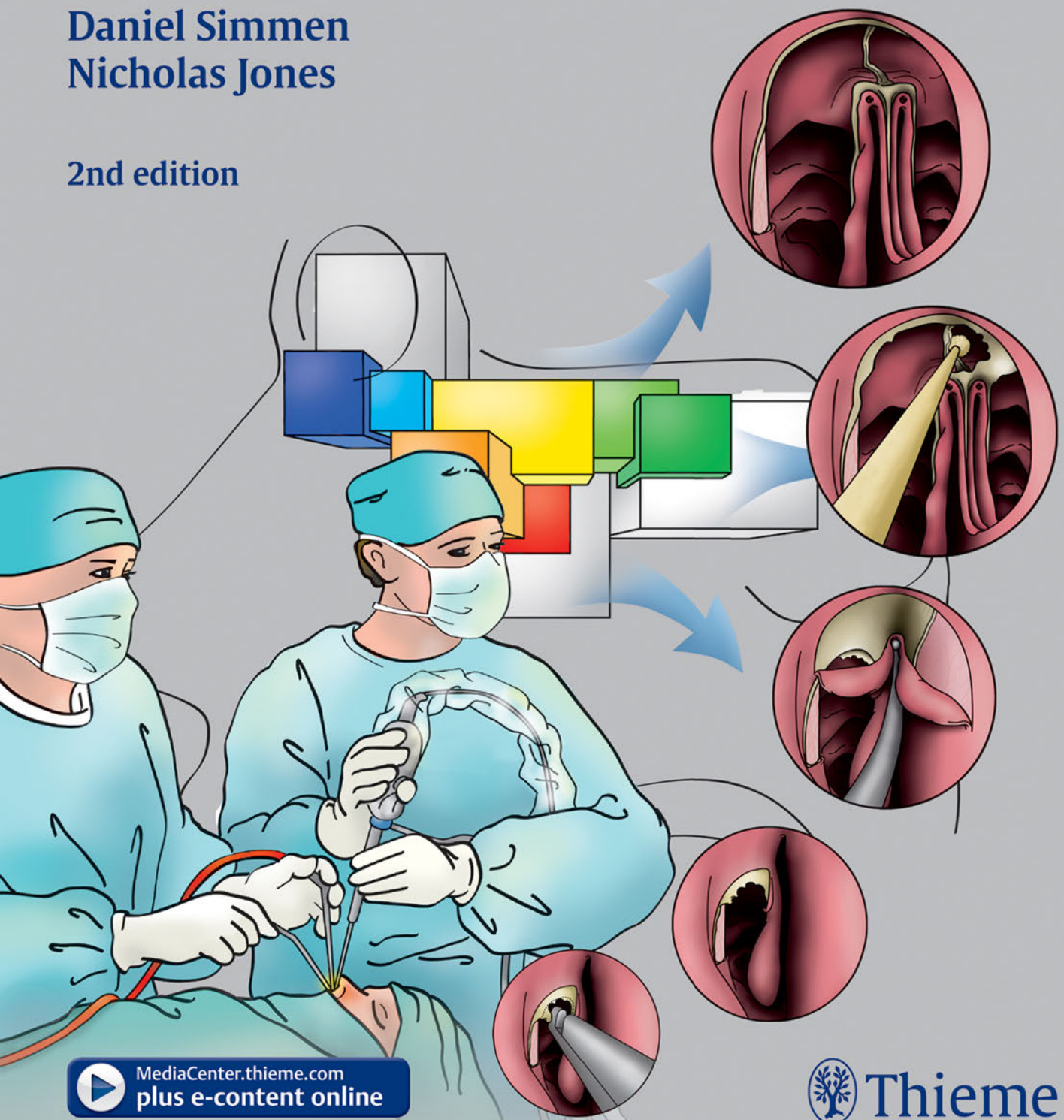
Manual of Endoscopic Sinus and Skull Base Surgery

Daniel Simmen
Nicholas Jones

2nd edition



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Manual of Endoscopic Sinus and Skull Base Surgery

Second edition

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1,442 illustrations

Thieme
Stuttgart · New York

Library of Congress Cataloging-in-Publication Data
Simmen, D. (Daniel), author.
Manual of endoscopic sinus and skull base surgery / Daniel Simmen, Nick Jones. -- Second edition.
p. ; cm.
Preceded by: Manual of endoscopic sinus surgery and its extended applications / Daniel Simmen, Nick Jones. c2005.
Includes bibliographical references and index.
ISBN 978-3-13-130972-3 (hardback : alk. paper)
I. Jones, Nick, 1953- author. II. Simmen, D. (Daniel). Manual of endoscopic sinus surgery and its extended applications. Preceded by (work): III. Title.
[DNLM: 1. Paranasal Sinuses--surgery. 2. Endoscopy--methods. 3. Perioperative Care--methods. 4. Skull Base--surgery. WV 340]
RF421
617.5'23--dc23
2013030980

Illustrator: Sandra Schuhmacher

Cover design: Thieme Publishing Group
Typesetting by Prepress Projects, Perth, UK
Printed in China by Everbest Printing Ltd., Hong Kong

ISBN 978-3-13-130972-3

Also available as an e-book:
eISBN 978-3-13-173012-1

Preface

> If you go to the mountain often enough, you ***will*** meet the tiger.
>
> Ancient Chinese proverb

This book not only represents our individual experience, but it is also the result of what we have each learnt from one another working together. We hope that it is of practical use as it is a distillation of our clinical practice.

We have become more and more convinced that patient selection is one of the main keys to successful surgery, and we have tried to explain how we go about this.

Much of the book focuses on how to improve the surgical approach, but we have also placed emphasis on what matters to the patient. The final chapters of the book involve advanced techniques and reflect the current direction in endoscopic sinus and skull base surgery.

Daniel Simmen, Zurich, Switzerland
Nick Jones, Nottingham, UK

Acknowledgments

To Dr. Hans Rudolf Briner, Lecturer in Rhinology, ORL-Zentrum – Center for Rhinology and Skull Base Surgery, The Hirslanden Clinic, Zurich, Switzerland.

To Dr. Kris S. Moe and Dr. Karthik Balakrishnan, Facial Plastic and Skull Base Surgery, University of Washington School of Medicine, Seattle, Washington, USA.

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To Dr. Aldo Cassol Stamm and Dr. Leonardo Lopes Balsalobre Filho, Federal University of São Paulo and Hospital Edmundo Vasconcelos, São Paulo, Brazil.

To Dr. Jean Shaw for her comments on the manuscript.

To Professor Jan Ullrich, Director of the Department of Anatomy, Institute of Anatomy, University of Zurich, and Dr. med. M. Vich and Dr. M. Manestar, also of the Department of Anatomy, for their help in supporting our courses and in preparing the anatomical specimens.

To Sandra Schuhmacher for all illustrations used in this edition.

Finally, we are grateful to our teachers, colleagues, and trainees who have all helped to educate us.

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Table of Contents

4 When? Medical Treatment and Timing of Surgery 49

5 Preserving and Improving the Sense of Smell 60

6 Why? The Goals of Surgery 67

7 Applied Anatomy for Endoscopic Sinus and Skull Base Surgery 80

8 How? Operative Procedures: A Step-by-Step Safe and Logical Approach 91

11 Preoperative Checklist 196

12 Patient Consent and Information 212

13 Peroperative Aids 217

14 Hints 239

15 Preventing and Dealing with Complications 242

16 Postoperative Management 260

17 Extended Endoscopic Procedures 266

List of Videos

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1 Principles of Practice

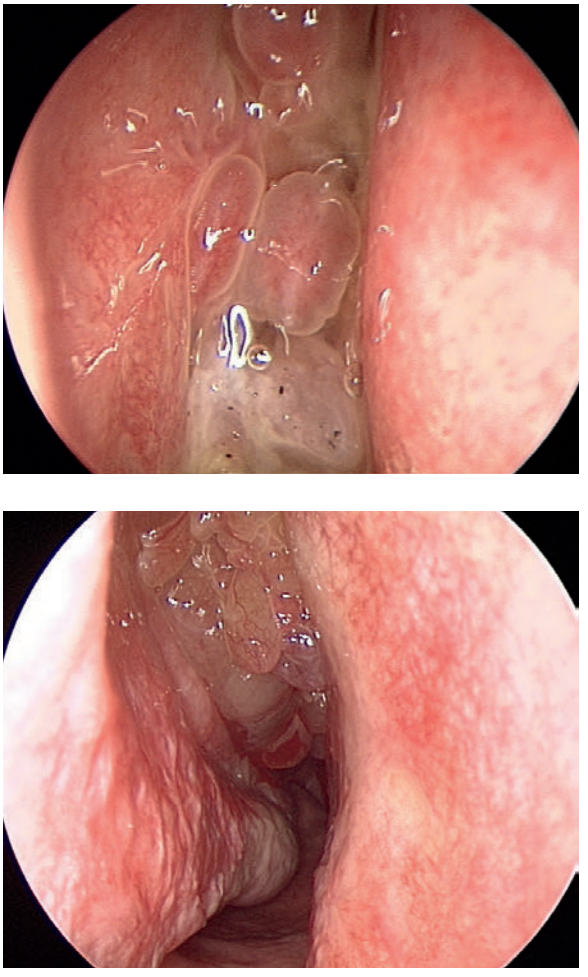
Accurate Diagnosis Is the Key to Success

A good surgeon should also be a good physician. The best results are often obtained by optimizing medical treatment both preoperatively and postoperatively (**Fig. 1.1a,b**). Optimizing medical treatment before surgery makes it less traumatic, reduces the chances of complications, and helps to preserve olfactory mucosa. To optimize medical treatment, the surgeon needs to have an understanding of the pathology of the mucosal disease. Postoperative medical treatment is frequently required to maintain the improvement that surgery has produced. A good surgeon may avoid or delay the need for surgery and obtain good symptomatic relief by medical means. The surgeon may decide, after discussion with the patient, that surgery will not help the particular symptoms that they have and advise against surgical intervention. It is often more difficult to convince a patient that surgery is not in his or her interest, rather than proceeding, but in the long run it is to everyone's benefit.

History and examination should allow basic categorization of the disease, but these are often insufficient to make an accurate diagnosis. It is often necessary to undertake other investigations or to have a trial of medical treatment to clarify the underlying pathology.

Each of the clinical appearances shown in **Table 1.1** can be associated with different pathological processes (**Figs. 1.2a–f, 1.3a–d, 1.4a–d, 1.5a–d**). Try to arrive at a diagnosis that fits into one of the broad groups that are used for the classification of rhinosinusitis. These groups are shown in **Table 1.2**.

In the light of history and examination, along with the relevant investigations, the physician can obtain an idea of the underlying pathology. Based on this, medical and surgical treatment can be maximized.



a

b

Fig. 1.1a,b Nasal polyps.

a Before medical treatment.
b After medical treatment.

Focus on the Patient's Main Complaint

The patient may mention any of a large array of symptoms in nasal disease. There are four primary symptoms that are always worth asking about:

1. Nasal obstruction
2. Sense of smell
3. Secretions
4. Pain or pressure.

It is important to rank these symptoms in their order of priority to the patient. This not only helps to make a diagnosis, but it focuses the surgeon's mind on how best to meet the patient's needs. In the notes underline the patient's main complaint.

Dealing with the Patient's Expectations

The patient's priorities may differ from what the surgeon can achieve. For example, the patient's main concern might be their postnasal discharge, but the surgeon may only be able to improve the symptoms of obstruction with little alteration to the postnasal drip (**Fig. 1.6a–d**). It is therefore vital that the surgeon is forthcoming and makes it as clear as possible to the patient which symptoms can and cannot be improved or resolved. When the

Table 1.1 Simplified table of macroscopic clinical appearance in rhinosinusitis and criteria to classify length of history along with possible diagnoses based on clinical appearance

Observation	Length of history			Possible diagnosis—NOT diagnostic on its own
	Acute	Subacute	Chronic	
	< 3 weeks	> 3 weeks, < 3 months	> 3 months	
Erythema				Infective, allergic, nonallergic, chronic rhinosinusitis
Edema				Active or postinfective, allergic, nonallergic, chronic rhinosinusitis
Hyperplastic mucosa				Chronic rhinosinusitis
Polyposis				Idiopathic polyposis, allergic fungal polyposis
Granular mucosa				Wegener granulomatosis, sarcoidosis
Purulent secretion				Infective rhinosinusitis
Dry mucosa				Postsurgical, environmental, rhinitis medicamentosa

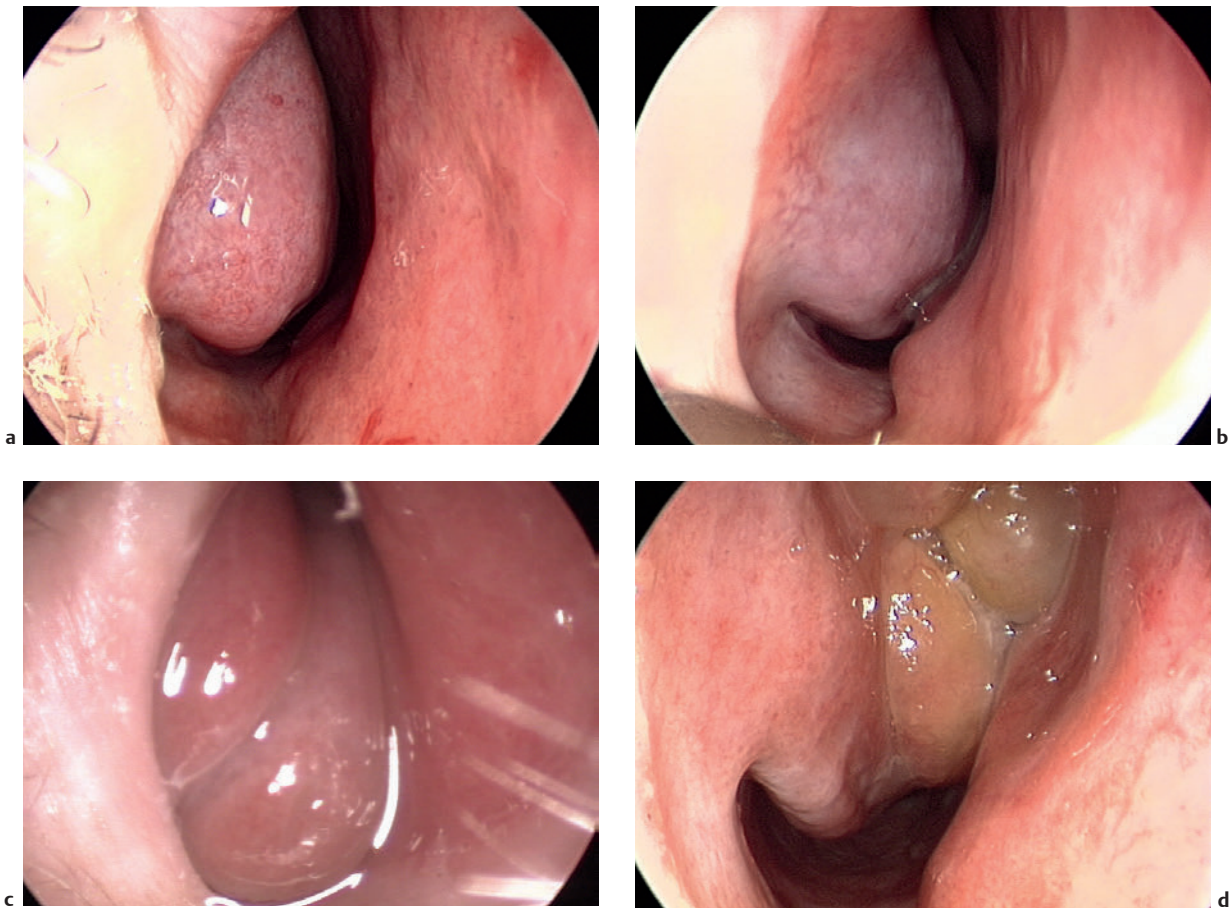
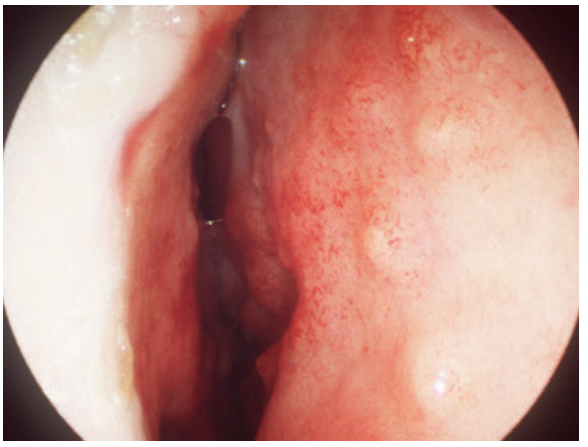


Fig. 1.2a–f Endoscopic appearance of a range of pathological conditions.

a Idiopathic rhinitis with erythema.
b Hyperplastic mucosa due to allergic rhinitis.
c Severe hypertrophy with edema.
d Polyposis in a nonatopic patient.

Table 1.2 Classification of rhinosinusitis

Type of rhinosinusitis	Comments
Infectious	
Viral	
Bacterial (including tuberculosis, leprosy, syphilis)	
Fungal	
Noninfectious	
Allergy	Seasonal (intermittent); perennial (persistent)
Chronic rhinosinusitis	
Chronic rhinosinusitis with polyposis	
Nonallergic rhinitis with eosinophilia (NARES)	
Hormonal	High-estrogen contraceptive pill; pregnancy
Autonomic/neurogenic	Primary symptom is rhinorrhea often reduced by ipratropium bromide; few other nasal symptoms; patients often elderly. Important not to include in idiopathic group
Sarcoidosis	
Vasculitis	Wegener granulomatosis
Drug induced	β-blockers
Rhinitis medicamentosa	Excessive use of local sympathomimetic agents
Occupational	
Atrophic	
Entopy	No systemic markers of atopy (skin prick test negative, no raised specific immunoglobulin E) but challenge/washings/biopsy show local immunoglobulin E in mucosa
Idiopathic	Some overlap in nomenclature as the cause of NARES is unknown



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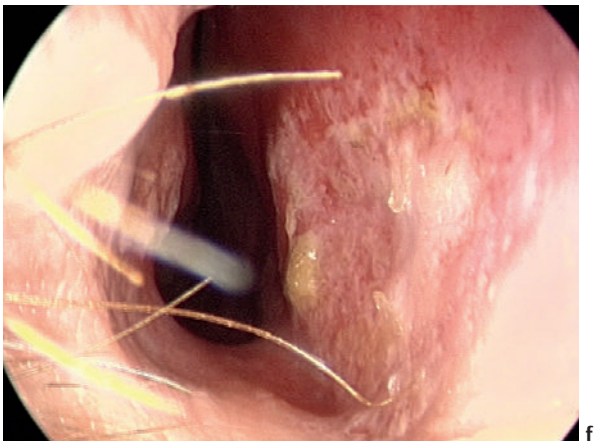

f

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e Granular mucosa.

f Dry mucosa.

