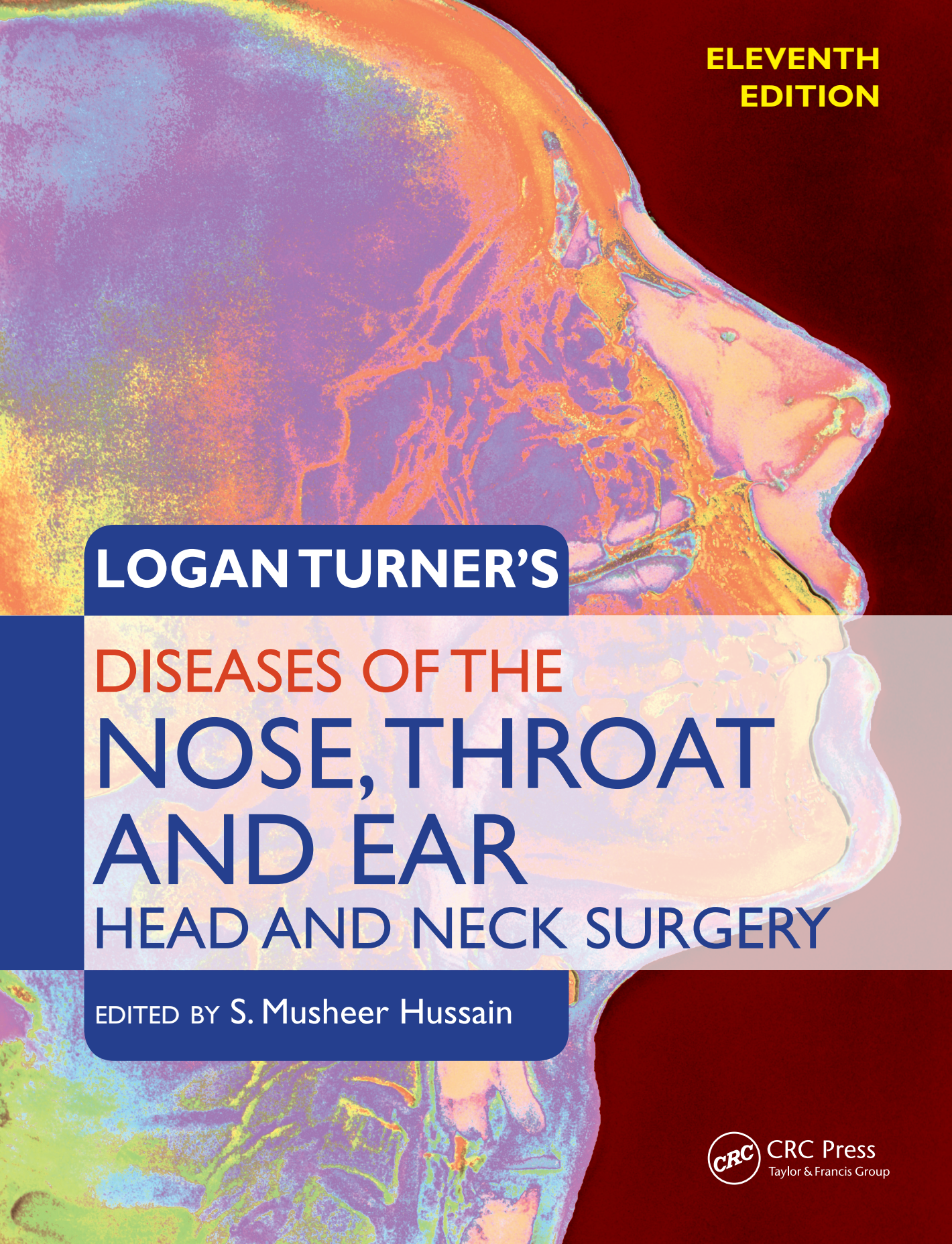
ELEVENTH EDITION
LOGAN TURNER'S
DISEASES OF THE
NOSE, THROAT AND EAR
HEAD AND NECK SURGERY
EDITED BY S. Musheer Hussain
CRC Press
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Edited by

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Foreword

Two years before the First World War began in 1914, Dr Porter, an Edinburgh otolaryngologist, wrote a book that became a 'best-seller'. We do not know how his career would have progressed, nor how his book would have developed, because he, like so many other young professional men, was killed before the end of the war.

Fortunately his Edinburgh colleagues revived the book the year after the war ended in 1919 under the editorship of Dr Arthur Logan Turner, who became President of the Royal College of Surgeons of Edinburgh. Between then and 1982 there were nine editions; the contributing authors traditionally were the consultants in post at the time in the Edinburgh department of otolaryngology. The tenth edition in 1988 was edited by Professor Arnold Maran who, like Dr Logan Turner, became a President of the Royal College of Surgeons of Edinburgh, and the edition he edited was also written by Edinburgh laryngologists.

Many plans for further editions were discussed, but they were in the years during which written works were being replaced by electronic publishing, and there was doubt about the value of an 'old-fashioned' text book. During this period of uncertainty there were many 'false starts', and for a time it looked as if this historic book would disappear. However, we are grateful to Musheer Hussain, who has had the perseverance to create a team of contributors to complete this eleventh edition.

So much time has passed and so many changes have occurred in the delivery of health care in Scotland that the authors of the chapters are now no longer confined to Edinburgh. That in itself is a marker of the development of the delivery of health care in Scotland, where now every centre has a standard of excellence that was once confined to the big cities.

This new edition reflects the huge change that has occurred in the specialty since the last century. Although otolaryngology was originally a specialty that was created in order to remove pus from bony boxes in the skull to avoid intracranial complications, it has morphed into a form that would be unrecognisable to the original innovators. These changes are elegantly presented in this book, which now covers neuro-sensory deafness, head and neck cancer, paediatric airway disease, skull base surgery and the rhinological revolution brought about by the endoscope.

Should there still be a place for the printed word? My answer would be in the affirmative, because electronic publishing has not reached the point where instant access to a single topic is as easy as it is with the printed word.

Is there still a place for a single work on a whole specialty? My answer would again be in the affirmative, because 95% of that specialty is between these hard covers. The more specialised areas are the domain of single-subject volumes, but in this book, the jobbing otolaryngologist will find most of the answers to everyday problems.

Professor Arnold A G Maran

Preface

When I was asked to edit the eleventh edition of *Logan Turner's Diseases of the Nose, Throat and Ear: Head and Neck Surgery,* I felt deeply honoured but did not quite grasp the magnitude of the task. It is a great privilege to edit this famous textbook in its centenary year. Not many medical books have been in continuous publication for 100 years.

The response I received from the chapter authors, all leaders in their field who un-hesitantly completed their chapters, was overwhelming. Many mentioned their feelings about the textbook. 'I still keep my copy of the ninth edition', said one. 'It was the book that got me through the exams', said another. 'It was my introduction to otolaryngology' was another comment. The last three editions of this book were popular not only in the British Isles, but also in South Asia, the Far East and the Middle East. The fifth edition was sold in North America and was well received.[1]

This edition has been completely revised to meet the needs of aspiring otolaryngologists. All four sub-specialities are represented in the chapters, along with a fifth section on radiology.

I must acknowledge the previous editors of the book: Dr Proctor, who first published the book in 1914 but died in the service of his country; and Dr Logan Turner, who edited Dr Proctor's book and contributed additional material.

Several editions followed. The ninth and tenth editions were prepared by Professor Arnold Maran, an acknowledged leader in field of otolaryngology who has very kindly written the Foreword to the book.

I would like to thank my former colleague, Robin Blair, and my colleague Brian Bingham for their trust.

If there are errors in the text, the responsibility is entirely mine. The publishers Taylor and Francis, and in particular Henry Spilberg and Linda Van Pelt, were unfailingly helpful, and this book would not have been completed without their support.

S Musheer Hussain
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[1] Hussain SM: Three textbooks and the 'Edinburgh Brand'. ENT and Audiology News 2010; 19 (3): 52-53.

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SECTION I

The Nose

1

Anatomy and physiology of the nose and paranasal sinuses

TAWAKIR KAMANI AND ANSHUL SAMA

EMBRYOLOGY OF THE NOSE AND PARANASAL SINUSES

The nose and paranasal sinuses are interlinked during development. At the end of the gestational fourth week, branchial arches, branchial pouches and primitive gut make their appearance. This is when the embryo gets its first identifiable head and face with an orifice in its middle known as the stomodeum (primitive mouth). The stomodeum is surrounded bilaterally by mandibular and maxillary prominences, which are derivatives of the first arch. The stomodeum is limited superiorly by the presence of the frontonasal eminence and inferiorly by the mandibular arch. Inferiorly, the frontonasal process gives two projections, the nasal placodes. These fuse to form the nasal cavity and primitive choana. The primitive choana forms the point of development of the posterior pharyngeal wall and the various paranasal sinuses.[1]

DEVELOPMENT OF THE NOSE AND NASAL CAVITY

Development of the nasal cavity

The primitive nasal cavity forms as the maxillary process of the first branchial arch and grows anteriorly and medially, fusing with the medial nasal folds and the frontonasal processes. The bucconasal membrane initially separates the primitive nasal cavity from the mouth, but it eventually breaks down, forming the primitive choanae. The palatal processes derived from the lateral maxillary mesoderm grow medially, fusing in the midline with each other and the septum to separate the nasal and oral cavities anteriorly. Posteriorly, this midline floor separates the nasopharynx and oral cavities and forms the soft palate.[1]

Development of the external nose and nasal septum

The lateral nasal folds form the nasal bones and the upper and lower lateral cartilages during the tenth to eleventh weeks. The nasal septum arises from a dorsal extending midline ridge from the posterior end of the frontonasal processes. This is continuous with the partition of the primitive nasal cavities anteriorly. The superior and posterior part of the primitive nasal septum ossifies to form the perpendicular plate and vomer, respectively. The anterior and inferior portions remain cartilaginous to form the quadrilateral cartilaginous septum.

DEVELOPMENT OF THE PARANASAL SINUSES

Development of the maxillary sinus

The maxillary sinus is the first to develop, appearing as a shallow groove in the primitive ethmoidal infundibulum into the maxilla at 7–10 weeks. At birth it measures 7 × 4 × 4 mm. It shows a biphasic growth at 3 and 12 years of age.

Development of the ethmoid sinus

The fetus develops six to seven folds in the lateral nasal wall at the ninth and tenth weeks. These folds fuse, forming crests termed *ethmoturbinates* from which the permanent ethmoidal structures develop. The first crest gives rise to the agger nasi and the uncinate process. The ethmoidal bulla arises from the second crest. The third crest is the basal lamella of the middle turbinate that divides the ethmoidal air cells in the anterior and posterior groups. The other structures that arise from these crests include the middle, superior and supreme turbinates, and all are seen to be attached to the lateral nasal wall by their basal lamella.

Development of the sphenoid sinus

The sphenoid sinus appears at the third intrauterine month as an evagination from the sphenoethmoidal recess and is about 2 × 2 × 1.5 mm at birth, to reach adult size at the age of 12–18 years.

Development of the frontal sinus

The frontal sinus starts developing at the fourth intrauterine month. The frontal sinus, the anterior ethmoidal complex and the complex array of the frontoethmoidal cells develop from five or so pits that lie between the first and second ethmoturbinates. The frontal bone is very poorly pneumatized at birth and the frontal sinuses not distinguishable from the anterior ethmoid complex. The fetal pits start to pneumatize the frontal bone and can be noted at the end of the first year of life. One of these fetal pits continues to pneumatize both plates of the frontal sinus such that by the twelfth year of life the frontal sinuses have largely developed.

ANATOMY OF THE NOSE AND PARANASAL SINUSES

THE EXTERNAL NOSE

The external nose is a pyramidal structure with a framework of bone and cartilage, covered by connective tissue and skin. The bony structure is made of the nasal bones, which unite superiorly with the frontal bone at the nasion and laterally with the frontal processes of the maxilla. The distal two-thirds of the nose is formed by the upper and lower lateral cartilages, which overlap each other at the margins. The upper lateral cartilages fuse medially with the quadrilateral cartilage, forming the cartilaginous part of the nasal dorsum. The lower lateral cartilages, also called the *alar cartilages*, are each composed of the medial and lateral crus connected by the intermediate crus. The latter forms the dome of the nostril and the tip-defining points. The medial crus contributes to the columella attached posteriorly with the membranous septum. The lateral crus forms the alar or nostril rim.

Branches of the facial artery supply the alar region, and branches of the ophthalmic and maxillary artery supply the dorsum and lateral walls. The venous drainage is to two units: the angular

vein and the ophthalmic veins. The latter interlinks with the anterior ethmoid system and thence into the cavernous sinus. The submandibular and submental nodes provide the main lymphatic drainage to the external nose.[2]

THE NASAL CAVITY AND PARANASAL SINUSES

The nasal cavity extends from the nostrils anteriorly to the choanae posteriorly, where it becomes continuous with the nasopharynx. The nasal cavity is divided in the midline by the nasal septum, which forms the medial wall of the nasal passages. The roof is formed anteriorly by the undersurface of the upper lateral cartilages and nasal bones, and posteriorly by the cribriform plate, which houses the olfactory epithelium. The rest of the nasal cavity is lined by respiratory epithelium. The nasal floor is made up of the palatine process of the maxillary bone anteriorly, fused with the horizontal process of the palatal bones posteriorly.

Nasal septum

The nasal septum is composed of a small anterior membranous portion, the cartilaginous portion called *quadrilateral cartilage* and the bony portions of the perpendicular plate of the ethmoid, the vomer and the nasal crests of the maxilla and palatine bones. These components articulate as shown in Figure 1.1. The medial crura of the lower lateral cartilages attach to the thin membranous septum anteriorly, forming one of the major tip support structures. At its upper margin, the quadrilateral cartilage is connected to the upper lateral cartilages, contributing to the projection and height of the mid-third of the nose. The keystone area represents the attachment of the quadrangular cartilage to the bony septum and nasal bones at the rhinion. Continuity and fixation at this point are important both aesthetically and functionally because it supports the projection of the upper third to mid-third of the nose.

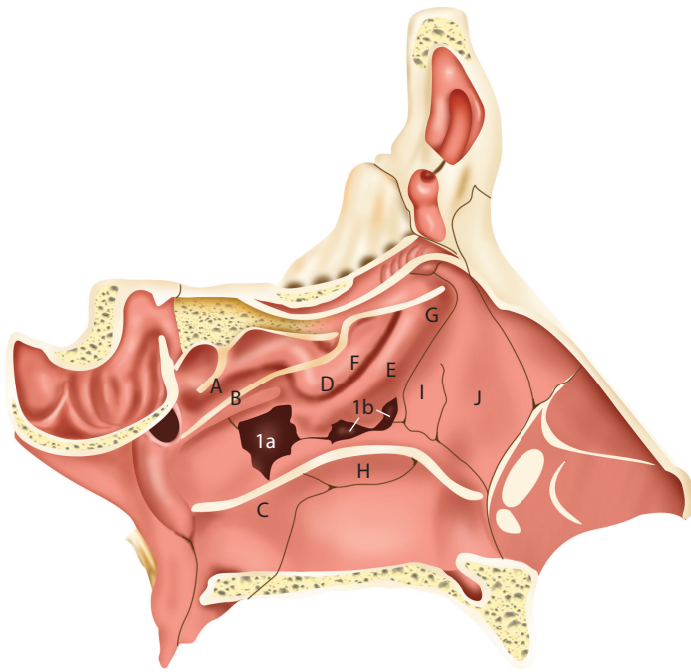

Figure 1.1 Left lateral nasal wall on removal of the turbinates. A, sphenoethmoidal recess; B, basal lamella; C, palatine bone; D, ethmoid bulla; E, uncinate process; F, hiatus semilunaris; G, aggar cell; H, maxillary process of inferior turbinate; I , lacrimal bone; J, frontal process of maxillary bone; 1a, posterior fontanel; 1b, anterior fontanels.

Lateral nasal wall

The lateral wall of the nasal cavity is predominantly composed of the maxilla, with contribution from the perpendicular plate of the palatine bone and the medial pterygoid plate posteriorly. The anterior aspect of the lateral wall has contributions from the nasal bones and upper lateral cartilage, the latter forming the internal nasal valve at its junction with the nasal septum. There are three prominences on the lateral nasal wall termed *inferior, middle,* and *superior turbinates*, respectively. Occasionally, there may be a fourth turbinate called the *supreme turbinate*. The spaces lateral to the turbinates are called *meati*. Figure 1.2 illustrates this anatomy.

The inferior turbinate develops as a separate bone whilst the middle, superior and supreme turbinates are medial projections of the ethmoid complex. Lateral to the inferior turbinate is a recess called the *inferior meatus* into which the nasolacrimal duct opens.[3]

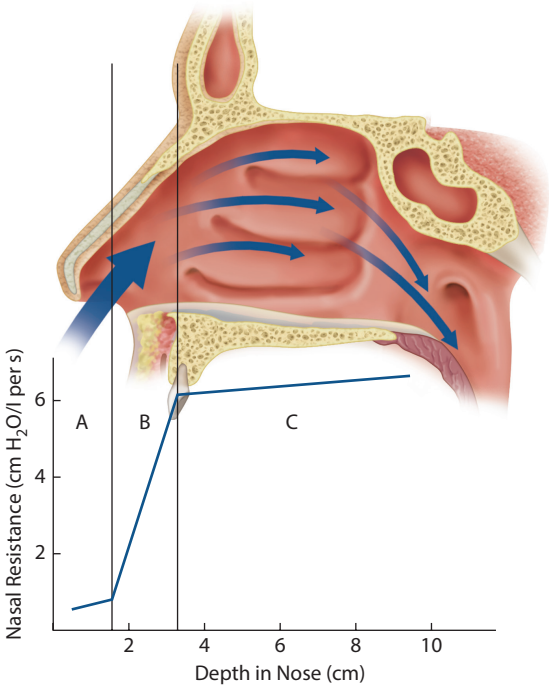

Figure 1.2 Nasal resistance and laminar airflow within the nasal cavity.

