<sup>4</sup>“George Emil Palade” University of Medicine and Pharmacy, Targu Mures, Romania

## ABSTRACT

Woakes' syndrome is a rare entity, defined as severe recurrent chronic rhinosinusitis with nasal polyps, which has as a consequence the deformation of the nasal pyramid, produced by the continuous pressure and the inflammation maintained by the size of the polyps.

This paper reviews the main features of Woakes' syndrome by describing a clinical case.

The patient presented to the ENT Outpatient Clinic for aggravation of an obstructive nasal respiratory syndrome, associated with rhinorrhea, headache, anosmia and deformity of the nasal pyramid. The ENT clinical examination, the nasal endoscopic examination and the craniofacial CT scan established the diagnosis of Woakes' syndrome. Functional endoscopic sinus surgery was performed. The follow-ups performed at 10 days and later, a month after surgery, showed a favourable evolution, without signs of local recurrence and a narrowing of the nasal pyramid compared to the preoperative moment. Proper management and proper adherence to the therapeutic protocol are factors that can prevent the onset of this syndrome.

**KEYWORDS:** Woakes' syndrome, functional endoscopic sinus surgery.

## INTRODUCTION

Nasal polyposis is a common entity in otorhinolaryngological practice, which may be associated with Woakes' syndrome. This very rare condition consists in the deformation of the nasal pyramid occurring as a consequence of the extensive growth of polyps in the paranasal sinuses and in the nasal cavity<sup>1</sup>. Few cases related to this syndrome are reported in the literature. According to PubMed, 77 articles and clinical cases related to this syndrome have been published in the last century (1920-2021), most of them being described in children.

Endoscopic sinus surgery is currently considered the gold standard in the treatment of Woakes' syndrome, as well as of frequent recurrences.

Surgical treatment must be accompanied by pre- and postoperative drug treatment, both topical and

systemic, with a role in limiting the associated allergic phenomena and reducing the rate of relapses.

## CASE REPORT

We present the case of a 60-year-old patient who addressed the ENT Outpatient Clinic of the Braila County Emergency Hospital for complete bilateral nasal obstruction, nasal pyramid deformity (Figure 1), posterior rhinorrhea, anosmia, closed rhinophony, frontal headache.

The patient's history of the disease begins more than 30 years ago. Between 1989 and 2005, the patient underwent 3 surgeries for ablation of the intranasal polypoid masses by the classic technique with polypectomy loop, the complete nasal obstruction reappearing approximately 4 years after



**Figure 1.** Deformation of the nasal pyramid at admission in the hospital compared to the appearance of the nasal pyramid at the age of 20.

each intervention. The patient relates the onset of nasal obstruction in youth, in the evolutionary context of nasal polyposis. We mention that there is no family history of nasal polyposis or allergic disease.

At the ENT clinical examination, the deformation of the nasal pyramid could be observed, with its significant widening (Figure 1). Anterior rhinoscopy and nasal endoscopy revealed translucent polypoid mass lesions that completely occupied the nasal fossae, reaching the level of the bilateral nasal vestibule.

Cranio-facial CT scan identified complete opacification of the nasal and paranasal cavities, with areas of osteitis and osteolysis, with the thinning and partial disappearance of the ethmoid intercellular bony septa and the partial lysis of the intersinus-nasal walls of the maxillary sinuses (Figure 2).

Preoperatively, injectable treatment with antibiotic and corticosteroid, oral antihistamine and corticosteroid nasal spray was established, obtaining a decrease in the volume of polypoid masses in both nostrils and drainage of mucopurulent secretions on the left side (Figure 3).

