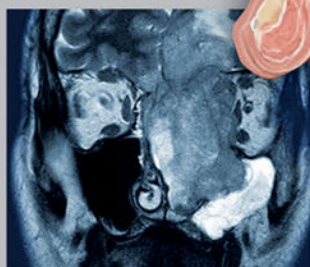
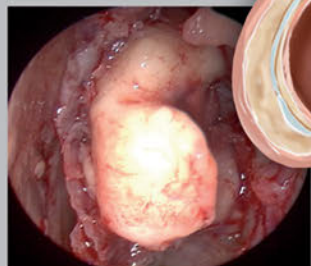
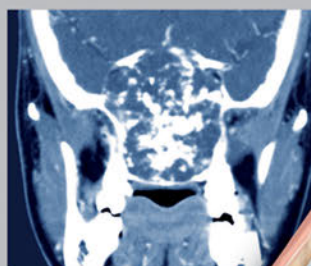


Tumors of the Nose, Sinuses and Nasopharynx

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¹Since writing his chapter, Professor Kian Ang has very sadly died. We would like to pay tribute to his tremendous contribution to radiation oncology throughout the world.

Foreword

This volume is an exhaustive, masterly crafted storehouse of invaluable information on the tumors of the nose, paranasal sinuses, and nasopharynx. In the spirit of completeness and thoroughness that characterized the original book, *Tumors of the Upper Jaw*, authored by Professor Valerie Lund and the late Sir Donald Harrison, this up-to-date text goes into extensive detail on all the common and virtually all the unusual pathological entities that may involve this complex and difficult area.

The introductory chapter by Professor David Howard, based on an earlier chapter by Professor Philip Stell, is a fascinating, often humorous, and extremely informative chapter describing the history behind the development of surgery of the maxilla and paranasal sinuses. Many of the myths attributing the original diagnoses of certain pathological entities and the origins of certain operative procedures to specific individuals, beliefs held by many of us throughout our professional careers, are “blown out of the water” by Professor Howard’s historical revelations.

Each individual chapter in this book is a treasure trove of valuable up-to-date information on each pathological entity. The format of each chapter—naming the disease process, listing the other common synonyms for the tumor or inflammatory processes, discussing the etiology and pathophysiology, and reviewing diagnostic procedures, particularly radiography, all followed

by treatment, complications, and results, and finally by citations of survivals from the world literature—is excellent. Regarding complications and survival results, the authors have had the good fortune of being able to draw from an extensive personal experience of their own, augmented by that of their mentors.

The section on the treatment of malignancies of the paranasal sinuses is extremely instructive. The techniques of endoscopic resection, both subcranially and intracranially, is well detailed. It is quite easy to then contrast these techniques with the subcranial and combined transfacial–intracranial open approaches to sinus malignancies with and without skull base involvement.

The section on the management of nasopharyngeal lesions, especially nasopharyngeal carcinoma by Professor William Wei is especially valuable. His extensive experience with this tumor, coupled with having practiced in the same institution for most of his career, has enabled him to follow his patients for long periods of time and formulate a best treatment strategy.

This volume will be an important addition to the libraries of residents, (registrars), and fellows in both endoscopic sinus surgery and head and neck oncological surgery and consultants in otolaryngology, plastic surgery, and oromaxillofacial surgery, as well as the ancillary services of therapeutic radiology, medical oncology, diagnostic radiology, and pathology.

Paul J. Donald, MD, FRCSC

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1 Introduction and Acknowledgments

When we three, Valerie, David, and William, decided to undertake this project 3 years ago, we knew that it was going to be a major undertaking. *Tumours of the Upper Jaw* had been published by Valerie with Professor Sir Donald Harrison (Fig. 1.1) in 1993 and in the intervening period there had been a significant number of changes in the evaluation and management of tumors in the sinonasal and nasopharyngeal areas. Unfortunately, tumors in these areas still present late and their cure provides huge challenges. In addition to our own substantial experience, we have conducted an extensive review of the literature to identify all significant contributions since the last book and have tried to use the best evidence available.

We recognize that level I or II randomized placebo-controlled trials are absent in the literature for virtually any treatment used for these tumors due to a combination of ethical considerations and the rarity of the conditions. Even the acquisition of large prospective cohorts of tumor patients has proved difficult, although there are now several centers, including our own, that have published data that are adequate for statistical analysis. Nonetheless, there are a large number of pathologies that are represented only by case reports or small retrospective series. It is also worth noting that journals willing to publish these types of report are now few because of the pursuit of scientific credibility and improved impact factors. Thus it may become increasingly difficult to gather information on the rarest pathologies but this situation also supports the view that there should be a degree of centralization of many sinonasal, skull base, and nasopharyngeal neoplasms. This is particularly underlined by the proliferation of endoscopic techniques and has led to the development of collaborative multicenter networks and prospective data collection.¹ Along with accurate records of long-term follow-up, this will greatly facilitate future evaluation of results. Similarly, the majority of the tumors covered in this book are now managed by multidisciplinary teams which include input from neurosurgeons, ophthalmologists, plastic and maxillofacial surgeons, radiation oncologists, and a range of other experts.

We have included a few additional clinical conditions that strictly speaking do not come under the definition of neoplasms but are difficult to manage, may mimic tumors, and at times are life-threatening in their own right.

While odontogenic tumors are normally the province of oral pathologists, oral surgeons, and maxillofacial surgeons, they are also seen and treated by ENT and plastic surgeons in many parts of the world and accordingly have been included.

In undertaking this project we have been helped by a large number of individuals, not only through the excellent contributions from Professors Zinreich, Ang, Chua and

their colleagues but also from friends and coworkers. This list is by no means complete but we would like to thank our long-suffering secretaries, Angela Constantinou, Trisha Holness, and Anne Oliphant; colleagues including Tim Beale, Lloyd Savy, Gita Madani, Anne Sandison, Peter Clarke, Dawn Carnell, Simon Stewart, Richard Welfare, Geoff Rose, and other colleagues at Moorfields; and Fellows and trainees Humera Babar-Craig, Ed Chisholm, Jo Rimmer, Adin Selcuk, and Matteo Trimarchi.

Finally we would like to pay tribute to several individuals whose unique clinical expertise and support have enormously enhanced the treatment of our patients: Tony Cheesman, Sir Donald Harrison, Glyn Lloyd, Leslie Michaels, and Margaret Spittle (Figs. 1.2, 1.3, 1.4, 1.5).

Reference

1. Lund VJ, Stammberger H, Nicolai P, et al. European position paper on endoscopic management of tumours of the nose, paranasal sinuses and skull base. *Rhinol Suppl* 2010;(22):1–143



Fig. 1.1 Professor Sir Donald Harrison.



Fig. 1.2 Tony Cheesman.



Fig. 1.3 Glyn Lloyd.



Fig. 1.4 Leslie Michaels.



Fig. 1.5 Margaret Spittle OBE.

2 Historical Aspects of Surgery in the Sinonasal Area

D. J. Howard and P. Stell

In an earlier book entitled *Tumours of the Upper Jaw* written by Valerie Lund and Sir Donald Harrison and published in 1993,¹ Professor Philip Stell, then Professor of Otolaryngology at the University of Liverpool, was asked to contribute a chapter on the history of the surgery of the upper jaw. I have chosen to reproduce and add further to that chapter here for several extremely good reasons. Following his appointment to the Chair in Liverpool in 1979, Philip Stell confined his practice largely to laryngology and all aspects of head and neck malignancies. He developed his own extensive computerized database and was a strong advocate of statistical analysis of all surgical results. He established the journal *Clinical Otolaryngology and Allied Sciences* in 1978 and founded the UK Otorhinolaryngological Research Society. He published more than 300 peer review papers and was a superb linguist, being fluent in Spanish, French, German, and Dutch. He lectured extensively in Europe and was honored by many European countries.

Following his early retirement at the age of 57 on health grounds in 1992, he subsequently moved to York where he enrolled for an MA in history at York University and his thesis was entitled “Medical care in late Medieval York.” He achieved his degree with distinction and in 1996 commenced further research in the Centre for Medieval Studies in the University of York, establishing a unique database for medieval Yorkshire wills, names, and other early documents. He was subsequently honored with invitations to become a Fellow of the Society of Antiquaries and also of the Royal Historical Society. He received an MBE for his contributions to medical history shortly before his death in 2004.

For all of the above reasons, it would be extremely difficult to improve on his superbly researched historical chapter which follows, but as there have been substantial and wide ranging developments since the 1980s, I have taken the liberty of adding an overview of the more recent developments. For consistency of style, the spelling and other conventions of the present volume have been adopted in the reproduced text.

History of Surgery of the Upper Jaw

(PM Stell. History of surgery of the upper jaw. In: Harrison DFN, Lund VJ. *Tumours of the Upper Jaw*. Edinburgh, London, Madrid, Melbourne, New York, Tokyo: Churchill Livingstone; 1993:1–15)

An historical introduction to major articles or textbooks has become commonplace. Sadly, most of these

historical vignettes are, for various reasons, inaccurate, the commonest reason being that the author failed to read the original articles. For example, it is often stated that Patrick Heron Watson did the first laryngectomy for syphilis, in 1865. But, the original paper shows that Watson described the larynx, trachea, and bronchi of a patient who had syphilis; the only operation performed during life was a tracheotomy.² A second source of error is an inability to read languages other than English. It is often said that adenoid cystic carcinoma was first described by Billroth in 1859 by the term “Zylindrome.”³ This is untrue: the tumor was first described by two Frenchmen, Robin and Laboulbène, as “tumeur heteradenique” in 1853.⁴

A third source of error is to ignore the context of the historical events. It is stated repeatedly⁵ that cancer of the ear was first discussed ca. 1775 by Wilde and Schwartz. Apart from the fact that Wilde was born in 1815 and Schwartz in 1837, this statement ignores the fact that in 1775, histopathological diagnosis still lay almost a century ahead, so that no such discussion was possible.