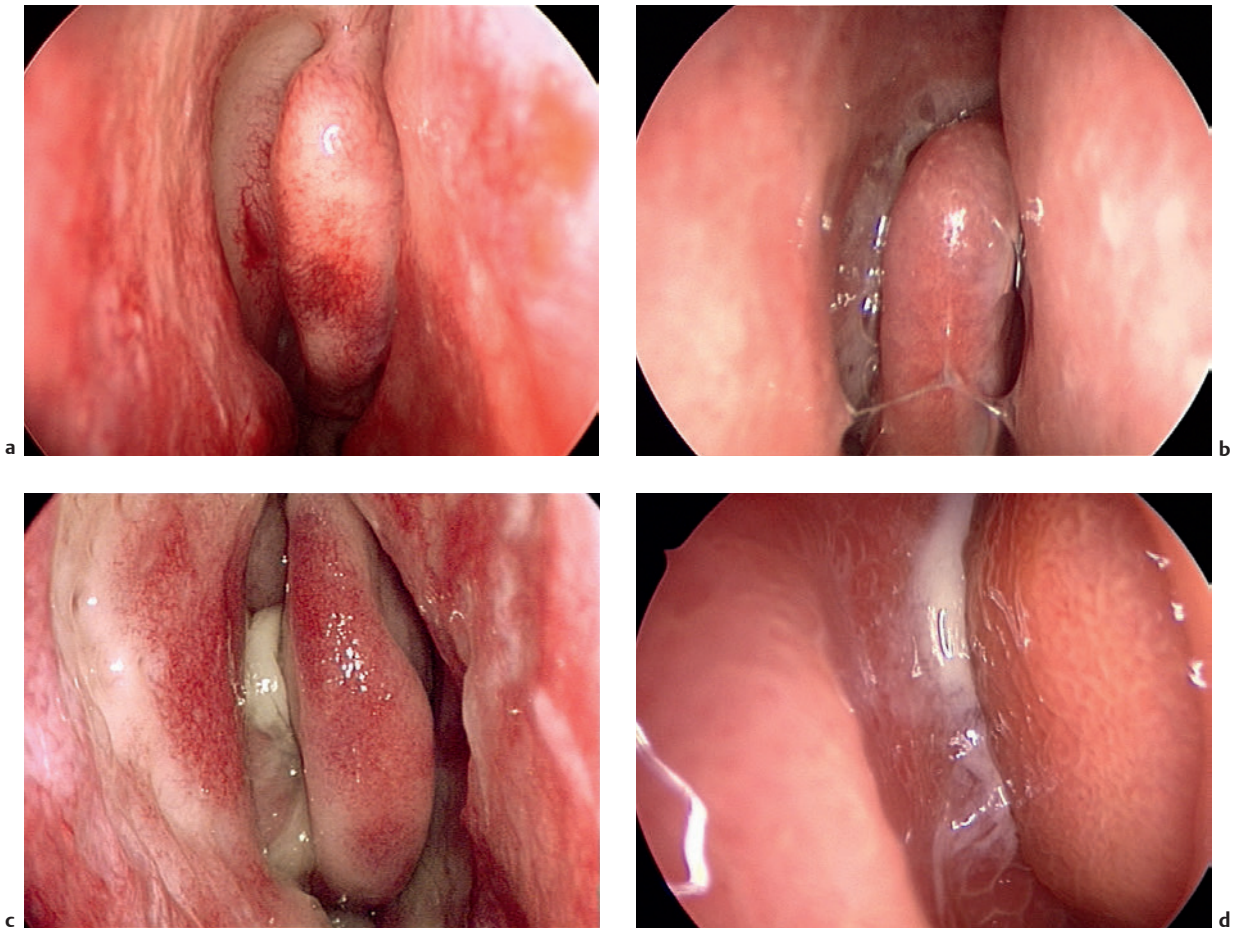


Fig. 1.3a–d Endoscopic appearance of a range of pathological conditions.

a Normal middle meatus.
b Serous secretions in marked allergic rhinitis.
c Purulent bacterial secretions.
d Purulent fungal secretions.

physician overlooks these reservations, the patient is likely to be disappointed with the outcome. Be aware that some patients may believe that even symptoms that they have not mentioned will be cured by any surgery.

Some patients will have well-formed ideas before they attend, whereas others will come with an open mind. It is worth finding out what the main motives for the visit are before examining the patient and particularly before embarking on any treatment. Many patients are seeking reassurance that they do not have cancer or a life-threatening illness and that is all they want.

Patients might reasonably be expected to want a diagnosis, a prognosis, an explanation of their symptoms in the light of the disease process, and a treatment plan. However, they may have a preconception about the cause of their symptoms that differs from the medical diagnosis. This is often the case in patients with "sinus headache," facial pain, or catarrh. It is worth taking time to discover what the patients' understanding of their disease process is so that their ideas about their symptoms and disease can be taken into consideration when explaining the cause of these processes.

Reassurance may be readily received after a clear explanation that recognizes the symptoms that the patient is experiencing, outlines their cause, and addresses the patient's concerns. However, in a small proportion of anxious individuals, the effect of firm reassurance after a thorough examination and explanation may fade with time. It is often counterproductive to ask these individuals to return because this may reinforce their concerns that the doctor may have some ongoing doubt.

Some patients come hoping, if not expecting, a cure for their symptoms. Many find it difficult to accept that there is no cure.

Many patients have tried alternative therapy before seeking specialist advice. Many have browsed the Internet looking for advice, but it is not easy for anyone without knowledge in the area in question to decipher what is good information. If a patient attends with printed extracts it is worth studying these in front of the patient,

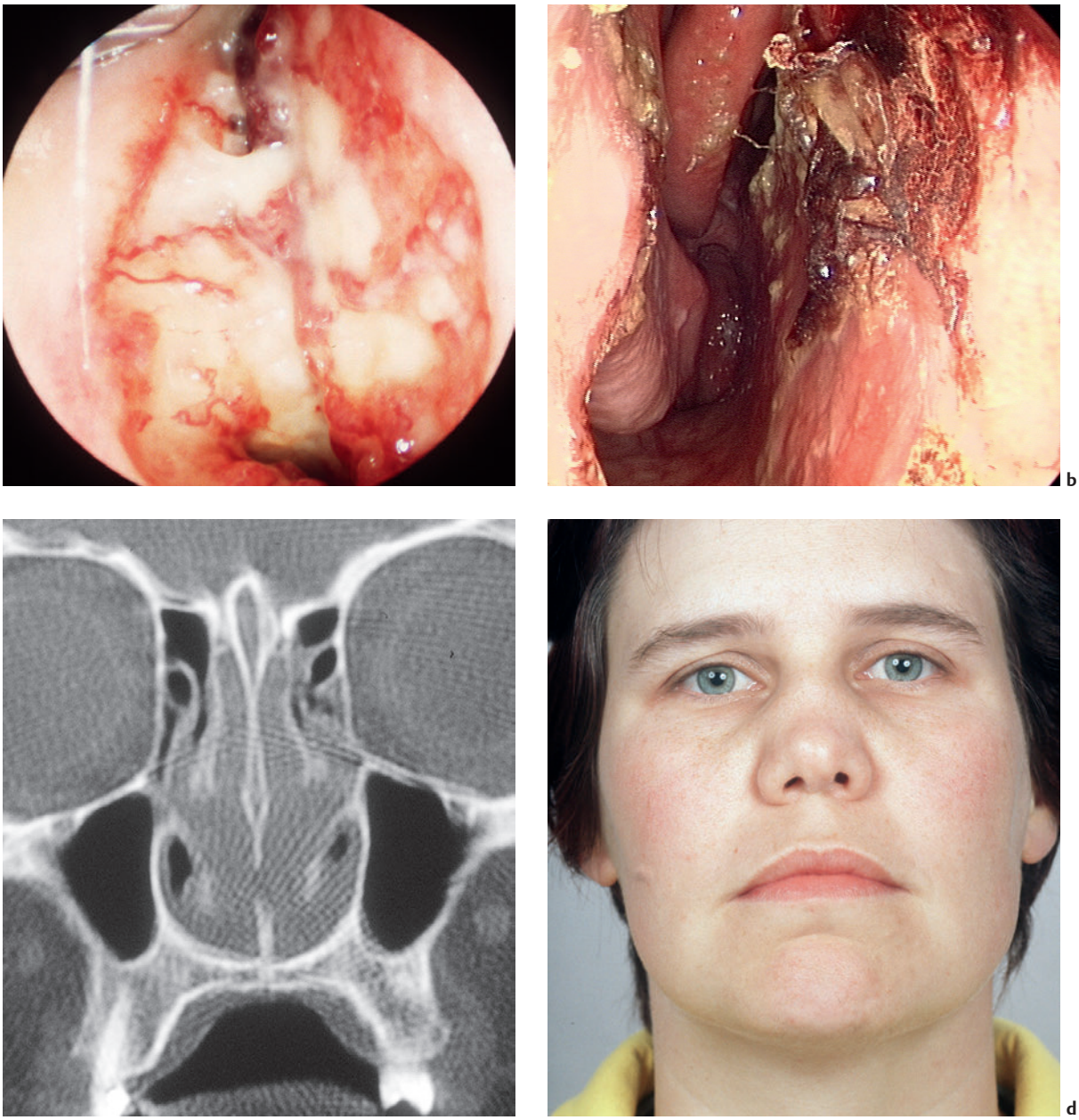


Fig. 1.4a–d Endoscopic appearance of a range of pathological conditions.

- **a** Endoscopic appearance of granulations.
- **b** Crusts in Wegener granulomatosis.
- **c** Coronal computed tomography scan showing the mucosal changes consistent with Wegener granulomatosis; there is often bony erosion as well.
- **d** Collapse of the nasal dorsum often seen in nasal Wegener granulomatosis.

time allowing, so that he or she can see that your advice is given having considered this information.

Anyone offering medical advice should not overplay the benefit of any treatment they propose. To be over-enthusiastic or excessively optimistic runs the risk of the patient being disappointed. That does not mean that all sense of hope for an improvement should be dashed but it is wise to qualify what the patient might realistically expect. This situation is not helped by claims that a treatment "works" or has been shown to produce a "statistically significant improvement," when in reality the studies on which these are based only show that the mean patient symptom score has reduced from say 8 to 5 out of 10 in severity; in other words the patient still has a mean

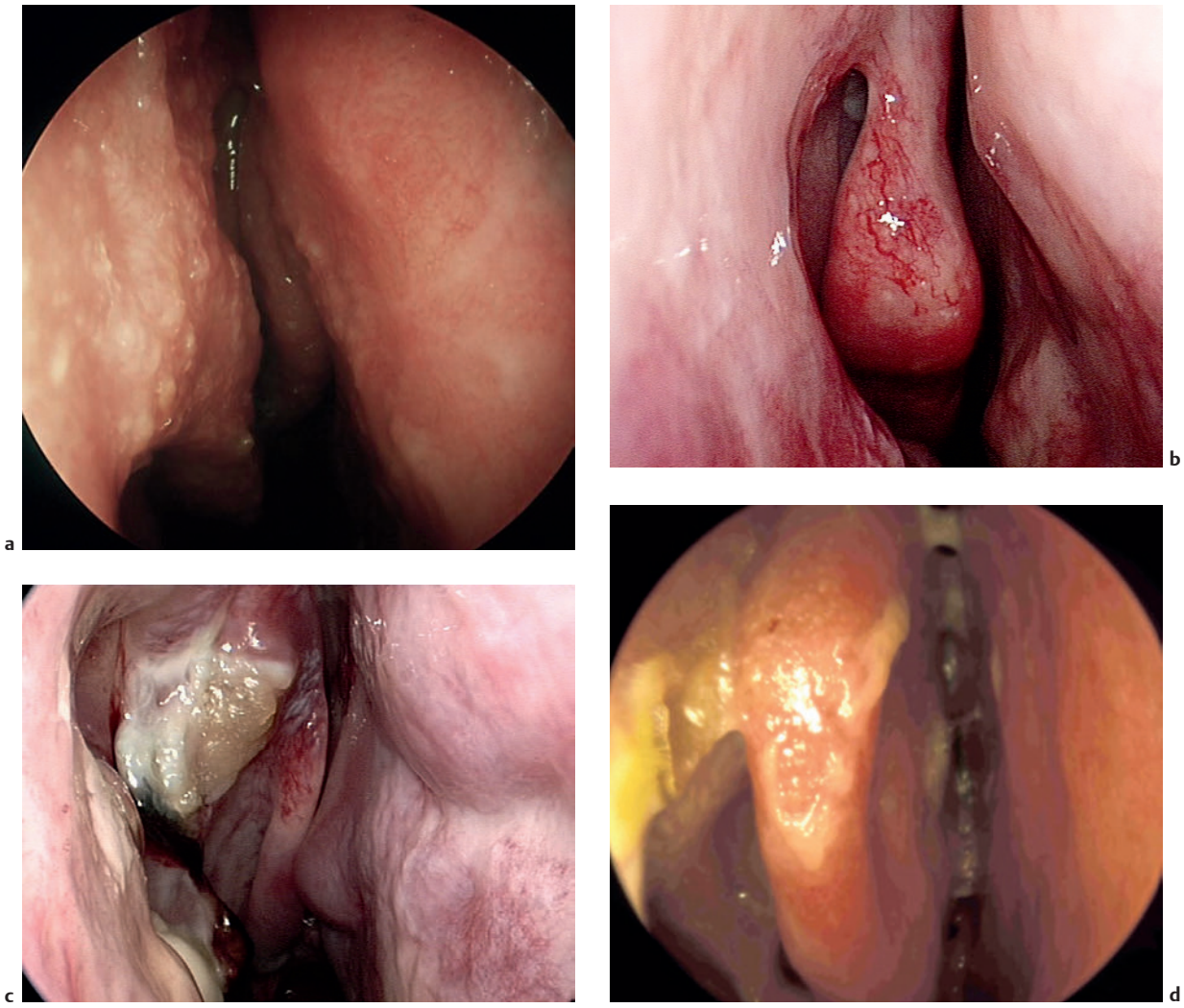


Fig. 1.5a–d Endoscopic appearance of a range of pathological conditions.

a Pale cobblestones in sarcoidosis.
b Hyperemic mucosa on the middle turbinate—a variation of normal.
c Purulent secretions in cystic fibrosis.
d Dry mucosa with stagnant mucus in the right nasal airway.

symptom score of 5 out of 10. Many rhinological studies show that, after treatment, high residual symptom scores remain.

The patient's expectations should ideally coincide with the surgeon's prognosis. Hence, it is worth communicating to him or her which specific symptoms will not be helped. It is often worthwhile making this clear in any correspondence to the referring doctor and sending the patient a copy.

Some Psychological Aspects of Nasal Symptoms

In rhinology many patients have symptoms that do not appear to be due to organic disease and a proportion of these patients are somatizing or have medically unexplained symptoms. Somatizing patients attribute their distressing symptoms to a physical illness that cannot be fully accounted for by organic disease. Often the distress caused by the symptoms is out of proportion when compared with that of someone with organic disease and there may be symptoms of depression and anxiety. Approximately 25% of patients attending an ear, nose, and throat surgeon are somatizing. Typical symptoms include fullness in the head or ears, dizziness without vertigo, catarrh or a dry sore throat. There are often many symptoms that change in nature over time. It is common for these patients to have seen many doctors.

It is important to exclude any organic pathology that can present with these symptoms by performing a thorough examination and any relevant investigation.

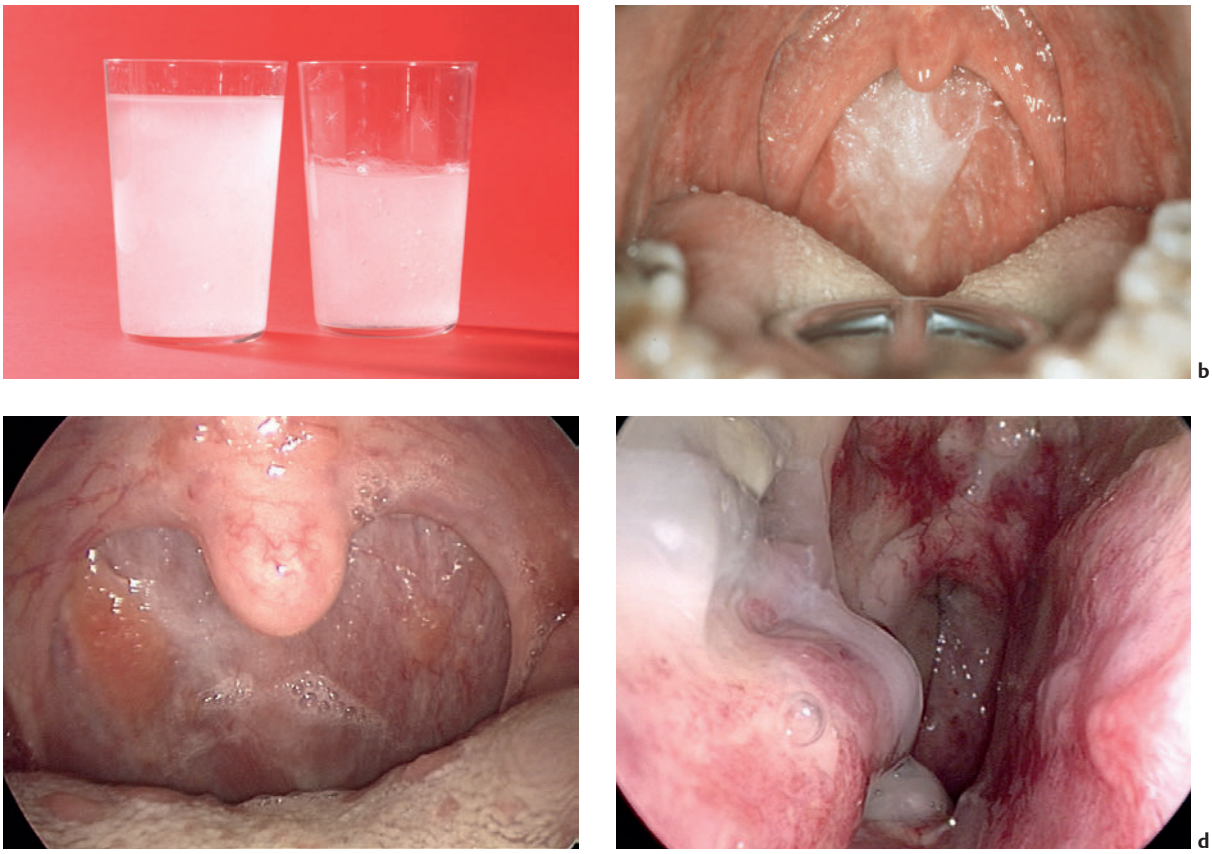


Fig. 1.6a–d Endoscopic appearance of a range of pathological conditions.

a The amount of secretions produced per day from the upper and lower respiratory tract.
b Thick postnasal mucus in the pharynx.
c Thin tenacious mucus in the pharynx.
d Purulent secretions from the middle meatus tracking over clear secretions.