The middle turbinate has a complex configuration and attachment. Anterosuperiorly it is attached to the skull base, and posteriorly it curves laterally to attach to the lamina papyrecea forming a lamella termed the *basal lamella*. The basal lamella divides the ethmoidal complex into an anterior and a posterior group. Lateral to the middle turbinate is a recess termed the *middle meatus*. The middle meatus is a recess into which drain the frontal, maxillary and anterior ethmoid sinuses.

Two distinct bony prominences are seen laterally on removal of the vertical portion of the middle turbinate: the uncinate process and the ethmoid bulla. The uncinate process is a thin, boomerang-shaped bone anterior to the ethmoid bulla. It attaches to the lacrimal bone anteriorly and to the inferior turbinate inferiorly. The posterior margin of the uncinate process is unattached and forms a sickle-shaped cleft between its free margin and the anterior face of the ethmoid bulla, called *hiatus semilunaris*. The hiatus semilunaris communicates laterally with a three-dimensional space, the ethmoidal infundibulum, bounded anteriorly by the aggar nasi and frontoethmoidal cells, medially by the uncinate process, posteriorly by the bulla ethmoidalis and laterally by the lamina papyracea.[4] See Figure 1.3.

The blood supply to the lateral nasal wall and septum is mainly provided by the branches of the sphenopalatine artery with contribution from the anterior and posterior ethmoid artery, the greater palatine artery and the facial artery. Nerve supply is from the nasociliary branch of the anterior ethmoidal nerve, branches of the pterygopalatine ganglion and anterior palatine nerves. Lymphatic drainage is to the submandibular nodes anteriorly and to the retropharyngeal and upper deep cervical nodes posteriorly.

The ethmoid sinus

The ethmoid sinuses are the most variable of the sinuses and develop from pneumatization of the ethmoid bone. When this pneumatization extends to the middle turbinate, it is termed a *concha bullosa*. Occasionally, the pneumatization of the ethmoid bone can extend beyond the ethmoid bone. These extra-ethmoidal pneumatizations

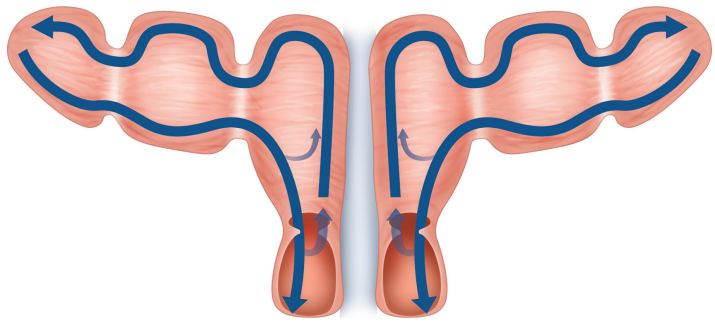

Figure 1.3 Direction and movement of mucociliary activity within the frontal sinus.

are often allocated with specific terms: orbit bone (supraorbital cell), roof of the maxillary sinus (Haller cell), floor of the frontal sinus (frontoethmoidal cell) and superolateral to the sphenoid sinus (onodi cell).

The ethmoid roof is seen to slope medially and posteriorly. Medially, it forms the fovea ethmoidalis and the cribriform plate. The lamina papyracae separates the ethmoid sinus laterally from the orbit. The sphenoid sinus forms the posterior boundary of the ethmoid sinuses.

A number of bony septa divide the ethmoidal sinus into up to 18 air cells, and these are grouped to form the anterior and posterior ethmoid air cells depending on their relationship to the basal lamella. The attachment of the basal lamella of the middle turbinate divides the anterior and posterior ethmoid systems.

The largest of the anterior ethmoid air cells is the ethmoid bulla. This is the most consistent surgical landmark. It may be unpneumatized in 8 per cent of patients. The ground lamella constitutes the posterior boundary of the bulla ethmoidalis. Suprabullar and retrobullar recesses may be formed when the ethmoid bulla does not extend to the skull base. The suprabullar recess is formed when there is a cleft between the roof of the ethmoid bulla and the fovea. The retrobullar space is formed when there is a cleft between the basal lamella and the bulla. These spaces are collectively called the *lateral sinus*. The ostia of the anterior ethmoid cells open into the ethmoid infundibulum in the middle meatus. The most anterior ethmoid air cell is called the *agger nasi cell*. The face of the bulla attaches to the skull base immediately anterior to the anterior ethmoid artery. The anterior ethmoid artery exits the orbit through the lamina papyracae and courses horizontally across the roof of the ethmoid sinus in a thin bony mesentery (dehiscent inferiorly in 40 per cent of patients) to enter the cribriform plate and anterior cranial cavity through the fovea ethmoidalis. It then vertically penetrates the most anterior aspect of the cribriform plate to enter the nasal cavity to supply the septum and the anterosuperior nasal cavity as the terminal septal branch. The posterior cells drain into the superior or supreme meatus. The most posterior ethmoidal air cell may extend superiorly and laterally to the sphenoid in 10 per cent of cases (the onodi cell) and may have the optic nerve and internal carotid artery bulging into it.

The blood supply to the ethmoid sinus is from branches of the anterior and posterior ethmoidal and sphenopalatine arteries. Venous drainage follows the arterial supply. Lymph drainage is to submandibular and retropharyngeal nodes. The innervation is from the supraorbital, anterior ethmoidal and orbital branches from the pterygopalatine ganglion.

The maxillary sinus

The maxillary sinus lies within the maxillary bone and is pyramidal in shape, with its base forming part of the lateral nasal wall and its apex pointing towards the zygomatic process. The roof of the sinus constitutes the orbital floor and contains the infraorbital nerve, which may be dehiscent in 14 per cent of cases. The alveolar process of the maxilla and the hard palate form the floor of the

maxillary sinus. The thinnest part of the anterior wall corresponds with the canine fossa. The posterior wall separates the maxillary sinus from the pterygomaxillary fossa and its contents, i.e. the internal maxillary artery, sphenopalatine ganglion and greater palatine nerve.

At the superior aspect of the medial wall of the sinus, opening into the region of the inferior aspect of the ethmoid infundibulum, is the natural ostium. This is normally hidden from view by an intact uncinate process and therefore cannot be visualized endoscopically. The nasolacrimal duct runs 4–9 mm anterior to the ostium. The medial wall of the maxillary sinus has areas of bony dehiscence usually covered by mucosa called the *anterior and posterior fontanels*. In up to 30 per cent of cases these may be patent, usually at the posterior fontanel, to form an accessory ostium. The accessory ostia are nonfunctional.

Branches of the internal maxillary artery, i.e. the infraorbital and greater palatine arteries, provide the blood supply, and venous drainage is through the pterygoid plexus and facial vein. Lymphatic drainage is to the submandibular lymph nodes. The infraorbital, greater palatine and superior alveolar nerves provide the nerve supply to the maxillary sinus mucosa.

Frontal sinus

Frontal bone pneumatization to develop the frontal sinus is variable, with approximately 5 per cent of the population demonstrating no frontal sinuses. Being a midline sinus, the two sides are usually separated by intersinus septa, with both frontal sinuses draining independently at the lowest medial portion of the cavity. The frontal sinus drains into the superior aspect of the ethmoid infundibulum via the frontal recess.

The anterior and posterior walls of the frontal sinus are composed of diploeic bone. The posterior wall separates the frontal sinus from the anterior cranial fossa and is much thinner. The floor of the sinus also functions as a portion of the orbital roof. The supraorbital and supratrochlear branches of the ophthalmic artery provide its blood supply, but venous drainage is to the cavernous sinus via the superior ophthalmic veins and to the dural sinuses through the posterior wall venules. The supraorbital and supratrochlear nerves provide its innervation.

Sphenoid sinus

The sphenoid sinuses are the deepest of the paranasal sinuses pneumatizing the sphenoid bone. The sinus can be absent in about 1 per cent of the population. Like the frontal sinus, sphenoid sinuses are divided, often asymmetrically, by a septum in the paramedian position. Anteroinferiorly, the sphenoid rostrum articulates with the perpendicular plate and vomer of the nasal septum. The sphenoid sinus ostium lies in the anterior medial wall of the sinus and drains medial to the superior turbinate into the sphenoethmoidal recess.

Several important vascular and neural structures lie in the walls of the sphenoid sinus and can indent the walls to a variable degree depending on the degree of pneumatization. The posterior wall is formed by the sella turcica superiorly and clival bone inferiorly. The clival carotid artery traverses vertically in the clival portion of the posterior wall and loops forward in the cavernous portion. Together with the optic nerve, these are prominences evident in the superolateral wall of the sphenoid sinus, with an indentation separating the two called the *lateral oculo-carotid recess*. The maxillary part of the trigeminal nerve traverses the lateral wall and the vidian nerve of the floor of the sphenoid sinus. Some bony dehiscence over structures such as the optic nerve (6 per cent of the population) and carotid arteries (8 per cent of the population) are not infrequent. Intersinus septa can often be in continuity with the carotid and the optic canal, and uncontrolled avulsion can result in a catastrophic bleed or blindness.[4]

Apart from the roof, which is supplied by the posterior ethmoid artery, the rest of the sphenoid receives its blood supply from the sphenopalatine artery. Venous drainage is via the maxillary veins to the jugular and pterygoid plexus systems. The nasociliary nerve supplies the roof, whilst the floor receives its innervations from branches of the sphenopalatine nerve.

PHYSIOLOGY OF THE NOSE AND PARANASAL SINUSES

The two main functions of the nose are respiration and olfaction.

RESPIRATION

The nose is solely responsible for the warming and humidification of air that reaches the lungs. Humidification is facilitated by evaporation of the secretions of the numerous serous glands found in the mucosal blanket and condensation of expired air at the anterior nose. Air is humidified to 75–80 per cent.

Warming occurs as cool, inspired air creates a temperature gradient as it comes into contact with the counterflowing rich arterial blood from the sphenopalatine artery, especially the inferior turbinate mucosa, such that the air reaching the postnasal space is approximately 31°C. Although this countercurrent exchange creates a more efficient heat exchange, this process remains imperfect, with as much as 10 per cent of total body heat being lost through the nose with expired air. Filtration of inspired air occurs first in the nose. The vibrissae filter the largest particles, and the mucous blanket filters the rest. The mucous membrane is enriched with immunoglobulin A (IgA), providing the first line of immunological defence.[5]

OLFACTION

Olfaction is the perception of smell and is a primal sense for humans and animals, allowing receptors to identify food and mates, and provide warnings of danger (such as fires or chemical dangers) as well as sensual pleasures (such as perfumes). For an odorant molecule to reach the olfactory area, turbulent airflow from the anterior nares is required. About 15 per cent of inspired air reaches the olfactory area within the nose, where it interacts with the mucus secreted by the Bowman's glands in the lamina propria and respiratory epithelium. To reach the olfactory receptors, the odorant molecules must be soluble in the mucus and need high water and lipid solubility. These molecules react with the lipid bilayer of the receptor cells at specific sites, which causes an efflux of K+ and CI–, resulting in depolarization of the cells. This process is mediated by G-protein coupled receptors in the cells that interact with a specific adenyl cyclase within the neuroepithelium.

Olfactory responses show variations in both thresholds and adaptation depending on the chemical nature of the stimuli. Thresholds of perception are lower than identification, because smells are sensed before they are recognized. Changes in the composition of the mucus can influence the diffusion time required for odorant molecules to reach the receptor sites. The beta-adrenergic, cholinergic, and peptidergic agents' effect on the sensory perception of smell is through their effect on the secretory activities of the mucosal layer.

These functions are facilitated by a number of physiological systems, which are described in the following sections.

Airflow

The biomechanics of airflow and nasal resistance is described by Bernoulli's and Raymond's equations, given below. Based on laminar flow equations, decreases in *r*, or the nasal airway radius, cause four-fold decreases in flow, as described by Bernoulli's equation.

$$Q = (\Delta P \pi r^4)/(8 \eta L)$$

Reynolds number = $2rQ \rho/\eta$ (Reynolds number greater than 2000 is equated with turbulent flow)

where L = length, r = radius, P = pressure, η = viscosity, and ρ = density.

The presence of laminar or turbulent flow is pertinent to the physiology of air exchange. The cross section within the nose is variable, and this continuously alters the pressure and velocity within the system. During inspiration, the airflow is directed upwards and backwards from the nasal valve mainly over the anterior part of the inferior turbinate, below and over the middle turbinate and then into the posterior choana. Air reaches the other parts of the nose to a lesser degree. The

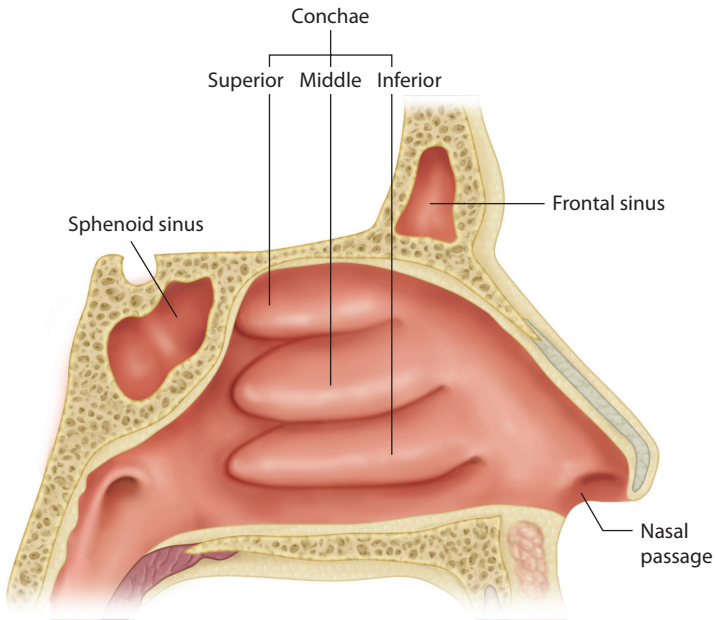

Figure 1.4 Lateral nasal wall with the superior, middle and inferior turbinates.

velocity at the anterior valve is 12–18 m/s during quiet respiration, and it is considered laminar, although in practice it is turbulent even in quiet respiration, producing eddies in the olfactory region. Turbulent flow results when the velocity of nasal flow is increased. Expiration lasts longer than inspiration, and flow is more turbulent. A sniff is required to facilitate olfaction with turbulent airflow. Narrowing or alterations at the nasal valve area (septal perforations, septal deviations) result in turbulent flow within the nose, which can cause a sensation of obstruction regardless of nasal passage patency.[6] See Figure 1.4.

The nasal cycle

This feature of normal nasal physiology is a cyclical alteration in nasal resistance between the two nostrils secondary to alteration in vascular activity that regulates the volume of venous sinusoids (capacitance vessels) in the nasal erectile tissue (located primarily in the inferior turbinate and to a lesser extent in the anterior septum). These changes occur between 4 and 12 hours and enhance humidification, warming and mucociliary clearance. The nasal cycle is affected by factors such as allergy, infection, exercise, hormones, pregnancy, emotions, sexual activity and recumbent position.

The mucociliary system

The mucociliary system is a vital component of the normal sinonasal function of humidification, filtering of inspired air, and elimination of secretions and debris from the paranasal sinuses and nasal airway. This system consists of three components:

a. *Ciliated, pseudostratified columnar epithelium lines nasal and paranasal sinuses.* Cilia on the surface of these cells propel mucus backwards in the nose towards the postnasal space. Each cilium has a surface membrane and encloses an organized ultrastructure of nine paired outer microtubules that surround a single inner pair of microtubules. Outer-paired microtubules are linked together by nexins and to the inner pair by central spokes. Outer pairs also have inner and outer dynein arms.
b. *Double-layered mucous blanket* has a less viscous, watery layer (sol phase) in which cilia move freely as well as a superficial, more viscous mucous fluid (gel phase) into which the tips of the cilia enter to move it.

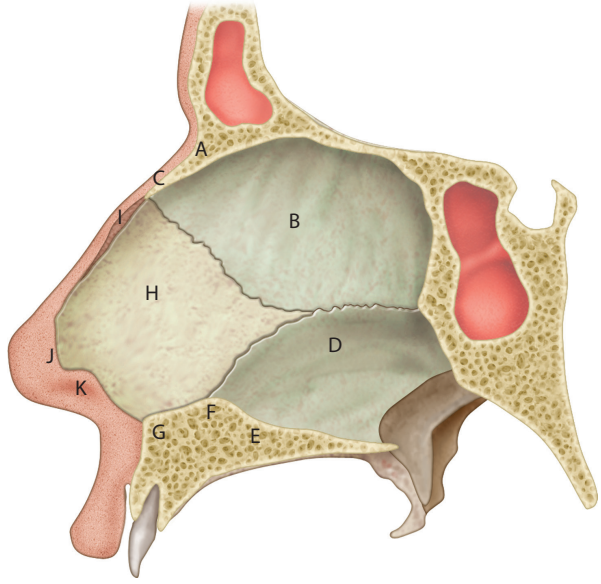

Figure 1.5 Nasal septum and its components. A, nasal spine of frontal bone; B, perpendicular plate of ethmoid; C, nasal bones; D, vomer; E, palatine process of maxillary bone; F, maxillary crest; G, anterior nasal spine; H, quadrilateral cartilage; I, upper lateral cartilage; J, lower lateral cartilage; K, columella.