A specific example of historical inaccuracy is the large monograph on malignant tumors of the maxilloethmoidal region written by Oehngren in 1933.⁶ His extensive historical introduction is marred by two facts: first it is obvious that he did not personally read all the original reports for he misquotes names, e.g., Lizzard for Lizards; second, he gives no references to the authors he quotes.

In compiling this account of the development of upper jaw surgery, I have read and searched widely through the available literature, attempting to resolve these writings to the technology and politics of the relevant times. The development of the single-lens microscope by Antonie van Leeuwenhoek around 1665 and the discovery of aniline dyes ca. 1856 by Perkin, an Englishman working in Germany, eventually made histopathology possible.⁷ Normal histology developed mainly in Germany, throwing up such well-known names as Schwann and Henle. They were followed by Virchow, who laid the foundations of histopathology with his book *Cellular Pathology*. Virchow was the first to emphasize that classification based on external appearances was arbitrary and rather it should be based on normal cellular structures. He was one of the first to use terms such as epithelioma for squamous carcinoma.⁸

By around 1860 economic and technological advances initiated a German surgical school led by famous men such as Conrad Langenbeck, his nephew Bernard Langenbeck, Billroth, Thiersch, Kocher, Trendelenburg, Czerny, Mululicz, and others (Figs. 2.1 and 2.2). This was the

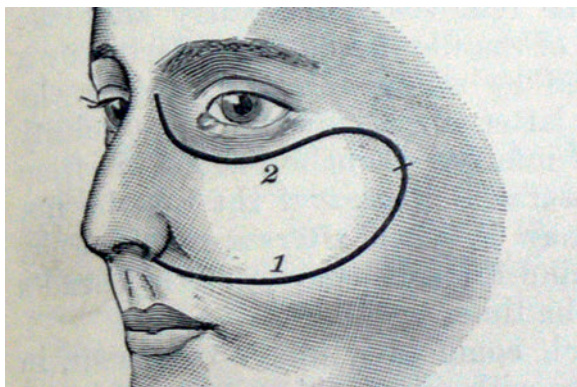


Fig. 2.1 B. Langenbeck's osteoplastic flap procedure described in 1861 to treat a 15-year-old boy with an angiofibroma. The object of this approach was to hinge the maxilla on the lateral aspect of the nose without interfering with the alveolar and palatine tissues or the floor of the orbit.

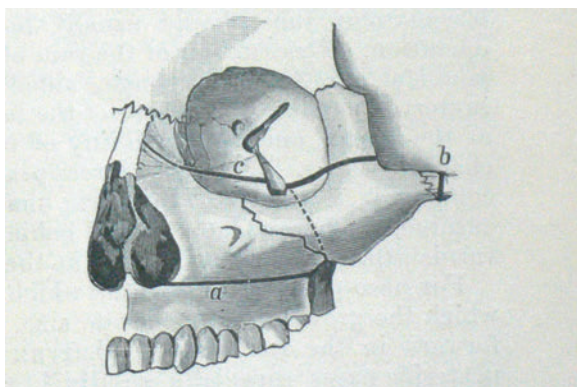


Fig. 2.2 This drawing shows the lines in Langenbeck's operation for the saw cuts through the upper jaw and zygoma followed by elevation of the bone by means of an instrument introduced under the malar bone.

"The growth was completely exposed and removed. The operation took one hour, was attended with much hemorrhage, most of which stopped spontaneously. The wounds healed well."

golden age of excisional surgery, and all the first major excisions of the internal organs such as gastrectomy, total laryngectomy, glossectomy, and so on were described at that time in Germany.

Between 1825 and 1875 when maxillectomy was developed, the main contributors came from the French and three British schools: Edinburgh, London, and Dublin. The outstanding names were those of Gensoul and Dupuytren in France; Syme, Liston, and Lizars of Edinburgh; the English school led by Fergusson; and the Dublin school under Stokes and Butcher. The German schools only contributed toward the end of this period. The school that flourished in Dublin from ca. 1800–1830 produced such well-known names as Wilde, the father of modern otology (and of Oscar Wilde), Stokes, Adams,

Colles, Corrigan, Cheyne, Graves, and the now forgotten Butcher, who in his time was preeminent in the field of maxillectomy. Why this school sprang up where and when it did is an historical mystery. Dublin society had been greatly depleted by the departure for London of diplomats and politicians after the Act of Union in 1800. Furthermore, Ireland had an entirely agricultural economy, whereas Europe was rapidly becoming industrialized and therefore more prosperous.

Surgical Practice in the 19th Century

It is virtually impossible to imagine how primitive were the conditions under which the pioneers of upper jaw surgery worked. Despite the incredible differences in pathology, anesthesia, instrumentation and, not least, operating facilities, between the early part of the 19th century and today, almost all the fundamental concepts of surgery in this region were during a period of 50 years.

Operating Conditions

If we could step back in time to 1830, perhaps the first difference that would be apparent would be the surgeon's dress. At that time it was usual to wear an old frock coat that hung on the back of the theater door: the more encrusted with blood and pus the more honorable it was. Surgical gowns and gloves belong to this century: even after the introduction of asepsis by Lister in 1867 it was usual for surgeons to operate with bare hands until at least 1895.⁹

Resection of the upper jaw was classified as a "capital operation," so called because the patient could, and often did, die during or immediately after the operation. It was usual to give notice, not only to the medical profession but also to society in general, of forthcoming operations of this type. Thus the audience included not only medical students but curious bystanders: the famous violinist Paganini performed a tour of England and Scotland in 1831–32, and attended a maxillectomy performed by Earle in 1831. At the end of the operation, Earle came forward and was greeted by "deserved applause." This operation had to be postponed because the rumor of it had brought together such a multitude that even after an adjournment to the anatomy theater, it was impracticable to continue with the operation.¹⁰ The operation performed by Liston in 1835 attracted several hundred spectators. The operating theater was so called because the seats were arranged in ascending rows, as in an ordinary theater, for easier viewing. For similar reasons, it was called an amphitheater in France (Fig. 2.3).

Pathology

The second enormous difference between the present day and the 50-year period from 1820 to 1870 during



Fig. 2.3 A photograph showing the operating theater of the old St Thomas' Hospital, London, which was used between 1822 and 1861 (and which can still be visited). From PM Stell. History of surgery of the upper jaw. In: Harrison DFN, Lund VJ. *Tumours of the Upper Jaw*. Edinburgh: Churchill Livingstone; 1993:1–15. With permission from Elsevier.

which maxillectomy was developed, was in pathological diagnosis. Surgeons had no histopathological description of the tumor on which to base their classification until ca. 1855, and the terms used were merely descriptions of the macroscopic appearance of the tumor. Histopathology was not placed on a firm basis until the publication of Virchow's book in 1856. He was the first to describe tumors on the basis of their cellular appearance.⁸ Gross descriptive terms were still being used as late as 1863 when Barton stated that maxillary tumors may be divided into medullary, scirrhus, melanoid, or epithelial.¹¹ Furthermore, even after histopathology became generally available around 1860, histological examination was restricted to postoperative examination.¹² The examination of a biopsy belongs to this century: indeed, as late as 1923 Ochsner was vigorously maintaining that preoperative biopsy with a knife led to the setting up of metastases.¹³

A patient described by Dickson in 1840, though not subjected to operation, gives useful insight into the appreciation of gross pathology at that time. The patient was described as having a "fungus of the antrum with lymphatic contamination, but not visceral taint." The lymphatic contamination was a lymph node enlarged to the size of an almond in the digastric space. It was clearly appreciated that this was a malignant tumor which had spread to the lymph nodes so that the tumor was therefore incurable. A necropsy was performed 12 hours after death showing no "visceral taint," that is, no distant metastases.¹⁴

In 1833 there were three indications for resection of the upper jaw:¹⁵

- "Malignant disease"
- "Augmented growth of the bony parts"
- "A sort of dropsy"

The terms used for "malignant disease" were often so vague that it is now impossible to know what they described. They included tumor, intumescence, malignant disease, medullary disease, sarcoma, and so on. The term carcinoma was not used in relation to the maxilla until 1878 by a German, Koerte.¹⁵ However, it is clear that the lesions were often carcinoma, and that the terms medullary tumor and sarcoma were squamous carcinomas. The word sarcoma was often used in its literal Greek meaning, that is a "fleshy tumor." It did not take on the connotation of a tumor of mesodermal origin until the latter part of the 19th century. Virchow in his classic *Cellular Pathology* tells us that the term medullary disease arose from the idea that it originated in the nerves and resembled nervous matter. This tumor was originally thought to arise from the body of the sphenoid bone or other bones of the base of the cranium, but Heath showed that it did indeed originate in the maxillary antrum itself.¹⁶ A review of 160 maxillectomies published between 1830 and 1880^{11,14,15,17–145} shows that ~50 of the 160 were done for carcinoma, although it was said that "cancer is certainly a very unusual form of growth to occur in the upper jaw."⁷¹

"Augmented growth of the bony parts" may refer either to bony tumors or to fibrous dysplasia. Of the first

160 cases, 19 were described as being bony tumors of some sort, such as exostosis, and many are described as osteosarcoma. Of 14 patients described by Dieffenbach, 6 were classed as osteosarcoma.⁴⁷ This description was not based on histology, and it is unlikely that almost half his patients suffered from so rare a disease. However, some of the bony tumors described were clearly osteomas; for example, the antral tumor described by Bickersteth in 1857, which could only be examined by sawing it in half.²⁵ It is interesting to speculate why osteomas (or exostoses) appear to have been so common in the early part of the 19th century.