If the patient is thought be somatizing then it is important to minimize the number of investigations, avoid unnecessary treatment, and address any psychological problems, while fully acknowledging the patient's symptoms. Explanations that recognize the symptoms and offer a tangible and involving explanation are more likely to satisfy patients and empower them (Dowrick et al 2004). By legitimizing the patient's symptoms, yet not colluding with the patient by acquiescing with their explanation, and offering a tangible mechanism that the patient 'owns' and gives him or her an opportunity for self management, the patient is less likely to make further demands on health care. The presentation of normal test results accompanied by reassurance has little effect on patients' doubts and anxieties. Central to making a difference to the patient is finding out what his or her fears are (Balint 2000). Cognitive behavior therapy, the treatment of comorbid depression and anxiety, and, in patients with moderate psychological problems, psychiatric help are the mainstays of treatment (van der Feltz-Cornelis et al 2012). If the patient's ability to function is impaired by the condition it may be necessary to seek psychiatric help.

Optimize Medical Treatment

While it is accepted that medical treatment will complement surgery in making the mucosa as healthy as possible, it is less well recognized that it can be a useful predictor of what can be achieved by surgery. For example, in a patient with anosmia and nasal polyposis, the use of oral and topical steroids can indicate the patient's remaining olfactory potential. If the patient has no sense of smell after a course of oral steroids (**Fig. 1.7a–d**), not even temporarily, then the surgeon must be very guarded in promising the patient that his or her sense of smell will be improved by surgery.

Tailor the Surgery to Fit the Extent of the Problem

There is a price to be paid for extensive tissue removal. That price may include the loss of olfactory mucosa, frontonasal stenosis, altered sensation, dryness, and an

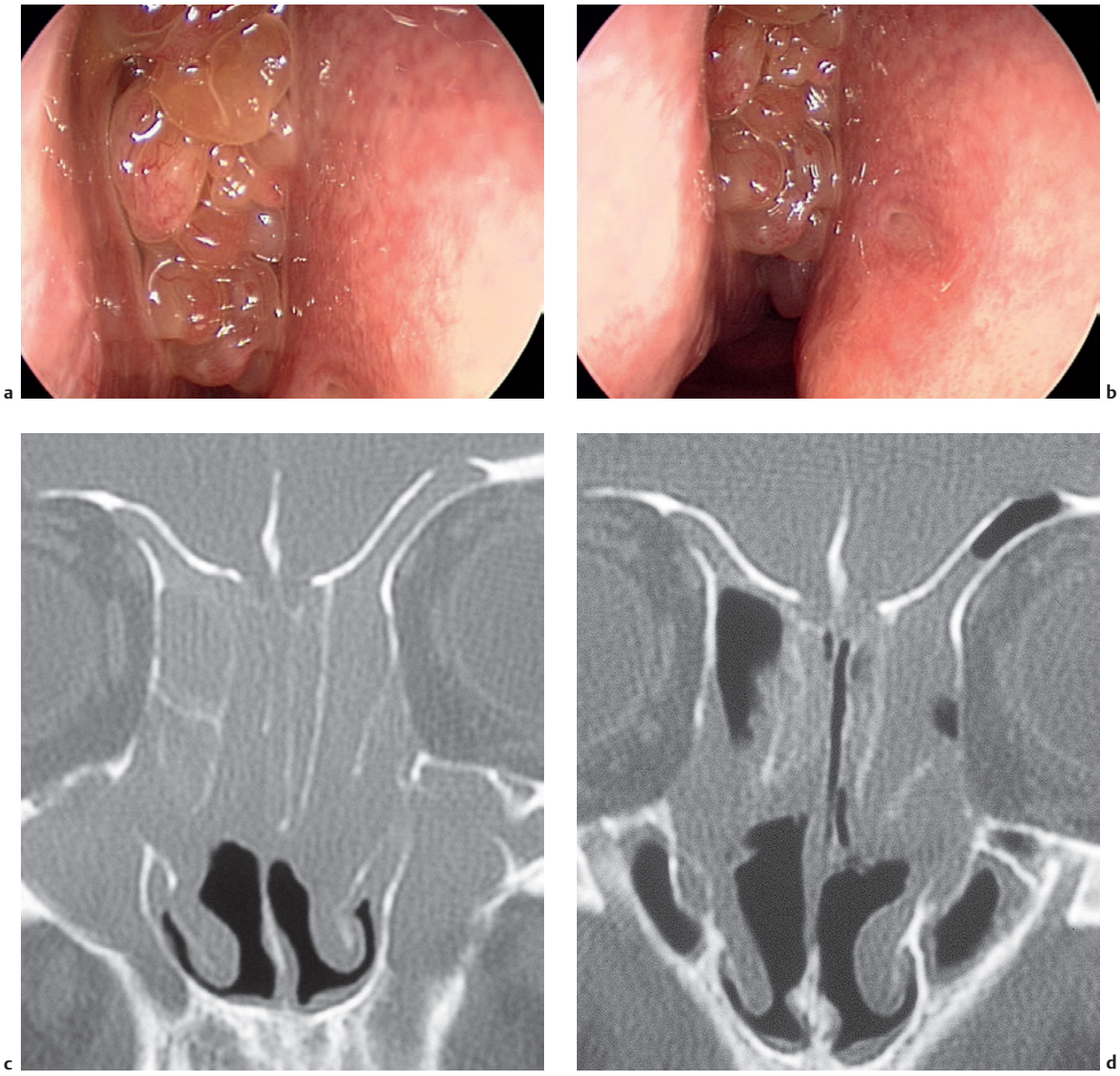


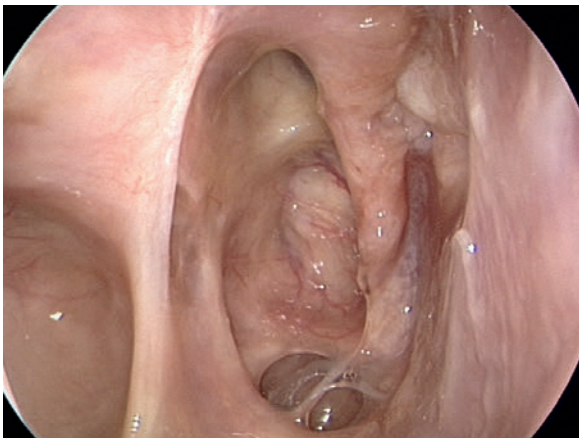
Fig. 1.7a–d Endoscopic and computed tomographic appearance before and after medical treatment.
a,b Endoscopic views before and after oral steroids. **c,d** Computed tomographic images before and after oral steroids.

increased risk of violating the boundaries of the paranasal sinuses (**Fig. 1.8a,b**). If a full thickness mucosal defect is created there will be persistent crusting until the mucosa has regenerated and it may take up to 1 year for cilia to return to normal so mucus can stagnate and dry causing debris that can collect for many months.

Surgery is primarily aimed at improving ventilation of the sinuses and restoring mucociliary clearance. Removal of tissue alone does not cure mucosal disease. After a trial of medical treatment, it is possible to estimate the extent of surgery that will be of most benefit. This means that it is often possible to preserve valuable tissue, such as mucosa in the olfactory cleft, that might otherwise be removed (**Fig. 1.9a–d**). Far less surgery is needed if medical treatment has been given preoperatively.

Minimize Surgical Morbidity

Morbidity can be caused by poor surgical technique, and in particular may arise from excessive tissue removal. Good surgical technique is based on setting explicit goals and achieving these with the minimum amount of tissue trauma.



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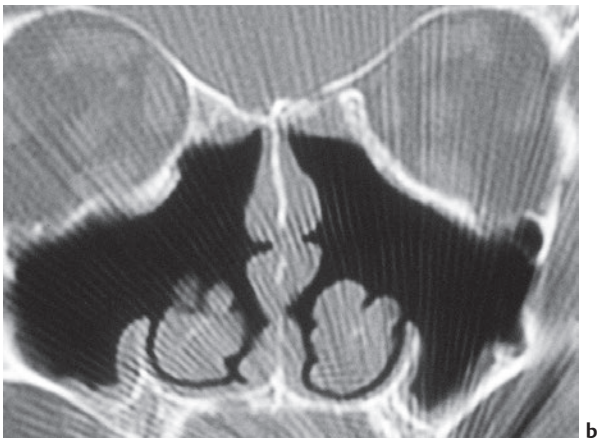

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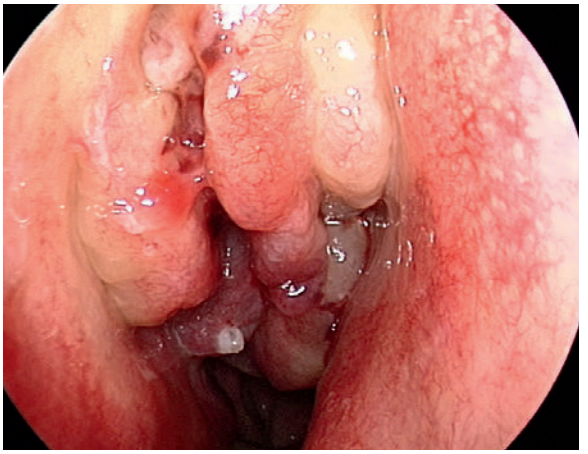
Fig. 1.8a,b Endoscopic appearance after extensive surgery.

a Endoscopic views after overzealous removal of olfactory mucosa.

b Computed tomographic images after overzealous removal of olfactory mucosa.



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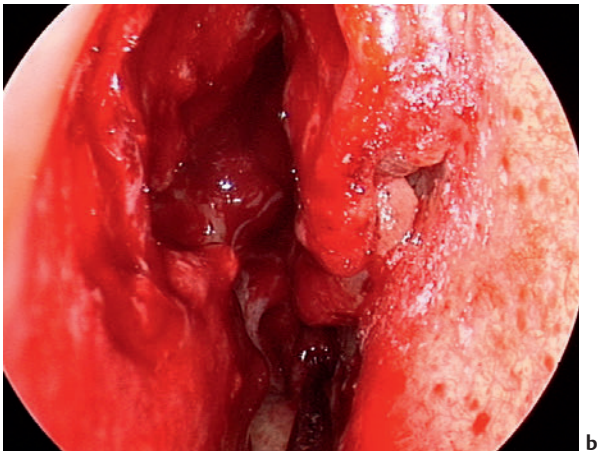

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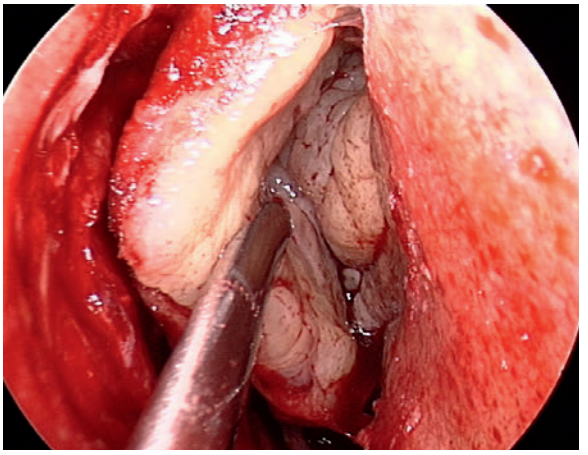

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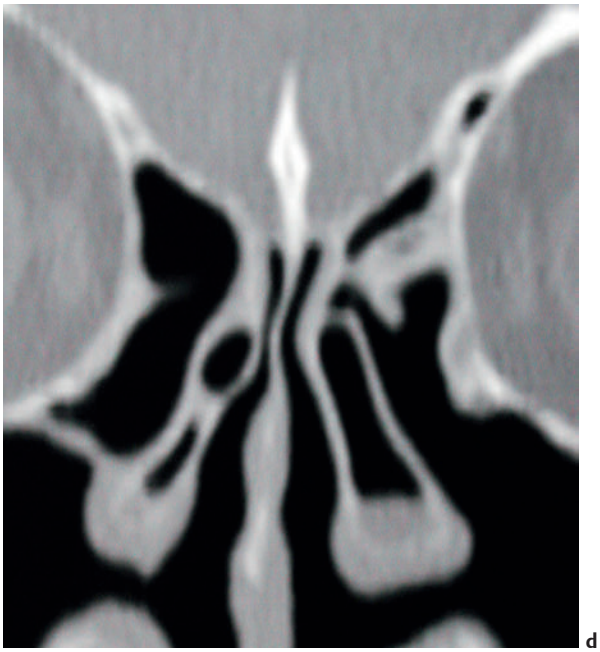

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Fig. 1.9a–d Endoscopic appearance of a range of pathological conditions.

a Right nasal airway showing severe nasal polyposis after oral steroids just before surgery.

b Perioperative view after ethmoidectomy.

c Perioperative gentle lateralization of the middle turbinate (note preservation of olfactory mucosa).

d Postoperative computed tomographic view to show open olfactory cleft.

How Can This Be Achieved?

Work out what surgical steps are needed and then address them systematically. This strategy will not only avoid unnecessary tissue removal but is also very time efficient. Progress is made step by step rather than by exploring the sinuses in a haphazard way. It is clear to the experienced surgeon when observing surgeons who are unfamiliar with operating because the purpose in their movements is limited and they often spend a lot of time aimlessly sucking and prodding. You must decide what needs to be done, and in particular which step needs to be done next, and then do this as atraumatically as possible. This means:

- Punching tissue rather than tearing it
- Preserving mucosal integrity in the frontonasal recess
- Respecting olfactory mucosa
- Avoiding mucosal damage to adjacent surfaces (**Fig. 1.10a,b**).

The surgeon must be aware of the variations that can occur in anatomy and the potential to cause damage to the surrounding structures.

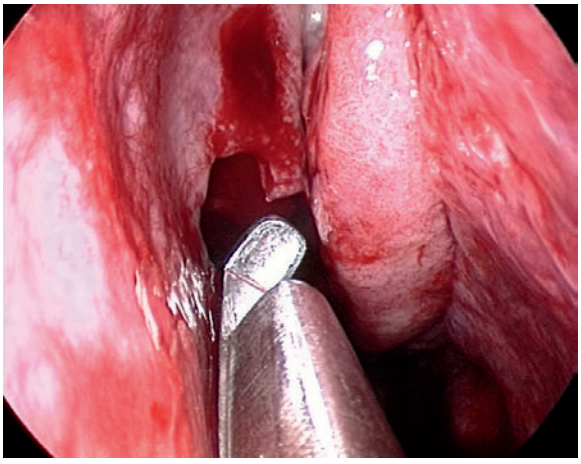
Sense of Smell Should Be Preserved at All Costs

Surgeons unfortunately often underestimate the importance of sense of smell to the patient. It is a sense that is all too often forgotten and may escape the notice of both surgeon and patient. The reason may be that the loss of this sense often creeps up on the patient slowly or that the patient does not recognize that this loss is responsible for the reduced enjoyment of food. In any case, the rewards for preserving or restoring their sense of smell are enormous. The surgical strategy to achieve this goal centers on maximum preoperative medical treatment to minimize trauma to the olfactory mucosa, and to respect all olfactory mucosa whenever possible by leaving it alone, even if it is polypoid.

The Importance of Postoperative Treatment

Unfortunately, surgery on its own cannot achieve or maintain healthy nasal mucosa in most patients with chronic rhinosinusitis. Accompanying medical treatment takes on a central role. During the operation, diseased mucosa is removed that has not recovered during preoperative medical treatment, thereby optimizing the drainage zones from the sinuses. Surgery may be able to overcome mucosa–mucosa contact and restore mucociliary clearance, remove diseased tissue, and allow access to topical nasal treatment, but surgery in and of itself cannot cure intrinsic nasal disease (**Fig. 1.11a,b**).

Patients need to be made aware of the need for continuing treatment to achieve the best possible result and an improved quality of life. One way of getting this message across to your patients with intrinsic mucosal disease is to tell them that it is like “asthma of the nose,” and they will need to keep the lining under control by regular medical treatment. This will help to prevent disappointment.



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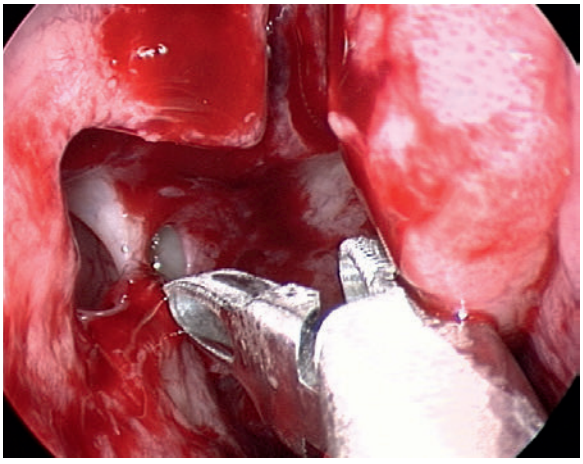

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Fig. 1.10a,b Punches and through-cutting instruments minimise mucosal trauma.

a Use of Hajek forceps to neatly remove the mucosa and bone of a right uncinate process.

b Through-cutting forceps joining natural and accessory ostia.