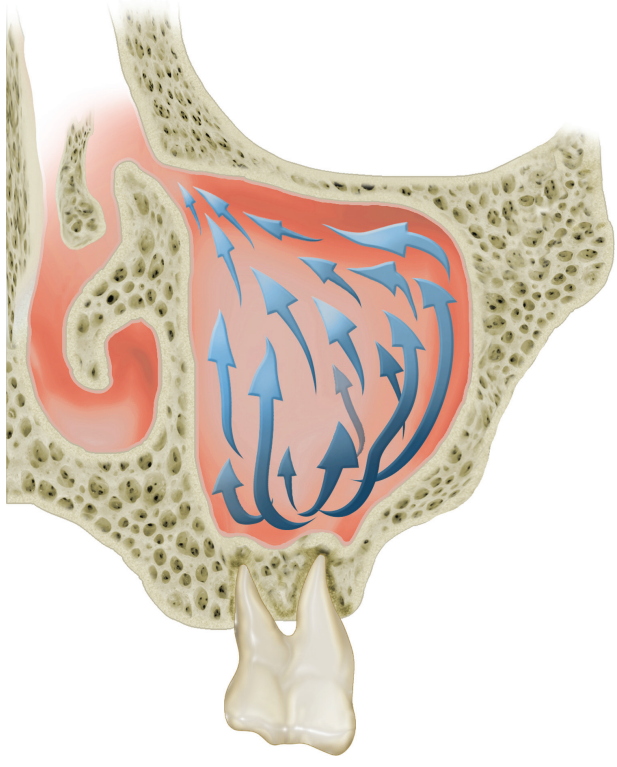

Figure 1.6 Direction and movement of mucociliary activity within the maxillary sinus.

c. *Mucous-producing glands* include goblet cells, seromucinous glands and intraepithelial glands. The major composition of nasal mucus is water (95 per cent), glycoproteins or mucin (3 per cent), salts (2 per cent), immunoglobulins (IgA), lysozymes (bacteriolytic) and lactoferrin (bacteriostatic).

The ciliated cells beat in a specific direction, resulting in a pattern of mucus flow. The cilia actively beat to propel mucus to the nasopharynx. This mucociliary flow occurs at 1 cm/min, and mucus stasis as a result of decreased beat frequency may allow noxious substances to penetrate the mucosa, resulting in disease. Pathological processes such as bacterial and viral infections impede ciliary beating by altering the ultrastructure or the viscosity of the mucous blanket.[7]

In the paranasal sinuses, the cilia beat such as to move material towards the sinus ostia. This means that cilia often move mucus against gravity to drain the sinus at its ostium. See Figure 1.6.

KEY LEARNING POINTS

- Continuity and fixation at the keystone area are important because this supports the projection of the upper third to mid third of the nose.
- The frontal, maxillary and anterior ethmoid sinuses drain into the middle meatus.
- Intersinus septa within the sphenoid sinus can often be in continuity with the internal carotid artery and the optic canal.
- Turbulent airflow within the nose can cause a sensation of obstruction regardless of the patency of the nasal passage.
- The nasal cycle is affected by allergy, infection, exercise, sexual activity and recumbent position.

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2

Investigation of nasal diseases

CARL PHILPOTT

INTRODUCTION

Rhinological diseases are very common, and whilst an array of investigative tools exists for them, many of these are not common practice in routine ear, nose and throat (ENT) clinics. The advent of the endoscope and the significant improvements seen in the imaging and image capture technology have allowed us to better see inside the nose and beyond. This not only gives the clinician a much more comprehensive assessment of the nose and sinuses, but also facilitates education and engagement of the patients themselves as they too can see the effects of their disease and the response to any treatments. The 'European Position Paper on Rhinosinusitis and Nasal Polyps' (EPOS) has been a key document for guiding clinicians involved in managing chronic rhinosinusitis, and it brings guidance as to the logical flow of investigation in this condition. This chapter will lay out the full armoury of investigations available for rhinological disease that are suitable both in clinical practice and in the research domain, or even in tertiary centres. It will also give some direction as to how these fit with the EPOS guidance and a framework for investigating rhinological disease.

HISTORY AND EXAMINATION

As with any part of medicine, history taking is the crucial step that should ideally precede all

investigations, but symptom scores filled in by patients in the waiting room can certainly help to focus both the patient's and clinician's mind on the key symptoms that have brought the patient to the clinic. These questionnaires are discussed in further detail later, but the key is to utilize a validated questionnaire that is relevant to the patient's presenting complaint. Use of open questions initially will allow patients to address their concerns and the reason they believe they are sitting in front of you (which may not always be the reason they were referred). Following this, closed questions may be needed to checklist key rhinological symptoms that the patient may not have covered. These include:

- Congestion/blockage/obstruction
- Rhinorrhoea – anterior and posterior
- Olfactory disturbances – quantitative (reduced or absent) and qualitative (distorted)
- Facial pressure/pain
- Epistaxis and crusting

Other related symptoms include sneezing, itching, cough, fever, dental pain, otalgia/aural fullness, snoring, visual disturbances and nasal deformity, but this list is not exhaustive. It is also crucial to explore what patients actually mean because 'congestion' and 'facial pressure' often overlap for many, and it is common for patients to say, 'I've got sinus, doc' or something to that effect. Because the symptoms and their duration are the centre point to making the diagnosis of both rhinitis and rhinosinusitis, it is important to have focused on this in order to proceed with the appropriate investigations. When faced with olfactory disorders, it is equally important to listen to the patient carefully and explore exactly what his or her prime complaints are. Many patients will talk about loss of taste without there being any true gustatory disturbance. Also, patients may be significantly affected by olfactory distortions such as parosmia and phantosmia, which may even outweigh any actual loss of olfactory acuity.

The social and occupational history of many patients will also be relevant, especially in the presence of atopy, and may influence the subsequent management of the patient. Thus exploration of the presence of any animal contacts, work environment and smoking status is needed, and alcohol intake may also influence symptoms and may give a key to salicylate sensitivity. This leads to key factors in the medical history such as any adverse effects of aspirin and nonsteroidal anti-inflammatory drugs (NSAIDs) as well as other drugs that may influence nasal physiology such as β-blockers. Many patients will have concomitant respiratory disease, and the severity of this should be quantified along with an understanding of any key areas of interaction between the two (e.g. common triggers, exacerbations linked to upper airways).

Examination of the external nose can easily be achieved by carefully viewing the nose from in front of the patient as well as from above and underneath and finally from the side (profile view). Traditionally, using a head mirror and Mills lamp along with a Thudicum's forcep allowed a clinician to perform anterior rhinoscopy. Most modern ENT clinics provide a battery-operated headlight, and this can be used to perform anterior rhinoscopy, which is useful in assessing the nasal vestibule, nasal valve and anterior nasal septum, especially the position of the columella. Internal examination of the nose is best achieved with a rigid nasendoscope connected to a monitor and image capture device, and this should be considered the gold standard. In the face of most rhinological symptoms, this is the only sure way to carefully assess the nasal cavity and the relevant meati. Use of a flexible nasendoscope will reduce the ability to use suction or take biopsies. A common approach with the rigid endoscope is to use the three-pass technique, working from inferior to superior in the nose and identifying the three meati, the three turbinates and the postnasal space (Fig. 2.1). In this method a 0° endoscope is passed first along the nasal floor towards the postnasal space and if possible underneath the inferior turbinate. The second pass is into the middle meatus lateral to the middle turbinate allowing identification of the ethmoid bulla and uncinate process. The third pass is medial to the middle turbinate and into the sphenoethmoidal recess to identify the superior turbinate, superior meatus and sphenoid ostium. However, an alternative strategy is to use a 30° endoscope inserted into the nose with the bevel facing superiorly and insert the scope into the middle meatus. By rotating the light post, the endoscope can then be turned

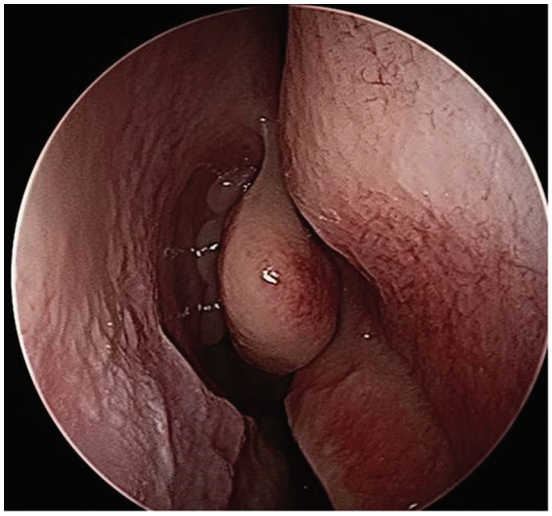

Figure 2.1 Endoscopic view of nose.

to visualize the middle meatus both laterally and then inferiorly and then passed under the middle turbinate towards the postnasal space. Here, rotation of the light post again will allow visualization of the superior meatus and sphenoid ostium. In a post-operative patient (after sinus surgery), the 30° endoscope should be mandatory to allow visualization of the sinus cavities, with a 70° endoscope in reserve if needed.

There are two schools of thought on the use of decongestant preparations prior to endoscopy. Some clinicians will routinely apply a spray such as co-phenylcaine to the nose before any examination, whereas others will choose to examine the nose without the influence of any vasoconstrictor. The latter approach gives the physician the advantage of being able to view the diseased mucosa in the nose in its current state as relevant to the patient's symptoms; this may be distorted by decongestants. Endoscopy requires a careful hand to avoid discomfort to the patient but can be seen as an extension of endoscopic sinus surgery where avoiding contact with the nasal mucosa reduces trauma to the mucosa and the need to clean the endoscope tip frequently. Nonetheless, some patients will need a local anaesthetic to tolerate the procedure or decongestant to enable visualization where anatomical variations such as a septal deviation are present.

MICROBIOLOGY

In chronic sinusitis without nasal polyps (CRSsNPs) identification of mucopus on endoscopy is one of the two ways in which the diagnosis based on the symptoms can be confirmed, and in the current EPOS guidelines indicates the potential use of long-term antibiotics (macrolides). However, retrieval of this material can also help to guide the clinician further in the appropriate choice of antibiotics because it will yield information on the bacterial sensitivity and resistance profiles. In the case of chronic sinusitis with nasal polyps (CRSwNPs), retrieval of thick eosinophilic mucin may be important in the process of defining a case of allergic fungal rhinosinusitis. This material should be sent for fungal smear and culture alongside the usual culture and sensitivity. Collection of such material can be achieved either by using a swab or by using one of a variety of suction traps such as a Leukens trap or a Xomed sinus secretion collector (Fig. 2.2). The latter has a more significant advantage in a post-operative patient in whom use of a curved suction tip can allow retrieval of mucus from sinus cavities. Although the evidence for the use of topical antibiotics is at present limited with formal randomized control trials (RCTs) lacking, culture-directed management can still be implemented so that any antibiotics given either orally or topically can be based on material sampled directly from the site of the disease process. This

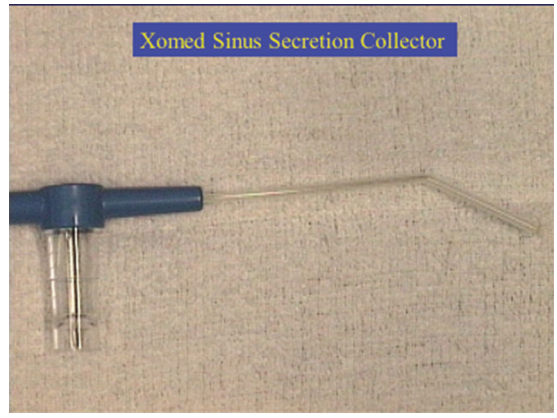

Figure 2.2 Xomed sinus secretion collector.

has the advantage of ensuring that antibiotic use is appropriate and helps to avoid resistant strains in common pathogens such as methicillin-resistant *Staphylococcus aureus* (MRSA). Liaison with colleagues in microbiology will also help to ensure that they understand your requirements, especially with respect to fungal specimens, for which it can be notoriously difficult to yield positive results.

BLOOD AND ALLERGY TESTS

The choice of chemical pathology testing will be largely influenced by the history. Investigation of atopy can be undertaken by either skin prick testing (SPT) or radioallergosorbent test (RAST), with the former considered the gold standard. However, this is dependent on staff availability to perform the skin prick. RASTs, on the other hand, are less involved in terms of staff resources in the clinic but are more costly to perform owing to the analysis required in the laboratory. RASTs do provide the option to test for allergens not available in routine sets of aeroallergens available for SPT, which may be useful for specific patient exposures or patients with suspected allergic fungal rhinosinusitis. The prevalence of atopy in chronic rhinosinusitis (CRS) varies in the literature but probably accounts for 50 per cent of CRS with nasal polyps (CRSwNPs). Detection of atopy in rhinitis will influence medical management and should be checked for if the history is unclear and initial medical measures have failed; in fact rhinitis cannot ultimately be classified as allergic or non-allergic without formal testing.

When considering blood tests, a total IgE level may indicate a patient's overall level of allergic response; in the latest EPOS guidelines, this is recommended in CRS without nasal polyps (CRSsNPs) to determine a patient's suitability for the use of macrolides. Other blood tests to be considered include:

- Full blood count (eosinophilia seen in allergic fungal rhinosinusitis and Churg-Strauss Syndrome, anaemia from epistaxis)
- Clotting screen – for epistaxis where anticoagulants used or deficiency suspected from history
- Immunoglobulin screening
- Blood markers of systemic diseases such as anti-neutrophil cystoplasmic antibody (ANCA), angiotensin-converting enzyme (ACE)
- Exclusion of potential underlying medical comorbidities (e.g. renal/hepatic dysfunction, hypothyroidism, Cushing's disease, sarcoidosis) in cases of suspected idiopathic anosmia

In patients with recurrent sinusitis or with CRS refractory to combined medical and surgical therapy, immunodeficiency should be considered (e.g. common variable immunodeficiency). An array of specific and complete immunodeficiencies is possible, with the most common ones found in the immunoglobulin G subclasses such as 2 and 3, which confer protection against common encapsulated nasal pathogens such as *Haemophilus influenzae*. The relevance of specific deficiencies should be assessed by referral to an immunology specialist; this may simply result in additional vaccinations being given. Severe combined immunodeficiencies will need much greater input from an immunologist.

If factors in the history or clues from examination suggest the possibility of systemic diseases such as sarcoidosis, these should be investigated on their own merit and, where appropriate, further corroborated with nasal biopsies.

CYTOLOGY AND HISTOLOGY

NASAL BRUSHINGS

Patients with rhinitis can grossly be divided into those with atopy (persistent or intermittent allergic rhinitis) and those who are not atopic. In a subset of those with non-allergic rhinitis, entopy may be present. This implies a local allergic response in the nasal mucosa that is not associated with a systemic atopic response seen on positive SPT/RASTs. This can be determined by taking a nasal brushing and sending it for cytological examination to look for eosinophilia. A positive result suggests a diagnosis of non-allergic rhinitis eosinophilia syndrome (NARES), which can be treated much the same as

allergic rhinitis, although it may require a nasal allergen challenge to confirm this. A nasal brushing can be performed easily with a bronchial brush rubbed along the anterior inferior turbinate.

HISTOLOGICAL EXAMINATION

Unilateral nasal masses should always be viewed with suspicion, especially in the older patient, and consideration should be given to performing a biopsy. This decision will be taken on a case-by-case basis influenced by any sinister symptoms in the history such as unilateral epistaxis, crusting, orbital symptoms, numbness or cranial neuropathies. In certain cases it may be prudent to arrange imaging before a biopsy is undertaken, as follows:

- Possibility of the lesion arising from the skull base with intracranial extension (e.g. meningo-encephalocoele)
- Posterior unilateral nasal mass in a juvenile male patient (juvenile angiofibroma)

Other circumstances for taking a biopsy will include suspicion of systemic diseases such as Wegner's granulomatosis or as part of investigation of a septal perforation. The location in which the biopsy is performed will be influenced by patient suitability for local anaesthesia, equipment available in the clinic and the nature of the lesion/condition being investigated, but biopsy of many lesions can be appropriately performed in the clinic.

RADIOLOGICAL INVESTIGATIONS

In the modern era the use of plain film radiography is negligible, and it has no real place in the investigation of rhinological diseases, with perhaps the occasional exception of a lateral film in the case of a young child in whom adenoidal hypertrophy is suspected but who will not tolerate endoscopic examination. The use of computed tomography (CT) imaging in patients who are due to undergo any form of nasal polypectomy or sinus surgery should be considered mandatory because this allows the clinician to assess any unfavourable anatomic features and provide a targeted approach to the affected sinuses as part of careful preoperative planning. CT scans in the presence of nasal polyposis can also give a clearer picture of the likelihood of an adequate response to medical and surgical treatment. For instance, a scan showing a Lund-Mackay score of 24 out of 24 and double-density signs would alert the clinician to the possibility of allergic fungal rhinosinusitis. A guide to the timing of imaging in the management of CRS can be found in the EPOS guidelines. As mentioned earlier, any case of suspected neoplasia will require imaging to assess and stage the lesion and will often require both CT and magnetic resonance imaging (MRI). Asymmetrical symptoms, even in the absence of endoscopic findings, usually warrant radiological investigation to ensure that no sinister pathologic organism is responsible. Liaison with radiological colleagues for unusual cases and neoplasia is important to ensure that the imaging is relevant and, when performed, answers the clinical question intended. However, clinicians should always aim to review all imaging personally to ensure that patient symptoms are carefully correlated with the findings. In cases proceeding to surgery, review of the imaging should be mandatory.

NASAL AIRWAY MEASUREMENTS

SPATULA MISTING

A simple test of the nasal airway is to place a cold metal spatula underneath the nose to look for any signs of unequal misting relating to each nasal cavity. It is a crude test but will demonstrate any gross asymmetry.

PEAK INSPIRATORY NASAL FLOW

This is performed using a peak flowmeter similar to the expiratory one used for lower airway function. The device has a mask that is placed over the nose and mouth. With the mouth closed, the patient is asked to take a deep breath in through the nose. The measurement is repeated twice more, and

the best of the three readings is taken as the final result. Limitations to the test include the presence of alar collapse and lower respiratory tract disease. A guide to interpreting the results is as follows:

<50 L/min = severe nasal obstruction
50–80 L/min = moderate nasal obstruction
80–120 L/min = mild nasal obstruction
>120 L/min = normal

ACOUSTIC RHINOMETRY

In what is effectively sonar of the nose, acoustic rhinometry relies on the reflection of a sound wave to measure the cross-sectional area of the nose and estimate volume. A frequency of 150–10,000 Hz in the form of an audible sound pulse is used by propagating the sound from a click in a tube positioned under the nostril. The differing cross-sectional areas of the nose created by the anatomical structures of the nasal valve and beyond create reflected sound waves that are picked up by the microphone. These are then plotted by the attached computer software as a trace. This is largely a research tool because it is cumbersome to perform and the user needs practice to get a meaningful reading. However, a standardization committee has written guidelines for the application and interpretation of the results.