"A sort of dropsy" was almost certainly expansion of the antrum and erosion of its bony walls by an expanding retention cyst. A patient who had undergone resection of the entire upper jaw for a cyst via a Fergusson type of incision on the face was shown at a meeting of the Medical Society of London in 1874.⁷⁰ The surgeon was strongly criticized for carrying out an operation that was more serious than the disease required. Another member pointed out that many of these cysts were of dentigerous origin, and were cured by the removal of the offending teeth, an opinion which would hold good to this day. Another dentigerous cyst of the antrum was described by Bryant, who said that doubtless upper jaws had been removed in former times for this affection, its true pathology not having been understood.¹⁴⁶

Another common diagnosis not included in Guthrie's classification¹⁵ was fibroid tumor or fibroplastic disease. Thirty of the first 160 maxillectomies were for such tumors. From detailed descriptions this tumor had a firm consistency with thin adherent bone, that had been eroded by pressure. Many of these tumors presented with a nonulcerated swelling of the palate; some later histological studies described fibrous tissue. Many of these tumors are clearly what would now be called angiofibromas, and a series of them is described by the Irish surgeon Butcher.³¹⁻³⁴ However, not all these tumors were angiofibromas as they appear to have arisen in both sexes and at all ages. Microscopy of one of these fibrous tumors showed elongated cells forming fibers, calcareous matter deposited along the course of the fibers, and a central hard portion infiltrated by earthy salts converting it into a stony mass.⁹⁰ This tumor was almost certainly an ossifying fibroma. A fibrous tumor, described as a fibrosarcoma, was removed from the upper jaw of a man of 58 by the well-known Irish surgeon Stokes in 1873.¹⁴⁷ The histological appearances showed tough fibrous tissue with a few small blood vessels.

At least a dozen maxillectomies were performed for what was described as necrosis or caries of the upper jaw, due to syphilis,^{27,117} typhus,¹⁴⁸ or occupational exposure to phosphorus.¹⁴⁹

Microscopical examination of tissue removed surgically began in the 1850s: Brainard in 1852 in the United States was one of the earliest practitioners describing

the tumor as presenting no trace of cancerous tissue and said that no cancer cells were detected under the microscope.¹⁵⁰ This examination was not necessarily based on examination of a section, because Craven in 1863 comments "under the microscope the juice scraped from the cut surfaces exhibited no fibrous element but simply a confused mass of broken up cells and granular matter."³⁹ Thus, in the early stages it appears that some form of cytology was practiced on cells scraped from the tumor. Furthermore, examination of sections as we know it did not develop until the end of the 19th century. Until then specimens for histology were preserved in alcohol²⁰ and cut by hand, but in ca. 1866 His made his sliding microtome, which was improved in the following decade. Automatic machines began with Threlfall's, made in 1883. These demanded rigid embedding of the specimen in substances such as paraffin wax.⁷

Early histological reports include that by Clark in 1856, who described a tumor "under the microscope as presenting cells and nuclei of every size and shape. A few commencing characteristics of epithelial cancer were present sufficiently distinctly to show positively that the tumor belonged to that class."³⁶ Another tumor³⁴ was encased in true bone, and histologically showed "oily globules compressed together but rather more irregular and oval in form, ~1/300th inch in the long diameter. The walls were made of closely packed cellules 1/2000th inch in diameter. No true bony cells could be found. When examined under polarized light at a power of 400 it showed a structure similar to that of horn or ivory."

Histological descriptions then followed rapidly: of a fibronucleated tumor,¹⁵¹ "a section of the mass hardened in spirit showed bundles of fibrous tissue but not arranged so as to form a cancerous stroma; several simple round cells and masses of spindle shaped cells";⁵¹ a "globular epithelioma";¹⁵² a small round cell carcinoma;¹⁵³ and a myxosarcoma including a woodcut of the histological appearances.¹¹⁶ Heiberg in 1861 described an adenoid cystic carcinoma under the then current term of cylindroma; this view was based on histological examination.¹⁵⁴ Other probable adenoid cystic carcinomas were soon described.^{49,138} In the latter case histology showed "an epitheliomatous epulis resembling an adenoma of the breast."¹² In another case recorded as a cubular epithelioma,²² histology showed "the ground work to consist of well-developed fibrous tissue with large groups of cells arranged in some parts like a racemose gland and in other parts like tubular glands. In the center of most of these groups there was a lumen." This was probably an adenoid cystic carcinoma, although it might have been an adamantinoma as it was said to resemble identically a "multilocular epithelioma" previously described in the upper jaw.

Sir William Fergusson must have had an interesting career, spanning as it did the development of both pathology and anesthesia. The circumstances when he

performed his first operation in 1842⁵³ must have been very different from those when he removed a maxilla in 1872.^{67,155} On the latter occasion histology was available to show that the tumor was composed of fibrous tissue with islandlike and spindle-shaped nuclear bodies with a calcareous nodule in its center, which was probably an ossifying fibroma. He was certainly using chloroform by 1863.⁶¹

Anesthesia

In the early days the patient was held or tied down. Nobody records “dulling the patient with alcohol” but this must surely have been common; laudanum (i.e., morphine) appears to have been given only after the operation. Chloroform was introduced in 1847 and ether in 1842, but chloroform was usually the sole agent used for maxillectomy (Fig. 2.4).

Chloroform was in frequent use by the 1860s, even in the provinces. Unfortunately, the patient was often conscious during the greater part of the operation: “chloroform was administered to its full extent to begin with, but its inhalation was not continued afterwards.”⁵² Another report from Australia in 1868 tells us that it “was tried but abandoned.”¹⁵⁶

It was often necessary to allow the patient to wake up during the operation if he or she was bleeding profusely. As late as 1870 it was customary to fix patients in the armchair as a precaution in case they did wake up.¹¹¹ Patients often recovered from the chloroform and “spat the blood as is often the case on all bystanders.”¹⁵⁷

Some thought that chloroform was dangerous because “the irritability of the glottis is weakened if not wholly lost, so that there must be the danger of the trickling of blood from the mouth into the glottis without the excitement of a cough for expelling it from the windpipe.”⁷⁶ It was often necessary to suspend the administration of chloroform and allow the patient to recover consciousness because of the “danger of strangulation from the great amount of blood poured out.” For this reason Rose recommended carrying out the operation with the head hanging.¹⁵⁸ Some surgeons remained unwilling to use chloroform until late in the century.¹⁵⁹

In the early days chloroform was administered by sprinkling it on a piece of lint,⁷⁹ but by 1860 it was being administered by a tube passed through one nostril.⁵⁸ Later a special tubular inhaler was developed to be passed through one nostril⁴⁵ but this method was rapidly displaced by Trendelenberg’s cannula. Trendelenberg had introduced a tracheostomy tube with a cuff in 1870.¹⁶⁰ This cannula had been used for the administration of an anesthetic through a tracheostomy to the first patient to undergo total laryngectomy by Billroth in 1873,¹⁶¹ and it was used for a maxillectomy for a cylindroma by Heiberg in Germany in 1872.¹⁵⁴ This method is the obvious way to avoid the dangers of hemorrhage during the

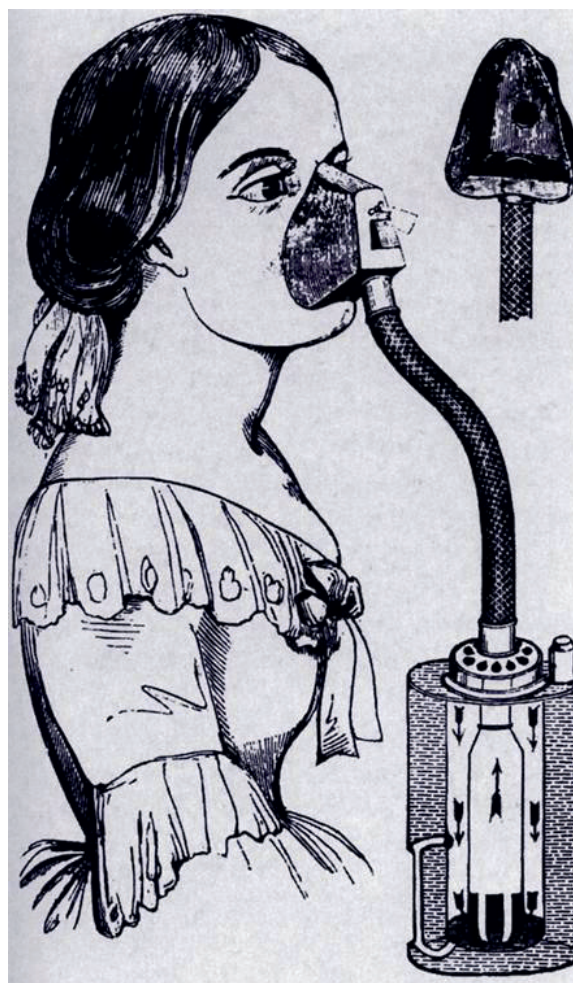


Fig. 2.4 An illustration from John Snow’s book “On chloroform and other anaesthetics” (1858) on anesthesia showing an apparatus for inhaling the vapor from liquid chloroform. By 1831 ether, nitrous oxide gas, and chloroform had all been discovered and in 1842 ether was first used medically. In 1853 Queen Victoria was given chloroform by John Snow for childbirth and its use spread worldwide within months!

administration of chloroform, and had become established by the 1880s: Bellamy in 1883 said “I was first inclined to do a prophylactic tracheotomy and to use Trendelenberg’s tamponade apparatus.”¹⁶²

A further means of preventing pain was to freeze the line of incision with ether.¹⁵⁶

Instruments

Although surgical instruments of many kinds had been available and used for centuries (Fig. 2.5) there were nonetheless some instances of great differences between the early 19th century and the present day. Two examples might suffice to emphasize this: first, the only form of illumination was natural daylight. Only one paper in the

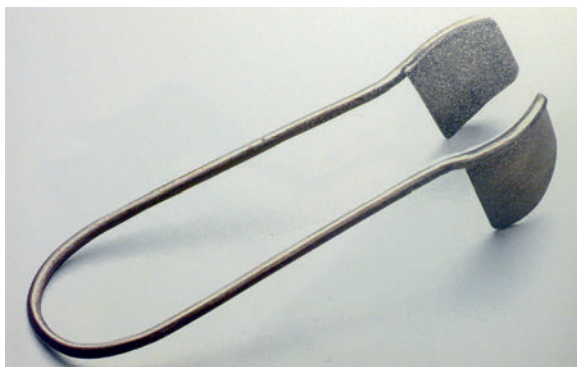


Fig. 2.5 Ludwig Johann Thudicum (1829–1901 from London) designed his speculum in 1868 and it remains in use in many ENT departments worldwide for initial anterior rhinoscopy.