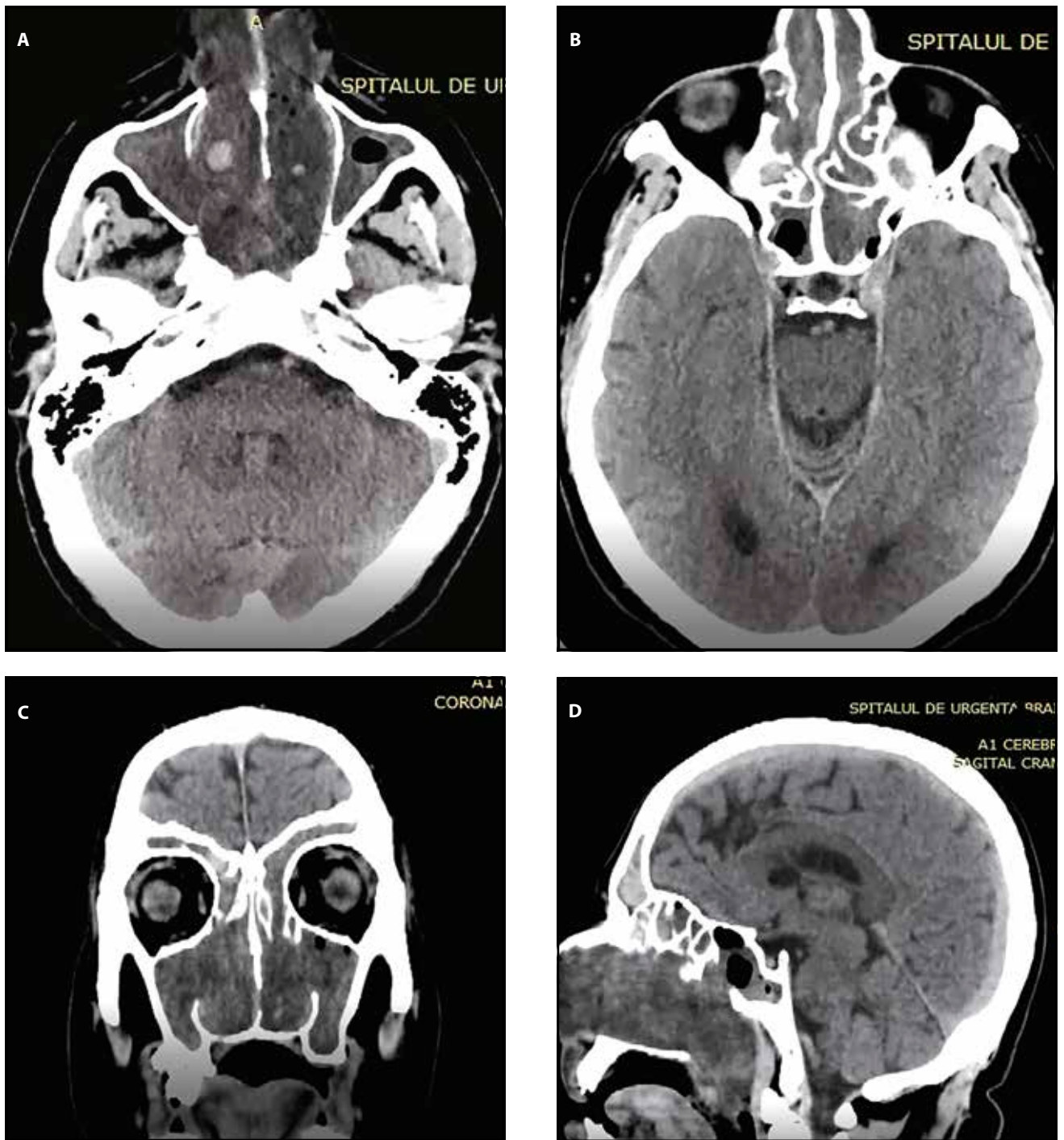
Surgical treatment consisted of endoscopic sinus surgery, performing in a first operative time a bilateral polypectomy with the microdebrider (Figure 4A). Intraoperatively, significant changes in the rhinosinus anatomy were found: complete absence of

the middle nasal turbinates and partially of the left lamina papyracea, multiple polypoid degenerations of the nasal septal mucosa. Subsequently, a bilateral maxillary antrostomy was performed, with drainage of a mucocele from the left maxillary sinus (Figure 4B), antero-posterior ethmoidectomy and bilateral sphenoidotomy with removal of the inflamed mucosa from this level (Figure 4C). The Draf type I frontal drainage allowed the permeabilization of the fronto-nasal duct.