RHINOMANOMETRY

Similar to acoustic rhinometry, rhinomanometry is generally performed in a research setting but may provide a useful tool in selected patients who complain bitterly about nasal obstruction in the absence of objective blockage. Nasal airway resistance is measured using a combination of flow and pressure readings from the nose. The technique can be performed actively or passively via anterior or posterior approaches. Typically, active anterior rhinomanometry is the choice. One nostril is blocked, and a mask is placed over the mouth and nose with a firm seal created on the face. One nostril is blocked using a sponge seal or tape, but within this is a catheter that will measure the pressure. With the patient breathing in and out moderately through the other nostril and with the mouth closed, the mask will measure the flow rate in the attached pneumotachograph. The combined readings produce a sigmoid-shaped curve on the attached computer software. This is repeated on the opposite nostril by switching the catheter over. Following the standardization protocols of the European committee, the resistance is measured at a fixed pressure of 150 pascals. Taking measurements before and after nasal decongestion is standard. Although results can vary significantly, rhinomanometry may still provide an objective before-and-after comparison in an individual subject.

RHINOSTEREOMETRY

This is a rarely performed investigation involving examination of the nasal cavity before and after an allergen challenge or mucosal moderator to assess the two-dimensional change in the inferior turbinate.

SPIROMETRY

This is traditionally a measure of lower respiratory tract function using peroral exhalation to measure forced expiratory volume; however, spirometers can be adapted with nasal prongs to provide the same function for measurement of the nasal airway, in the same way that peak inspiratory nasal flow (PINF) achieves the equivalent of peak expiratory flow rate (PEFR). Standard spirometry will usually be obtained by a respiratory department or a cardiorespiratory assessment unit and will provide measurements of forced expiratory volume in one second (FEV_1) and forced vital capacity (FVC) as well as the ratio of these two measurements.

OTHERS

PEFR is a well-recognized test of pulmonary function and simply requires a best of three peroral exhalations using a handheld flowmeter, which can be readily available in the clinic. Patients with concomitant lower respiratory disease may already be well versed in this measurement, but, nonetheless, it can provide a useful measure of the impact of upper respiratory tract (URT) disease on the lower airways.

NASAL PHYSIOLOGICAL MEASUREMENTS

MUCOCILIARY CLEARANCE TIME

A simple measure of the mucociliary train inside the nose is the saccharin test. This relies on the normal function of the nasal cilia, which pass the mucus blanket posteriorly towards the postnasal space. To perform the test, the patient is asked to identify when a sweet taste in the back of the mouth is detected. This moment is considered the end point of the test. To begin, a saccharin tablet is placed just underneath the anterior end of the inferior turbinate on one side of the nose, and a stopwatch is started. The stopwatch is then stopped when the patient experiences the sweet sensation. Normal mucociliary function is considered to be a duration of less than 20 minutes; however, the test may be used to compare function before and after treatment by looking for a decrease in the duration of the time interval. Those patients with a duration of 30 minutes or more should certainly be considered for further investigation for underlying ciliary dysfunction or dyskinesia. Clues in the history to primary ciliary dyskinesia include middle ear effusions and situs inversus, which can be seen in Kartagener's syndrome.

CILIARY BEAT FREQUENCY

This is a very specialized test to investigate ciliary dyskinesias and is performed in only three centres in the UK: Leicester, Royal Brompton (London) and Southampton. A normal-functioning sinonasal epithelium will beat at a frequency of 12–16 Hz, but if lower than 11, it is considered in keeping with ciliary dyskinesia, although ciliary beat pattern analysis may pick up some cases with a normal frequency. Abnormal responses will usually be correlated with electron microscopy of the tissue itself.

ELECTRON MICROSCOPY

As an adjunct to ciliary beat frequency and an abnormal mucociliary clearance test, electron microscopy (EM) allows detailed examination of the microstructure of the sinonasal epithelium, which may be relevant to any findings of poor function. Typical abnormalities detected include microtubule array anomalies in the cilia, such as incorrect number of pairs of microtubules, compound cilia and dynein arm irregularities.

NITRIC OXIDE

As with many of the airway parameters already discussed, measurement of nitric oxide (NO) remains largely a research tool. NO levels in the sinuses appear to have an optimum level; levels above or below can be associated with disease states. The ostiomeatal complex is believed to act as an interface controlling NO levels between the nose and sinuses. Nasal nitric oxide can be measured by inserting a probe into one nostril, and, while the patient holds her breath, a reading is taken by the attached gas analyzer. Normal levels are considered to be 450–900 parts per million (ppm); a level above 900 ppm infers inflammation, one less than 450 ppm is associated with blockage and one less than 100 ppm indicates ciliary dyskinesia. The measurement is performed for both nostrils and the mean value derived.

SWEAT TEST AND OTHER CYSTIC FIBROSIS TESTS

Many cases of cystic fibrosis (CF) are now detected perinatally, but if the family history or presentation of a paediatric patient with nasal polyps suggests the need to investigate for CF, the patient should be referred for a formal sweat test. Other clues to the diagnosis may be found in the medical history (lower respiratory tract or gastro-intestinal symptoms) or from CT imaging, because patients with CF typically have hypoplastic frontal and sphenoid sinuses in addition to the opacification associated with the apparent rhinosinusitis. Most units will have the facility to refer locally to their paediatric colleagues for sweat testing. Should this test be positive, patients will also require further testing for the CF transmembrane regulator (CFTR) gene and genetic counselling for future implications. A finding of two CFTR mutations in association with clinical symptoms is diagnostic. The pilocarpine iontophoresis sweat test remains the gold standard

and will produce a chloride concentration of greater than 60 mmol/L in CF. However, this result should be found on two separate occasions when the individual is not suffering any infective exacerbations before a diagnosis of CF is confirmed. Although nasal potential difference across the epithelium is a sensitive test for CF, the equipment to perform this is rarely available; hence few centres perform this test as part of a diagnostic workup.

OLFACTORY TESTS: PSYCHOPHYSICAL

Although olfaction is a key function of the nose that most of us take for granted, it is rarely tested as a matter of routine. There are a number of commercially available kits for olfactory testing with a variety of prices, and they vary in the aspects of olfaction that they address. Validation in a local population is key to the relevance of an olfactory test because cultural influences have a significant impact on their suitability. This is particularly the case when employing an identification test, owing to the need for a subject to verbalize the odour names and to have sufficient familiarity with them for the test to be relevant. Hedonics also have a role to play in this context, because what may be a pleasant odour to one cultural group may be considered repulsive to another.

Olfactory threshold testing

This is the olfactory equivalent of determining hearing thresholds in audiology. However, in contrast to audiology, olfactory threshold testing starts by using small odour concentrations and working towards greater ones as the test progresses. Odours typically used for this purpose include phenethyl alcohol (PEA) and 1-butanol (1-BUT); the former has a rose-like smell, but the latter is a more nonspecific solvent-like smell. However, both are considered to be pure olfactory stimulants in the concentrations used for testing and therefore avoid the issue of trigeminal pathway activation. Recognition of the odour here is not relevant, because subjects are required to simply detect the presence or absence of an odour stimulus. To perform this test, the internationally recognized format is the alternative forced-choice format using a ladder progression. This can include two alternative choices, as found in the Connecticut Chemosensory Clinical Research Centre Test (CCCRCT), or three alternative choices, as found in the Sniffin' Sticks test. Subjects are asked to choose which of two or three stimuli contains an odour, even if the subjects cannot detect anything. Some subjects struggle to understand why they must name a choice regardless of definitive detection, but this avoids allowing a subject to skew the test when close to his or her threshold and tempted to give a null response. Starting with the smallest odour concentration, subjects are given two or three sets of stimuli (two for three alternative forced-choice and vice versa for two), and any incorrect response will automatically mean that the next stimulus will be from a level up on the odour concentration chart. In Sniffin' Sticks, this means a jump up of two levels during the first part of the test, allowing quicker focus on the threshold level relevant to the subject. Once a subject detects two or three correct sets for one odour concentration, the level is then moved down to a smaller concentration. This ladder approach is repeated until seven reversals have occurred; the first three reversals are excluded. The average of the final four reversals is then taken as the threshold. This test is time consuming and at present usually requires a member of staff to perform the test. However, it is considered the most sensitive indicator of overall olfactory function, especially because it avoids recognition and verbalization of odours in the test.

Olfactory discrimination

Tests of discrimination examine a subject's ability to distinguish one odour from another. For example, in the Sniffin' Sticks test, subjects are asked to identify which of three stimuli represents the 'odd one out', where two of the three stimuli are the same odour. All odours presented are suprathreshold to avoid an additional dimension to the test.

Olfactory identification

One of the most widely recognized formats of olfactory testing is identification. Its greatest advantage is that it can often be performed by subjects with

little or no supervision, such as is seen with the University of Pennsylvania Smell Identification Test (UPSIT). The disadvantage is that this form of testing is culturally specific and requires verbalization of the odour in question, although research has shown some good correlation with threshold testing. In essence it is dependent on individual experience and familiarity with the odours being tested. Common formats for the test include scratch-and-sniff booklets, pens and diskettes, such as seen in the Zurich Smell Diskettes test. All identification tests include a four alternative forced-choice format with three possible distractors listed alongside the correct answer for each stimulus presented.

Retronasal testing

All of the previously discussed test modalities are orthonasal tests. In daily life we smell in two ways: orthonasally when something is in the environment around us, and retronasally when we are eating food. This is why many patients with olfactory loss may complain of taste disturbances in the absence of any true gustatory dysfunction. If there are specific issues around perceived loss of flavour, then this can be better evaluated using a retronasal test kit. This involves placing a selection of food powders on the back of the tongue using a dispenser and then asking the subject to perform an identification test, much as in the aforementioned format. This may be relevant in subjects with pathologic disorders causing blockages to the spheno-ethmoidal recesses or postnasal space.

OLFACTORY TESTING: OBJECTIVE

Most objective forms of testing currently lie in the research domain owing to the costs, manpower and time involved in performing them. A summary of those tests follows.

Olfactory event-related potentials

Akin to the electroencephalogram of smell, the olfactory even-related potentials (OERP) test is conducted using electrodes placed on the scalp, measuring a cortical response to an odour stimulus. The key apparatus involved is an olfactometer, which delivers a continuous stream of humidified air to one nostril with pulsed bursts of one of two odours that are released into the stream at varying intervals and durations. Airflow presented during the inter-stimulus interval must be odour free and warmed to body temperature and have more than 80 per cent relative humidity. Odours typically used are phenethyl alcohol (PEA) and hydrogen sulphide, to give both a pleasant and an unpleasant odour. For OERPs three midline electrodes are placed on the scalp (FZ, CZ, and PZ) referenced to linked earlobes (A1A2). In most circumstances, obtaining reliable OERPs requires the recording and averaging of 10–30 consecutive trials. The waveforms recorded are analyzed for the presence of negative-positive complexes consisting of an initial negative peak (N1) followed by a positive peak (P2) that have characteristic latencies and amplitudes. In simple terms, this is an all-or-nothing test where waveforms either exist or do not exist, but various factors can confound the result, such as signal noise and facial movements. Typically, subjects are given a distraction task and made to wear headphones producing white noise to reduce the influence of such factors. Testing takes 30–45 minutes and requires trained personnel, not to mention the cost of the olfactometer, which is typically in excess of £80,000.

The same setup can be used to deliver carbon dioxide to the nose, which produces a mildly painful response to trigger the trigeminal pathway. Then the responses can be measured as mentioned previously. The presence of an OERP implies that the olfactory apparatus is functioning, but its absence does not definitely imply a complete absence of function, although it is much less likely. Thus, in medicolegal work, it does have a limitation in dealing with malingerers.

Olfactory bulb volume and functional MRI

Using standard MRI sequences that include the entire olfactory bulbs (OBs), volumetric measurement of the right and left OB usually is performed by manual segmentation of the coronal slices through the OB. Usually, a 1.5 Tesla MRI with planimetric contouring of the bulb surface is performed, and the volume is calculated by

multiplying the obtained surface by slice thickness. The OB volume has been shown to correlate well with psychophysical testing and can also indicate recovery of olfactory function when sequential imaging shows an increase in volume.

Giving a topographical representation of higher neural centres activated by odour stimuli, functional MRI (fMRI) has been correlated with both orthonasal psychophysical testing and OERPs. By using the large olfactometer needed for OERPs, subjects can be provided with stimuli whilst in an MRI scanner. Activity in the orbitofrontal and entorhinal cortex as well as the cingulate gyrus can be mapped and measured, with a ratio to total functioning brain area calculated. For obvious reasons, this remains in the domain of research in specialist centres.

Electro-olfactogram

Electro-olfactograms (EOGs) show the electrical potentials of the olfactory epithelium that occur in response to olfactory stimulation. The EOG represents the sum of generator potentials of olfactory receptor neurons. In combination with nasal endoscopy and air-dilution olfactometry, the EOG can help to provide a comprehensive picture of olfactory processing. Again, because of the intricacies involved as well as costs and resources, it remains in the research domain.

PATIENT REPORTED OUTCOME MEASURES AND SYMPTOM SCORES

A number of symptom scores have been developed for rhinological disease internationally, and all of them typically address disease-specific quality-of-life complaints. These symptom scores are now referred to as patient reported outcome measures (PROMs) and have been validated in both healthy and diseased subjects in several countries as a means of evaluating the response to interventions in affected patients. In the United Kingdom, the most widely used PROM is the SNOT-22, which is the third iteration of the original SNOT-16 questionnaire. It contains specific symptom questions that can be divided into nasal and ear/facial pain and quality-of-life topics divided between mood/affect and sleep disturbances/concentration. Each item has a scale of 0–5, with 0 indicating no problem and 5 indicating the worst problem ever. Patients are asked to rate these symptoms for the preceding 2 weeks. A score of less than 10 is considered to be normal, and the maximum possible score is 110.

Other commonly used symptom scores for CRS include the Rhinosinusitis Outcome Measure (RSOM-31), Rhinosinusitis Disability Index (RSDI) and Chronic Sinusitis Survey (CSS). For rhinitis other questionnaires are available that are more disease-specific, including the Rhinoconjunctivitis Quality of Life Questionnaire (RQLQ) and related questionnaires developed by Professor Juniper in Montreal. All these have been validated and, where appropriate, can be used for routine clinical and research purposes to monitor outcomes.

KEY LEARNING POINTS

- A careful history is crucial to accurate diagnosis of inflammatory nasal/sinus diseases.
- Objective measures often correlate poorly with subjective symptoms.
- Patient reported outcome measures such as SNOT-22 are recommended for monitoring response to treatment.
- A range of objective measurements and investigations are available for selection on a case-by-case basis.

3

Epistaxis

THUSHITHA KUNANANDAM AND BRIAN BINGHAM

BACKGROUND

Epistaxis, derived from the Greek term *epistazein,* is defined as bleeding from the nose. It is one of the most common emergencies dealt with by otolaryngologists, although its severity and management can vary significantly.

The overall incidence of epistaxis in the general population is difficult to determine because most cases are unreported minor self-limiting episodes or those controlled with simple first-aid measures. Fewer than 10 per cent of patients seek medical attention for this condition and, again, fewer than 10 per cent of those requiring hospitalization require surgical intervention for control of bleeding. However, because many cases involve the elderly population, epistaxis is a significant cause of morbidity and even mortality in general otolaryngology practice.

ANATOMY

The nose has an extremely rich blood supply with contributions from both the internal and external carotid arteries. Vessels from both these sources anastomose extensively within the nasal cavity, including the lateral wall, the septum and also across the midline. The Kiesselbach plexus, or Little's area, is the most frequently associated anastomotic site for epistaxis and is located on the anterior cartilaginous septum. Woodruff's plexus is a confluence of vessels on the lateral wall posterior to the inferior turbinate and is often implicated in 'posterior' bleeds.

The external carotid artery's contribution to the nasal cavity is via the facial and maxillary arteries. The superior labial artery is one of the terminal branches of the facial artery, and it supplies the anterior nasal septum. The lateral nasal artery and ascending palatine artery are two further branches that supply the nasal vestibule and a small area of the nasal cavity. The maxillary artery supplies the nose via the greater palatine branch to the anterior nasal floor and septum. The sphenopalatine branch of the maxillary artery is the most recognized and is a significant contributor to the vascular supply of the nasal cavity. It enters the nasal cavity through the sphenopalatine foramen and then divides into

posterior, lateral and septal branches. The lateral branches give rise to the arteries for the middle and inferior turbinates whilst the septal branch supplies the posterior septum and then terminates in the Kiesselbach plexus anteriorly on the septum.

The internal carotid artery contributes to the nasal vasculature through both the anterior and posterior ethmoidal branches of the ophthalmic artery. The larger anterior ethmoidal artery and the smaller posterior one run below and above the superior oblique muscle, respectively, and exit the orbit through their named foramina. They both cross the ethmoid roof and enter the nasal cavity through the cribiform plate and then divide into branches to supply the lateral nasal wall and nasal septum.

EPIDEMIOLOGY

Epistaxis can occur at any age, but there is a notable bimodal age distribution with peaks in childhood and then in the more elderly population, between the ages of 60 and 80. It is rarely seen in infants and tends to be less common in early adulthood. There is a slight male preponderance (male:female ratio, 55:45).

CLASSIFICATION

Epistaxis can be classified in several ways. The pronounced age distribution, for example, lends itself to the classification of adult and childhood epistaxis. Descriptive parameters can also be used for classification such as recurrent or acute/severe epistaxis. The anatomical site of bleeding has also been proposed as a basis for classification into anterior and posterior epistaxis. This system, however, requires a consistent definition of the terms *anterior* and *posterior* epistaxis for it to be universally applicable, whereas in practice, these terms vary from author to author.

Perhaps the most useful classification is by aetiology, but rather than employing an exhaustive list, a broader division into primary and secondary epistaxis can be used. This has the advantage of guiding management, which can be quite different between the two groups.