first 50 years comments on illumination. In Irving's case in 1824 the patient was placed in an armchair opposite a window.¹⁶³ The question of illumination is not otherwise discussed. Lighting must have been very difficult as efficient illumination, either by gas or electric light, did not come into general use until the 1880s. Second, artery forceps for the control of hemorrhage were not invented until the latter part of the century, although ligatures were available for the arteries, and indeed were used by Nivison in 1824.¹⁶³ Fergusson's textbook of 1870 shows that the vessel was held by a forceps and the ligature was applied.⁶⁶

A common instrument was the cautery, of which there were two types: the actual cautery and the potential cautery. The actual cautery was a hot branding iron, whereas the potential cautery consisted of caustics of different sorts. Division of cautery into these two types was of ancient origin, and their use was described by Parey in the 17th century.¹⁶⁴ The actual cautery was used to deal with carious bone. Parey felt that it was more effective than potential cauteries such as sulfuric acid, scalding oil, and molten sulfur, because it could be used more precisely, but that the potential cautery often had to be used because of the pain produced by the actual cautery! In the 19th century discussions as to the relative merits of the two continued: Liston (1821) felt that the actual cautery was preferable in maxillectomy because it was effective and the pain it produced was greater but momentary, whereas the pain of potential cautery persisted for several days.¹⁰⁵

The term "actual cautery" continued to be used into this century: Ochsner wrote a paper entitled "Treatment of cancer of the jaw with the actual cautery" in 1923.¹⁰ The cautery he used was a simple soldering iron heated to red heat in a gas flame. He felt that it was important to hold the iron in place for at least a minute to destroy tissue up to 2 cm away. Also, he thought that the necrotic tissue thus formed stimulated the production of antibodies that attacked the cancer, a concept which reemerged

some 50 years later with the cryosurgical probe. However, by 1926 the diathermy had almost completely replaced the use of soldering irons, as it requires no protection for the surrounding tissues, and may be employed with greater facility.¹⁶⁵ The electrocoagulation was produced by a bipolar high-frequency current of the d'Arsonval type.¹⁶⁶ A further extension of the principle of the actual cautery is cryosurgery, which was first used for maxillary carcinoma around 1970.¹⁶⁷

An interesting illustration of the use of potential cautery is provided by a patient from Wales with a tumor of the palate who was eventually subjected to excision of the jaw in 1843, but who for some time had been under the care first of a "wild wart" doctor and then a wild wart doctress. These two practitioners had treated the tumor with external applications consisting of a mixture of clay, French brandy, and a caustic fluid, probably sulfuric acid.¹⁶⁸ The Welsh wild wart doctors survive to this day and still have a successful practice for the treatment of basal cell carcinomas of the skin using mixtures of this kind.

There was much discussion about the best way of removing the bone; one of the common methods was the use of the lion-jawed forceps designed by Sir William Fergusson (Fig. 2.6).²⁰ The use of the "chain saw" (i.e., Gigli's saw) was popularized by Davies in 1858⁴⁶ and Heyfelder in 1857.¹⁶⁹ The latter devised a blunt needle passed into the sphenomaxillary fissure to emerge in the zygomatic fossa, allowing a chain saw to be pulled through for division of the malar bone. He pointed out the advantages of the chain saw over the ordinary saw: the greater ease and rapidity with which the bones can be divided; the avoidance of splintering; and the facts that the parts are cut from behind forward, avoiding unintentional division by the saw, that corners can be rounded, and that the division of the bony parts can be effected in a very small space. He strongly criticized Desault's procedure of boring a hole into the antrum with a punch and enlarging it with a short curved knife ("instrument tranchant en forme serpette") because the walls of the antrum in many cases are not thinned. He clearly understood the principle of total excision for cancer when he stated that

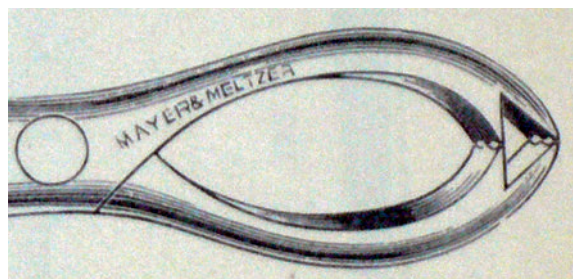


Fig. 2.6 A drawing from Meyer & Phelps's catalogue in 1931 of the distal end of Sir William Fergusson's lion-jawed forceps.

"all pathologists and operators on the upper jaw seem with one consent to deprecate the removal of tumors and especially cancerous with a sparing or niggardly hand, their usual counsel in practice being the extirpation of the whole jaw." Another commonly used means of dividing bone was the Hayes saw.¹⁵⁶

The speed with which these operations were performed can but leave us breathless. The length of the operation is only rarely recorded, but Hancock resected the entire upper jaw in 8 minutes in 1847,⁷⁸ and Key in 20 minutes in 1833.⁹⁶

Development of Surgery for Maxillary Cancer

This surgery developed in three phases: first, piecemeal removal of tumors, a phase lasting until 1825; second, the establishment of formal excision of the upper jaw beginning about 1825; third there followed the development of more refined procedures such as lateral rhinotomy in the latter part of the 19th and early 20th centuries.

The controversy as to who performed the first maxillectomy was most aptly summed up by Butcher: "the operations on the upper jaw may, in reality, be classed under two heads, that of exsection and that of disarticulation of the bone."³¹

Localized Removal

The first recorded partial removal of a maxillary tumor was that performed by Wiseman, surgeon to Charles II, reported in 1676:¹⁷⁰

A man about twenty-eight years of age came out of the Country recommended to me with a Cancer of his left Cheek, stretching itself from the side of his nose close under the lower Eye-lid to the external Canthus, so making a compass downwards. It was broad in its basis, and rose copped like a Sugarloaf. It gleeted, and was accompanied with Inflammation and much pain. He had also some scirrhus glands under that jaw. The extirpation of this Cancer had been attempted in the Country; but it growing afterwards bigger and threatening his Eye lately with inflammation, he hastned up, and importuned me to undertake it. I complied with his desire, and four or five days after having prepared all things ready, viz actual Cauteries, Digestives, Defensatives, Bandage, etc. Doctor Walter Needham and my Kinsman Jaques Wiseman being assisting. I pulled the Tumour toward me with one hand, during which I made my Incision close by the Eye-lid, and cut it smooth off as close to the Os jugulare as I could doe it, avoiding the Periosteum. The blood at first spurt out forcibly from many capillaries besides two considerable Arteries: we permitted them to bleed awhile. The lesser Vessels stopped

of themselves, and we cauterized the greater afterwards. Then viewing our work, and observing some relique of the Cancer remaining above the external Canthus, we consumed it by actual cautery, and dressed up the Wound with our Digestive, with Embrocations, Desensatives, and moderate bandage to retain them. The third day we took off Dressings, saw it well disposed to digest, and dressed it as before. The second day after, dressing it again, the Cancer appeared rising from the side of the nose and Eye-lid; it also overspread the Cheek-bone. I dressed it as I had done the time before, and the next time came prepared with actual cauteries, and consumed it all, then dressed it up with Lenients. From that time the Ulcer healed daily, and contracted in ten days space to the half; yet since then it begins to bud again here and there, which will put me upon a necessity of using the actual Cautery: and what account to give of it I yet know not.

According to Butcher a part of the upper jaw was removed by Acoluthus, a physician at Breslau in 1693. A woman had a turnout on the jaw after the extraction of a tooth. He enlarged the mouth with a cut, removed part of the swelling, together with four teeth, but was unable at once to get completely round it; "he attacked it several times at intervals of a few days, sometimes with cutting instruments, and sometimes with the actual cautery, and at last succeeded in curing his patient."³¹ In 1770, White described a turnout of the antrum of two years' standing. He removed it by a semicircular incision in the face, scooping away "matter like rotten cheese and many fragments of rotten bones"; the bony walls of the orbit were already destroyed. He preserved the eye, the optic nerve and part of the alveolus, but stopped at the dura which he could see and feel!¹⁷¹

Operations for tumors of the upper jaw were thus rarely attempted before 1800, and they are not mentioned at all in the standard 18th-century texts such as those by Bell, Heister, Hunter, and Pott.¹⁷²⁻¹⁷⁵ However, between 1800 and 1820 sporadic attempts were made with increasing frequency at localized excision of diseased tissue.

Localized removal of a turnout after elevating skin flaps, was performed by Dupuytren in 1818,¹⁷⁶ by Liston in Edinburgh in 1821,¹⁰⁵ by Irving a surgeon in Annan, Scotland on November 1, 1822,^{88,163} by Rogers in 1824 in New York,¹⁷⁷ by Ballingal in 1827 in Edinburgh,²¹ and by Velpeau of Paris in 1829 and 1830.¹⁷⁸ In all of these cases an incision was made in the face, the soft tissues of the cheek were elevated, and a tumor of the antrum was removed by traction on the tumor itself. No deliberate attempt was made to divide the bony attachments of the maxilla and such cases could not really qualify as maxillectomies.

Butcher also tells us that the scooping operation was practiced by Desault, Garengot, Jourdain, Plaque and others, and has been “in modern times more especially brought under notice by Dupuytren, in 1820, and since by many surgeons.”³¹ Although Dupuytren argued that the greater part of the jaw might be excised, he did not do the operation himself.

A similar operation was being performed as late as 1837 in Germany: Dieffenbach described 17 cases, but only one of these could be classed as a subtotal maxillectomy, the remainder being localized resections of tumors affecting the hard palate or alveolus. The exception was an osteoma probably arising in the ethmoid sinuses which he removed preserving the alveolus and hard palate.⁴⁷

Formal Maxillectomy

Guthrie in 1835 said that maxillectomy was one of the great improvements in modern surgery over the previous 16 years for which we were mainly indebted to the French.¹⁵ This statement suggests that the operation began to develop around 1820.