At the end of the intervention, Meroce<sup>®</sup> haemostatic sponge packing was performed on both maxillary sinuses, the bilateral inferior, middle and upper nasal meatus, and also the packing of both nasal cavities with gauze strips. It should be noted that, although the inferior turbinates with hypoplasia due to the compression exerted by the polypoid masses have been preserved, the remaining postoperative intranasal volume was significant, reason for which the type of packing described above was performed. The nasal packing removal was performed 48 hours postoperatively, without complications in the following days.

The histopathological examination identified respiratory epithelium, with edema and an inflammatory lymphoplasmacytic and focal infiltrate with eosinophils, suggestive of nasal polyposis (Figure 5).

The postoperative treatment consisted of nasal



**Figure 2.** Cranio-facial CT scan: **A** – axial slice – partially homogeneous tissue mass that completely occupies the paranasal sinuses with partial bone lysis in the medial walls of the maxillary sinuses and partial destruction of the ethmoid intercellular bony septa; **B** – axial slice – sphenothmoidal sinuses almost completely occupied by tissue mass; **C** – coronal slice – complete opacification of the fronto-ethmoido-maxillary sinuses, with partial lysis of the bilateral intersinuso-nasal-maxillary medial wall; **D** – sagittal slice – tissue mass occupying the frontal sinus, antero-posterior ethmoid, sphenoid and nasal fossa with partial lysis of the ethmoid intercellular bony septa.

saline irrigation and topical intranasal corticosteroid spray.

The follow-up performed at 10 days postoperatively revealed rhinosinusal cavities partially lined with crusty secretions, without signs of local recur-

rence (Figure 6). There was also a narrowing of the nasal pyramid compared to the preoperative aspect (Figure 7).

Patient reassessment was indicated at one and three months postoperatively, respectively.



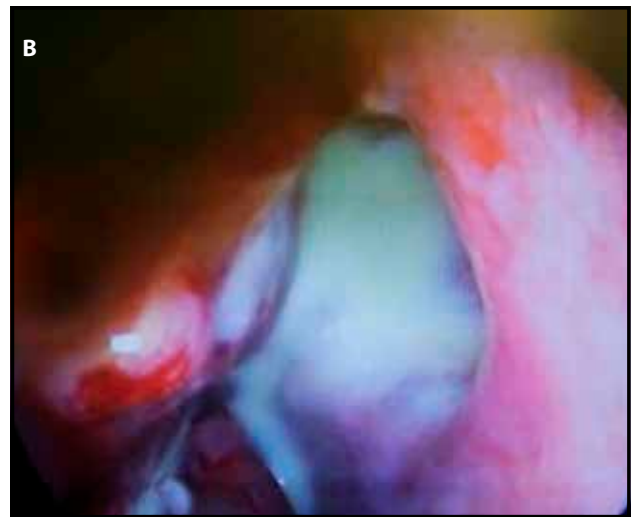
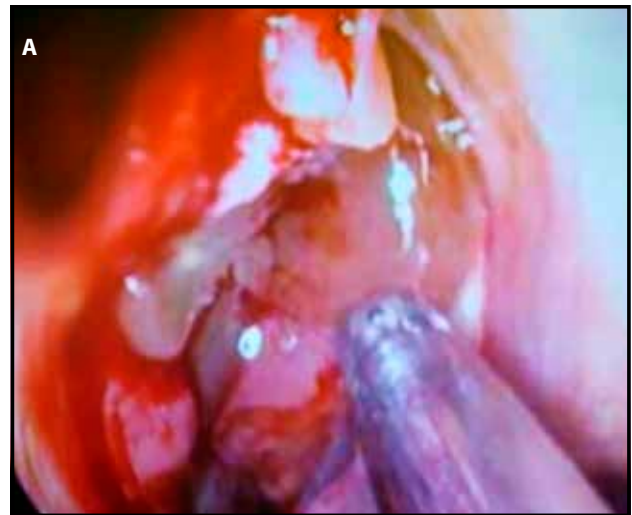
**Figure 3.** Nasal polyposis status after 3 days of postoperative treatment.