AETIOLOGY

The majority of epistaxis cases (80 per cent), across the age groups, will be primary or idiopathic (Table 3.1). It is quite possible that with

Table 3.1 Table of aetiological factors and associations

Aetiological factors	
Idiopathic	(80%)
Trauma	Digital, acute facial/nasal, iatrogenic
Coagulopathy	Idiopathic thrombocytopaenia (ITP) Disseminated intravascular coagulopathy (DIC)
Drugs	Warfarin, aspirin, clopidogrel, nonsteroidal anti-inflammatories (NSAIDs)
Chronic granulomatous disease	Wegener's, sarcoidosis
Neoplastic	Angiofibroma, inverted papilloma, squamous cell cancer
Hereditary	Hereditary haemorrhagic telangiectasia (HHT), haemophilia, Von Willebrand's factor deficiency
Aetiological associations	
Septal abnormalities	Spurs, perforations
Alcohol	
Hypertension	

advancement in our knowledge of the aetiological factors involved, this percentage will decrease. Minor trauma may be implicated but not identified in many cases that are classified as idiopathic. Currently, only 20 per cent of cases will be classified as secondary epistaxis with a determined local or systemic underlying cause. Significant physical trauma, either acute maxillofacial or surgical injuries, is an important cause of secondary epistaxis. The majority of other cases of secondary epistaxis are due to underlying coagulopathy induced by either medication or systemic disease (haematological disorders, liver disease).

In a case of epistaxis it is important to thoroughly examine the nose. Such a nasal examination locates the source of bleeding and helps exclude sinister causes such as benign or malignant tumours and granulomatous conditions. Juvenile angiofibroma of the postnasal space should be considered in cases of unilateral epistaxis in the young/adolescent male population. Nasal endoscopy is required for a competent and complete examination of the nose.

Hereditary haemorrhagic telangiectasia (or Osler-Rendu-Weber disease) is an important hereditary cause of epistaxis and is discussed at the end of this chapter.

AETIOLOGICAL ASSOCIATIONS

Septal deviations and spurs may disrupt the normal nasal airflow, leading to dryness and epistaxis. The bleeding site is usually located anterior to the septal spur. The margin of a septal perforation may not be covered by epithelium. This lack of epithelium promotes granulation and crusting and is a common source of epistaxis. Alcohol will cause prolongation of bleeding time, although platelet counts and coagulation factor activity can be recorded as normal. Epistaxis patients are more likely to consume alcohol than matched control patients and, also, more likely to have consumed alcohol within 24 hours of admission for epistaxis.

The relationship between hypertension and epistaxis is often misunderstood. Patients with epistaxis commonly present with an elevated blood pressure. Epistaxis is more common in hypertensive patients, perhaps owing to vascular fragility from long-standing disease. Hypertension, however, is rarely a direct cause of epistaxis. More commonly, epistaxis and the associated anxiety cause an acute elevation of blood pressure.

Children with migraine headaches have a higher incidence of recurrent epistaxis than children without the disease. The Kiesselbach plexus, which is part of the trigeminovascular system, has been implicated in the pathogenesis of migraine.

MANAGEMENT

PRIMARY EPISTAXIS

Effective management of this condition should follow a logical sequence from simple first-aid measures and resuscitation to targeting and treating the bleeding point (Figure 3.1).

Simple first-aid measures should always be employed first because often the bleeding point is anterior and, therefore, amenable to such techniques. The soft cartilaginous portion of the nose should be pinched continuously for 10 minutes to exert pressure internally on the anterior nasal septum. The patient should sit down with head hanging forward breathing through an open mouth. If this is unsuccessful some cotton wool soaked in xylometazoline (Otrivine) can be inserted gently into the anterior nose and the 10-minute compression technique repeated. This correct technique should be reiterated to all patients seeking medical attention for epistaxis.

Any patient requiring further medical attention for epistaxis should be resuscitated appropriately after establishing intravenous access. Initial investigations should include a full blood count, coagulation screen, and group and save sample. A detailed and targeted history should be taken to elucidate the pattern and severity of bleeding and to take into account any underlying aetiological factors.

Direct therapies

The effective management of epistaxis relies on treating the underlying bleeding point, and key to this is close and careful examination with the

appropriate equipment following adequate preparation of the nose (Figure 3.2). A combined local anaesthetic/vasoconstrictor solution should be applied topically to the nose and anterior rhinoscopy performed with a headlight and Thudichum's speculum. The nasal cavity should be cleared of all clots and then inspected with a view to identifying an anterior bleeding point. Chemical cautery with silver nitrate sticks or electrocautery with bipolar diathermy can then be applied to coagulate the appropriate vessel. If an anterior source is not visualized, rigid nasendoscopy should then be performed to try to identify a bleeding point more posteriorly in the nasal cavity. Even following successful identification of anterior bleeding points, nasendoscopy should be performed to exclude other posterior sources or underlying sinister pathology. Electrocautery rather than chemical cautery should ideally be used to coagulate these more posterior bleeding points to avoid inadvertent cauterization of normal nasal anatomy. Cautery by either method should always be performed cautiously and with precision to avoid complications such as chemical burns to the lips/nares or longer-term septal necrosis/perforation. Occasionally, a small localized pack (e.g. sinus pack) or an absorbable haemostatic agent such as Surgicel can be used to apply direct pressure to an identified bleeding point that is not amenable to cautery.

Indirect therapies

If after adequate inspection of the nasal cavity a bleeding point is not identified, then indirect therapies are employed to control the epistaxis. Nasal packs are the most commonly employed indirect techniques, but others such as hot water irrigation and anti-fibrinolytics are occasionally indicated. There are a variety of anterior nasal packs available; these include ribbon gauze, nasal tampons (Merocel) and balloon catheters (Brighton). Posterior packs may be required if bleeding persists despite anterior packing. In an awake patient this is most easily achieved with a Foley catheter advanced nasally past the posterior choanae of the nasal cavity in the nasopharynx. Formal posterior packing requires a general anaesthetic.

Once in place, nasal packing should be left i*n situ* for 24–48 hours; in patients with prosthetic heart valves, antibiotic cover should be provided. Local complications of packs include sinusitis, septal perforation and alar necrosis.

More recently, topical haemostatic compounds have been proposed as an alternative, indirect treatment option for epistaxis. Floseal, a compound consisting of gelatin granules and human thrombin, is such an agent, and it has been shown to be effective in cases refractory to packing. These compounds are relatively easy to use and are associated with low morbidity. They are, however, much more costly when compared to simple chemical cautery, although they become much more cost effective when compared to surgical intervention.

Surgical management

Surgical intervention is required if indirect therapies fail to control the bleeding. There are several procedures that can be performed under a general anaesthetic in the management of refractory epistaxis, and they can be either facilitatory or more targeted. A general anaesthetic may be required to perform adequate packing of the nose, to apply anterior packing in an uncooperative patient or to insert formal posterior packing. In the uncooperative individual, a general anaesthetic may allow adequate endoscopic inspection and effective nasal cautery. Correction of a deviated nasal septum, under general anaesthesia, can facilitate good access to the bleeding site and permit cautery, effective packing or surgical arterial ligation.

In recent times, surgery has evolved in epistaxis management towards more formal arterial ligation. Arterial ligation should be performed at the most distal (nasal) point with a progression to more proximal ligation if the initial procedure is unsuccessful. Endoscopic sphenopalatine artery ligation is, therefore, the most commonly employed procedure. Internal maxillary artery and, thereafter, external carotid artery ligation are infrequently used. Anterior ethmoidal artery ligation is employed in cases of traumatic epistaxis (particularly nasal ethmoid fracture) or as an adjunct in the management of refractory epistaxis in combination with a sphenopalatine or internal maxillary artery ligation.

Embolization

Arterial embolization has a greater than 80 per cent success rate for severe epistaxis, but its usage depends on the availability and experience of an interventional radiologist. Arterial embolization is indicated either after failure of ligation techniques or in patients who are assessed to be unsuitable or for whom general anaesthetic carries a high risk. Transfemoral angiography is used to demonstrate the bleeding point with subsequent selective embolization of the maxillary or facial arteries performed with materials such as Gelfoam or microcoils. Complications from this technique, although relatively unusual, are more common than following surgical ligation and include facial skin necrosis, paraesthesia, cerebrovascular accident and groin haematoma.

RECURRENT EPISTAXIS

Nonsevere recurrent (primary) epistaxis is commonly seen in the paediatric population. In the adult population, recurrent epistaxis can be primary but is often secondary; therefore, a detailed history is vital to elucidate any causative factors. Examination in adults should always include nasendoscopy to fully visualize any potential bleeding points but also to rule out sinister underlying pathology. Any identified bleeding point can be treated with cautery, and areas of vestibulitis are treated with topical antiseptic creams. The efficacy of chlorhexidine and neomycin creams (Naseptin) is well established as an important part of treatment in paediatric recurrent epistaxis. These chlorhexidine and neomycin creams, together with instillation of Vaseline, are commonly used

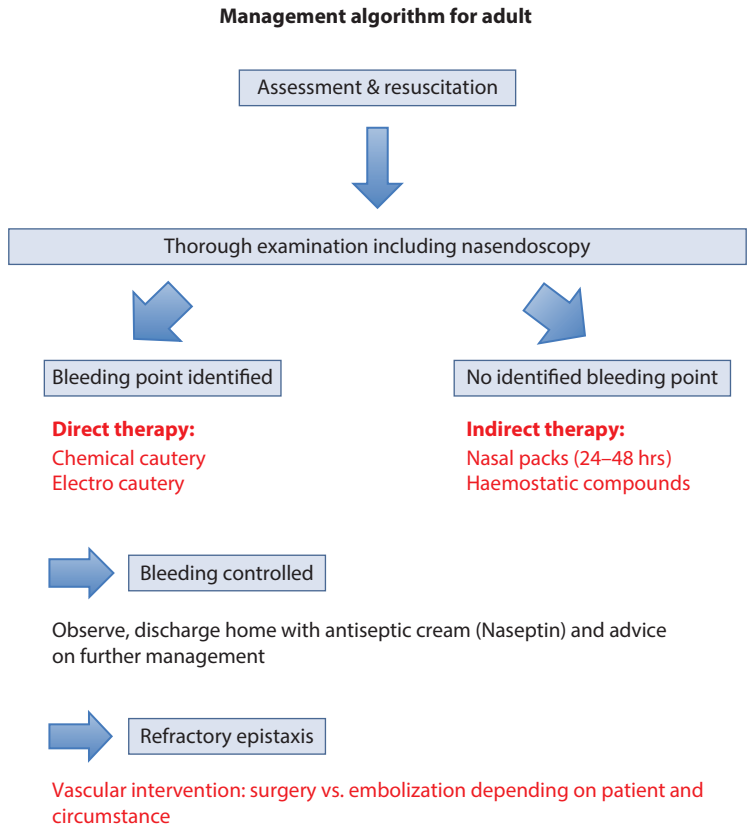

Figure 3.1 Management algorithm for adult acute idiopathic epistaxis.

in the adult population to reduce the incidence of recurrent epistaxis. In all consultations for recurrent epistaxis, it is worthwhile to reiterate the simple first-aid measures that should be employed to control each new episode.

SECONDARY EPISTAXIS

Coagulopathies

In coagulopathic cases, indirect therapies may be indicated in preference to direct measures to control the bleeding. The reason for this course of action is that the bleeding can arise from several sites and instrumentation can lead to further mucosal trauma and exacerbate the bleeding. In these coagulopathic cases haemostatic compounds and nasal packs form the mainstay of treatment, in addition to close collaboration with the haematology services to guide anticoagulant therapy or reversal. If antiplatelet therapies are implicated as a cause of epistaxis, there can be benefit in temporary cessation of this medication. In a patient using warfarin tablets, the international normalized ratio (INR) needs to be considered when managing the epistaxis. If the bleeding is easily controlled with nasal packs, then the warfarin dose may be continued. A warfarin dose can be omitted if the INR is above the

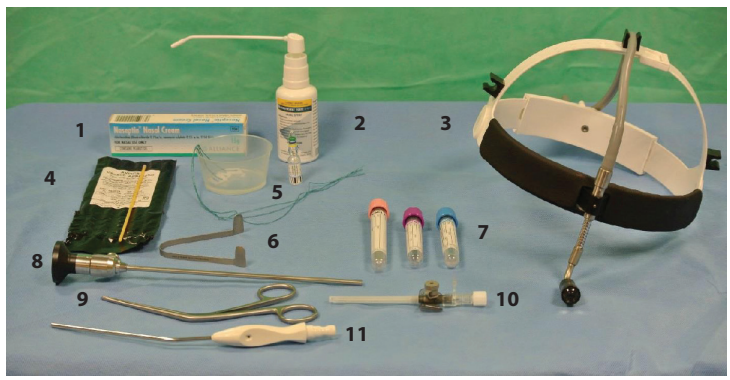

Figure 3.2 Epistaxis management: equipment and medicines.

1. Nasal cream (Naseptin).
2. Topical anaesthetic + decongestant (Co-phenylcaine).
3. Head light.
4. Silver nitrate cautery sticks.
5. Adrenaline vial + patties for application.
6. Nasal speculum.
7. Blood bottles: full blood count, coagulation, group and save.
8. Rigid nasal endoscope.
9. Tilley's dressing forceps.
10. Large bore cannula.
11. Suction.

Cautery principles: examination and silver nitrate cautery steps.

Suction clots.
Apply topical local anaesthetic/vasoconstrictor (topical spray or cotton pledget).
Identify bleeding point; anterior rhinoscopy and nasendoscopy.
Apply silver nitrate cautery (no more than 5 seconds).

therapeutic range. Unfortunately, if the bleeding is not controlled, then intravenous vitamin K can be administered to reverse the anticoagulant effect of warfarin. The INR will require close monitoring in this often difficult time for the patient.

Septal perforation

Crusting of the margin of a septal perforation should be treated with chlorhexidine and neomycin creams (Nasetin), followed by regular instillation of Vaseline. If bleeding persists despite simple measures, then options include (1) cautery of granulations and bleeding spot, (2) insertion of septal button to allow re-epithelialization of the margin or (3) surgery to the margin to trim back bare/granular cartilage and re-epithelialize the margin with a local flap.

Trauma

Epistaxis following trauma or surgery may require nasal packing but may also be amenable to direct electrocautery or arterial ligation techniques.

HEREDITARY HAEMORRHAGIC TELANGECTASIA

Hereditary haemorrhagic telangectasia (HHT) is an autosomal dominant condition associated with recurrent bleeding from vascular anomalies. The disease affects vessels – ranging from capillaries to arteries in the skin – mucous membranes and viscera, leading to the formation of telangiectasia, arteriovenous malformations and aneurysms. Although the disease is almost universally associated with recurrent epistaxis, the severity can be extremely variable. The need for blood transfusion can help to quantify the severity of HHT, which in turn can be a guide to the management of this condition. Milder cases may be simply and effectively managed with topical emollients and oestrogens with interval laser photocoagulation as required. The more severe cases warrant consideration of other measures such as septodermoplasty, arterial ligation and arterial embolization. In the most severe cases (those necessitating significant volumes of blood transfusion), an obturator or surgical closure of the nostrils (Young's procedure) should be considered. Surgical closure of the nostrils is effective in reducing the need for blood transfusion, but the patients find the loss of the sense of smell and taste a difficult side effect of this procedure.

KEY LEARNING POINTS

- Epistaxis is a common ENT emergency.
- Primary idiopathic epistaxis should be managed using a stepwise algorithm.
- Definitive treatment of primary epistaxis involves therapy directed towards an identified bleeding point.
- Refractory epistaxis may require surgery or embolization to control bleeding.
- Recurrent idiopathic epistaxis, commonly occurring in children, can be more successfully treated with topical antiseptic creams and rarely requires surgery.
- Secondary epistaxis management requires treatment of the underlying cause.

4

Acute rhinosinusitis and its complications

ANDREW C SWIFT AND ADAM J DONNE

DEFINITIONS OF ACUTE RHINOSINUSITIS

Rhinosinusitis is specifically defined in the current European Position Paper on Sinusitis and Nasal Polyps 2012 (EPOS 2012).[1] Because rhinitis and sinusitis usually coexist, the correct and accepted terminology is now rhinosinusitis. The more stringent definition includes symptoms as well as endoscopic or computed tomography (CT) findings. However, the criteria have to be adaptable to wider groups beyond otorhinolaryngologists.

The following features are necessary for a diagnosis of acute rhinosinusitis (ARS) in adults to be acceptable for epidemiological/primary care purposes where there is no ear, nose and throat (ENT) examination or imaging:

- Sudden onset of two or more symptoms, one of which must include nasal blockage/obstruction/congestion, or anterior nasal discharge/posterior nasal drip
- + Facial pain/pressure
- + Reduction of loss of smell for less than 12 weeks

This differs in children in that they should have sudden onset of two or more of the following symptoms:

- Nasal blockage/obstruction/congestion
- Or discoloured discharge
- Or cough (daytime and nighttime)

Children with adenoiditis or adenoidal enlargement/hypertrophy can have identical symptoms, but these are normally long-term and in excess of 12 weeks once seen in an ENT clinic.

Most acute episodes occur during the common cold: The duration of symptoms for ARS during

the common cold/acute viral rhinosinusitis should be less than 10 days.

Extension of symptoms beyond 10 days is defined now as post-viral ARS; a small percentage of such patients will develop acute bacterial rhinosinusitis (ABRS). The criteria for ABRS require at least three of the following symptoms/signs:

- Discoloured mucus, predominantly from one side, with mucopus within the nasal cavity
- Severe local pain, mainly on one side
- Pyrexia >38°C
- Elevated C-reactive protein (CRP)/erythrocyte sedimentation rate (ESR)
- 'Double sickening', referring to deterioration after an initial milder phase of illness

THE COMMON COLD

Most people will have experienced ARS during the course of a viral upper respiratory tract infection or common cold. The common cold is in fact the most common infectious disease known to man: Adults are likely to suffer from two to five episodes of ARS per year and children seven to 10 colds per year. The prevalence of ARS is high, affecting 6–15 per cent of the population, and increases in winter months, climatic variations, damp environments and air pollution. There is good evidence to support the hypothesis that allergic inflammation and cigarette smoke are predisposing factors (EPOS 2012).