Lizars of Edinburgh, in 1826, proposed the entire removal of the superior maxillary bone, and described the procedure.¹⁰⁸ Speaking of “polypi, or sarcomatous tumors, which grow in the antrum,” he says:

All the cases which have come within my knowledge (with the exception of one) wherein these sarcomatous tumors have been removed by laying open the antrum, have either returned or terminated fatally. I am, therefore, decidedly of opinion, that unless we remove the whole diseased surface, which can only be done by taking away the entire superior maxillary bone, we merely tamper with the disease, put our patient to excruciating suffering, and ultimately to death. An incision should be made through the cheek, from the angle of the mouth backward or inwards, to the masseter muscle, carefully avoiding the parotid duct, then to divide the lining membrane of the mouth, and to separate the soft parts from the bone, upwards to the floor of the orbit; second, to detach the half of the velum palati from palate bone. Having thus divested the bone to be removed of its soft coverings, the mesial incisive tooth of the affected side is to be removed; then the one superior maxillary bone to be separated from the other, at the mystachial and longitudinal palatine sutures, and also the one palate bone from the other at the same palatine suture, as the latter bone will also require to be removed either by the cutting pliers or a saw; third, the nasal process of the superior maxillary bone should be cut across with the pliers; fourthly, its malar process, where it joins the cheek bone; fifthly the eye, with its muscles and cellular

cushion, being carefully held up by a spatula, the floor of the orbit is to be cleared of its soft connections, and the superior maxillary bone separated from the lacrymal and ethmoid bones with a strong scalpel. The only objects now holding the diseased mass are, the pterygoid processes of the sphenoid bone, with the pterygoid muscles. These bony processes will readily yield by depressing or shaking the anterior part of the bone, or they may be divided by the pliers, and the muscles cut with a knife. After the bone with its diseased tumor has been removed, the flap is to be carefully replaced, and the wound in the cheek held together by one or two stitches, adhesive plaster, and bandage. In no other way do I see that this formidable disease can be eradicated.

Lizars attempted the operation in December 1827 “for a medullary sarcomatous tumor of the antrum, from a miner or collier”, but had to abandon the operation because of bleeding. He tried again on August 1, 1829 and this time succeeded. He first tied the trunk of the temporal and internal maxillary arteries, and also the external jugular vein which had been divided in the first incision. He cut through the alveolar process and bony plate on the left side of the palatine suture, and completely separated the upper jaw with the saw, Liston’s forceps, and strong scissors, but the orbital plate was separated from the eyeball by the handle of the knife. The tumor was medullary sarcomatous, and a portion of it, attached to the pterygoid process of the sphenoid bone, could not be detached, but part of the malar bone involved in the disease was removed. On the 16th day the wound had healed and she left the house on that day. Three days after “she expired suddenly, but no examination was permitted.” He performed a further successful operation on 10 January 1830.¹⁷⁹

A very similar procedure was performed by Syme, also at the Edinburgh Royal Infirmary on May 15, 1829.¹³¹ He made a cruciate incision and, after exposing the tumor, divided the malar bone with a saw and pliers, divided the nasal process of the maxillary bone, and cut through the hard palate using cutting pliers after having extracted one of the incisor teeth. He therefore probably did the operation a few weeks before Lizars, although Lizars gave the first description.

The early French literature is reviewed very thoroughly by Gensoul in his monograph of 1833.¹⁸⁰ He describes operations performed by Garengot, Desault, and Dupuytren up to 1824. He records the great pains he took to find out what operations were actually performed, both by reading the contemporary accounts and by talking to those present at these operations. His research can be summarized as showing that all the procedures performed to 1827 consisted of an incision on the face followed by piecemeal removal of diseased

tissue; no formal excision had been attempted. Gensoul then described his own patient, a 17-year-old boy with a 2-year history of a swelling in the superior part of the canine fossa, which he described as a hyperostosis (Fig. 2.7). The tumor measured $7\frac{3}{4} \times 7\frac{1}{2}$ inches (197 mm $\times$ 190 mm) with a circumference of $16\frac{1}{4}$ inches (413 mm). After much thought and consultation with colleagues he embarked on an operation on May 26, 1827, at the Hotel Dieu in Lyon. After making three incisions in the skin of the face he elevated skin flaps. Then he used a mallet and chisel to divide the lateral wall of the orbit close to the frontozygomatic suture, passed the chisel as far as the pterygomaxillary fissure, and divided the frontal process of the zygoma. Next he applied a very large chisel to the inner canthus and passed it through the lacrimal bone. He divided the ascending process of the nasal bone in a similar manner. He used the knife to divide the soft tissues of the nasal ala from the maxilla, removed the first upper incisor on the left side, and divided the hard palate. Finally, to detach the maxilla from the pterygoid process,



Fig. 2.7 Pre-operative illustration of Gensoul's patient in 1827. The operation was performed without anesthesia. This was almost certainly the first total maxillectomy and was undertaken for an osteosarcoma.

he plunged the chisel through the orbit and through the tumor, dividing the superior maxillary nerve, and used the chisel to bevel the specimen into the mouth. Shortly afterwards the patient fainted, but ultimately recovered! This is clearly the first account of a deliberate excision of the upper jaw.

Gensoul also did at least six other similar procedures, some for cancer, one with a 5-year cure. Unusually for that time, he followed his patients for upwards of 5 years and also recorded the size of the tumors, and at one point frankly admits a diagnostic mistake! Even more unusually, he deliberately delayed publication for 6 years to assess the long-term effects. His monograph runs to 77 pages, and in addition describes excision of the lower jaw. Gensoul also acknowledged Lizars' claim to have done the first operation, an apparent reference to Lizars' *System of Anatomical Plates*, published in 1826.

In the early 1850s there was a fairly vicious correspondence under pseudonyms such as "studens chirurgiae" or "chirurgus"^{130,181} in the medical press about the question of who did the first maxillectomy: Lizars, Syme, or Gensoul. Who it was is of little consequence, except perhaps to Lizars, Syme, and Gensoul at the time! Such claims for scientific precedence are common: they tell us that the procedure was not a "maverick" out of its time, but rather that surgery had progressed to the point where the operation was feasible and several surgeons in different countries had decided to try it, indicating that the topic was one of general interest. The main countries contributing to this development were France and Great Britain and, to a lesser extent, Germany. The US Surgeon General's Catalogue tells us that the procedure did not spread to other European countries until the second half of the 19th century.¹⁸² It was first performed in Belgium in 1845 by Heylen,¹⁸³ in Poland in 1852 by Klose,¹⁸⁴ in Italy in 1857 by Gianflone¹⁸⁵ (a previous resection for necrosis had been reported in 1850 by Moretti),¹⁸⁶ in the Netherlands in 1857 by Leonides van Praag,¹⁰² in Austria in 1857 by Dehler,¹⁸⁷ in Portugal in 1862 by Barbosa,¹⁸⁸ in Russia in 1862 by Kade,⁹⁵ in Spain in 1864 by Rosa¹⁸⁹ (one case for necrosis had been performed by Toca in 1858¹⁹⁰), and in Finland in 1873 by Estlander.¹⁹¹

Resection of both upper jaws was first performed by Heyfelder in Erlangen, Germany in 1844.⁸⁴ A report was given in English by his son Oscar in 1857 in the *Dublin Journal of Medical Science*; Dublin being one of the main centers for this procedure, notably under Butcher, a name now forgotten. The operation was performed for a large "pseudo-plasma" arising from the palate, pushing the nose forward. He raised a large bilateral flap up to the inferior orbital margin and then formally excised both maxillae, preserving the nose. No attempt was made to provide a prosthesis. The patient returned to work but died 15 months later of a recurrence in the frontal

bone.¹⁶⁹ Oscar Heyfelder stated that the indications for the removal of both upper jaws included the following:

- Necrosis and caries
- Benign tumors including enchondroma and osteosarcoma
- Malignant tumors including epithelial cancer (canceroid of Virchow), cancer gelatiniforme, carcinoma medullare, and cystocarcinoma.

Incision

Many incisions have been described for maxillectomy, but they can be divided into two main types. The first is an incision passing from the outer canthus to the angle of the mouth. This was used in the early years—by Ballingal in 1827,²¹ Lizars in 1829,¹⁷⁹ Velpeau in 1832,¹⁷⁸ Key in 1834,⁹⁶ and Liston in 1835¹⁰⁶—but appears to have been abandoned by about 1840. The second is an incision passing down the lateral side of the nose. This was first used in 1827 by Gensoul¹⁸⁰ and has become the standard incision. Gensoul brought the incision through the upper lip at the level of the first incisor tooth, and Fergusson, in 1842, brought it through the upper lip in the midline.⁵³ The French school also developed a similar incision without division of the upper lip for partial operations on the upper part of the maxilloethmoidal complex, first described in 1865 by Legouest.¹⁹² A further lateral limb through the lower eyelid was soon added. Farabeuf ascribes to Blandin of Paris an incision running from the inner to the outer canthus at the level of the infraorbital margin,¹⁹³ to join the incision running down the side of the nose, but this incision is *not* included in Blandin's original paper of 1834.²⁶ An incision passing from the inner to the outer canthus within the lower eyelid and through the conjunctiva at the oculopalpebral fold was described by Michaux in 1854, the purpose being to prevent retraction of the lower eyelid.¹⁹⁴ The incision through the external surface of the lower eyelid just below the lashes is usually ascribed to Weber. However, the source of this attribution is a mystery: a careful search has failed to reveal any record of a description of this incision by Weber, and the reference to his work¹⁹⁵ relates to fractures of the jaws. The so-called Weber–Fergusson incision would be more accurately termed the Blandin–Gensoul incision (Fig. 2.8). The incision described by Dieffenbach, splitting the patient down the midline, did not catch on!⁴⁷

Ligation of the Carotid Arteries

In the earlier operations it was customary to ligate the common or external carotid artery before the operation. For example, Earle, in 1831, ligated the common carotid artery on the affected side, apparently with no ill effects,¹⁰ whereas Scon in 1830 tied the external carotid artery.¹⁹⁶ Heath, in his textbook *Injuries and Diseases of the Jaws*, tells us that this practice had been quite abandoned by the time of writing.¹⁶



Fig. 2.8 A drawing showing the so-called Weber–Fergusson incision, which should probably be more accurately termed the Blandin–Gensoul incision.