## DISCUSSIONS

Woakes' syndrome is a severe recurrent nasal polyposis, which can cause significant deformities of the nose and face by the continuous pressure exerted by the extensive growth of nasal polyps in the nasal cavity and in the paranasal sinuses.

Over time, nasal polyposis has been described by numerous authors, the first reference to this pathology dating back to antiquity, in the time of Hippocrates and Galen<sup>1,2</sup>. In 1885, Woakes describes the syndrome that bears his name as a necrotic ethmoiditis with mucous polyps that may be associated with enlargement of the nasal pyramid in some patients<sup>3</sup>. The high incidence of nasal polyposis in patients with aspirin intolerance is reported for the first time by Widal in 1922, and in 1968 Samter introduces the term Samter's Triad, associating the triad "bronchial asthma, rhinosinusitis inflammation and recurrent nasal polyposis", refractory to treatment. These authors have shown that there is a close link between nasal, bronchial and aspirin susceptibility<sup>4</sup>.

In 1924, Appaix and Robert mention the four characteristics of Woakes' syndrome: deformation of the nasal pyramid caused by hypertrophic processes, bilateral nasal polyposis (eosinophilic or

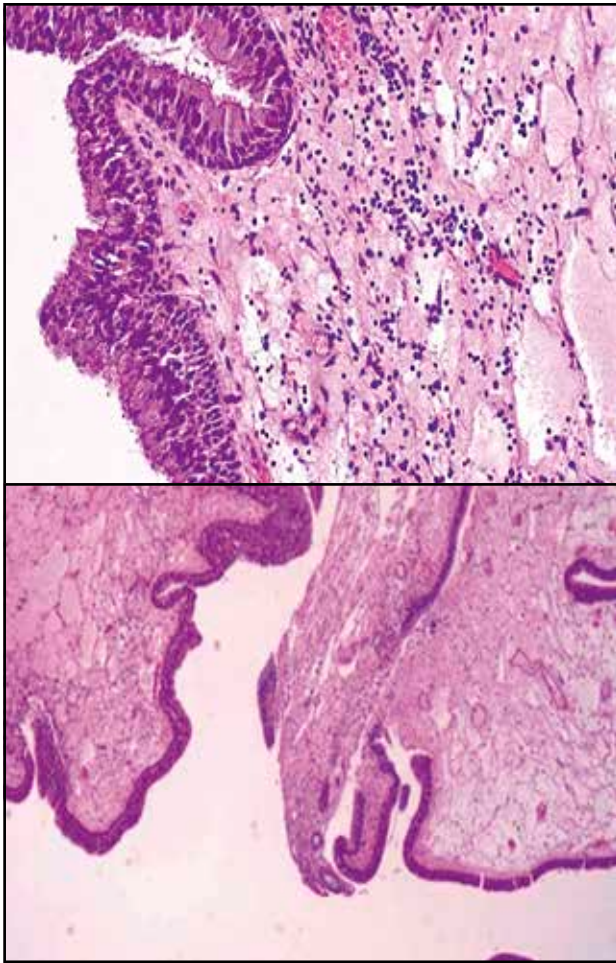


**Figure 4.** Intraoperative nasal endoscopy:

**A** - nasal polypectomy with the microdebrider;

**B** - left maxillary antrostomy and mucocele drainage from the left maxillary sinus;

**C** - left maxillary sinus (MS), sphenoid sinus (SS), choanal orifice (CO).



**Figure 5.** Histopathological examination - respiratory epithelium, with edema, an inflammatory lymphoplasmacytic and focal infiltrate with eosinophils, suggestive of nasal polyposis.



**Figure 7.** Postoperative appearance on day 10 - narrowing of the nasal pyramid.



**Figure 6.** Endoscopic examination 10 days postoperatively - rhinosinusal cavities partially lined with crusty secretions, without signs of local recurrence (maxillary sinus (AM), sphenoid sinus (SS), choanal orifice (CO)).

non-eosinophilic) with childhood onset, ethmoiditis and treatment failure with multiple recurrences<sup>5</sup>. 55 years later, Kellerhals and De Uthemann redefine the syndrome by associating four other elements: widening of the nasal pyramid, frontal sinus aplasia, bronchiectasis and production of extremely viscous mucus<sup>6</sup>.