The common cold is caused by airborne viruses, mainly rhinoviruses and coronaviruses. However, other viruses include adenovirus, influenza and parainfluenza viruses, respiratory syncytial virus and enterovirus. The viruses attach to the host cells: Normal ciliary action is disrupted, the mucosa becomes very congested and mucus glands over-secrete. The mucosal congestion extends to the paranasal sinuses, and sinus ostia are likely to become blocked.

The familiar features include nasal obstruction, a mucous nasal discharge and a sense of pressure or heaviness over the cheeks. This is one condition where self-diagnosis is reasonable because we are all familiar with the symptoms and virology is not feasible. The symptoms generally start to subside after a few days, but it may take 2 to 3 weeks to resolve. Investigations such as plain radiographs and culture swabs are normally unnecessary.

The first-line treatment is generally to await natural resolution and alleviate symptoms with medication when necessary. Oral analgesics will alleviate facial discomfort; nasal obstruction is relieved by topical and/or systemic decongestants; and excess mucus discharge is treated with saline rinses or sprays.

Antibiotics are generally unnecessary, and recent Cochrane research has shown that they offer a minimal advantage and are more likely to induce a number of unwanted side effects such as nausea and vomiting, abdominal pain, diarrhoea and the risk of bacterial resistance.[1]

Topical steroid sprays have been shown to aid resolution of symptoms in moderate and severe episodes of viral ARS and in bacterial ARS in combination with antibiotics.

PURULENT RHINOSINUSITIS

Undoubtedly, some episodes of acute viral rhinosinusitis will progress to acute bacterial sinusitis. This has been estimated to occur in 0.5–2.0 per cent of patients.

The clinical features include a purulent nasal discharge and worsening of facial discomfort, often accompanied by dental neuralgia, general malaise and pyrexia.

The usual bacteria are *Streptococcus pneumonia* and *pyogenes*, *Haemophilus influenza*, *Moraxella catarrhalis* and *Staphyloccocus aureus*. Although patients with immune deficiency may well develop acute purulent bacterial sinusitis, often with more unusual organisms such as pseudomonas, there is normally an underlying chronic sinusitis.

The pathogenesis of bacterial infection has been studied for the maxillary sinus. Ostial obstruction leads to the development of low-pressure mucosal congestion and a hypoxic environment within the maxillary sinus, thus favouring bacterial proliferation and increasing ciliary disruption.

Antibiotics are certainly justified in this scenario, but resolution will often progress at the same rate even without antibiotics (EPOS 2012).[1] Amoxicillin or erythromycin is a suitable choice, but there is a lack of evidence showing a significant difference between short 5-day courses and longer

courses of a week or more. Although amoxicillin is the most commonly prescribed antibiotic, bacterial resistance, particularly to *S. pneumonia* and *M. catarrhalis,* is increasing.

ACUTE RHINOSINUSITIS IN CHILDREN

Most ARS cases in children are viral and self limiting. Some understanding of sinus development is helpful in understanding the management of children with severe infection and complications of ARS.

The ethmoid and maxillary sinuses are present in small children: The frontal sinuses only begin to enlarge from the age of 4 years, and growth continues into the late teens. After the twelfth year, the floor of the maxillary sinus descends below the level of the nasal floor as the secondary dentition erupts. The sphenoid is very small at birth but has pneumatized in 85 per cent of children by the age of 8 years and continues to grow until the age of 15 years, although further aeration may occur into adulthood.

In children with ARS, it is important to exclude a foreign body and unilateral choanal atresia, and, in recurrent infection, underlying CRS and adenoiditis may be difficult to differentiate. Children with cystic fibrosis may present with exacerbations of acute sinusitis but normally have an underlying chronic sinus disorder. Examination may include rhinoscopy with an otoscope and large-diameter speculum, headlight or endoscope, according to preference and availability. A CT sinus scan is occasionally required should there be prolongation of symptoms or a suspected complication.

The main bacteria that have been implicated are *S. pneumonia, H influenza, M. catarrhalis, S. pyogenes*, and anaerobes. Although resolution will occur without antibiotics, it is likely to be faster with an antibiotic, but the advantage is modest. Antibiotics should, however, be prescribed for children with a complication of ARS, or those with concomitant chest disease such as asthma or bronchitis. There is evidence to support adding an intranasal steroid, although compliance will be an issue with younger children. There is a lack of evidence to support the use of topical and systemic decongestants, antihistamines and saline irrigation.

RECURRENT ACUTE RHINOSINUSITIS

Some patients are prone to developing recurrent episodes of ARS. It is important to differentiate these patients from those with chronic rhinosinusitis with recurrent exacerbations of their condition. By definition, patients with recurrent acute sinusitis are asymptomatic between episodes. Such episodes can become clinically significant, owing to either their severity or their frequency, and lead to frequent courses of antibiotics.

The assessment of such patients should include a search for predisposing factors such as allergic rhinitis or exposure to nasal irritants. Underlying dental disease should also be considered. Typically, nasal endoscopy shows narrow nasal cavities, especially in the region of the middle meati, and a CT scan of the sinuses normally shows osteomeatal complex obstruction with generally clear paranasal sinuses or mild mucosal thickening of the maxillary sinuses.

Daily application of a topical nasal steroid, either as a spray or sometimes as nasal drops in a head-down position, is often an effective strategy for preventing recurrent infection. If medical management fails and the CT scan confirms osteomeatal complex obstruction, endoscopic sinus surgery to open the middle meatal drainage channels is often beneficial. However, the latter can be challenging and must not be underestimated owing to the technical difficulties of operating in a narrow nose.

COMPLICATIONS OF ACUTE RHINOSINUSITIS

Fortunately, most cases of ARS resolve without the development of complications. The incidence of serious complications is low, but they are more likely in the winter months. It is also of interest to note that oral antibiotics do not necessarily protect patients from developing complications.

Acute infective complications can arise from acute or chronic rhinosinusitis: The former is more common in children and the latter in adults. Complications may affect the orbit (60–75 per cent),

the intracranial tissues (15–20 per cent) or the frontal bone (5–10 per cent). In a recent UK survey, the orbit was most commonly affected (76 per cent of 78 patients), followed by intracranial infection (9 per cent) and bone infection (5 per cent).[3] Occasionally, multiple sites can be affected simultaneously.

ORBITAL COMPLICATIONS

Orbital infection secondary to sinusitis is often referred to as periorbital cellulitis, but this term lacks precision. Orbital infections have been classified into five subgroups of worsening severity.[4] Although this classification is still in common use today, it is not strictly true, and the increased escalation of infection is not necessarily sequential. However, the classification does provide a clinical aide de memoir to the possible complications that may arise with severe infection (Table 4.1).

Infections of the orbit are initially contained by the fibrous sheet of the orbital septum, and it is much more helpful to refer to preseptal cellulitis and orbital cellulitis according to whether infection is anterior or posterior to this septum.

Infection is most likely to spread from the ethmoid sinus, by direct or vascular spread across the thin lamina papyracea, although some may arise from the maxillary, the frontal or, rarely, the sphenoid sinus.

Any age group can be affected, but the incidence is much greater in children. Most cases in children are preseptal cellulitis that is relatively mild, may not be referred for an ENT opinion and may occur in the absence of sinusitis: 60–90 per cent are reported to be secondary to acute rhinosinusitis.[5] In contrast, orbital cellulitis is always secondary to acute rhinosinusitis.

Table 4.1 Complications of periorbital cellulitis as modified and described by Chandler et al.[4]

Preseptal cellulitis
Orbital cellulitis
Subperiosteal abscess
Orbital abscess
Cavernous thrombosis

Clinical features

PRESEPTAL CELLULITIS

Preseptal cellulitis presents with unilateral swelling of the eyelids, erythema, local pain and sometimes pyrexia (Figure 4.1a). There should be no proptosis and no limitation of eye movement.

Identifying clinical features in the acute stage of infection may be challenging, particularly in children, and tender, swollen eyelids may cause significant difficulty in performing a proper examination of the eye and globe. Normally, a scan is unnecessary unless there is diagnostic uncertainty or clinical progression despite adequate treatment (Figure 4.1b).

It is important to be aware that infection can progress rapidly, and spread beyond the orbital septum could lead to abscess formation and threaten vision.

ORBITAL CELLULITIS

The following are important clinical features that should be identified, documented and monitored: conjunctival oedema (chemosis), limitation of eye movement (ophthalmoplegia), painful eye movements, proptosis, pupillary reaction, visual acuity and colour vision. Colour vision as assessed by an Ishihara chart is typically impaired first, affecting particularly red colour perception, and any element of doubt should instigate an urgent opinion from an ophthalmologist. In the pre-antibiotic era, blindness following periorbital cellulitis was not uncommon, the possible causes being retinal artery occlusion, compression or inflammation of the optic nerve, panophthalmitis and corneal ulceration.

It is of paramount importance to determine whether the infection is complicated by the development of a subperiosteal abscess. These abscesses typically arise adjacent to the lamina papyracea, although some extend or arise superiorly beneath the thin floor of the frontal sinus. Displacement of the globe, proptosis and ophthalmoplegia can all arise from orbital cellulitis or a subperiosteal or orbital abscess. However, it can be very difficult to fully examine the eye in a sick, unwell, uncooperative child, and the presence of painful eye movements, chemosis and/or proptosis should instigate an urgent CT scan.

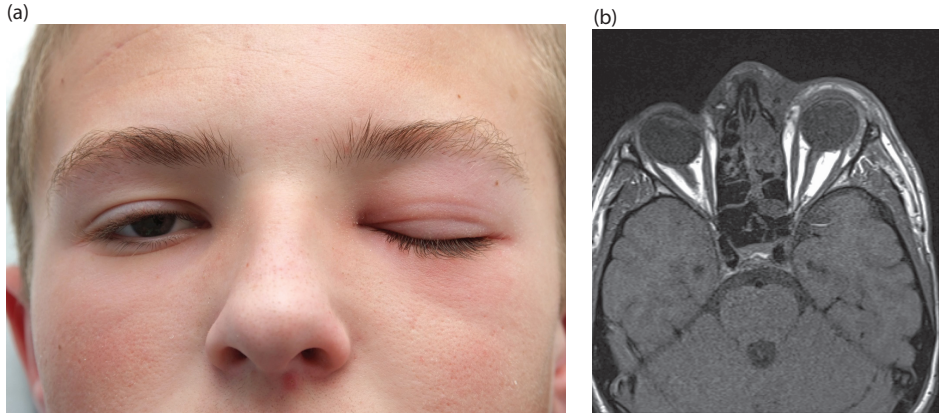

Figure 4.1 **(a)** Preseptal cellulitis in a 16-year-old male. **(b)** Axial CT scan showing ethmoid opacity and preseptal opacity in soft tissues.

Initial management

An early opinion from an ophthalmologist is wise at this stage.

Although oral antibiotics may be effective for the early stages of periorbital cellulitis, intravenous antibiotics are essential for patients with significant infection. It is generally not possible to determine the bacterial profile at the time of instigating treatment, but prior to starting an antibiotic, a blood culture should be considered, particularly if the patient is pyrexial or has rigors. Similar to acute bacterial sinusitis, the spectrum of cover should include the likely pathogens such as *Streptococcus spp.*, *Pneumococcus spp.* and *Haemophilus influenza*. It should be noted that anaerobic bacteria may cause severe infection, and the antibiotic profile should cover these virulent organisms. Early advice from a microbiologist should be sought.

Supplementary treatment with decongestant nasal drops is often practised, but there is no evidence base to support this.

Imaging

A scan is not essential in patients with preseptal cellulitis unless the condition worsens despite treatment with intravenous antibiotics.

If a subperiosteal abscess is suspected, an urgent CT scan of the sinuses and orbits should be obtained (Figure 4.2a and b). It is wise to include brain settings with this request to avoid missing a coexisting intracranial complication.

A magnetic resonance image (MRI) will provide supplementary information if there is diagnostic uncertainty, a subperiosteal abscess or suspected intracranial infection (Figure 4.3a and b).

The specific features to note on scans in patients with orbital infection include a subperiosteal abscess, oedema or bowing of the medial rectus muscle, displacement of the globe, loss of clarity of posterior extraocular muscles and optic nerve and orbital abscess formation. Gas bubbles in the soft tissues develop in anaerobic infection owing to gas-producing bacteria (Figure 4.4).

Surgical intervention

Surgery is indicated in patients who fail to improve on intravenous antibiotics after 24–48 hours, or in those with a subperiosteal or an orbital abscess. A small or early subperiosteal abscess may respond to non-operative treatment, but if this strategy is chosen, the response to treatment should be closely monitored and abscess drainage instigated if the patient fails to respond.

The standard way to explore and drain a subperiosteal abscess is by an external incision, but in the day of the endoscope, there is much discussion about endoscopic drainage. However, the latter approach is likely to be challenging in the acute

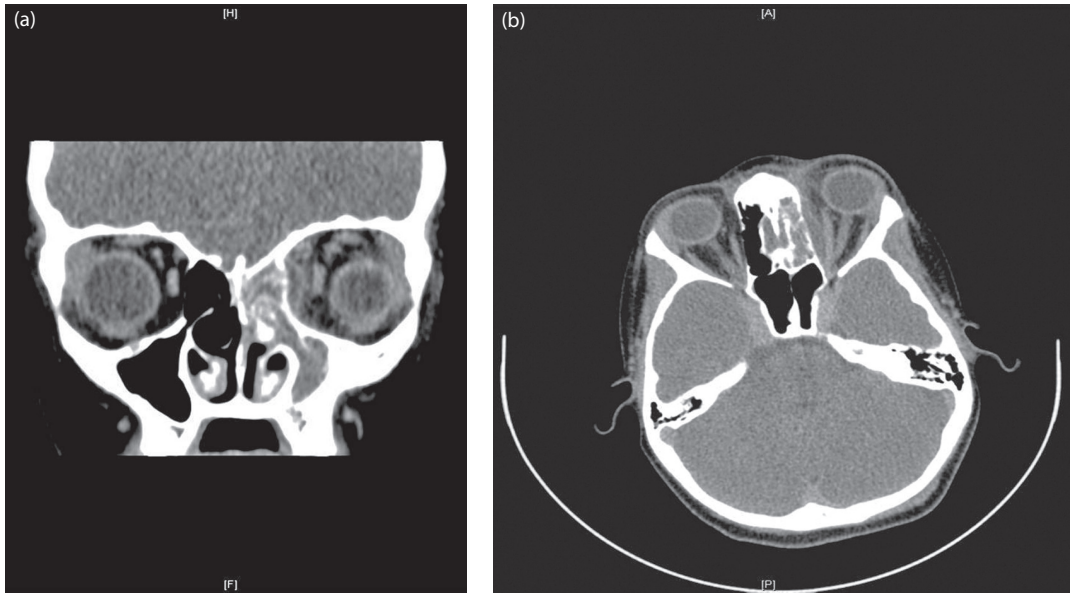

Figure 4.2 CT scan showing a left subperiosteal abscess secondary to acute sinusitis. Coronal image **(a)**, axial image **(b)**.

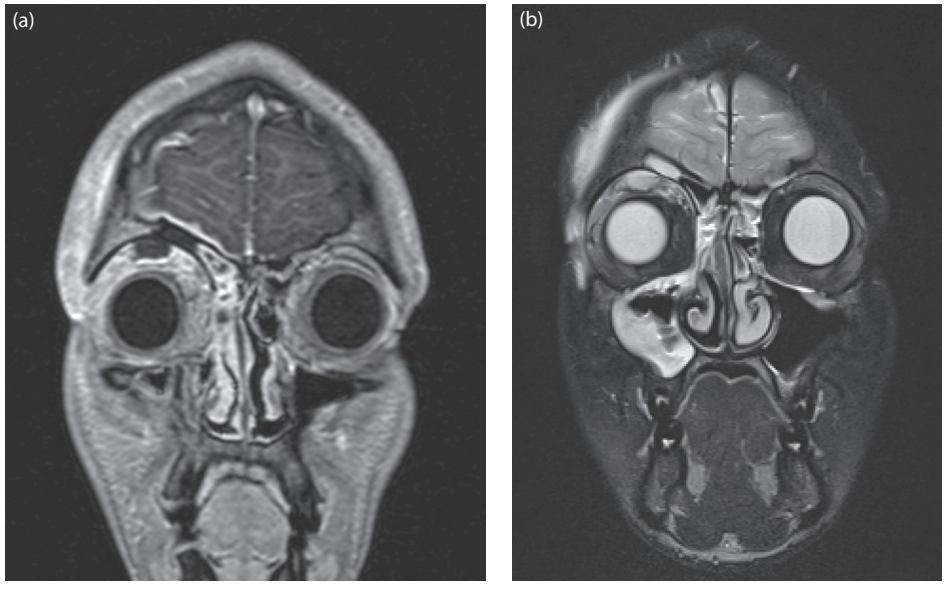

Figure 4.3 MR scan of a superior periorbital abscess with frontal sinusitis and dural inflammation secondary to localized encephalitis. T1 image **(a)**, T2 image **(b)**.

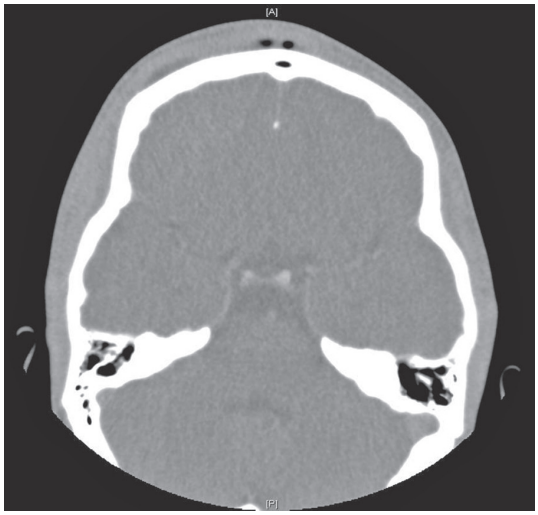

Figure 4.4 Axial CT scan showing gas bubbles in soft tissues in a child with periorbital cellulitis and frontal sinusitis.

situation, particularly in young children, and has not gained popularity within the United Kingdom. External drainage facilitates complete drainage of multilocular abscesses and access to an abscess sited superiorly above the globe (Figure 4.3a and b).