Partial Resections

Surgery of the upper jaw continued to be developed for a further century, almost exclusively by the French, who introduced the concept of surgery “*a la demande des lesions*.”³⁹

In 1925 Comet reviewed this development dividing the upper jaw into three stages: superstructure (the ethmoido-maxillo-orbito-malar complex), a naso-sinus mesostructure, and a palatal infrastructure. He discussed the embryological basis of this division and the main histological tumor types, pointing out that about half are squamous carcinomas. He then discussed the anatomical origin of these tumors—from the ethmoids, the nasal cavity, from the antrum itself, and from the hard palate and alveolus—and the route of spread.¹⁹⁷ He performed experiments with Sebileau on the cadaver, demonstrating that it was impossible to clear the ethmoids via a buccal or transantral approach, and therefore recommended the paralateronasal rhinotomy described by Moure as the operation of choice for tumors of the suprastructure. Sebileau in 1906 described the clinical forms of maxillary cancer (neoplastic, suppurative and putrid) and gave a description of the routes of spread of maxillary cancer into the cheek, nose, and mouth; into the nose through the inferior meatus; into the canine fossa through the anterior walls; through the socket of an extracted tooth to appear on the upper alveolus; into the orbit through the superior wall; and into the pterygomaxillary or pterygoid fossa through the posterior wall.¹⁹⁷

Superstructure

In the early years it was thought necessary to resect all of the upper jaw, because the methods of investigation had not allowed the exact point of origin and extent of the malignant tumor to be determined. However, as early as 1848 Michaux was questioning whether it was necessary to excise the hard palate when it was healthy.¹⁹⁸ He described partial operations for ethmoidal tumors and also preserved the floor of the orbit to maintain the function of the eye.¹⁹⁴ He also stressed the need to exclude extension of the tumor into the cranial cavity. In his monograph of 1854 Michaux described seven different procedures:¹⁹⁴

- Ablation of the maxilla and malar bones
- Ablation of the maxilla alone
- Removal of the upper portion of the maxilla preserving the hard palate
- Removal of the inferior portion of the maxilla conserving the floor of the orbit and the ascending process of the maxilla
- Removal of the palatine arch
- Resection of the upper alveolus
- Removal of both maxillae.

In 1865 Legouest made an incision from the inner canthus descending along the nasal ala as far as the center of the upper lip. He then opened the left nostril widely and retracted the nose toward the healthy side after having divided the articulation of the nasal bone with the ascending ramus. Finally, he turned the internal wall of the maxillary sinus outward using scissors, after dividing the ascending ramus and the external and inferior part of the anterior opening of the nostril.¹⁹² He did this operation for a boy with a nasopharyngeal polyp, presumably an angiofibroma. Until then these tumors had been treated by total maxillectomy with sacrifice of the orbit, but Legouest made a plea for a more conservative approach.

Cornet tells us that Michel of Nancy in 1869 modified the operations of Michaux and Legouest by omitting the resection of the maxilla itself and by adding an incision perpendicular to the vertical incision to allow partial excision of the orbital rim.¹⁹⁷

The next development was a lateral rhinotomy described by Moure (Fig. 2.9) in 1902 as a radical intervention for malignant tumors arising from the upper part of the nasal fossa or from the ethmoid. He tells us that the orbital route had been previously recommended for approaching tumors of the upper jaw but advocated a different approach as follows. The nose was turned aside after making an incision from the lower part of the frontal bone to the nostril, the nasal bone being exposed using a periosteal elevator. He divided the ascending process of the maxilla and a part of the lacrimal bone after first elevating and retracting the “membranous nasal canal” so as not to damage it. He then divided the nasal bone



Fig. 2.9 Emile-Jean Gabriel Moure (1855–1914), inaugurator of the lateral rhinotomy.

within the nose, and finally the nasal spine of the frontal bone. To avoid opening the cranial cavity he passed a gouge parallel to the cribriform plate as far as the anterior wall of the sphenoid sinus, removing the ethmoids with a large curette working from below upwards. This step was performed using illumination from a forehead mirror. The operation finished with curettage of diseased areas of the septum, the orbit and sphenoid sinus. He counseled conserving the ridge of the nasal bone to preserve the shape of the nose, and advised packing the postnasal fossa at the start of the operation to prevent inhalation of blood. However, he said that bleeding usually stopped after removal of the tumor, and packing was then no longer necessary. He pointed out that it is possible to reach the sphenoid sinus by this route.¹⁹⁹

His first patient, a cooper of 55, underwent the operation described above on July 9, 1901. One year later the patient was alive and well with no recurrence. Histology showed an “epithelioma cylindrique.” Moure did not describe what he meant by this term, but the tumor is described fully in a later French paper by Hautant in 1933. From this description, including the fact that it arises from the olfactory mucosa, and from the accompanying

woodcut, it is clear that “epithelioma cylindrique” would now be called an olfactory neuroblastoma. Indeed this tumor was fully described by the French.⁵

Sebileau further refined this procedure under the name of paralateronasal rhinotomy, emphasizing certain technical details and dividing rhinotomy into high, low, or total.²⁰⁰ Moure's lateral rhinotomy was extended further by Hautant²⁰¹ in 1933 using an incision beginning above at the same point as Moure's incision, but then extending laterally beneath the eye. He used chisels to excise all the anterior part of the wall of the maxilla plus the floor of the orbit. This monograph was the first to include a description of the radiological appearances of these tumors.

The French were also the first to point out that sacrifice of the floor of the orbit is excessive if it is not invaded, because the physiological suspension of the eye is lost; “an eye not lying in its correct place is an eye lost.”

It is interesting that St. Clair Thomson said in 1937 that lateral rhinotomy had quite replaced excision of the upper jaw,²⁰² and yet by 1977 it was described as a neglected operation.²⁰³

Mesostructure

Cornet in 1925 described unusual tumors that destroy the nasoantral wall and early invade the maxillary antrum.¹⁹⁷ These cases, he says, are suitable for a procedure that preserves both the floor of the orbit and hard palate. He describes a procedure that he terms an extended Caldwell–Luc antrostomy: the initial incision in the gingivobuccal sulcus between the canine and second molar tooth is extended and the entire anterior wall of the sinus is resected. Thereafter, the tumor is removed by careful and meticulous curettage of the antral cavity, whose other walls are assessed for erosion.

This was only a minor modification of the operation described by Denker in 1909.²⁰⁴ After retracting the upper lip, Denker made an incision in the upper buccal sulcus of the affected side and for 2–3 cm on the opposite side. The soft tissues were then elevated from the face up to the orbital rim. He opened the maxillary antrum through the canine fossa and removed the lateral nasal wall with Luer's forceps and chisel. The lower part of the nasal bone and the nasal process of the maxilla were also resected. If necessary, he cleared the ethmoidal labyrinth and removed the anterior wall of the sphenoid.

Cornet also described a similar procedure¹⁹⁷ for the excision of malignant tumors arising from the inferior part of the nasal fossa (the septum and the inferior turbinate), which he ascribes to Rouge but sadly gives no reference. A careful search has failed to reveal where this procedure is recorded. The steps are as follows:

- A horizontal sublabial incision extending from one first molar to the other in the gingivolabial groove.
- Exposure of the nose: the curved periosteal elevator is used to denude the bone toward the bony orifice of the

nostrils, which it exposes on the lateral part of their circumference, exposing the anterior and inferior nasal spine.

- Division of the quadrangular cartilage from below upward using the scissors, and of the nasal spine allowing the superstructure of the nose to be elevated.
- Pterygomaxillary disarticulation, using a special shears curved on the flat.
- Extraction of the block held by a Farabeuf's forceps.

Infrastructure

Partial procedures for palatal tumors were described in 1854 by Michaux.¹⁹⁴ He describes resection of the upper alveolus, which he divided above the roots of the teeth with small scissors. He also describes in some detail an operation described by Nelaton of Paris, but unfortunately does not give a reference. Nelaton first elevated the palatal mucosa, providing of course that it was not diseased, and next made several holes at the anterior end of the hard palate. He introduced one blade of the scissors through this hole, dividing the hard palate and inferior edge of the septum.

Farabeuf generally made a transverse incision on the face to expose the upper alveolus. He perforated the anterior wall of the antrum to allow scissors to be introduced to cut the attachments of the alveolus.⁵² He too ascribes to Nelaton the midline incision in the hard palate, but gives no reference to where Nelaton's work may be found.¹⁹³

Barwell in 1873 removed the hard palate and alveolus from within the mouth, without opening into the nasal cavity.²³ He said that he could find no account of such an operation in any surgical work or journal, completely ignoring the fact that Michaux had given a very full description of this procedure in an extensive monograph 20 years earlier. Barwell's procedure of transoral palatotomy appears to have been empirical, rather than a systematic development based on the study of anatomy and pathology as was the case with the developments described by the French. The final development was that described by Cornet for tumors of the infrastructure. This operation was redescribed in the English literature a quarter of a century later by Wilson, who gave no credit to the French either for describing the operation or for describing the anatomical and pathological principles upon which it is based.¹³⁹

Osteoplastic Procedures

The only other contribution from the German school, apart from resection of both maxillae and Denker's sublabial approach, was the development of an osteoplastic approach, originally described by Langenheck in 1859.²⁰⁵ His patient was 18 years old, with two fibrous polyps, probably an angiofibroma of the nasopharynx. He first made a skin incision, much like that used for lateral rhinotomy, and divided the nasal cartilages from their

attachments. Using a cutting bone forceps he divided the nasal bone close to the septum up to the nasal process of the frontal bone. A second incision divided the bone of the nasal process and continued into the maxillary sinus, ending at the attachment of the nasal process of the maxilla where it forms the lower border of the orbit. He used an elevator to turn the bone back on to the forehead.