In today's medical world, the term Woakes' syndrome associates recurrent severe nasal polyposis with consequent deformity of the nasal pyramid, appeared as a consequence of the permanent and prolonged pressure exerted by the polypoid formations<sup>1</sup>. The vast majority of cases reported in the literature are children or young adults<sup>5-10</sup>. Taking into account the plasticity of the facial bone structures that are developing and growing in these age groups, we can explain the pathological changes in the facial bones with the local evolution of the disease.

Although chronic allergic inflammation of the nasal and sinus mucosa is considered to be the main starting point of nasal polyposis, a genetic mechanism could be responsible for an isolated

part of the cases. In the case of Woakes' syndrome, heredity has been discussed by some authors, who have described cases of synchronous occurrence in siblings<sup>6,8</sup>. Despite insufficient data and the absence of important studies regarding the family context, it is possible that certain forms of nasal polyposis may be inherited<sup>11</sup>.

Considering the important allergic component in Woakes' disease, the therapeutic management of this disorder must be done in team with the allergist, thus helping to improve the body's general biological parameters. It would be advisable to perform repeated allergological tests with the detection of a possible atopic field by the Phadiatop test and skin-prick tests, as well as the investigation of a possible patent asthma by bronchial hyperreactivity tests such as the methacholine test. Hyposensitisation or desensitisation schemes could decrease the severity and frequency of acute allergic episodes and, consequently, the recurrence of nasal polyps<sup>12</sup>.

Our patient had no risk factors for nasal polyps and no positive family history, aspirin intolerance or multiple allergies. The allergological evaluation and allergic skin-prick tests did not show the coexistence of an atopic disease.

Severe recurrent nasal polyposis, by the extensive growth of nasal polyps in the nasal cavity and paranasal sinuses, exerts continuous pressure, which can cause significant deformities of the nose and face. When the onset of the disease occurs in adulthood, initially the bone component is normal, affecting only the cartilage, which appears deformed, enlarged and hypertrophied. Over time, a disjunction occurs on the midline of the nasal bones, with the widening of the nasal bone pyramid, as in our case. We cannot specify the reason why in some of the patients the nasal pyramid is deformed, while in others there are limited changes only in the paranasal sinuses or they are completely non-existent in most cases. In our patient, the development of the severe recurrent nasal polyposis, over a period of more than 30 years, led to a distinct deformity of the nose. Considering the evolution and characteristics of the rhinosinusal pathology in the presented case, it can be included in a Woakes' syndrome as defined by Appaix and Robert<sup>5</sup>. This evolution of the disease cannot be explained either by a certain histopathological feature of nasal polyposis in Woakes' syndrome.

In addition to the treatment of chronic rhinosinusitis with nasal polyps, Abbud-Neme et al. described rhinoplasty as part of the treatment for Woakes' syndrome<sup>10</sup>.

In the case presented, the deformation of the nasal bones could have been corrected by osteot-

omy. The patient is to decide the opportunity for rhinocorrection, although at only 10 days post-surgery a degree of narrowing of the nasal pyramid could be observed. This simple and efficient procedure would lead to the adjustment of the nasal shape but also of the impressive volume of the nasal cavity, thus reducing the risk of "empty nose" syndrome.

The case presented in this paper raised therapeutic difficulties given the long history of the disease, with multiple surgeries, the modification of the normal rhinosinusal anatomy with the disappearance of some normal anatomical landmarks (such as the middle nasal turbinates) and locoregional destructions, left lamina papyracea, multiple areas of osteitis). These changes involve a significant intraoperative risk of orbito-ocular or endocranial complications (orbit penetration, dural tear with or without cerebrospinal fluid fistula).