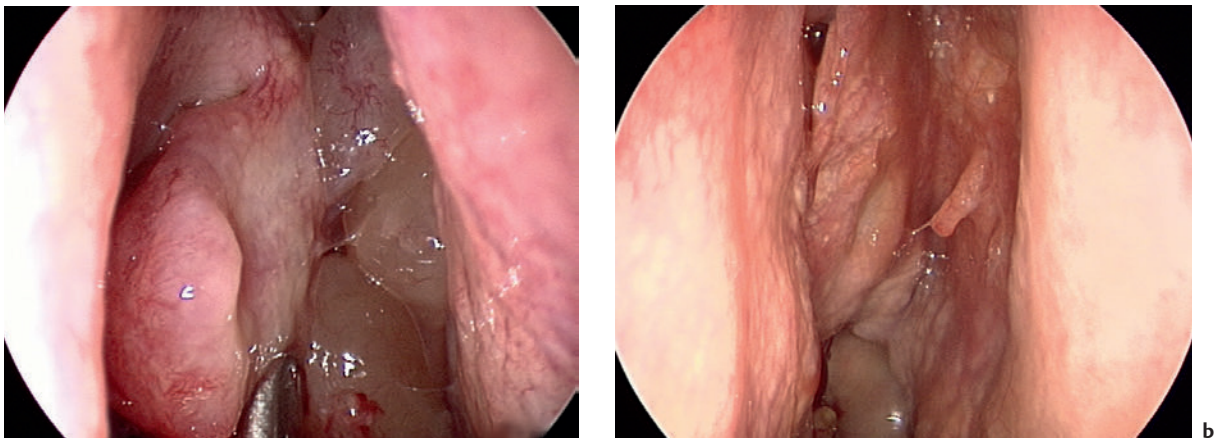


Fig. 1.11a,b Preserve olfactory mucosa where possible.

a Nasal polyps in the olfactory area medial to the middle turbinate, deliberately not removed at surgery.

b The superior turbinate can now be seen after lateralization of the middle turbinate along with 2 months of topical nasal steroids.

2 Pathophysiology of Rhinosinusitis

A key aim for both surgeons and physicians is to arrive at a diagnosis. This is not always straightforward given the poor specificity and sensitivity of nasal symptoms and signs in making many rhinologic diagnoses.

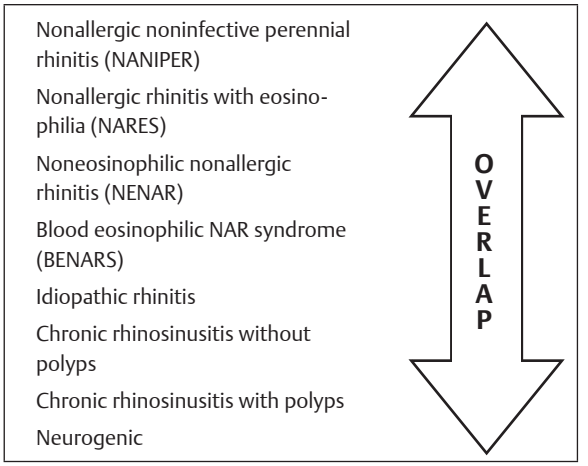
However, making a diagnosis helps to determine which treatment strategy is most likely to be effective. Implicit in this is the belief that the treatment will provide symptomatic help for the attributed diagnosis.

Treatment works well for many specific diagnoses as is described later in this book, e.g., allergic aspergillosis, a frontoethmoidal mucocele or inverted papilloma. However, there are many rhinologic conditions that pose both a diagnostic and a treatment challenge because we have a limited understanding of their pathophysiology, e.g., chronic rhinosinusitis and idiopathic rhinitis. This chapter tries to face up to what we do not know about these conditions and put this problem into a clinical context. Fortunately, although we may not fully understand the pathogenesis of some of these conditions, classifying these patients using clinical characteristics has enabled

Table 2.1 Classification of rhinosinusitis

Type of rhinosinusitis	Comments
Allergic rhinosinusitis	
Seasonal	Intermittent (IgE-mediated)
Perennial	Persistent (IgE-mediated)
Allergic fungal rhinosinusitis (IgE-mediated)	Diagnostic criteria: Antigen-specific IgE/IgG raised, eosinophil count raised, total serum IgE raised. Mucin—eosinophils, Charcot–Leyden crystals, scanty fungal hyphae (Gomeri methenamine silver stain, periodic acid Schiff, Fontana–Masson stain, calcofluor white method)
Entopy or local allergic rhinitis (local allergy in the absence of atopy-negative skin prick tests and normal specific IgE)	Supported by a positive nasal challenge with a negative reaction to a control challenge, IgE in washings, or immunocytochemical evidence of local IgE production
Nonallergic rhinosinusitis	
Rhinitis medicamentosa	Drug induced (history, and stopping medication with resolution support diagnosis)
Infections	
Viral	
Acute bacterial	
Chronic bacterial	
Fungal	
Cystic fibrosis	
Vasculitus	Wegener granulomatosis, Churg–Strauss syndrome
Sarcoid	
Atrophic	
Hormonal	Estrogen related: stopping the contraceptive pill or hormone replacement therapy improves symptoms, or the end of pregnancy resolves symptoms
Occupational	Usually irritant, care should be taken as it perhaps provokes allergic hyper-reactivity, occasionally allergic. Removal from environment improves symptoms. Challenge helps confirm allergy if there is an allergic basis
Ozena	
Empty nose syndrome	
Autonomic rhinitis	Rhinorrhea as a primary symptom responsive to ipratropium bromide spray four times daily
Structural	Adenoidal hypertrophy, post laryngectomy
Chronic rhinosinusitis	
Chronic rhinosinusitis with polyposis	

Fig 2.1 Where there is controversy about classification



some progress to be made in treating these groups (**Table 2.1 and Fig. 2.1**). It is useful to try to address what we know about the etiology of mucosal disease to place what we are doing in context.

The term rhinosinusitis is better than rhinitis because almost all nasal conditions are not solely limited to the mucosa of the sinuses or the nasal airway.

Chronic Rhinosinusitis

It is thought that chronic rhinosinusitis (CRS) is not one uniform disease but the result of many different pathogens, allergens, or foreign proteins on hosts with different gene expression at the mucosal level. This results in different immune responses, inflammatory mechanisms, and regulation of these responses.

At present the clinical definition of CRS is very broad and is based on clinical symptoms, computed tomography, and endoscopy but *not* on pathology.

Definition of CRS

Inflammation of the nose/paranasal sinuses with two or more of the following symptoms (Fokkens et al 2012):

- Blockage/congestion
- Anterior or postnasal discharge
- Facial pain/pressure (on its own rarely due to rhinosinusitis)
- Reduction in or loss of smell

and either

- Endoscopic signs of polyps/mucopurulent discharge/edema

or

- Mucosal obstruction of the middle meatus

and/or

- Computed tomographic mucosal changes within the ostiomeatal complex and/or sinuses.

There are many theories and studies of CRS in different population groups and as a result there is little consensus. One of the problems is that, understandably, most studies center on one area of investigation. A holistic picture that analyzes clinical groups along with pathologic groups has been difficult to achieve.

Whatever the mechanism that causes mucosal thickening, there appears to be a balance between the upregulators of inflammation and/or reduced apoptosis, and the downregulators of these processes (**Fig. 2.2**). This is akin to the dynamics that are found in the fibrinolytic and thrombogenic system that exists in blood. Each individual has a different ability to express certain cytokines or have a mechanism to downregulate inflammatory or neurogenic mediators that will influence whether any insult to the nasal lining has a limited or persistent effect. To illustrate the dynamic and complex nature of this balance, ~ 15% of an asymptomatic population have positive skin prick tests and 5% have raised specific immunoglobulin E (IgE) but have no symptoms or signs of allergic rhinitis.

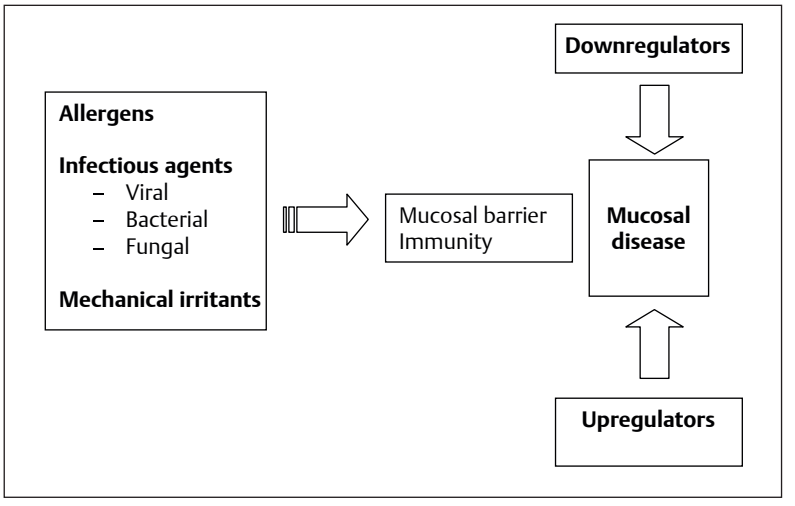


Fig. 2.2 Mucosal homeostasis: a dynamic system.

The analogy to the blood's fibrinolytic and thrombogenic system is pertinent but it is important to appreciate that in CRS it is not a simple summative system. For example all the upregulators listed below do not simply add up together to be balanced against the downregulators. The interaction of different cytokines and chemokines, and the influence of mast cells and lymphocytes, are more complex. This is illustrated by the lack of direct evidence that allergy is a major source of sinonasal inflammation in CRS (Pant et al 2009).

It is important to appreciate that most individuals with CRS do not have an infective basis for their disease. However, in those who do have persistent or recurrent infection as the initiator of their disease, it is vital to consider their immunity.

Upregulators (Many Influenced by Genetic Factors)

Cellular Basis of IgE-mediated Inflammation

- Mast cells and basophils express interleukin-4 (IL-4), IL-13, and CD40 ligand and therefore have the potential to augment/amplify IgE synthesis in a nonspecific manner.

Cytokines Involved in Mast Cell Maturation and Survival Include IL-4, IL-5, IL-6, IL-8, IL-9, IL-13, and Interferon-γ

- Basophils are detectable in the bronchial mucosa in atopic asthma, and they increase in numbers and express IL-4 during allergen-induced late responses.
- Eosinophils produce lipid mediators, including leukotriene C4 and platelet-activating factor. They also produce a range of cytokines, including IL-4, IL-5, and granulocyte–macrophage colony-stimulating factor (GM-CSF). Chemokine receptor 3 and very late antigen 4 (VLA-4) occur in response to chemokines, particularly eotaxin and RANTES (regulated upon activation, normal T-cell expressed and secreted).
- Adhesion of eosinophils to the vascular endothelium is dependent on binding of vascular cellular adhesion molecule-1 (VCAM-1) produced from the vascular endothelium to its ligand VLA-4 on the eosinophil surface.
- Tumor necrosis factor-α.
- GM-CSF.
- Reduced apoptosis (Powe et al 2009).

Bacterial

- Biofilms that harbor bacteria which produce toxins. Those with *Pseudomonas* spp. are associated with more severe disease (Foreman et al 2011)
- Bacterial toxins
- Osteitis (it is uncertain whether this is a reaction to inflammation or a cause of persistent disease) (Lee et al 2006).

Fungal

- A non-IgE-mediated, mixed T helper type 1/type 2 cytokine response to fungi. A 60-kDa component of *Alternaria* degranulates eosinophils. Eosinophils release major basic protein. *Alternaria* proteins are recognized by antigen-presenting cells and present them to T cells whose response attracts and activates eosinophils. Cytokines with a nonspecific protease response are released. It is not clear whether this is a disease-specific response.

Superantigen Hypothesis

- Staphylococci secrete superantigen toxins, which stimulate T cells. T cells produce cytokines and local polyclonal IgE and eosinophils are recruited. Specific IgE directed against toxins in polyp tissue is found in 50% of patients with idiopathic polyposis (not found in CRS without polyposis). A staphylococcal protein A has been found to cause mast cell degranulation. Staphylococci are present in many patients with nasal polyposis and it is not clear whether they are causative or the result of colonization of the mucus that tends to stagnate because of the dysfunctional ciliary clearance that occurs with nasal polyps (Bachert et al 2001).

Protease Activity

- House dust mite allergen, Der p 1 (Kauffman et al 2006; Furmonaviciene et al 2007)
- *Alternaria*
- *Staphylococcus aureus.*

Neuronal

- Sensory nerves themselves may produce inflammation by an antidromic axon reflex, which causes sensory nerves to release neuropeptides such as substance P and neurokinin A.
- Nerve growth factor, responsible for maturation and development of sensory nerves, is present in nasal fluids of patients with chronic allergic rhinitis and is increased after allergen challenge.

Other Factors

- Damaged epithelium may amplify any response
- Mechanical irritation
- Chemical exposure
- Free light chains are raised in both allergic and nonatopic rhinitis mucosa suggesting a role in nasal hypersensitivity (Powe et al 2010a).

Rare Conditions

- Primary ciliary dyskinesia
- Immunodeficiency
- Cystic fibrosis.

Limited Evidence Associated with Anatomical Factors

Downregulators (Many Influenced by Genetic Factors)

Cellular Mechanisms

- IL-10 can suppress inflammation
- Transcription factor Fox P3
- IL-6 role in inhibitory local innate immune responses
- Transforming growth factor-b (TGF-b) (also has some proinflammatory effects)
- Sialic acid-binding, immunoglobulin-like lectin-8 (siglec-8) receptor
- "Functional" IgG antibodies may downregulate antigen-specific T-cell responses
- Regulatory T cells might also downregulate T-cell responses through cell–cell contact, as well as through the direct effect of these cytokines

Innate Immunity

- Nitric oxide
- Defensins
- Pattern recognition of damage associated with foreign proteins that induce a reaction to help maintain the barrier (Tan et al 2010)
- SPINKS5, a polyvalent antiprotease that encodes LEKT1, which is a protease inhibitor and helps to protect gap junctions
- S100 proteins with direct antimicrobial and antifungal properties
- IL-22 activates epithelial cells in the innate response
- Mucociliary clearance
- Toll-like receptors (Vanhinsbergh et al 2007; Tan et al 2010)

Mast Cells

- Mast cells can cleave IgE with a protease and may play a role in limiting the late phase reaction (Rauter et al 2008; Hakim-Rad et al 2009).

Mucosal Barrier

- Pseudostratified columnar epithelium
- Pattern recognition receptors recognize damage and initiate a response, e.g., Toll-like receptors
- Tight junctions
- Mucociliary clearance
- Defensins
- Lysosymes
- Mucus

Comments on Specific Conditions

Allergic Rhinosinusitis

Allergic rhinitis and seasonal or childhood asthma appear to share a unified disease pathway attributed to an inflammatory cell cascade involving T helper type 2 cells with the production of allergen-specific IgE antibodies from activated B cells (plasma cells). $CD4^+$ T cells, also called T helper cells, IL-4, IL-5, and IL-13 are all involved in the allergic response. IgE is taken up by receptors on mast cells and when cross-linked by allergen, degranulation occurs with the release of inflammatory mediators. Mast cells are abundant in the nasal mucosa of allergic rhinitis mucosa and submucosal lung tissue, consistent with their role in maintaining inflammation in allergic airway disease.

Much is made of the balance or dynamics of the predisposition toward either a T helper type 1 (defense against pathogens) or a T helper type 2 (important in parasitic immunity and allergy) pattern of cytokine production, although, as our understanding progresses, this view appears to be simplistic. However, degranulation can be caused by IgG and immunoglobulin-dependent pathways that include Toll-like receptors, complement proteins, calcium-binding proteins, and some cytokines.

Entopy or Local Allergy

Approximately 20% of symptomatic people who are skin prick test negative and have normal IgE levels have a positive nasal challenge to house dust mite but not to saline. They also have local IgE in their secretions. There is compelling evidence that has shown that IgE is produced locally within the nasal mucosa in atopic subjects (Huggins and Brostoff 1975; Durham et al 1997; Kleinjan et al 1997; Cameron et al 1998; Kleinjanajk et al 2000; Smurthwaite et al 2001; Takhar et al 2005; Roden et al 2010).

Interestingly, some patients with apparently nonallergic rhinitis share similar histologic mucosal features characterized with increased numbers of mast cells and eosinophils and produce local IgE (Powe et al 2001, 2003, 2004, 2006, 2010b).

The diagnostic measures proposed in this group include the detection of specific IgE in the nasal secretions after exposure to the aeroallergen along with the local production of tryptase and eosinophil cationic protein (Rondón et al 2010).

Nonallergic Rhinitis

Listed above are the recognized causes of nonallergic rhinitis. However, this leaves a group whose etiology is unclear.

The terms nonallergic noninfective perennial rhinitis (NANIPER), nonallergic rhinitis with eosinophilia (NARES), noneosinophilic nonallergic rhinitis (NENAR), blood eosinophilic NAR syndrome (BENARS) and idiopathic rhinitis have all been used (see **Table 2.1**). There are no specific diagnostic tests for this group because they are largely a diagnosis of exclusion (e.g., no vasculitis, hormonal, or environmental causes, rhinitis medicamentosa). The cause of idiopathic rhinitis, or those with nonallergic rhinitis where no cause in the list above is present, has evaded medical science (Carney and Jones 1996). This is in part because most studies focus on one area of investigation and an analysis that examines this clinical group along with the pathology has yet to be done.

"It is likely that neuronal mechanisms, T cells, innate immunity and possibly auto-immune responses all play a role in nonallergic rhinitis and may also contribute to the symptoms of allergic rhinitis" (Bousquet et al 2008).

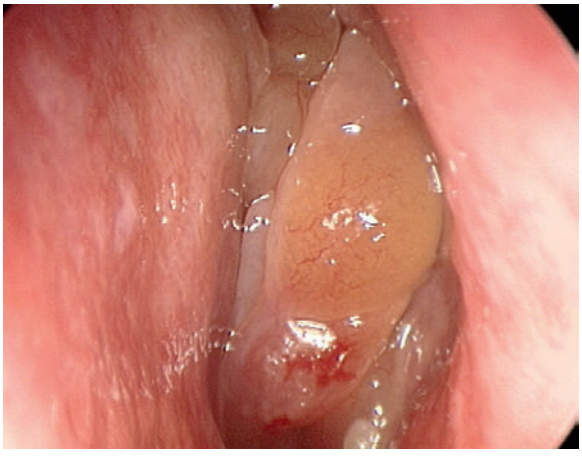
A Note on the Term "Nasal Polyposis"

The term nasal polyp is very nonspecific as it is not a diagnosis but a sign.