In 1863 Voelckers described an osteoplastic flap, pedicled superiorly, of the anterior wall of the antrum for removal of a tumor—again probably an angiofibroma—invading the antrum from the region of the sphenoid sinus.²⁰⁶

Combined Irradiation

In the early years of the 20th century maxillectomy was also combined with the introduction of radium into the cavity. This method was popular both in England and the United States.^{165,166,207,208} Jacketed tubes, steel points, or emanation seeds were used. The radium was applied directly to the tumor using a 50- or 100-mg tube within a silver tube for 15 to 20 hours.²⁰⁹ An alternative was radium needles contained in a dental plate molded to fit the cavity.²⁰⁷

Prostheses

It has been customary since the earliest days to fill the defect left by maxillectomy with some form of dental prosthesis. A prosthesis was made after one of the very first maxillectomies, performed by Syme in 1835: Nasmyth made an artificial plate and a set of teeth that rendered the patient's appearance, mastication, and articulation "hardly at all defective."¹³² Hart in 1862 tells us of a patient undergoing maxillectomy for scrofula for whom an artificial set of teeth was made by Hart's brother, a dentist.⁸¹ Baker, a surgeon/dentist to Dr. Stevens Hospital in Dublin, "arranged the palate and dental apparatus in a most satisfactory manner so that the patient was enabled to eat with comfort and to articulate with perfect distinctness."¹⁰⁹

By the early years of the 20th century the technique of dental restoration was well developed. Woodman describes how a plaster cast must be taken a few days before operation, to allow a temporary denture to be made with a bulbous extension to fit the cavity and prevent prolapse of the cheek. A permanent vulcanite splint, bearing teeth, is made a few months later (Fig. 2.10).^{210,211}

Summary

Despite the dramatic technical innovations developed during the last half of the 20th century within the fields of chemotherapy, anesthesia, illumination, and instrumentation, possibly the only new major surgical procedure relevant to upper jaw neoplasia has been the craniofacial resection.



Fig. 2.10 A photograph of a permanent dental obturator (after Woodman 1923²⁰⁹).

A search through the original European literature has shown that during a period of some 50 years (1825–1875) most of the operations viewed today as standard were developed without the assistance of adequate illumination, blood replacement, anesthesia, and so on by a small number of exceptionally gifted and determined surgeons within a small number of European countries. The fact that any patients survived these traumatic experiences is a tribute to these surgeons' skills as well as the patients' own forbearance.

Despite the best efforts of the early pioneering surgeons and radiotherapists, the majority of patients with malignant disease of the nose, paranasal sinuses, and nasopharynx subsequently succumbed to their disease with high rates of locally recurrent disease within the structures of the adjacent anterior and antrolateral skull base, infratemporal fossa, clivus, orbit, and anterior and middle cranial fossae. The anatomy of the sinuses and skull base had been extensively studied and described by the end of the 19th century, notably by Onodi (Fig. 2.11a, b). However, it was not until the second half of the 20th century, with its dramatic technical innovations within the field of anesthesia, illumination, instrumentation, Hopkin's rod telescopes (Fig. 2.12), and the operating microscope, that it became possible to develop new major skull base surgical procedures, most notably the wide variety of craniofacial approaches and resections.

These new operative procedures were considerably aided by improved understanding and preoperative evaluation of the extent of both benign and malignant tumors by means of computed tomography (CT), introduced in the 1960s, and magnetic resonance imaging (MRI) introduced in the 1980s. Improved understanding of the pathological process and natural history of each disease improved our understanding of preoperative considerations in regard to treatment of the individual pathologies.

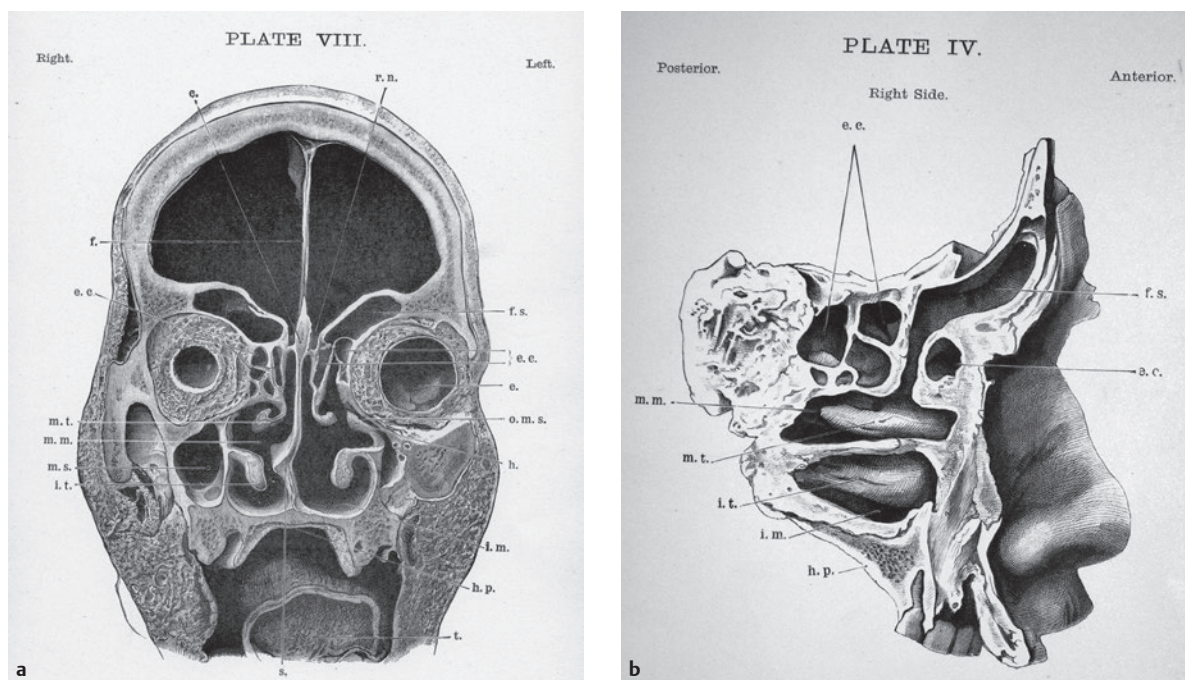


Fig. 2.11a, b

a Plate VIII Coronal section made vertically through the nose in its posterior third. (From "Dr. Onodi's Atlas of the Nasal Cavity and Sinuses" 1894, translated by St. Clair Thomson.)

b Plate IV Sagittal section through the right side of the nose external to the plane of the middle and inferior turbinates. (From Dr. A. Onodi's Atlas 1894, translated by St. Clair Thomson.)



◁ **Fig. 2.12** Harold Hopkins, Professor of Optical Physics at Reading University, who developed his solid rod lens system in 1954, which allowed the development of modern endoscopic sinus surgery by Professor Walter Messerklinger of Graz.

Skull Base Surgery

Pituitary Surgery

Sir Victor Horsley is believed to be the first person to undertake in 1899 a transfrontal approach to a substantial hypophyseal tumor.²¹² The technique of this operation remains the basis of the approach still used today. Just prior to Horsley's work, an Italian surgeon, Giordano,²¹³ approached the pituitary by turning aside the nose and frontal sinus as an osteoplastic flap and then removing the nasal septum, turbinates, and sphenoid to gain access. Schloffer in 1906²¹⁴ described the first purely transnasal approach, also removing the septum and turbinates to gain access through the sphenoid, but it was really Cushing in 1909²¹⁵ who developed this procedure over the following two decades. He described a sublabial approach coupled with a submucous resection of the nasal septum and preserved the nasal structure and function, developing the transeptal/transsphenoidal operation that is used by many to this day.

Dandy²¹⁶ is credited with the first reported anterior craniofacial resection which commenced as an

orbitocraniotomy through an anterior approach, but he entered the ethmoid block to achieve resection of the tumor. Smith, Klopp, and Williams described the anterior craniofacial excision of a frontal sinus carcinoma in 1954.²¹⁷ It became increasingly well-recognized that the poor prognosis associated with malignant tumors of the nose and paranasal sinuses was a consequence of local recurrence, often related to inadequate resection. The realization that every tumor affecting the inferior surface of the cribriform plate and the roof of the ethmoid could spread intracranially became even more apparent with the advent of CT scanning. Thus, the principle of a combined craniofacial procedure became of increasing interest and was subsequently developed, most notably by the report of Ketcham and colleagues in 1963.²¹⁸ This initial report was subsequently added to by Ketcham et al in 1966 and 1973,^{219,220} in Australia by Millar in 1973,²²¹ in the UK by Peter Clifford in 1977²²² and in the USA by Schramm, Myers & Maroon in 1979²²³ and Terz et al in 1980.²²⁴

Our own experience at the Royal National Throat, Nose and Ear Hospital commenced with Sir Donald Harrison, who had a life-long interest in tumors of the nose and paranasal sinuses, reporting their management notably in 1973 and developing his rationale with his great friend George Sisson from the United States, who reported his 15-year experience in 1976.²²⁵ Harrison's personal series of 85 patients with sinonasal tumors, which he reported in 1973,²²⁶ led him to believe that ~80% of all patients with antroethmoidal tumors would have extension of their neoplasm up to and beyond the limits of conventional total maxillectomy with or without orbital exenteration. He encouraged Tony Cheesman, who already had an extensive training in ENT, skull base, and neurosurgery, to commence craniofacial resection in 1978 (Fig. 2.13). Our 7-year experience of 60 patients was subsequently reported in 1986.²²⁷

While the craniofacial procedure that we developed involved the use of a small anterior window craniotomy (Fig. 2.14), minimal dural retraction, and resection under operative microscope control, other colleagues wished to improve the exposure of the posterior extent of the anterior cranial fossa without retraction of the frontal lobes and the fronto-orbital band was removed in a procedure developed from craniofacial surgery for congenital craniomaxillary anomalies introduced by Tessier et al.²²⁸ The extended anterior subcranial approach was developed by Raveh initially to manage intracranial trauma but subsequently to allow tumor resection, reported in 1993.²²⁹

There was an explosion of interest in the anterior approaches to tumors of the nose, paranasal sinuses, and skull base in the 1980s and many of the approaches to the skull base began to blend in with intraoral approaches and more lateral based approaches such as the infratemporal fossa approach pioneered by Fisch,²³⁰ and Sekhar and Schramm.²³¹ Many institutions formed multidisciplinary



Fig. 2.13 Tony Cheesman, who has been the main promoter of craniofacial resection in the United Kingdom.