Cases of recurrent nasal polyposis, with unusual stromal atypia and a potential malignant transformation, are cited in the literature<sup>13</sup>. In the case presented, contrary to the long evolution of an extensive and locoregional deforming rhinosinusal polyposis, no malignant transformation has occurred to date.

## CONCLUSIONS

The etiology of Woakes' syndrome remains uncertain, the disease generally showing a long-term evolution, with difficulty in terms of treatment and control of the disease.

The therapeutic attitude remains the medical-surgical one. Endoscopic sinus surgery, followed by topical treatment and, if necessary, even systemic, are required to prevent or reduce relapses.

The treatment should be performed in collaboration with the allergist, thus helping to improve the general biological parameters of the body and increase the patient's quality of life.

**Conflict of interest:** The authors have no conflict of interest to disclose.

**Contribution of authors:** All authors have equally contributed to this work.

---

## REFERENCES

1. Caversaccio M, Baumann A, Helbling A. Woakes' syndrome and albinism. *Auris Nasus Larynx*. 2007;34(2):245-8. DOI: 10.1016/j.anl.2006.09.030.
2. Hargrove SW. Discussion on the treatment of nasal polypi. *Proc R Soc Med*. 1954;47(12):1015-21.

3. Woakes E. Necrotizing ethmoiditis and mucous polyps. *Lancet*. 1885;61:619-20.
4. Picado C. Aspirin intolerance and nasal polyposis. *Curr Allergy Asthma Rep*. 2002;2(6):488-93. DOI: 10.1007/s11882-002-0089-8.
5. Appaix S, Robert J. Polyposé déformante et récidivante des jeunes (maladie de Woakes). *Revue de Laryngologie*. 1953;74:216-54.
6. Kellerhals B, de Uthemann B. Woakes' syndrome: the problems of infantile nasal polyps. *Int J Pediatr Otorhinolaryngol*. 1979;1(1):79-85.
7. Bailey DL. Deformity of the nose and face due to nasal polyposis. *The West Indian Medical Journal*. 1965;14(2):116-123.
8. Groman JD, Bolger W, Brass-Ernst L, Macek M Jr, Zeitlin P, Cutting G. Recurrent and destructive nasal polyposis in 2 siblings: a possible case of Woakes' syndrome. *Otolaryngol Head Neck Surg*. 2004;131(6):1009-11.
9. Triglia JM, Dessi P, Cannoni M, Pech A. Intranasal ethmoidectomy in nasal polyposis in children. Indications and results. *Int J Pediatr Otorhinolaryngol*. 1992;23(2):125-31. DOI: 10.1016/0165-5876(92)90048-t.
10. Abbud-Neme F, Reynoso VM, Deutsch Reiss E. Woake's syndrome. A case report in a teenager. *Int J Pediatr Otorhinolaryngol*. 1987;12(3):327-33.
11. Cutting GR. Genetics of rhinosinusitis. In: Kennedy DW, Bolger WE, Zinreich SJ. *Diseases of the sinuses: diagnosis and management*. B.C. Becker Inc. Hamilton, Ontario; 2001, p. 29-34.
12. El Fakiri MM, Lakhdar Y, Raji A. Woakes' syndrome. *BMJ Case Reports*. 2019;12(3):e229021. DOI: 10.1136/bcr-2018-229021.
13. Compagno J, Hyams VJ, Lepore ML. Nasal polyposis with stromal atypia. Review of follow-up study of 14 cases. *Arch Pathol Lab Med*. 1976;100(4):224-6.

© 2021 Lucian Lapusneanu, Cristina Andrei, Daniela Mihalache, Luciana Lapusneanu, published by Romanian Rhinologic Society



This is an open access article published under the terms and conditions of the Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International (CC BY-NC-ND 4.0) License (<https://creativecommons.org/licenses/by-nc-nd/4.0/>). CC BY-NC-ND 4.0 license requires that reusers give credit to the creator by citing or quoting the original work. It allows reusers to copy, share, read, download, print, redistribute the material in any medium or format, or to link to the full texts of the articles, for non-commercial purposes only. If others remix, adapt, or build upon the material, they may not distribute the modified material.