A minority can result from a bacterial infection or allergic fungal sinusitis and, if they are unilateral, neoplasia needs to be excluded. The most common type of nasal polyposis is associated with late-onset asthma and this patient group is 10 times more likely to have nasal polyposis than the rest of the population. The prevalence of idiopathic nasal polyps is ~ 4% in the population. The term idiopathic nasal polyposis is best used to describe these individuals where the cause is uncertain although both staphylococcal superantigens and an immunologic response to fungal spores have been implicated. The term idiopathic nasal polyposis is preferable at present because it helps to remind us to exclude other recognized causes such as allergic fungal sinusitis or polyposis secondary to bacterial infection. In idiopathic nasal polyposis high levels of IL-5, IL-13, eosinophil cationic protein, TGF-β, eotaxin, RANTES, matrix metalloproteinases 7 and 9, and eosinophils are characteristic in most western populations. Raised levels of B-cell attracting chemokine-1 and stromal cell-derived factor-1α and their receptors may be partially responsible for the increase in B cells and the eosinophilic inflammation seen in CRS with nasal polyps. The complexity of the dynamics at the subcellular level is shown by the finding that 192 genes were upregulated at least twofold, and 156 genes were downregulated by at least 50% in nasal polyp tissue (Liu et al 2004). Mucociliary clearance is dysfunctional in CRS with nasal polyposis but this is probably a secondary phenomenon (Mason et al 1996).

Chronic Rhinosinusitis Without Nasal Polyps

These patients have a tendency to a T helper type 1 cell cytokine expression (Daines and Orlandi 2010). Various cells and cytokines have been found to be raised in CRS without nasal polyps including macrophages, mast cells, eosinophils (although fewer than in CRS with nasal polyposis), IL-1, IL-6, IL-8, tumor necrosis factor-α, TGF-β_1, IL-3, GM-CSF, intracellular adhesion molecule-1, myeloperoxidase, and eosinophil cationic protein. Neither IL-5 nor VCAM-1 was raised and the levels of IL-22 were reduced.



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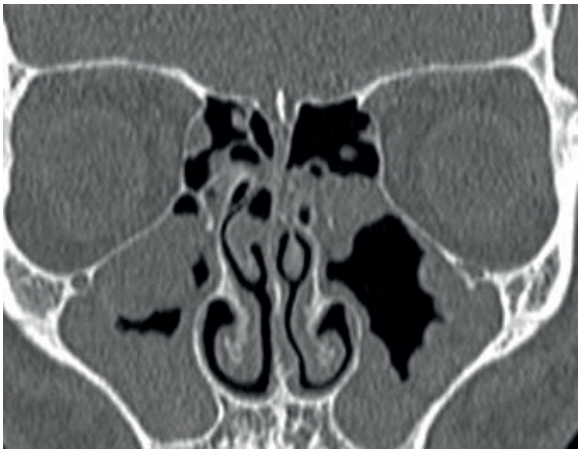


b

Fig. 2.3a,b Idiopathic nasal polyps and computed tomography scan.

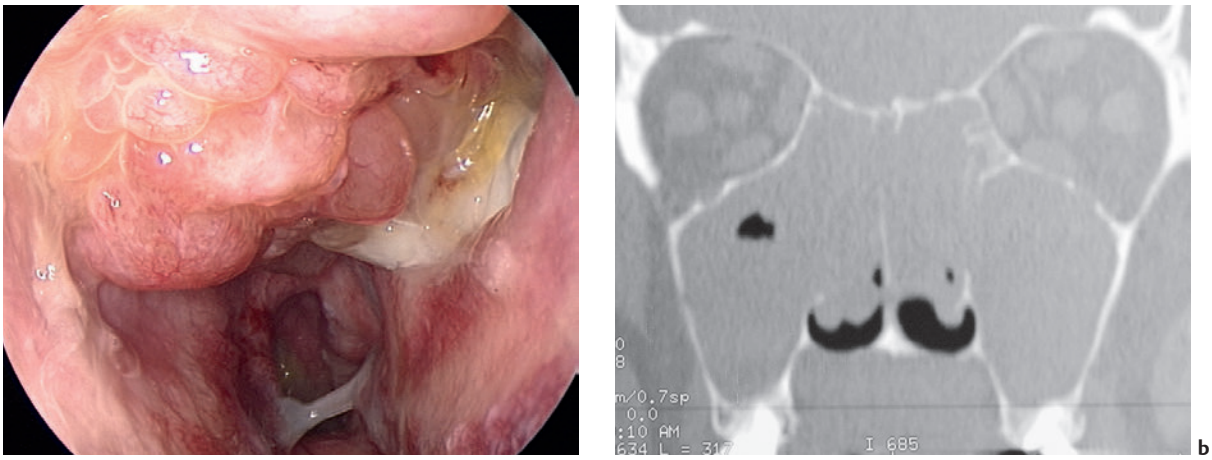


Fig. 2.4a,b Nasal polyps associated with aspirin sensitivity and computed tomography scan.

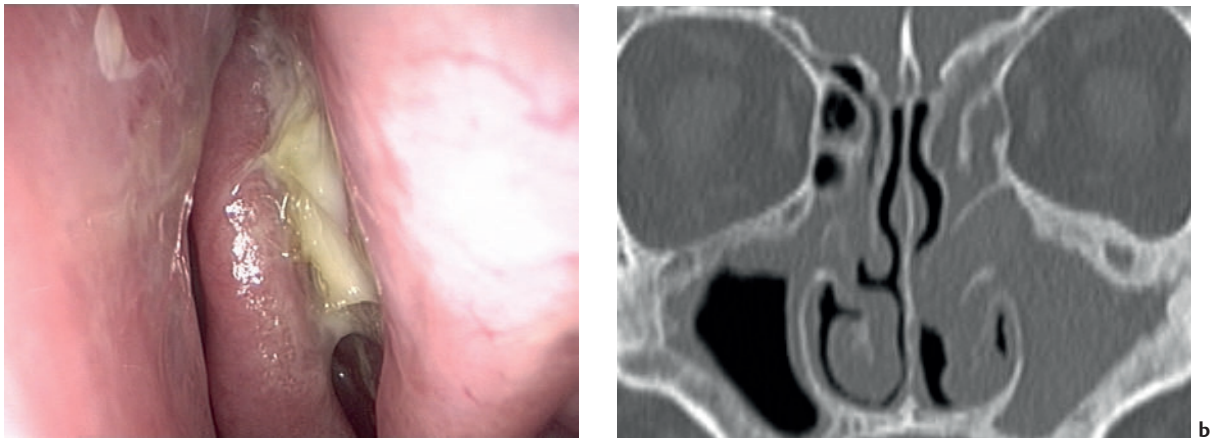


Fig. 2.5a,b Chronic rhinosinusitis with osteitis and computed tomography scan.

Practical Comments

Patients with CRS and polyps tend to do better than those without polyps after endoscopic sinus surgery.

Aspirin-sensitive patients with nasal polyposis often have worse disease as well as a poor response to steroids and all too frequently surgery provides only short-term benefit (**Figs. 2.3a,b and 2.4a,b**).

Patients with CRS and osteitis are a very difficult subgroup. In spite of medical and surgical treatment these patients tend to have a more resistant disease process (**Fig. 2.5a,b**).

Most patients with CRS have no evidence of atopy but those who do are very difficult to manage surgically. Thankfully most of these patients respond well to steroids (**Fig. 2.6**).

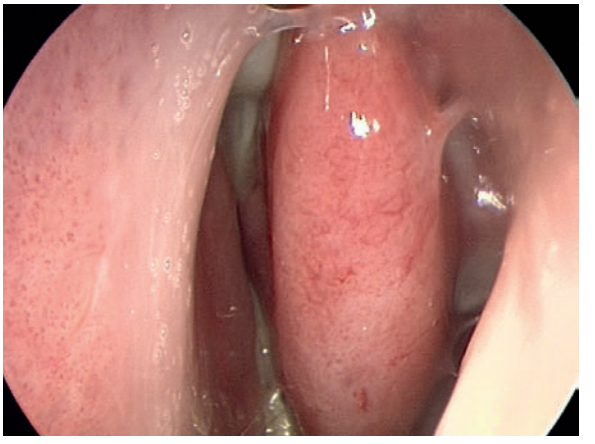


Fig. 2.6 Chronic rhinosinusitis with no evidence of atopy.

3 Who? Optimizing Diagnosis and Selection of Patients for Surgery

Symptom-oriented Patient Selection

The initial goal is to make a correct diagnosis. Many patients with rhinologic symptoms do not need surgery. The art of a good rhinologist is to select "who" will benefit from surgery. While surgery can provide an invaluable benefit in restoring patients' health and well-being, advocating surgery is not an appropriate response to all symptoms that our patients may report.

Which are the Cardinal Symptoms? What Criteria in the History are Best to Focus on?

A Summary of the Diagnostic Approach

History—Interpreting Nasal Symptoms

- Nasal obstruction
- Disorders of smell
- Rhinorrhea
- Crusting
- Facial pain and pressure
- Relevant medical history
  - Aspirin or nonsteroidal anti-inflammatory sensitive
  - Late-onset asthma
  - Response to medical treatment
  - Medication

Clinical Examination

- Examination of the nose
- Nasal endoscopy
- Posterior oropharynx
- Neck
- Eyes and cranial nerves

Investigations

- Skin prick tests
- Total and specific immunoglobulin E
- Immunity testing
- Other hematologic test
- Ciliary dysmotility/structural studies
- Sweat test
- Serum biochemistry
- Culture
- Radiology
- Histopathology

History—Interpreting Nasal Symptoms

Nasal Obstruction

This is the main reason for operating, as it is one symptom that surgery can almost be guaranteed to help in chronic rhinosinusitis with nasal polyposis (**Fig. 3.1**). However, results are disappointing in allergic rhinitis and chronic rhinosinusitis without nasal polyposis. However, be careful about operating on anyone whose primary symptom is not nasal obstruction—think twice!

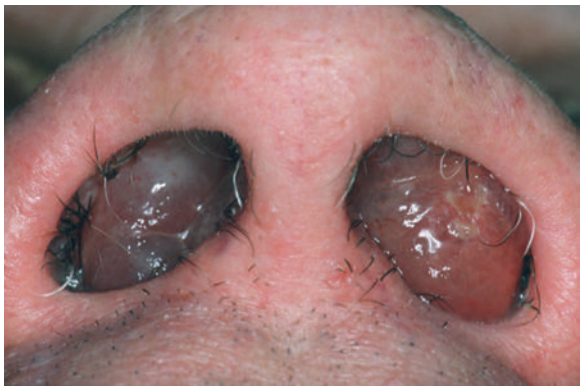


Fig. 3.1 Gross polyposis where the patient will appreciate the improvement in their airway after removal of the polyps.

Also be careful about operating on someone who complains about nasal congestion but in whom there is *no* objective sign of poor airflow. These patients may have a form of altered sensation due to a neurologic condition called midfacial segment pain. Another cause is a dry lining when the subjective sensation of airflow is reduced as the receptors lining the nasal mucosa do not work well under these conditions (**Fig. 3.2**).

If you are operating to help someone whose primary symptom is an obstructed airflow and they have idiopathic nasal polyps, it is imperative that you explain to them before surgery that they will not be cured and that they will need to comply with continuing postoperative medical treatment. Even then it is likely that they will need further surgery for recurrent polyposis in future. Otherwise you will have a disappointed and disaffected patient after surgery!

The wise words of Ian Mackay are worth reflecting on. "You want to be the last surgeon to operate on someone with idiopathic nasal polyps." This implies that

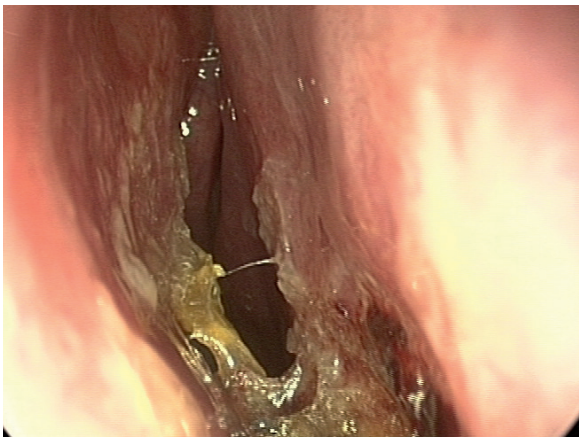


Fig. 3.2 Dry lining with mucous stagnation. The patient lacked a sensation of airflow.

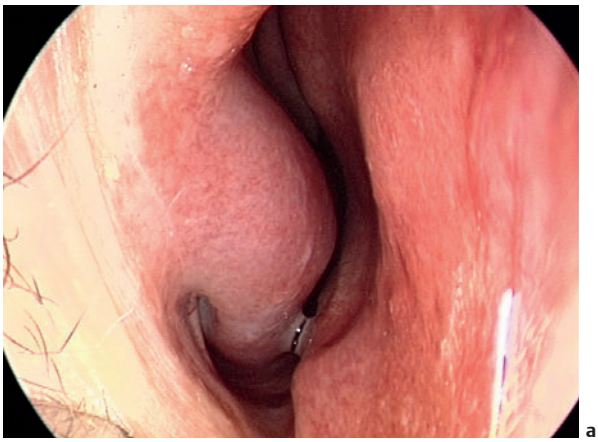
they usually recur, but often at some point in time that is impossible to discern, the patient's immunity changes and their polyps no longer return. The patient then thinks that it is the last surgeon that did a "proper" job, when it is their immunity that has corrected the situation!

Ask the patient whether they have unilateral obstruction or bilateral obstruction, or whether it alternates from side to side. Bilateral nasal obstruction is often associated with generalized rhinosinusitis, as is obstruction that alternates from side to side. If this is the case, it is likely that the patient has generalized swelling of the nasal mucosa from any cause that can produce bilateral rhinosinusitis.

The nasal cycle happens in ~ 80% of patients every 3 to 8 hours, with one side being congested while the other is clear (**Fig. 3.3a–c**). Any disease that causes a generalized swelling of the intranasal lining may lead to the nasal cycle being "disclosed" so that the patient notices that one side is partially blocked. In the disease-free individual, the amount of swelling of the nasal lining that occurs in the nasal cycle is usually insufficient to cause any symptoms.

If the patient has persistent unilateral nasal obstruction, the most common cause is septal deviation (**Fig. 3.4**). If there is any other pathology, such as a malignancy, there are often other associated symptoms in the history, such as a bloody mucous discharge, loose teeth, diplopia, or distortion of the cheek (**Fig. 3.5**).

Bilateral marked nasal obstruction associated with a reduction in taste and smell is often due to idiopathic polyposis. These patients often respond well to maximum medical treatment but when this has failed they are good candidates for surgery because their nasal obstruction can be helped. Nevertheless, the patient needs to be told that surgery is not a cure and they will need long-term medical treatment to maintain any improvement.



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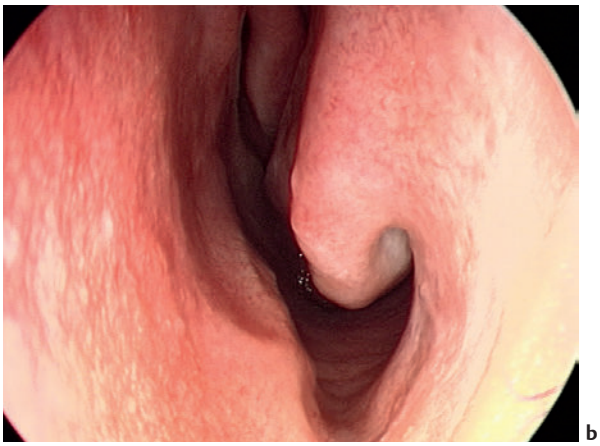

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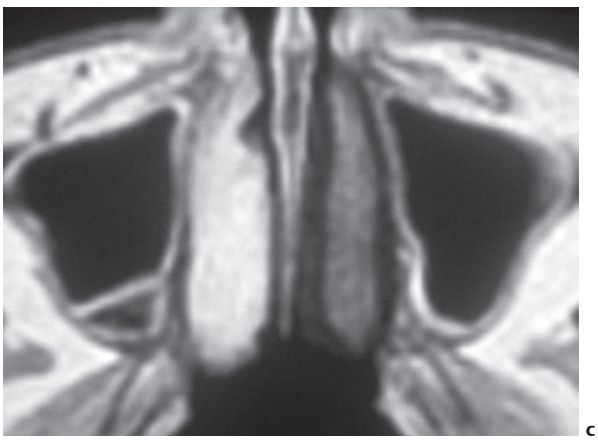


c

Fig. 3.3a–c Nasal cycle.

a,b The inferior turbinates.

c Axial magnetic resonance image showing one side congested at one point in the nasal cycle.

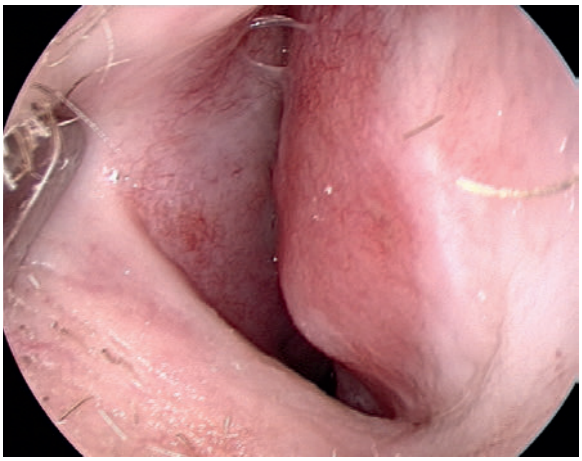


Fig. 3.4 A septal deviation in the right nasal airway.

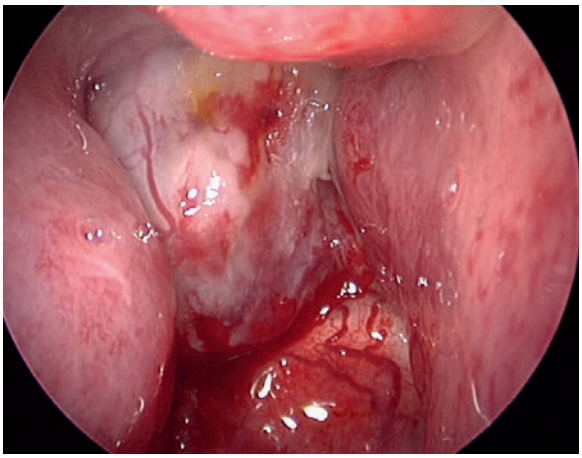


Fig. 3.5 A unilateral mass producing nosebleeds—an angiofibroma.