Orbital exploration should be accompanied by drainage of pus within the sinuses, by maxillary antral washout or frontal trephine. Simultaneous endoscopic drainage of the anterior ethmoid should also be considered.

INTRACRANIAL INFECTION

Fortunately, intracranial infection secondary to acute sinusitis is unusual, but the consequences are potentially very serious and include long-term morbidity: Death is reported in up to 19 per cent of cases. In contrast to otogenic intracranial infections that often develop in patients with chronic ear disease, rhinogenic brain abscesses are much more likely to occur during an episode of acute sinusitis.[6]

Infection may reach the intracranial cavity by haematogenous or direct spread. Intracranial complications of acute sinusitis include meningitis, cerebritis, extradural abscess, subdural empyema, cerebral abscess and superior sagittal and cavernous thrombosis.

Because intracranial infection is typically rapidly progressive, early recognition, rapid referral and effective treatment are essential to promote the likelihood of successful recovery.

The clinical features that should alert the clinician are an unwell patient with a persistent headache, altered level of consciousness, spiking pyrexia, nausea and vomiting. Other clinical features may include neck stiffness, photophobia, confusion, seizures, limb dysfunction, cranial nerve abnormalities and dysphasia.

MENINGITIS

Meningitis presents as a rapidly progressive illness in an acutely unwell patient who develops an incessant headache, photophobia and neck stiffness. Straight leg-raising may induce pain – Kernig's sign. The diagnosis is confirmed by performing a lumbar puncture and sending the aspirate for cellular analysis and culture. Lumbar puncture should be done only after exclusion of an intracranial abscess owing to the risk of coning.

Perhaps the most important consideration to remember in such patients is to look for a defect in the bony anterior skull base, particularly if there is a history of previous or recurrent meningitis. Significant head injuries with associated fractures of the skull or facial bones from many years ago may heal with areas of dehiscent bone and exposed dura that predispose to the development of meningitis.

INTRACRANIAL PUS

Most rhinogenic brain abscesses are sited in the cerebrum or frontal lobes, and the clinical features are determined by the site and stage of abscess formation. It is important to appreciate that some patients will have relatively few symptoms or the abscess may be silent, particularly if symptoms have been masked by the previous use of antibiotics. Eventually, meningeal irritation, raised intracranial pressure and focal deficits may occur.

In a subdural empyema, areas of the brain surface are covered by pus. The characteristics are a rapidly progressive, dramatic clinical presentation

in a very ill patient. Focal features will depend on which parts of the brain are covered at any one time. Haematogenous spread of infection may result in numerous intracranial sites of pus accumulation.

An epidural abscess is normally associated with frontal sinusitis and may not be recognized until shown by a CT/MRI scan of the head (Figure 4.5a and b).

Imaging

Once the possibility of intracranial infection is considered, an urgent scan should be requested. A CT of the head with brain settings and contrast is essential: It will be helpful to include simultaneous sinus images as well (Figure 4.3a). An MRI of the head will provide important supplementary information (Figure 4.3b). Once the diagnosis has been confirmed, urgent referral to a neurosurgeon is imperative.

Management

The management priority once an intracranial infection is recognized is to seek advice from a neurosurgeon. Treatment options include long-term high-dose intravenous antibiotics, burr-hole drainage, craniotomy or image-guided aspiration.

Otorhinolaryngological intervention is based on the principle of drainage of pus and eradication of the source of infection and may include antral lavage and frontal trephine if there is frontal infection. An alternative is to open the anterior ethmoid cells with the option of draining the frontal sinus endoscopically, expertise allowing, although bleeding is likely to be an issue in the acute stage. Frontal sinus ostial drainage may be facilitated by balloon dilatation if the equipment is available. These drainage procedures can often be combined with neurosurgical drainage during the same anaesthetic administration.

THROMBOPHLEBITIS

Superior sagittal vein thrombosis

This rare complication is likely to arise with epidural or subdural sepsis. The characteristics include a severe headache and focal deficits, especially affecting the legs, because venous thrombosis spreads

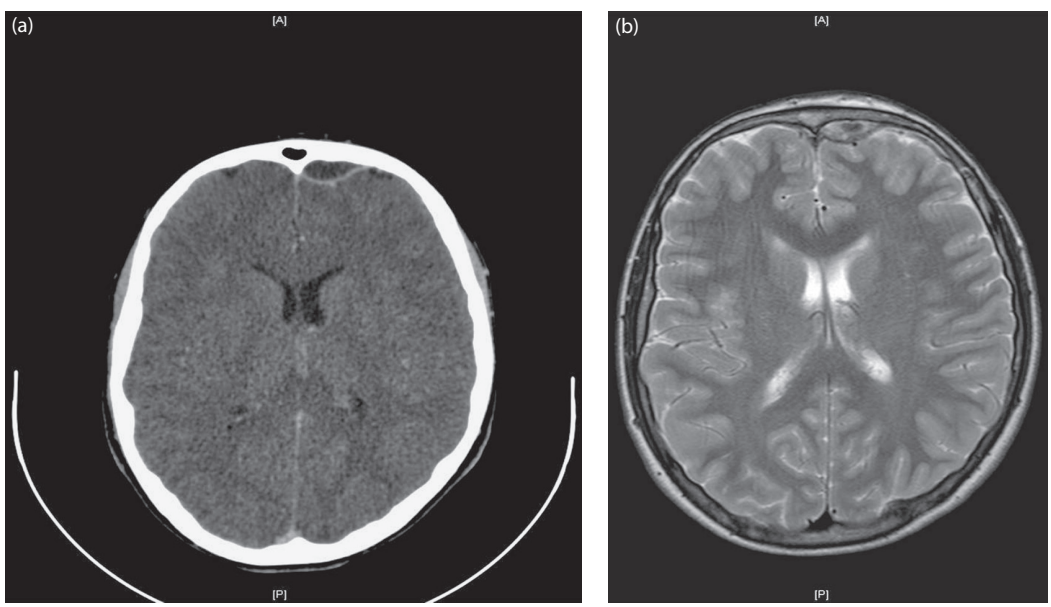

Figure 4.5 Axial images showing a left frontal epidural abscess secondary to acute sinusitis with periorbital cellulitis. CT image **(a)**, T1 MR image **(b)**.

and leads to cerebral infarction. Progression leads to raised intracranial pressure and coma.

Cavernous sinus thrombosis

Cavernous sinus thrombosis is fortunately a rare complication of acute sinusitis but is associated with a significant risk of death. It usually arises from extension of purulent sphenoid sinus infection and adjacent intracranial vein thrombosis. Cavernous sinus thrombosis secondary to acute sinusitis is associated with rapid onset of symptoms, beginning with swelling of one eye, to be followed by the other as the thrombosis progresses to the contralateral side. The clinical features include a spiking pyrexia, ptosis, deep retro-orbital pain, papilloedema and complete ophthalmoplegia due to involvement of the III, IV and VI cranial nerves. There may be corneal hypersensitivity of the affected eye to be followed by anaesthesia due to disruption of trigeminal sensation. Septic emboli may reach the brain, orbit, lungs, kidney, spleen and liver.

An MRI venogram should confirm the diagnosis by demonstrating absent venous flow from the cavernous sinus. High-resolution CT with contrast should also show a filling defect around the cavernous sinus.

Treatment includes high-dose intravenous antibiotics and drainage of purulent sinus collections, particularly from the sphenoid sinus. Systemic steroids and anticoagulants are often used, but their use remains controversial.

FRONTAL OSTEOMYELITIS

Frontal osteomyelitis is a condition that achieved notoriety when it acquired its well-known synonym of Pott's puffy tumour, described by Sir Percivall Pott, surgeon at St Bartholomew's Hospital, in 1760.

It develops as an acute frontal abscess that complicates an acute sinus infection and frontal sinusitis. The frontal bone that forms the anterior wall of the frontal sinus becomes osteomyelitic: Bone necrosis leads to a subperiosteal abscess that presents as fluctuant tender lump of the forehead. In some instances, the abscess points and bursts before surgical intervention can take place, and this may lead to a fronto-cutaneous fistula.

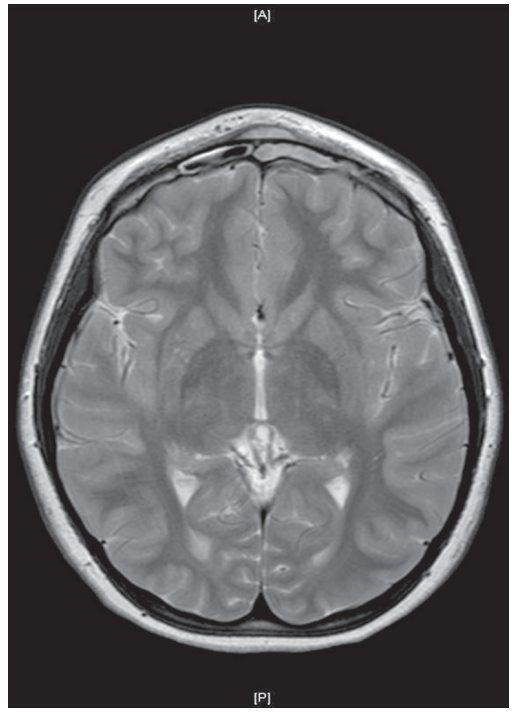

Figure 4.6 MR image showing soft tissue swelling and frontal sinusitis in acute frontal osteomyelitis (Pott's puffy tumour).

Although the condition presents as an acute purulent infection, patients often have underlying chronic rhinosinusitis, and this is important to appreciate to prevent a later recurrence.

A CT sinus scan, often combined with brain images, is necessary to assess the disease and plan operative intervention (Figure 4.6).

Treatment includes an intravenous antibiotic that is likely to cover the bacterial profile and also penetrate bone. Following microbiological advice, clindamycin is often recommended as the preferred antibiotic.

Operative intervention is normally required. Emergency procedures should be limited to simple drainage of pus. This can be achieved by placing a small incision directly over the abscess within a skin crease on the forehead. This facilitates curettage of necrotic bone and clearance of the purulent contents and loculi within the frontal sinus. A drain can be inserted into the frontal sinus to prevent recurrence of a purulent collection.

There is often infection within the other paranasal sinuses, and the maxillary antra should be checked and lavaged at the same time. Should endoscopic surgery be contemplated, this should be restricted to opening of the anterior ethmoid and frontal recess.

Oral antibiotics are often required for several weeks following acute infection. Once the patient has recovered from the acute infection, more definitive surgery can be undertaken to prevent recurrence and address any unsightly depression or scarring on the forehead. This may include performing a Draf type 3 sinuplasty median frontal drainage procedure in special cases.

KEY LEARNING POINTS

- Acute rhinosinusitis is most often viral.
- Recurrent acute rhinosinusitis may be secondary to allergy, irritants or immune deficiency.
- Complications arising from acute rhinosinusitis are uncommon but may be serious.
- Acute collections of pus within the orbit or intracranial cavity should be drained urgently.
- Purulent infections within the orbit or intracranial cavity are serious and generally require urgent additional specialist ophthalmic or neurosurgical care.

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5

Granulomatous conditions of the nose

JOANNE RIMMER AND VALERIE J LUND

A granuloma is an organized collection of macrophages, often referred to as epithelioid cells, which tend to fuse to form multinucleated giant cells. Granulomatous conditions of the nose and paranasal sinuses may be secondary to infection, inflammation, trauma or substance abuse, or may be due to an autoimmune or neoplastic condition. The disease process may be confined to the nose or be part of a systemic condition involving multiple organ systems.

GRANULOMATOSIS WITH POLYANGIITIS (WEGENER'S GRANULOMATOSIS)

HISTORY AND EXAMINATION

Granulomatosis with polyangiitis (GPA) is a systemic autoimmune disease of unknown aetiology, histologically characterized by granulomatous inflammation of the respiratory tract with necrotizing vasculitis of small and medium-sized blood vessels and focal or proliferative glomerulonephritis. It has previously been known as Wegener's granulomatosis, but its name has changed, initially to cANCA-positive vasculitis and most recently to GPA.[1] Classically, it affects the nose, lungs and kidneys, but it can affect any organ. It is also now recognized that more localized forms of the disease can occur, and up to 25 per cent of patients may only complain of sinonasal symptoms. The true incidence is therefore likely to be under-reported, but is estimated at approximately 10 per million per annum, with a prevalence of up to 100 per million in Europe. The average age at onset is 40–55 years, but it may occur at any age. Men and women are equally affected, but it is predominantly a disease of Caucasians (93 per cent).

The presenting symptoms are rhinological in 60–90 per cent of cases, including nasal obstruction, crusting, discharge, epistaxis, hyposmia or anosmia, pain, epiphora or change in shape of the nose.[2] Otological symptoms occur in up to 40 per cent, primarily conductive or sensorineural hearing loss. Approximately 16 per cent of patients have subglottic stenosis, which may prove fatal; this percentage is higher (23 per cent) in younger patients. Lower respiratory tract symptoms such as cough, haemoptysis and dyspnoea can occur, and patients may complain of constitutional symptoms such as fever, lethargy and weight loss.

Examination reveals friable granular mucosa, often with old blood and crusting or excessive adhesion formation (Figure 5.1a and b). There may be a septal perforation, and in advanced cases there may be loss of normal internal nasal architecture with a single large cavity. The classic 'saddle' deformity of the nose is seen in 5–20 per cent of patients with GPA (Figure 5.2); the only other condition that produces a similar deformity is relapsing polychondritis. Middle ear effusions may be present. The differential diagnosis of GPA includes all other nasal granulomatous conditions.

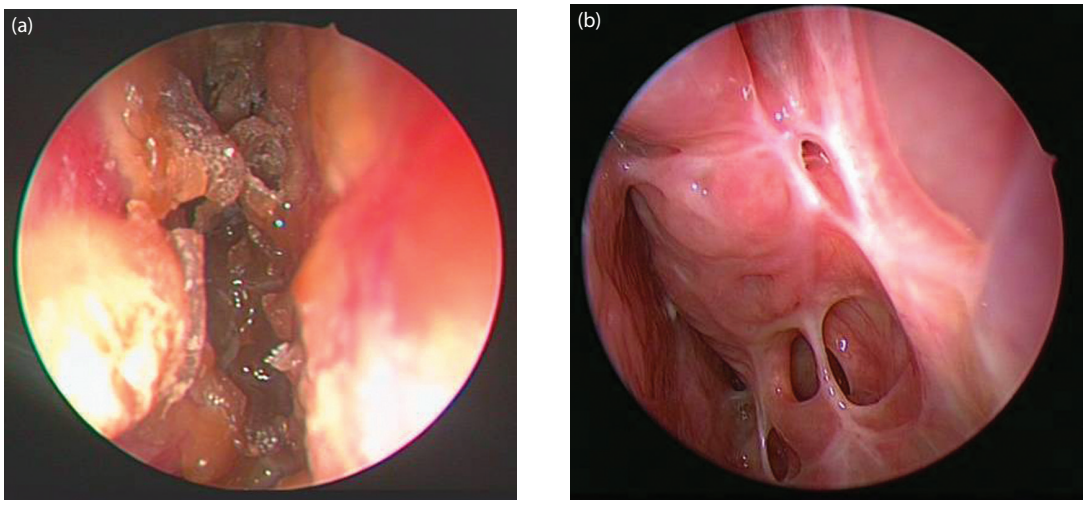

Figure 5.1 **(a)** Endoscopic photograph showing significant nasal crusting in GPA. **(b)** Endoscopic photograph showing marked adhesion formation in GPA.

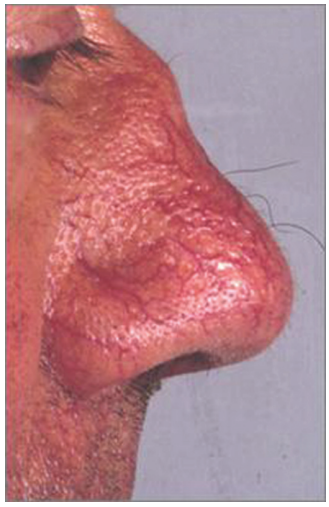

Figure 5.2 Clinical photograph showing collapse of nasal bridge in GPA.

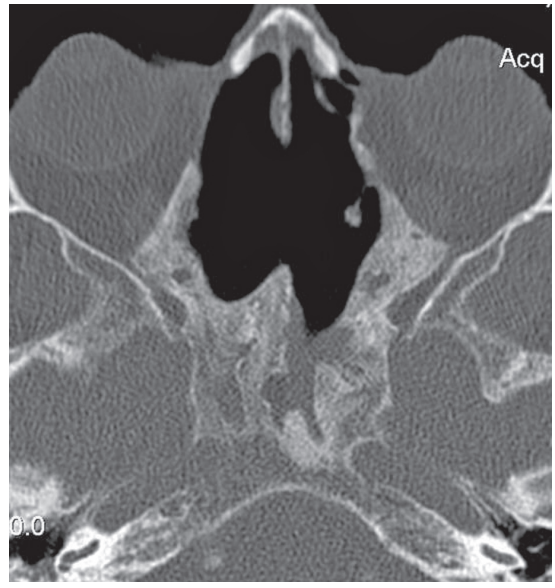

Figure 5.3 Axial CT scan of a patient with GPA showing loss of septum, ethmoids and anterior lamina papyracea together with marked sclerosis of residual bone.