"Blockage" without Airway Obstruction

Be specific in asking the patient whether the sensation is of "blockage" or if there is a feeling that their nasal airflow is impaired. There is a subtle but important distinction between these two symptoms. Someone with an impairment in their airflow normally has a mechanical obstruction, whereas a patient who complains of "blockage" without any airflow obstruction is less likely to have intranasal pathology that will benefit from surgery. Be careful about operating on someone who has a sensation of "blockage" but whose airflow is normal, because these patients may have a feeling of pressure under the bridge of the nose, on either side of the bridge of the nose, or behind the eyes or supraorbital and infraorbital margins. This sensation of pressure is often not related to the obstruction of the ostiomeatal complex, as is implied in most orthodox texts. These patients often have a variation of a tension-type headache called midfacial segment pain that affects the midface. It gives them a feeling of blockage in this area (Jones 2001a) and can produce a sensation of pressure or congestion rather than pain. These symptoms usually respond to low-dose amitriptyline (▶ Video 3).

There are several other different conditions that can cause a sensation of blockage when the airway is good and it is important to make sure that you diagnose the right one as the treatment for them varies a great deal.

For the receptors within the nose to feel airflow they need to register a change in humidity and temperature. If the lining is dry then the sensation of airflow is reduced. These patients often sniff to enhance the sensation of airflow and this in turn may exacerbate the problem by causing secondary nasal valve collapse. Sniffing menthol helps temporarily because it stimulates the receptors and relieves the lack of sensation. A lack of sensation with a dry lining is not uncommon in patients who have had a lot of nasal surgery and in particular turbinate surgery of any kind. At a glance, little may appear to be abnormal but on closer examination the septal mucosa does not have any moisture on it. Sometimes there will be obvious crusting but the patient may have cleared any debris before attending. An explanation that this is the problem along with advice on regular douching is required, but, more importantly, instruction is necessary that the prolonged application of petroleum jelly over several months is often needed to allow the mucosa to recover. Petroleum jelly is placed on the little finger inside the nostril margin and then sniffed up and "milked" up by squeezing the nostrils.

Collapse of the nasal valve area can cause nasal obstruction but it is often blamed as the cause when it is secondary to other factors. The external valve is made of the ala, the skin of the vestibule, the nasal sill and the contour of the medial crus of the lower lateral cartilages. The nasal valve area and internal nasal valve are two entities that should not be confused. The nasal valve area is the narrowest portion of the nasal passage. It is bounded medially by the septum and the tuberculum of Zuckerkandl; superiorly and laterally by the caudal margin of the upper lateral cartilages, its fibroadipose attachment to the pyriform aperture, and the anterior end of the inferior turbinate; and inferiorly it is made of the floor of the pyriform aperture (**Fig. 3.6**).

The nasal valve, on the other hand, is the specific slit-like segment between the caudal margin of the upper lateral cartilage and the septum and it is measured in degrees at ~ 15°.

The mucosal soft tissue changes that affect the inferior turbinate and septum are the commonest causes that narrow the nasal valve area and in turn this can cause secondary collapse of the nasal valve. Nasal valve problems can follow excessive resection of the lower lateral cartilages, or be the result of a long returning of upper lateral cartilages, the inherent concave shape of the lower

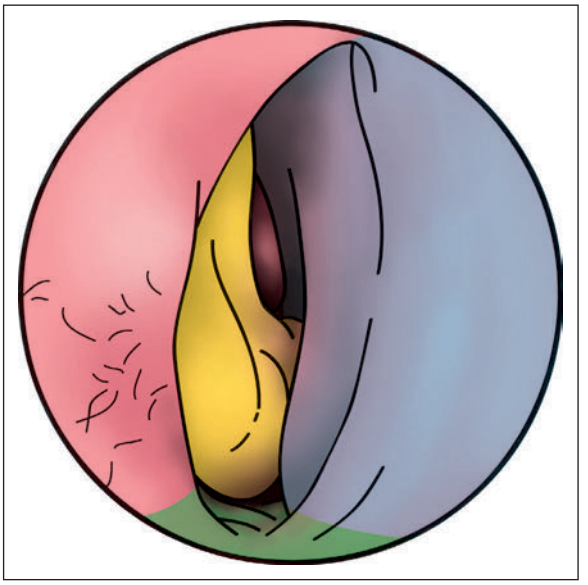


Fig. 3.6 A diagram of the nasal valve area. Yellow is the inferior turbinate, blue is the septum, green is the nasal floor, and red is the lateral nasal vestibule and upper lateral cartilages.

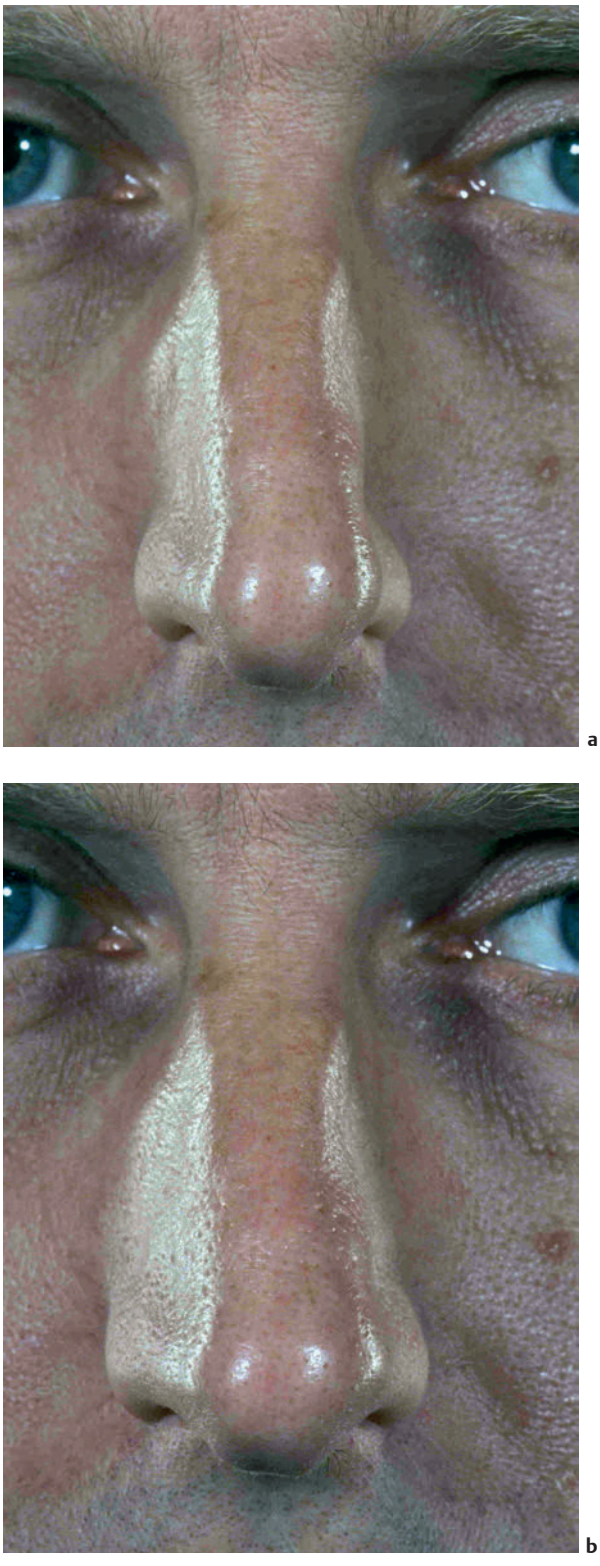
lateral cartilages, no overlap between the upper and lower lateral cartilages, inherently weak upper and lower lateral cartilages, soft tissue stenosis, a narrow pyriform aperture, or facial nerve palsy affecting the dilator muscles.

A diagnosis of nasal valve dysfunction can be made by simple inspection and watching the patient breathe at rest (**Fig. 3.7a,b**). Inspection during increasing rates of inspiration can reveal various degrees of collapse of the nasal valve. There are no reliable objective measurements. Using a Jobson–Horne probe it is possible to gently support the upper lateral cartilage and ask the patient if it provides symptomatic benefit (**Fig. 3.8**). If artificially supporting the nasal valve, or opening the soft tissues of the alar region along with the nasal valve with a Jobson–Horne probe, does not provide good subjective improvement in airflow, then valve surgery is unlikely to be of any benefit. The Cottle sign, distracting the nasal valve by pulling on the soft tissues of the cheek (**Fig. 3.9**), is a nonspecific sign and often provides symptomatic improvement in most primary and secondary causes affecting the nasal valve, and it is of little diagnostic use.

It is important not to overlook these problems because surgery to the paranasal sinuses will be unhelpful.

Disorders of Smell

The patient whose sense of smell returns after oral steroids, only to deteriorate thereafter, is the patient whose sense of smell may benefit from surgery. A patient with anosmia who has had previous surgery is unlikely to regain any sense of smell if systemic steroids have not helped. However, a patient with anosmia who has not



a

b

Fig. 3.7a,b Primary valve collapse occurs when a patient breathes gently **(a)** compared with at rest **(b)**.

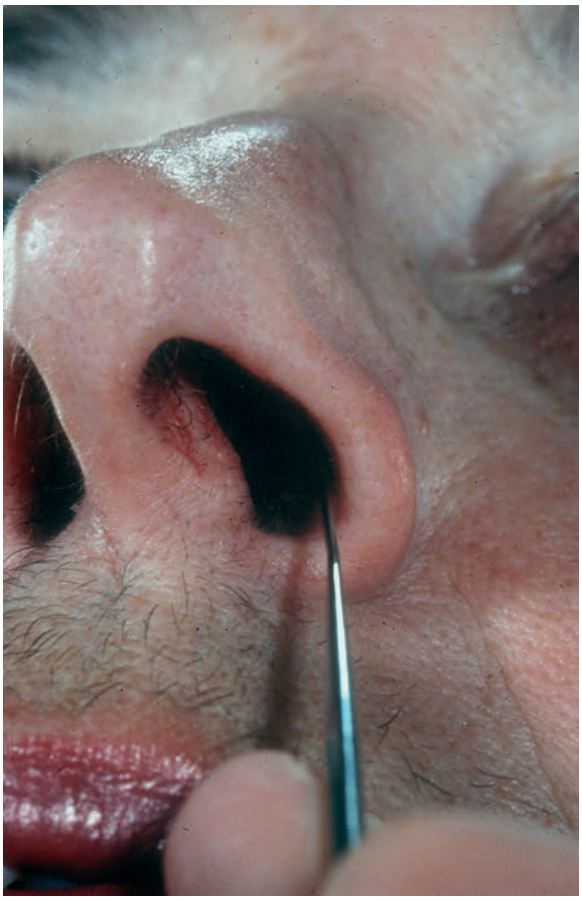


Fig. 3.8 A fine instrument gently supports the lateral aspect of the nasal valve area—if this provides good symptomatic benefit then this may be a factor in the symptom of nasal obstruction.

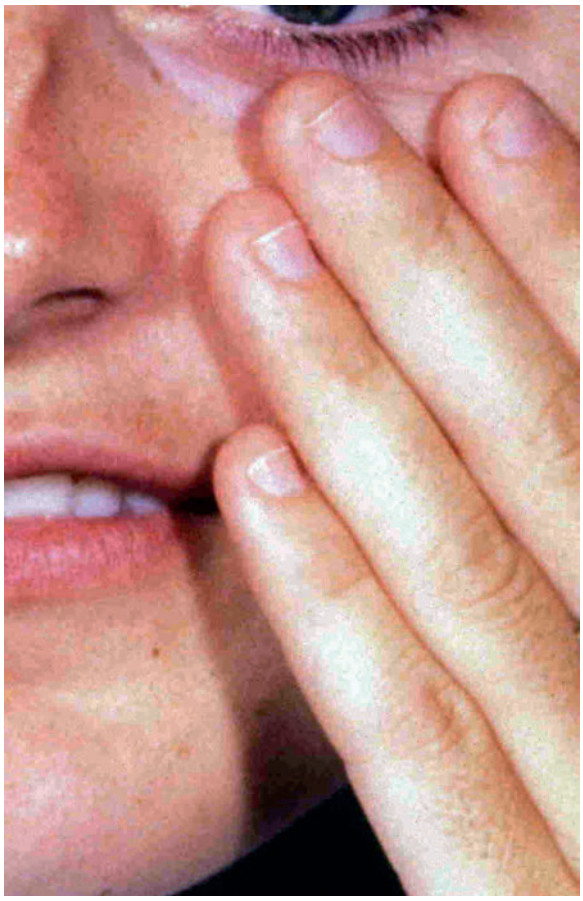


Fig. 3.9 Cottle sign is poor at defining the site of the cause of any nasal obstruction.

had previous surgery and did not respond to oral steroids still has a small chance of regaining his or her sense of smell through an ethmoidectomy and gentle lateralization of the middle turbinate. It is vital that the middle and superior turbinate are treated with absolute care in these patients when surgery is performed to open the olfactory cleft.

Hyposmia is the commonest disorder of smell and is normally caused by any inflammation of the nasal lining of any cause ranging from a cold, severe allergic rhinitis, and chronic rhinosinusitis, to sarcoid or a vasculitis (Jones and Rog 1998).

Partial anosmia (an ability to detect some, but not all, qualitative olfactory sensations) usually follows when there has been some damage to the olfactory mucosa or bulb. This may follow an influenza-like illness where the virus is neuropathic to the olfactory apparatus or following trauma with either a head injury or nasal surgery. The extreme end of this spectrum is anosmia. What we do not know is, from a cohort of people who have had a neuropathic virus or head injury causing severe damage to their olfactory mucosa, how many recover and after what length of time. In secondary care we see individuals whose sense of smell has only partially returned or failed to return and these may represent a minority who have not recovered as many may have done so in the first few weeks. It is difficult to predict the outcome in any individual and there are reports of an individual's sense of smell returning up to 7 years after these events, even though the basal cells that replace the neuroepithelium do so approximately every 40 days. Frontal blows are a common site of trauma that results in olfactory loss but occipital blows, in themselves much less common, are more likely to result in total anosmia. Amnesia for longer than 24 hours is an indicator of a poorer prognosis (Jones et al 1997b). However, although it is often premature to dismiss the possibility that some or all of their sense of smell may return, it is unwise to predict that it will improve because recovery occurs in fewer than 10%, most occurring within 6 months.

Congenital anosmia is associated with some patients with Turner syndrome and with patients with premature

baldness, vascular headaches, and other abnormalities. This presents remarkably late as the individual knows nothing different.

Patients with anosmia or severe hyposmia need to be advised about using smoke alarms. They may become fastidious about cleaning both themselves and their surroundings and overuse fragrances for fear of there being a bad smell that they cannot detect.

Parosmia or cacosmia (the presence of an unpleasant odor when a normal odor is presented), from the Greek *kakos* "bad" and *osme* "smell," can be even more disturbing than anosmia. These patients may also have had an influenza-like illness or head trauma but the neuronal pathways to the glomeruli in the olfactory bulb have become disrupted. Typically a particular substance such as coffee, perfume, or smoke will initiate another sense of smell such as a chemical-like sensation, or, even more distressing, a smell of feces. In some individuals all olfactory substances induce the same sensation. It is important to exclude other causes such as anaerobic organisms in the paranasal sinuses, diseased teeth, and occasionally organisms within the vestibular hairs.

The risk of mild hyposmia and anosmia following nasal surgery is ~ 30% and 1%, respectively (Kimmelman et al 1994; Briner et al 2003).

Nasal and paranasal surgery can affect the olfactory pathway by direct trauma to the olfactory epithelium on the middle or superior turbinate, septum or cribriform plate, or by obstructing the olfactory cleft with adhesions. The olfactory mucosa should be preserved if at all possible because to remove it will significantly affect an individual's quality of life.

Olfactory Hallucinations or Phantosmia

These terms apply to the perception of an odor in the absence of an olfactory stimulus. Olfactory hallucination can be a symptom of various non-nasal conditions associated with a head injury, epilepsy, migraine, cluster headache, schizophrenia, depression, bipolar mood disorders, eating disorders, substance abuse, iatrogenic causes, cerebral aneurysm, or tumors (**Fig. 3.10**). Migrainous olfactory hallucinations are rare and there is usually a clear temporal relationship between episodes of headache and olfactory hallucination. Epileptic olfactory auras are rare. Electroencephalogram changes during the olfactory hallucination indicate an epileptic origin of the aura. Phantosmia following head injury is uncommon. Iatrogenic olfactory hallucination is sometimes seen in epileptic patients on dopaminergic therapy in the early stage and these patients frequently also have a synchronous visual hallucination. Olfactory hallucination is relatively rare in psychotic patients. The presence of olfactory hallucination along with psychosis indicates serious psychopathology with a poor prognosis. Various modalities of treatments for idiopathic olfactory hallucinations have been reported in the literature and include surgical extirpation of the olfactory neuroepithelium and ablation of the olfactory bulb. Rhinosinusitis, viral infection of the upper respiratory tract, and head injury with cribriform plate fracture have been reported to be associated with phantosmia and sometimes with simultaneous parosmia.

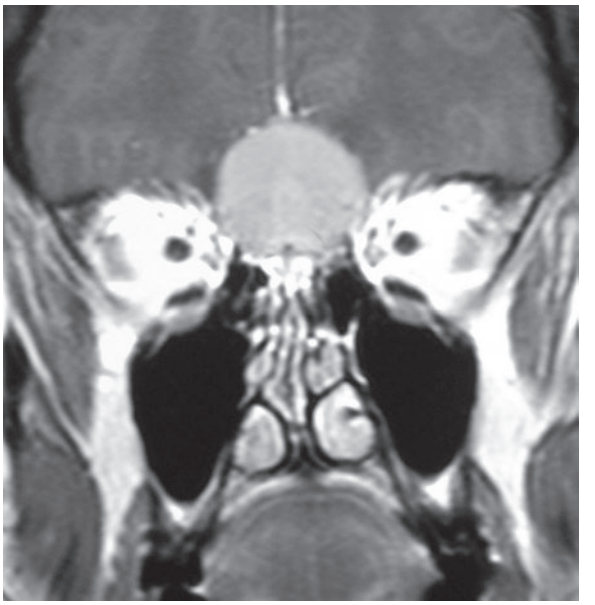


Fig. 3.10 Anterior skull base meningioma that caused a gradual deterioration of the patient's sense of smell.

Rhinorrhea

Anterior Rhinorrhea

Anterior rhinorrhea is usually secondary to viral or allergic rhinitis. The reason for anterior rhinorrhea in a viral rhinitis is not only an increase in mucus production but also paralysis of the cilia. The degree of cilial stasis that is needed to produce anterior purulent bacterial rhinorrhea is very marked in bacterial infections and normally occurs only in cystic fibrosis and ciliary dyskinesia. Unilateral clear rhinorrhea should be investigated to exclude a cerebrospinal fluid leak. A specimen of the discharge must be sent for analysis of β_2-transferrin by immunofixation (**Fig. 3.11**); this test has a high specificity and has superseded all other diagnostic techniques. Unilateral autonomic rhinitis can look very much like cerebrospinal fluid rhinorrhea, and it is essential that fluid be sent for β_2-transferrin analysis before surgery is contemplated. Other causes that can feign a cerebrospinal fluid leak are mucous retention cysts bursting or nasal secretions pooling in the maxillary or other sinus and draining when leaning forward or when the head is placed in a certain position.

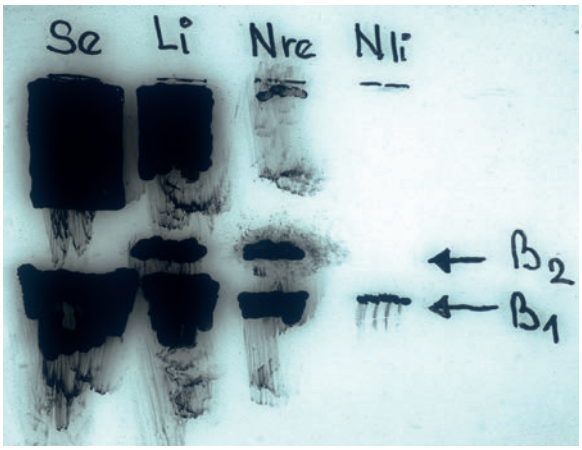


Fig. 3.11 Immunofixation of β_2-transferrin is specific and sensitive for diagnosing a cerebrospinal fluid leak.

Clear rhinorrhea caused by allergic rhinitis normally responds well to antihistamines. Topical nasal steroids can also help. When these measures are not enough then an ipratropium nasal spray taken four times a day along with an antihistamine and steroid spray will often suffice. Vidian neurectomy has been advocated but its effect lasts only 6 to 9 months even if fragments of bone are placed in the canal.

Posterior Rhinorrhea

As with facial pain, be very cautious about recommending surgery if posterior rhinorrhea is the patient's primary symptom (**Fig. 3.12a,b**). Surgery can help reduce the discoloration of the postnasal mucus by helping drainage, but it is important not to promise the patient a "cure" because the mucus secretion may be due to systemic mucosal disease. Because of this, ongoing medical treatment is often important. Patients with asthma can expect an improvement in their lower respiratory symptoms. It is also important to take time to explain to the patient the connection between the upper and lower respiratory tract.

Ask the patient about the color of the mucus. Is it clear, yellow, or green, or does it vary in color? Many who mouth breathe or snore when they sleep wake up with some discolored mucus which has collected in their nasopharynx or oropharynx and has dried in this area and become discolored with oropharyngeal commensals or smoke particles. It is therefore important to ask patients who complain of discolored mucus if it is just in the morning or whether it becomes clearer throughout the day. If they say it is always discolored it is useful to ask them to blow into a handkerchief and have a look! If they do blow out green mucus into their handkerchief, it is likely that they have a chronic infective rhinosinusitis. This is relatively unusual, but when it does occur a 2-week course of a broad-spectrum antibiotic that also

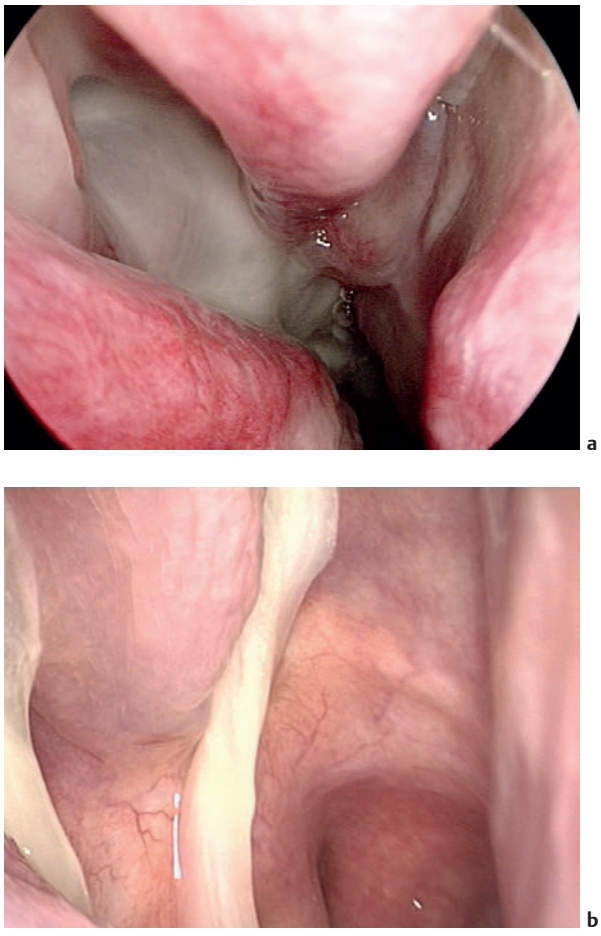


Fig. 3.12a,b Visible signs of mucus are uncommon and make it more likely that there is sinus pathology.

a Clear mucus from the accessory ostia.
b Mucus tracking back from the sphenoethmoidal recess from the middle meatus.

covers anaerobes usually clears the infection (Clifton and Jones 2007). Patients with nasal polyposis often produce a lot of yellow-stained mucus, which is due to the presence of eosinophils; this discoloration does not necessarily mean that it is infected. The mainstay of treatment for this condition is oral steroids followed by topical nasal steroids, and compliance is important.

A separate group of patients with primary ciliary dyskinesia, cystic fibrosis, or immunodeficiency will present with discolored secretions (**Fig. 3.13**) that they can blow out into a tissue, or that can be seen with an endoscope in the middle meatus or tracking down from the sphenoethmoidal recess. Surgery is disappointing in these groups because it does not address their underlying pathology.

Surgery can help to reduce the discoloration of the postnasal mucus in patients with genuine chronically infected rhinosinusitis when medical treatment has failed by aiding drainage. They may need to douche regularly if the cilia are not functioning or in abnormal mucus

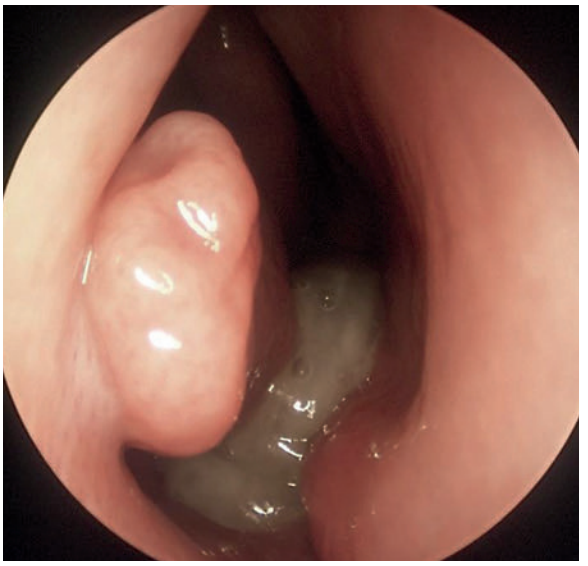


Fig. 3.13 Stagnant mucus in primary ciliary dyskinesia.

production such as in cystic fibrosis. In those with idiopathic nasal polyposis who often have associated late-onset asthma, it is important not to promise the patient a "cure" because mucus secretion is part of their systemic mucosal disease. Because of this, ongoing medical treatment is often important. Patients with asthma can expect a temporary improvement in their lower respiratory symptoms after sinus surgery but it is important to take time to explain to the patient the connection between inflammation of the upper and lower respiratory tract, and that surgery cannot cure the cause of the inflammation of their whole upper and lower respiratory tract.

Catarrh and Postnasal Drip

"Catarrh" means different things to different patients. You need to be quite specific in your history-taking to find out what they mean by it. Ask whether it is a sensation of mucus coming down the back of the throat, at the level of the soft palate, or if there "something" lower down around the level of the cricoid cartilage. Ask the patient to point with one finger to the area or level where they feel the sensation. Ask whether it is clear or mucky, and if it is mucky is that mainly in the morning, and clearer during the day? How much do they produce? Does it make them sniff, snort, clear their throat repeatedly, or hawk?

Normally, the paranasal sinuses produce a half a cupful of mucus a day, and this is swallowed along with 1.5 L of saliva. The sensation of an increase in mucus production felt in the back of the throat is sometimes called "postnasal drip." Patients often complain of a sensation of "something" in the back of the throat that they cannot clear and persistently attempt to clear their throat. Frequently, these symptoms are a result of a hyperawareness of normal mucus. It is particularly important to warn these patients that this symptom cannot be helped by surgery. Other strategies, however, may help these patients; for example, breaking a cycle of repeated throat clearing, snorting, or hawking by swallowing ice-cold sparkling water instead without doing any of the aforementioned for a week (see p. 440, "Regimen to Break the Cycle").

However, paranasal sinus disease can lead to more mucus being produced. This includes all the causes of chronic rhinosinusitis and in particular the subgroup with nasal polyposis. The presence of nasal obstruction and hyposmia complemented by endoscopic signs will differentiate these patients from those who have a hyperawareness of mucus. It is important that these problems are distinguished from habitual snorting, dry swallowing (swallowing without drinking or eating), or clearing of the throat: this is often part of a habitual cycle which accompanies a hyperawareness of normal mucus. The snorting and throat clearing appear to maintain, if not exacerbate, the sensation that mucus is present. Often the snorer whose uvula is edematous complains of a sensation of "something" around the soft palate, and they may use the term "catarrh" to describe this (**Fig. 3.14**). To further complicate matters, some patients with globus pharyngeus may complain of catarrh because they have a sensation of something (or mucus) at the level of the cricoid cartilage.

It is far more common for patients to have a hyperawareness of normal postnasal mucus, and through repeated clearing of their throat or snorting to have "sensitized" these areas to the half a cup of mucus that is normally produced from the paranasal sinuses each day, as well as the liter of saliva that is swallowed. In this context it is worth considering that, of the large number of people with allergic rhinitis who have a definite increase in their mucus production, few complain of catarrh (**Fig. 3.15**).

A strategy to break the cycle of repetitive throat clearing, dry swallowing, and hawking is to advise the patient to strictly avoid all of these for 1 week and instead swallow ice-cold water that will stimulate the back of the throat and take away the desire to clear the throat. They must be disciplined about doing this and rotate bottles of ice-cold water from the fridge regularly and have some by their bedside. An explanation that the secretions that they have become aware of are healthy and can be swallowed without causing any problems also helps. An audit of this strategy has shown that it works well in a large proportion of patients with these symptoms (Acharya et al 2007).

Postnasal Drip Syndrome

Some patients complain of "something" dripping down from the nasopharynx, a need to clear the throat, a tickle in the throat, and posterior nasal discharge, therefore overlapping the symptoms with catarrh. Postnasal drip syndrome has been described as one of the "pathogenic

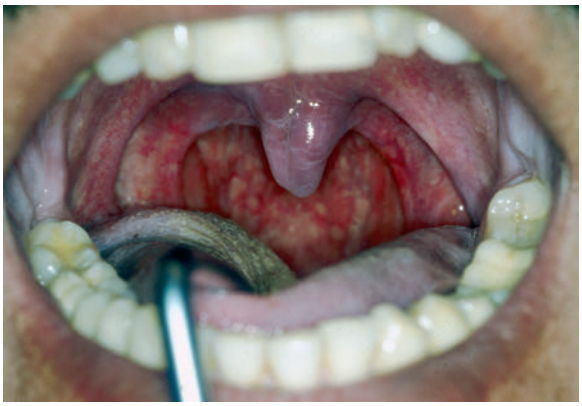


Fig. 3.14 Edematous uvula in a snorer.

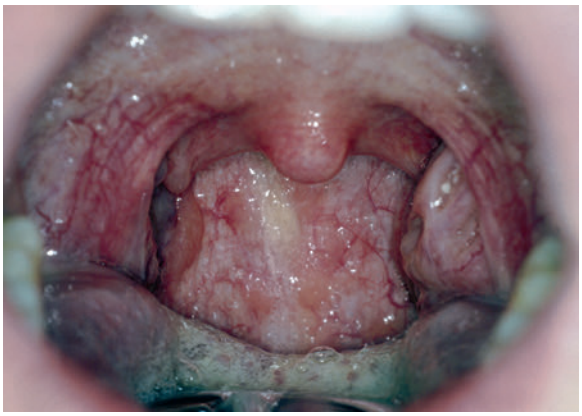


Fig. 3.15 A snorer who mouth breathes and has dried, discolored mucus in the morning and pharyngitis. During the day the mucus was clear, as were the sinuses.

triad of the causes of chronic cough" together with asthma and gastroesophageal reflux. It has been suggested that the symptoms are the result of mechanical stimulation in the upper airway and specifically due to secretions dripping into the hypopharynx. However, it has no clear definition and there is no physiologic reason why secretions should "drip" in this way—nasal mucus is normally tenacious and cannot drip like water from a tap. Most authorities in this field believe that nasal disease has a role in the production of chronic cough through a continuum or "global" airway inflammation affecting the upper and lower airway and *not* through any "drip."

Symptoms of heartburn, dysphagia, dysphonia, globus, acid regurgitation, and a bitter taste in the mouth suggest a diagnosis of gastroesophageal reflux disease but this condition has been shown to be asymptomatic or "silent" in many patients. It has also been implicated in chronic cough, dysphonia, globus, throat clearing, dysphagia, and excessive throat mucus. The two potential pathophysiologic mechanisms are aspiration of gastric contents irritating the larynx or tracheobronchial tree, and an esophagobronchial reflex via the vagus nerve. This condition is also worth considering in patients who throat clear. A trial of a proton pump inhibitor may be worth considering.

Chronic Cough

Respiratory tract infections are the commonest cause of an acute cough that, by definition, should have resolved within 2 months. Patients with a cough lasting longer than this period are defined as having a chronic cough. Studies of the etiology of chronic cough suggest that ~ 95% of symptoms in immunocompetent, nonsmoking patients with a normal or near-normal chest radiograph are caused by asthma, gastroesophageal reflux disease, chronic bronchitis, bronchiectasis, eosinophilic bronchitis, or the use of an angiotensin-converting enzyme inhibitor. Less common causes such as bronchogenic carcinoma, left ventricular failure, sarcoidosis, and tuberculosis may explain the remaining 5%.

Physiologically, cough is a defense mechanism protecting the tracheobronchial tree. Afferent receptors are believed to be innervated through the vagus nerve via the pharyngeal, superior laryngeal, and pulmonary branches. Innovation of the tracheobronchial region has a great role in the generation of cough as in the larynx, as evidenced by the poor or absent cough of lung–heart transplant recipients with absent pulmonary vagal innervation. Stimulation of the vagus can also initiate cough whether it is by stimulating the external auditory canal or by instilling acid into the lower third of the esophagus.

As mentioned in the last section, there is no good evidence to support postnasal drip as a cause of chronic cough. In the literature there is no diagnostic test to define those who are labeled as having postnasal drip syndrome other than a response to a first-generation antihistamine. Nasal disease is more likely to result in cough through the co-existing involvement of the lower airways through an as yet undefined pathway, and eosinophil and mast cell mediation appear a likely mechanism (**Fig. 3.16**). Studies have shown that 60 to 78% of people with asthma also suffer with rhinosinusitis. How gastroesophageal reflux disease may trigger cough is not entirely clear. The two potential pathophysiologic mechanisms are aspiration of gastric contents irritating the larynx or tracheobronchial tree, and an esophagobronchial reflex via the vagus nerve. A trial of a proton pump inhibitor for 2 months can be both diagnostic and therapeutic.

Sneezing

Most people sneeze, but more than four sneezes a day are almost pathognomonic of allergic rhinitis. If a patient sneezes a great deal in the morning, then you should be suspicious that they have an allergy to house dust mite, having been exposed to it over the previous few hours

Fig. 3.16 Diagrammatic representation of the concept of the whole respiratory tract being a single airway.

in their bedding (**Fig. 3.17a,b**). Itchy and watery eyes are in keeping with an allergic rhinitis, whether this is persistent or intermittent. The best treatment for this is nonsedative antihistamines that normally work well and a topical nasal steroid provides supplementary help in many, although a minority find that a nasal spray can initiate a sneezing bout. Sprays that contain benzalkonium chloride as a preservative tend to initiate sneezing bouts more than those that do not.

A small proportion of patients have incapacitating sneezing bouts that can last for hours. Most of these patients are young adolescents or children and psychogenic factors play a major role. Cognitive behavior therapy, treating anxiety, or suggestion therapy can help.

Surgery has no role in the management of sneezing.

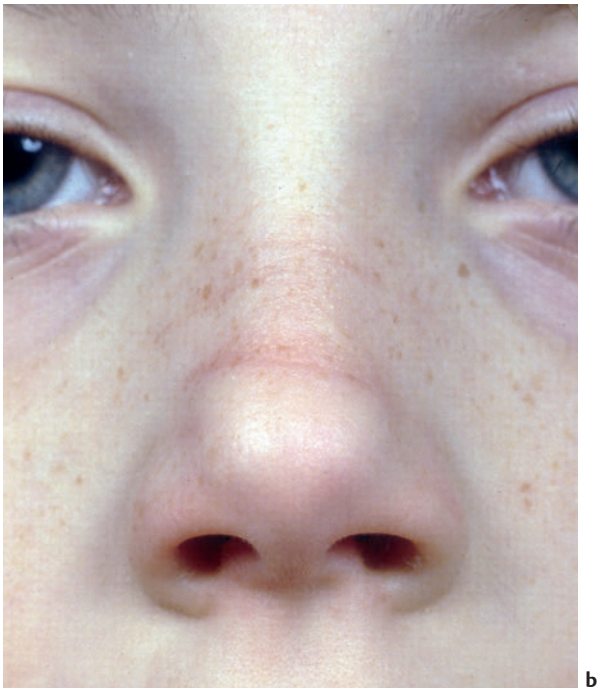
Crusting

This symptom should alert you to the possibiility of the patient having a systemic disease such as Wegener granulomatosis or sarcoidosis (Fergie et al 1999), and the relevant investigations should be undertaken to exclude these (Jennings et al 1998).

Facial Pain and Pressure

Facial pain and pressure are often wrongly attributed by patients and their primary care physicians as being due to rhinosinusitis (West and Jones 2001).

a



b

Fig. 3.17a,b An external clinical sign in allergic rhinitis.

a A nasal salute in allergic rhinitis.

b The crease over the bridge of the nose created by repeated rubbing.