INVESTIGATIONS

Anti-neutrophil cytoplasmic antibodies (ANCAs) are strongly associated with three vasculitides: GPA, eosinophilic granulomatosis with polyangiitis (Churg-Strauss syndrome) and microscopic polyangiitis (MPA). They are therefore not 100 per cent sensitive or specific for the diagnosis of GPA. However, cytoplasmic ANCA (cANCA) specific for proteinase 3 (PR3) is 90 per cent sensitive and 98 per cent specific for generalized GPA; the sensitivity falls to 50 per cent in localized disease. Ten per cent of patients with GPA will have a positive test for perinuclear ANCA (pANCA) against myeloperoxidase (MPO), and up to 30 per cent may have a negative ANCA test initially. This result may change over time, and cANCA may therefore be used to monitor disease activity.[3] It is often difficult to obtain a positive biopsy from nasal mucosa; lung and renal biopsies have a higher yield. Histological diagnosis of GPA requires the presence of granulomatous inflammation, vasculitis and necrosis. Computed tomography (CT) imaging of the paranasal sinuses shows non-specific mucosal thickening in more than 85 per cent of patients, but up to 75 per cent may have evidence of bony destruction, and new bone formation is seen in up to half of patients (Figure 5.3). However, sinus scans can also be normal. Chest x-ray (CXR) and urinalysis may be helpful in confirming the involvement of other organ systems.

MANAGEMENT

The European Vasculitis Study Group has classified GPA as *localized* (respiratory tract involvement only), *early systemic* or *generalized* disease.[4] If the disease is suspected, then a multidisciplinary approach with involvement of a rheumatologist or vasculitis specialist should begin at an early stage. Survival in generalized GPA was previously very poor, but the advent of corticosteroid treatment has reduced mortality rates to 50 per cent. The introduction of cyclophosphamide has further improved remission rates to 90 per cent, and steroid-sparing agents such as azathioprine, methotrexate and mycophenolate mofetil are then used to maintain remission. Monoclonal antibodies such as rituximab and infliximab have also been used recently with good effect. For disease localized to the nose, or for symptomatic relief of sinonasal involvement, nasal douching and topical nasal steroids are the mainstay of treatment. Nasal

lubricants may also be used. Sinus surgery should be avoided if possible, as post-operative scarring can make symptoms significantly worse, but may be required in selected patients. Surgical repair of septal perforations is unlikely to be successful, but a Silastic button may be inserted in addition to the use of topical treatments. Reconstruction of a saddle deformity requires the use of cartilage or bone grafts and generally has poor results if disease is active; however, if the disease has been quiescent for at least one year, it may be considered in selected cases. Grommets should be avoided owing to the risk of chronic discharge, and hearing aids are the treatment of choice for hearing loss. Subglottic stenosis can usually be managed with regular dilatation, intralesional steroid injection and endolaryngeal laser treatment. Mortality rates remain approximately 5 per cent despite immunosuppressant treatment, and up to 10 per cent of patients will never enter remission. Of those who do, up to 50 per cent will relapse.

EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (CHURG-STRAUSS SYNDROME)

HISTORY AND EXAMINATION

Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare but potentially fatal systemic necrotizing vasculitis of unknown aetiology, affecting small to medium-sized vessels. It is associated with asthma, eosinophilia and extravascular eosinophilic granulomas and was previously known as Churg-Strauss syndrome.[1] The asthma is characteristically late-onset and, together with allergic rhinitis and nasal polyps, may predate the vasculitis by several years. The average time from onset of asthma to diagnosis of EGPA is 12 years (range 1–34 years). It is less common than GPA, with an incidence of 2.4–4.2 per million per year in Europe. However, it has been reported in up to 67 per million asthmatics. The mean age at onset of the vasculitis is 50 years, but it has been reported in children. Men and women are equally affected, and most patients are Caucasian (up to 98.2 per cent).[5]

Three overlapping disease phases have been described: (1) a prodromal phase that may last for several years, during which time nasal and pulmonary symptoms tend to worsen; (2) the development of peripheral eosinophilia of greater than 10 per cent, which may be associated with other organ involvement as a result of eosinophilic infiltrates; and (3) the onset of systemic vasculitis and its associated symptoms. There are peripheral nervous system (PNS) symptoms in 50–78 per cent, primarily mononeuritis multiplex; skin involvement in 40–70 per cent, with purpura and nodules; gastro-intestinal symptoms in one-third; renal disease in 21–80 per cent; and cardiac involvement, usually myocarditis, in 10–49 per cent. It has been reported to occur after treatment with zafirlukast, a leucotriene receptor antagonist, but this is thought to be due to unmasking of the vasculitis by the reduction in oral steroid treatment that zafirlukast allowed.

Nasal obstruction and rhinorrhoea are the most common sinonasal symptoms, occurring in up to 95 per cent of patients, but anosmia (90 per cent), sneezing (80 per cent), crusting (75 per cent) and epistaxis (60 per cent) are also common.

The American College of Rheumatology 1990 criteria for diagnosis of EGPA (Churg-Strauss) require at least four of the following: asthma; eosinophilia of greater than 10 per cent differential; mono- or polyneuropathy due to vasculitis; non-fixed pulmonary infiltrates; abnormalities of the paranasal sinuses; and extravascular eosinophils on biopsy. This gives a sensitivity of 85 per cent and a specificity of 99.7 per cent. The differential diagnosis includes idiopathic hypereosinophilic syndrome, GPA, MPA, sarcoidosis, allergic bronchopulmonary aspergillosis and parasite infection.

INVESTIGATIONS

EGPA is another ANCA-associated vasculitis, but pANCA (against MPO) is only positive in 31–50 per cent of cases. There is less cardiac involvement in ANCA-positive cases, but ear, nose and throat (ENT), renal and PNS symptoms are more common.[5] There is eosinophilia, and acute phase proteins are often elevated. Histological diagnosis requires the presence of extravascular eosinophilic granulomas and necrotizing vasculitis, but nasal

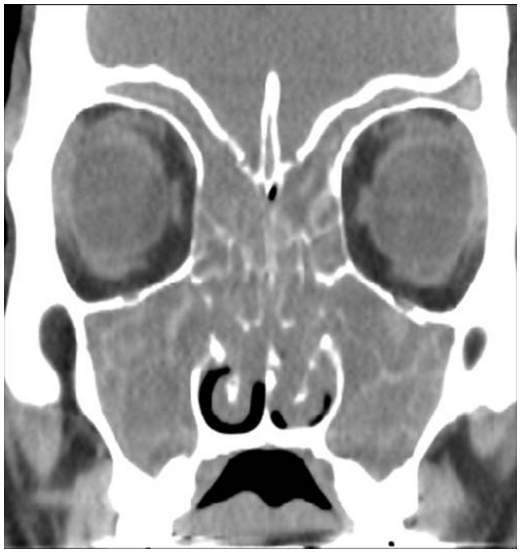

Figure 5.4 Coronal CT scan showing widespread chronic rhinosinusitis with nasal polyps in a patient with EGPA (Churg-Strauss).

biopsy is generally unhelpful. Skin, nerve, muscle or lung tissue has a higher positive yield. There is pan-opacification of the paranasal sinuses on CT, due to chronic rhinosinusitis (CRS) with nasal polyps, and there may be associated bony expansion and mucocoele formation (Figure 5.4). CXR may show nodular infiltrates.

MANAGEMENT

Again, this should be multidisciplinary, involving a respiratory physician or one with an interest in vasculitis. Outcomes have been shown to be better if patients are treated at an experienced vasculitis centre. Immunosuppression is the mainstay of treatment, with corticosteroids and steroid-sparing agents, but cytotoxic drugs such as cyclophosphamide may also be required. Rituximab has been used, and interferon-α has been tried in refractory cases with limited success. Sinonasal symptoms may be managed with topical treatments such as alkaline nasal douching and intranasal steroids, but surgery may be required for polyps or mucocoele formation. Remission is achieved in 81–92 per cent of patients, but more than a quarter will relapse. The French Vasculitis Study Group reported 5-year survival rates of 88.9 per cent, falling to 78.6 per cent at 10 years; survival was better with ANCA-positive disease.[5] Mortality rates can be as high as 46 per cent if there is cardiac involvement, which is the major cause of death in EGPA. The presence of cardiomyopathy at diagnosis, older age at diagnosis and a diagnosis of EGPA prior to 1996 were all found to be independent predictors of death in the French study.

SARCOIDOSIS

HISTORY AND EXAMINATION

Sarcoidosis is a chronic granulomatous disease of unknown aetiology, although it appears to be immune mediated.[6] It has a reported incidence of 1.2–19 per 100,000 per year but is more common in northern Europe and the southeast United States. It is significantly more common in African Americans, with an incidence of 36.5–81.8 per 100,000 per year. It can affect any age group, but its onset is most common in the third and fourth decades, and women are affected twice as often as men.

It is a multisystem disease that affects the lungs in more than 90 per cent, but cutaneous lesions are also common with a classic purple discolouration, known as lupus pernio (Figure 5.5), and

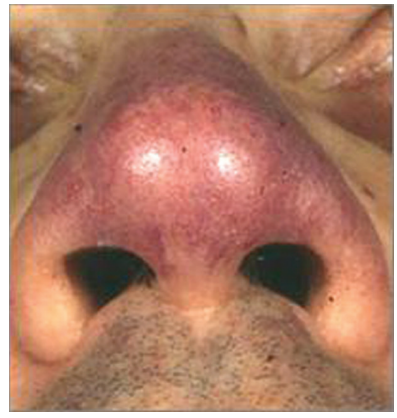

Figure 5.5 Clinical photograph showing lupus pernio affecting tip of the nose.

subcutaneous nodules. The upper respiratory tract is affected in up to 9 per cent of cases. Nasal obstruction, crusting, bleeding and facial pain are the most common symptoms. The ears and mouth may also be affected, as can the larynx, classically with supraglottic lesions. The nasal mucosa tends to have a characteristic 'strawberry' appearance of inflamed erythematous mucosa covered with pale granulomas. Crusting is often seen, and there may be an anterior septal perforation. The differential diagnosis includes all other granulomatous conditions of the nose.

INVESTIGATIONS

Sarcoidosis was previously diagnosed using the Kveim test, but this has been withdrawn in the United Kingdom due to health and safety reasons. Serum angiotensin-converting enzyme (ACE) level may be elevated, although this is also seen in other conditions such as tuberculosis (TB) and lymphoma and so is not specific. Serum calcium may be elevated in systemic disease, but only in 11 per cent of patients. Histology shows non-caseating epithelioid granulomas; nasal biopsy is usually negative if the mucosa appears normal, but a positive result is obtained in 91 per cent if it is clinically abnormal. CT of the paranasal sinuses may show changes consistent with CRS, and lacrimal gland enlargement is common. Sometimes there may be soft tissue infiltration of the nasal bones. CXR classically shows bilateral hilar lymphadenopathy.

MANAGEMENT

Management should be multidisciplinary with a respiratory physician. The Krespi staging classifies sarcoidosis into mild (stage I), moderate (stage II) or severe (stage III). The mainstay of treatment is systemic steroids and steroid-sparing agents. Topical treatment for the nose includes nasal douching, topical steroids and lubricants. Intralesional steroids may be used in cutaneous or laryngeal lesions. Surgery has only a limited role. Sarcoidosis may remit spontaneously, but severe pulmonary or cardiac involvement may be fatal.

COCAINE-INDUCED MIDLINE DESTRUCTIVE LESION

HISTORY AND EXAMINATION

Chronic intranasal cocaine abuse can cause granulomatous inflammation and destruction of the nose, sinuses and palate that may be clinically indistinguishable from GPA. This process is thought to be due to the marked vasoconstrictive effect of cocaine. A history of intranasal substance abuse should therefore be sought in all patients presenting with such symptoms.

INVESTIGATIONS

ANCA is often positive in this condition, with PR3 reactivity in more than 50 per cent, which compounds the similarity with GPA, but there are some subtle differences in capsase 3 and 9 expression and ANCA reactivity with neutrophil elastase that can distinguish the two conditions.

MANAGEMENT

Cessation of cocaine abuse is most important, and topical nasal treatments may provide symptomatic relief. Surgical reconstruction is often difficult with poor outcomes.

NATURAL KILLER/T-CELL LYMPHOMA

HISTORY AND EXAMINATION

Previously known as *midline destructive granuloma* or *midline lethal granuloma*, this tumour is now known to be a lymphoma but usually presents with midface destruction.

INVESTIGATIONS

Representative biopsy from beneath necrotic tissue is required for diagnosis. Imaging shows widespread bony and soft tissue destruction.

MANAGEMENT

Treatment is with radiotherapy plus or minus chemotherapy. Long-term follow-up is required because there may be late relapses.

GRANULOMATOUS INFECTIONS OF THE NOSE

Most of these infections are rare in the developed world, and treatment should be given by an infectious diseases specialist.

TUBERCULOSIS

HISTORY AND EXAMINATION

Nasal TB is uncommon, and primary nasal TB accounts for only one-third of cases. The causative organism is usually *Mycobacterium tuberculosis*. Ulcerative and nodular (lupus vulgaris) forms are seen in the nose, with an isolated granuloma occasionally found in the paranasal sinuses.

INVESTIGATIONS

Biopsy reveals caseating epithelioid granulomas, but acid-fast bacilli may not be seen and cultures may be negative. Skin testing for TB may prove infection, and serum testing such as QuantiFERON is now available.

MANAGEMENT

Extrapulmonary TB is usually managed with combination antituberculous chemotherapy.

LEPROSY

HISTORY AND EXAMINATION

Leprosy is a chronic granulomatous condition caused by the acid-fast bacillus *Mycobacterium leprae*. It is unusual in developed countries, but there are 12–15 million cases worldwide. There are two main forms of the disease, tuberculoid leprosy, which is localized, and lepromatous leprosy, which is systemic. The nasal skin is often involved with anaesthetic plaques, and nasal mucosa may be affected in systemic cases.

INVESTIGATIONS

Diagnosis is based on clinical findings with microbiological confirmation of the bacillus on biopsy.

MANAGEMENT

Long-term dapsone remains the standard therapy.

SYPHILIS

HISTORY AND EXAMINATION

Syphilis is a sexually transmitted disease caused by the spirochaete *Treponema pallidum*. It is classified into primary, secondary, tertiary and congenital forms. Primary chancre of the nose is rare but may be seen on the skin or within the vestibule. Secondary syphilis is an infectious systemic condition, with rhinitis the most common nasal symptom. Tertiary syphilis may cause a perforation of the bony septum or present as a subcutaneous nodule (gumma) which then ulcerates. Congenital syphilis most commonly presents with 'snuffles', a purulent rhinitis with eventual excoriation of the upper lip.

INVESTIGATIONS

Syphilis is usually diagnosed with serological tests, but smears from a lesion may show the organism.

MANAGEMENT

Parenteral penicillin is the antibiotic of choice for all stages.

MUCOCUTANEOUS LEISHMANIASIS

HISTORY AND EXAMINATION

Leishmaniasis, a parasitic disease caused by *Leishmania spp.*, is endemic in more than 80 countries, with 12 million cases worldwide. It may present in cutaneous, mucocutaneous or visceral forms. The mucocutaneous form can cause swelling and erythema of the external nose with erosion of the internal nose, septum, palate and trachea, and has been reported to mimic GPA.

INVESTIGATIONS

Diagnosis is difficult, but biopsy may show granulomatous inflammation without vasculitis. Biopsy of involved skin may be more helpful than nasal mucosa.

MANAGEMENT

Treatment is usually with amphotericin or a pentivalent antimony compound.

RHINOSPORIDIOSIS

HISTORY AND EXAMINATION

Rhinosporidiosis, a chronic granulomatous infection caused by *Rhinosporidium seeberi*, is most commonly seen in India and Sri Lanka. The nasal mucosa becomes granular and polypoidal with a strawberry-like appearance.

INVESTIGATIONS

Histological examination shows the organism's spores.

MANAGEMENT

Treatment typically involves surgical excision of the lesions followed by medical therapy such as dapsone, griseofulvin, amphotericin and diaminodiphenylsulfone, but none of these preparations has proved particularly successful.

RHINOSCLEROMA

HISTORY AND EXAMINATION

Klebsiella rhinoscleromatis causes this progressive granulomatous infection of the nose and upper respiratory tract, with laryngeal involvement in up to 50 per cent of cases. There are three stages: atrophic, with crusting similar to atrophic rhinitis; granulomatous or proliferative, with nodule formation; and cicatrizing, with stenosis and adhesion formation.

INVESTIGATIONS

The diagnosis is usually made in the later stages, based on the typical histological findings of granulomatous infiltrates and Mikulicz cells.

MANAGEMENT

Prolonged antibacterial treatment with ciprofloxacin or trimethoprim-sulfamethoxazole is required, but initial surgical debridement may be helpful.

KEY LEARNING POINTS

- Systemic granulomatous conditions often present with ENT symptoms. Although they are unusual diseases, it is important to have a high index of suspicion to avoid delay in diagnosis.
- Always suspect a vasculitis if there is general malaise disproportionate to the clinical findings.
- Nasal crusting and abnormal mucosa should prompt investigation for granulomatous disease, as should recalcitrant symptoms of chronic rhinosinusitis despite appropriate management.

- There is no one diagnostic test; diagnosis is based on the clinical picture together with examination findings, imaging, serological and histological tests.

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6

Chronic rhinosinusitis

ROBIN YOUNGS

CHRONIC RHINOSINUSITIS

Rhinosinusitis is the term used to describe inflammation and infection within the nasal cavity and paranasal sinuses. This condition frequently coexists with nasal polyps. Chronic rhinosinusitis (CRS) is very common and accounts for a large number of visits to both primary care physicians and specialists. It is estimated that chronic rhinosinusitis affects between 5 per cent and 15 per cent of the general population in Europe and the United States.

DEFINITION

The 2012 update of the European Position Paper on Rhinosinusitis and Nasal Polyps (EPOS) defines chronic rhinosinusitis in adults as the following:

- Inflammation of the nose and paranasal sinuses characterized by two or more symptoms, one of which should be either nasal blockage/obstruction/congestion or nasal discharge (anterior/posterior nasal drip)
- +/- Facial pain/pressure
- +/- Reduction or loss of smell

and either

- Endoscopic signs
  - Nasal polyps, and/or
  - Mucopurulent discharge primarily from middle meatus and/or
  - Oedema/mucosal obstruction primarily in middle meatus and/or
- Computed tomography (CT) changes
- Mucosal changes within the osteomeatal complex and/or sinuses

for at least 12 weeks without resolution.