In patients with chronic pain involving the face and/or head, it is important to see whether their symptoms are associated with, or exacerbated by, an upper respiratory tract infection and, furthermore, to see whether there is a temporal relationship with any purulent discharge. If they have no significant nasal symptoms and if their nasal endoscopy is normal, it is unlikely that their facial pain is due to rhinosinusitis. However, patients who have facial pain and purulent secretions at endoscopy do well with surgical or medical treatment—over 80% will be helped.

Beware if pain and pressure are the patient's main symptoms. The majority of patients with nasal polyposis have no facial pain or pressure from rhinosinusitis unless there are purulent secretions present and their is an acute episode with obstruction of the sinus ostia (Fahy and Jones 2001).

However, if patients have symptoms of pain or pressure in addition to nasal obstruction and a loss of sense of smell, especially if the pain and pressure get worse with a cold or when flying or skiing, then you can advise the patient that these symptoms may be helped by surgery.

It is important to be aware of other causes of facial pain that are much more common. Even in an ENT clinic only ~ 16% of those who have been referred with a provisional diagnosis of facial pain due to rhinosinusitis turn out to have pain that is related to paranasal sinus disease (West and Jones 2001).

Useful Generalizations

- The vast majority of patients who present with a symmetrical frontal or temporal headache, sometimes with an occipital component, have tension-type headache.
- Unilateral, episodic headaches are often vascular in origin.
- Symmetrical symptoms of facial pressure, particularly if it is long standing, is usually due to midfacial segment pain, a version of tension-type headache that affects the midface.

Sinogenic Pain

Sinusitis rarely causes headache, let alone facial pain, except when there is an acute bacterial infection when the sinus in question cannot drain. These patients usually have a history of a viral upper respiratory infection immediately before this, and they have pyrexia with unilateral nasal obstruction. The vast majority of patients with acute sinusitis respond to antibiotics. Patients with more than two episodes of genuine bacterial sinusitis in 1 year should be investigated for evidence of poor immunity. Patients with chronic bacterial sinusitis rarely have any pain unless the sinus ostia become blocked in an acute exacerbation and then their symptoms are the same as acute sinusitis (Clifton and Jones 2007). There are isolated reports of sphenoidal sinusitis that can cause headaches but these are extremely rare, there are usually systemic signs such as pyrexia, endoscopic signs, or there is a raised C-reactive protein, and most of these patients respond to antibiotics.

Patients with a headache often make a self-diagnosis of "sinusitis" because they know that some sinuses lie within the head. With the advent of nasal endoscopy and computed tomography (CT), along with the finding that many patients' symptoms of headache or facial pain persist after sinus surgery, it has become apparent that this is not the case. Also of note is that over 80% of patients with purulent secretions visible at nasal endoscopy have no headache or facial pain (Clifton and Jones 2007) (**Fig. 3.18**). Even if patients with intermittent symptoms of headache or facial pain, and who believe that it is due to infection, are asked to attend the clinic when they are symptomatic, the majority are found not to have any evidence of infection and another neurologic cause for their pain is often responsible (Stewart and Michael 2002; Tepper 2004).

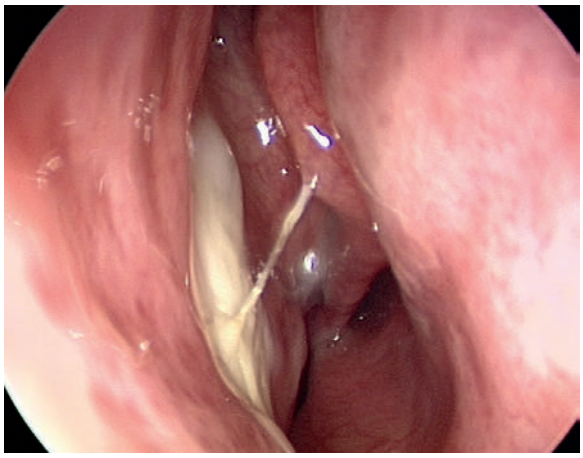


Fig. 3.18 Purulent secretions in the right middle meatus—the patient had no pain as is often the case.

Over 90% of self- and doctor-diagnosed sinus headaches meet the International Headache Society criteria for migraines. Many migraine sufferers had at least one unilateral nasal symptom of congestion, rhinorrhea, or ocular lacrimation, and redness or swelling during an attack that caused confusion and led them to think their sinuses must be the cause (**Fig. 3.19**).

One series showed that nearly 90% of participants with self- or physician-diagnosed sinus headache met the International Headache Society criteria for migraine-type headache and responded to triptans (Cady and Schreiber 2004). They note that during a migrainous episode there is engorgement and erythema of the nasal mucosa along with rhinorrhea, and after subcutaneous sumatriptan both the symptoms and endoscopic signs resolve. An

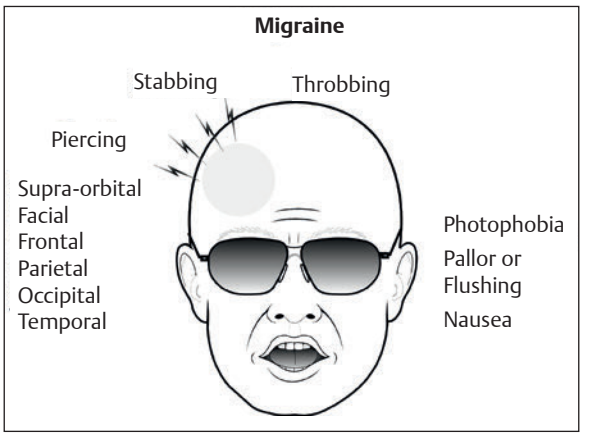


Fig. 3.19 Symptoms that can occur in migraine.

interdisciplinary consensus group recently agreed that "the majority of sinus headaches can actually be classified as migraines" and that "unnecessary diagnostic studies, surgical interventions, and medical treatments are often the result of the inappropriate diagnosis of sinus headache" (Levine et al 2006).

Chronic sinusitis is usually painless, with episodes of pain occurring during acute exacerbations which are often precipitated by an upper respiratory tract infection or when there is obstruction of the sinus ostia. An increase in the severity of pain on bending forward is traditionally thought to be diagnostic of sinusitis, but this is nonspecific as many types of facial pain and headache are made worse by this.

The key points in the history of sinogenic pain are an exacerbation of pain during an upper respiratory tract infection, an association with rhinologic symptoms, and a response to medical treatment.

Midfacial Segment Pain

A relatively recently described condition, which affects about a third of patients with facial pain seen in ENT clinics, is midfacial segment pain. This is a version of tension-type headache that affects the midface, although 60% have a coexisting headache, and has been shown not to be related to sinusitis (Jones 2007).

The definition of midfacial segment pain is:

- A symmetrical sensation of pressure, tightness, or "blockage" (but without airway obstruction).
- Can affect the nasion, under the bridge of the nose, either side of the nose, the periorbital or retro-orbital regions, or across the cheeks (**Fig. 3.20a–f**). Approximately 60% also have the symptoms of tension-type headache.
- There may be hyperesthesia of the skin and soft tissues over the affected area. Lightly touching the skin can cause discomfort.
- Nasal endoscopy is normal.
- CT of the paranasal sinuses is normal (note: a third of asymptomatic patients have incidental mucosal changes on CT).
- The symptoms may be intermittent (< 15 days/month) or chronic (> 15 days/month).
- There are no consistent exacerbating or relieving factors, although a short placebo effect may be reported after some medication. This is not consistent and quickly wears off.
- There are no nasal symptoms (but note that ~ 20% of most populations have intermittent or persistent allergic rhinitis, which may occur incidentally in this condition) (▶ Video 3).

Patients with midfacial segment pain describe a symmetrical feeling of pressure, heaviness, or tightness and they may say that their nose feels blocked when they have no airway obstruction. There are no consistent exacerbating or relieving factors and patients often take a range of analgesics but they have no, or minimal, effect. Patients may be convinced that their symptoms are due to sinusitis as they know that their sinuses lie under this area with the exception of the bridge of the nose; indeed, their primary care physician may have treated them as having sinusitis for many years.

Patients often describe tenderness on touching the tissue of the forehead or cheeks, leading them to think there is underlying inflammation of the bone. However, on examination there is hyperesthesia of the skin and soft tissues in these areas; gently touching these is enough to cause discomfort and there is no evidence of underlying bony disease. This is similar to the tender areas over the forehead and scalp seen with tension-type headache. They may say that the skin of the infraorbital margin region or cheeks swells up, but there are no objective signs—this symptom appears to be due to an alteration in sensation in this area.

Nasal endoscopy is normal. As around one in three asymptomatic people have incidental changes on their CT images, this may confuse the picture. The majority of patients with this condition respond to low-dose amitriptyline, but usually require up to 6 weeks of 10 mg (occasionally 20 mg) at night before it works (Agius et al 2013). Amitriptyline should then be continued for 6 months before being stopped, and the 20% of patients whose symptoms return when amitriptyline is stopped need to restart it if the pain returns. It is our practice to inform patients that amitriptyline is also used in higher doses for other conditions such as depression, but its effectiveness in midfacial segment pain is unrelated to its analgesic properties, which would take effect much more quickly and normally require 75 mg. It is often reassuring for patients to know the dose used for depression is some seven or more times the dose used in tension-type headache or midfacial segment pain and that no other antidepressant works for this condition. If amitriptyline fails, then relief may be obtained from gabapentin or pregabalin.

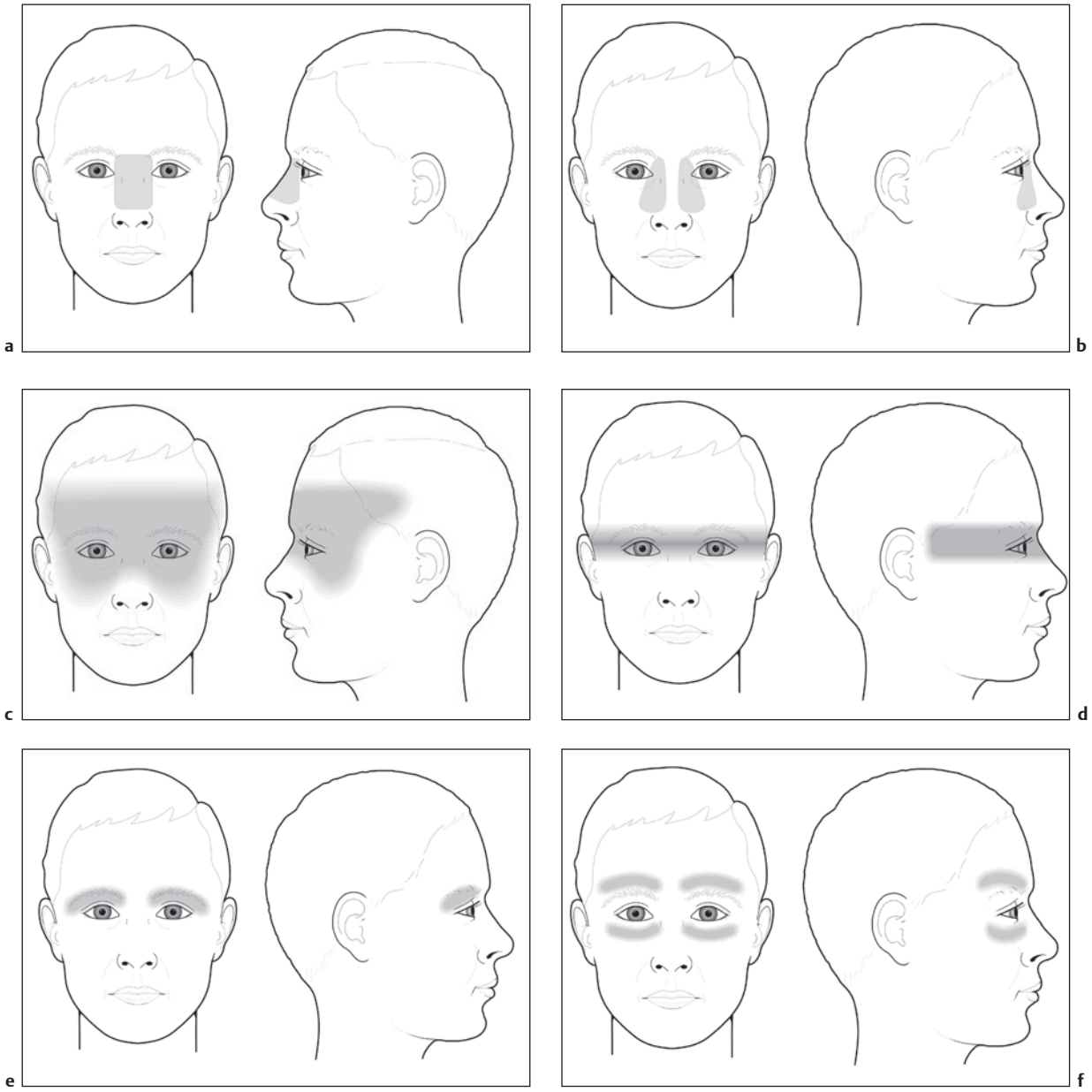


Fig. 3.20a–f The different patterns of pain distribution in midfacial segment pain.

If patients with midfacial segment pain undergo septal or sinus surgery it makes no difference in approximately a third, in a third it makes their symptoms worse, and in the remaining third it helps their pain but only for a few weeks and rarely more than a few months. It is as though the surgical stimulus alters the "balance" of neuronal activity in the trigeminal caudal nucleus for a short time. It is possible that the placebo effect or cognitive dissonance may also be responsible for a temporary symptomatic improvement (Homer et al 2000). The term midfacial segment pain avoids the use of the term "tension," which often results in a long and relatively unproductive discussion with the patient about the role of stress in their condition.

Facial Pain Due to Vascular Causes

Unilateral, Episodic Headaches, or Facial Pain

These are often vascular in origin. Vascular pain of various types can be associated with autonomic rhinologic symptoms such as nasal congestion and rhinorrhea and this has led to confusion in arriving at a correct diagnosis (Erros et al 2007). Vascular causes of facial pain include

atypical forms of migraine cluster headache, paroxysmal hemicrania, chronic paroxysmal hemicrania, hemicrania continua, and short-lasting neuralgiform pain with conjunctival injection and tearing.

Migraine

Migraine is typically episodic, lasting 4 to 72 hours and throbbing in nature. Although classical migraine often has a prodromal state and is usually preceded by an aura that frequently contains visual phenomena, facial migraine often has none of this. The pain is typically unilateral but may be bilateral. Nausea, vomiting, and photophobia often accompany the pain. The pain can affect the face as well as the head and a minority can have pain confined to the periorbital area, rarely affecting the cheek and nose alone (**Fig. 3.19**).

The treatment options for acute migraine attacks include the triptans (e.g., sumatriptan, naratriptan, rizatriptan, zolmatriptan), ergotamine or dihydroergotamine, aspirin, paracetamol, codeine phosphate, ibuprofen, or naproxen, with or without metoclopramide. Preventive therapy includes pizotifen (weight gain is a common side effect and reduces its acceptability), propranolol, and amitriptyline. Acute antimigraine therapy is most likely to be beneficial if it is started early in an attack (▶ Video 3).

Cluster Headache

Cluster headache is both severe and uncommon. It is characterized by recurrent, strictly unilateral attacks of headache that typically wake the patient and are retroorbital or centered at the medial aspect of the orbit, of great intensity, and last up to 1 hour. There is often ipsilateral rhinorrhea, nasal obstruction, and lacrimation (**Fig. 3.21**). It is called cluster headache because there are active or inactive bouts separated by clinical remission when the patient is completely pain free. Treatment includes sumatriptan and oxygen (▶ Video 3).

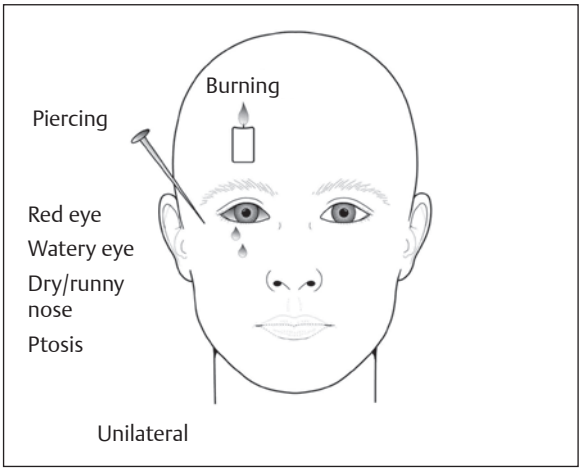


Fig. 3.21 Symptoms of cluster headache.

Paroxysmal Hemicrania

This has been described as an excruciating unilateral pain that is usually ocular and frontotemporal, with short-lasting (2–45 minutes), frequent attacks (usually more than five a day). At least one of the following autonomic symptoms should be present: nasal congestion, rhinorrhea, lacrimation, conjuctival injection, or, rarely, ptosis, eyelid edema, increased local sweating, and facial flushing. Approximately one in four patients develop a chronic form. The average age at onset is usually 30 to 40 years, but the age spectrum is wide. The condition's complete or rapid response to indometacin is said to differentiate paroxysmal hemicrania from cluster headache.

Hemicrania Continua

Hemicrania continua is a unilateral headache that is moderately severe without side shift, continuous but with fluctuations, with complete resolution of pain with indomethacin, and exacerbations that may be associated with autonomic features such as conjunctival injection, lacrimation, and photophobia to the affected side.

SUNCT

Short-lasting neuralgiform pain with conjunctival injection and tearing (SUNCT) is one of the rarest idiopathic headache syndromes. There is trigeminal pain, particularly in the orbital or periorbital areas, associated with autonomic symptoms in which conjunctival injection and tearing are the most prominent features. Attacks last between 15 and 60 seconds and recur between 5 and 30 times an hour. These attacks may be precipitated by chewing movements and ingesting of certain foods such as citrus fruits. Treatment is not always straightforward and a trial of lamotrigine, carbamazepine, or topiramate may help.

Other Symptoms That Are Difficult to Interpret

Facial Swelling without Any Objective Signs, or Tenderness of the Soft Tissues of the Face

It is not uncommon for patients to say that their cheeks swell when they have facial discomfort when there are no objective signs, even when they are symptomatic. They often turn to their partner to confirm this, yet they have no objective signs. Some patients with facial pain of vascular origin release neuropeptides in the distribution of their pain. This may be responsible for some sensation of facial swelling, and they may even have a facial flush. Another reason some patients complain of facial swelling without any signs, particularly when it is bilateral, is that they have an altered sensation in the affected area akin to the feeling of facial swelling that occurs after a dental