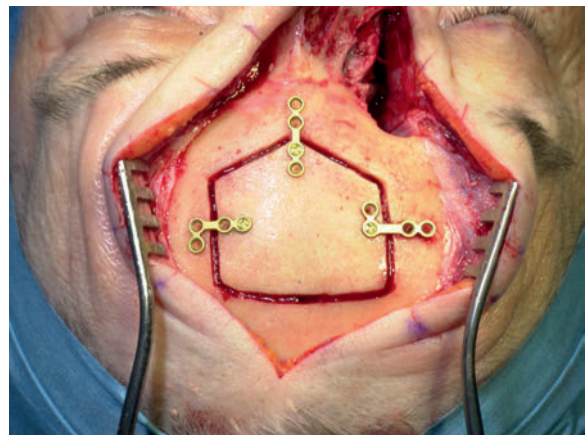


Fig. 2.14 Anterior craniofacial operation. Intraoperative photograph showing initial placement of 1.7 mm microplating prior to removal of the shield-shaped craniotomy window. This allows precise replacement of the window at the end of the craniofacial procedure.

teams around the world that continue with this work to the present day, and it becomes invidious to single out individuals and institutions as a wide variety of people continue to contribute significantly to this advancing area

of surgery. However, it would be remiss not to mention the contributions of Cocke, Derome, Donald, Jackson, Janecka, and Maniglia,^{232–237} who have all made substantial contributions to the wide variety of approaches now available for tumors in this region.

Endonasal and Endoscopic Surgery

Endonasal surgery has its main origins in Germany, notably with four generations of doctors in the Heerman family in the 20th century. Hans Heermann²³⁸ demonstrated the use of the binocular Zeiss operating microscope in 1957 for endonasal ethmoid surgery. By 1974 this endonasal microscopic technique was reported to have been used in 14,000 ethmoid, maxillary, sphenoid, and frontal sinus procedures.²³⁹

Hirshman was reported to be the first surgeon to attempt nasal and sinus endoscopy using a modified cystoscope based on the design of Nitze in 1897.²³⁸ This endoscope was made by Reiniger, Gebbert, and Schall in Berlin, Germany. The endoscope was initially used for diagnosis and minor procedures such as cautery and removal of foreign bodies, but essentially these rigid telescopes and other derivatives had proximal illumination with a small bulb and were used for diagnosis.

Harold Hopkins's invention of the rod endoscope, manufactured by Storz, facilitated the major developments that began in the 1960s and 1970s and this coincided with the development of endonasal surgery using the binocular operating microscope, first reported by Heerman in 1958.²³⁹ Draf also used the microscope but additionally combined this with an angled endoscope.^{240,241}

Endoscopic sinus surgery was first reported in the European literature in 1967 by Messerklinger,²⁴² Wigand,²⁴³ and Stammberger.²⁴⁴ The latter in particular further expanded and popularized the technique that Kennedy introduced into the United States in 1985.²⁴⁵ With an abundance of new instruments to complement the improved endoscopes, it was only a matter of time before benign and malignant tumors were approached using this method.

During the last three decades, the considerable improvements in detailed scanning by CT and MRI of the skull base and recently image guidance systems, have allowed skull base surgeons to approach deeply seated tumors using minimal-access endoscopic techniques. Extended endonasal endoscopic surgical procedures are increasingly described and employed and the feasibility and low morbidity of these extended approaches have now been well established and reported in numerous studies. These expanded endonasal approaches are continuously being used throughout major centers around the world as a new method for sinonasal and skull base surgeons tackling these tumors. It is now possible to gain access to anterior, middle, and posterior cranial fossa



Fig. 2.15 Operating theater set-up for endoscopic sinus surgery (for a meningoencephalocele).

structures. The use of extended endoscopic approaches instead of traditional open surgery is limited by the experience of the surgical team involved and the relationship of many tumors to critical neurovascular structures (Fig. 2.15).

As with the open approaches, it is invidious to try to single out all colleagues who have made contributions to the development of this surgery, but the recent European position paper on the endoscopic management of tumors of the nose, paranasal sinuses, and skull base gives an excellent overview of the subject and its development by colleagues Carrau, Castelnovo, Kassam, Lund, Nicolei, Stammberger, Stamm, and Wormald, who have made and continue to make contributions to this important area.²⁴⁶ Interestingly, the two-person, four-handed technique was first promoted by May in 1990²⁴⁷ but has been extensively popularized more recently.^{248–250}

Multiple centers have now concentrated on all aspects of skull base surgery with an increasing emphasis on endoscopic approaches but with the retention of the wide variety of open procedures that remain necessary in combination with new developments in radiotherapy, chemotherapy, and the recently introduced novel targeted pharmacotherapies that antagonize cellular proliferation by interfering with specific cellular processes. While much is hoped for these latter therapeutic agents, guided by our increasing understanding of molecular factors, history teaches us that the early demise of surgery as the main treatment of cancer is a considerable way off as a consequence of the complexity of malignant disease. Our hope is that this publication will continue to guide surgeons and promote an increasing understanding of these diseases in a multidisciplinary team, but our expectation is that the necessary skills will be required for the present generation of young surgeons.

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Section IA Nose and Paranasal Sinuses

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3 Surgical Anatomy

The complex anatomy of the nose, the paranasal sinuses, and the adjacent nasopharynx and intracranial and intraorbital structures has been explored in considerable detail from the seminal works of Johannes Lang and Heinz Stammberger in several recent publications¹⁻⁸ and it seems inappropriate to reinvent the wheel. However, an intimate knowledge of the anatomy (and physiology) of the area is a prerequisite to understanding the natural history of tumor pathology and its treatment.

Intrinsic areas of weakness exist throughout the area (Fig. 3.1). The lamina papyracea is well named and may be dehiscent in the young and old, though the orbital periosteum is fortunately resistant (Figs. 3.2 and 3.3). Unfortunately, even when the orbital periosteum resists tumor spread, disease can run extraperiosteally to the apex and thence into the middle cranial fossa. The superior and inferior orbital fissures also offer routes of tumor exit and entry. The inferior fissure communicates with the pterygopalatine fossa medially and the infratemporal fossa laterally while the superior fissure leads to the cavernous sinus.

Similarly, the dura is relatively robust even when tumor infiltrates the bone of the skull base. Areas of vulnerability are found, however, in the cribriform plate with the many fenestrations for the olfactory fibrils connecting to the bulbs and tracts, together with dural prolongations and emissary veins to the sagittal sinus. The length and depth of the cribriform niche varies considerably (length

15.5 to 25.8 mm; depth 0 to 15.5 mm). The roof of the ethmoids is largely composed of hard frontal bone (Fig. 3.4) but the lateral lamina of the cribriform plate is effectively an extension of the vertical attachment of the middle turbinate, the depth of which has been divided into three using the Keros-Kainz classification. As this bone is very thin, it represents an easy route into the anterior cranial fossa. This situation is further compounded by the route and foramina of the anterior and posterior ethmoidal neurovascular bundles offering access to the orbit. The anterior ethmoidal artery is most vulnerable, usually running across the anterior skull base posterior to a suprabullar cell, often in a mesentery of mucosa or in a dehiscent bony canal. The posterior ethmoidal artery is generally more protected, running within the bone of the roof.

The sphenoid has important relationships with the optic nerve, carotid artery, and pituitary (Fig. 3.5). These structures vary in their prominence in the sinus walls and in the thickness of bony covering, which is dependent on the shape of the sinus cavity and its degree of pneumatization. The cavernous sinus also lies laterally and the foramen rotundum (V2) and pterygoid canal may impinge on the sinus cavity, especially if well pneumatized. The intersinus septum is often asymmetric and can attach to the lateral wall in the region of the carotid. Posterior to the jugum of the sphenoid, lies the optic chiasm (mean distance 21 mm).² In 5 to 12% of the population,

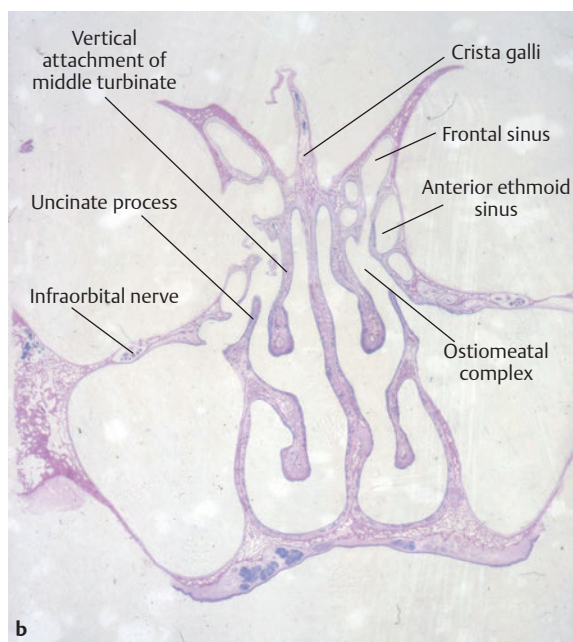
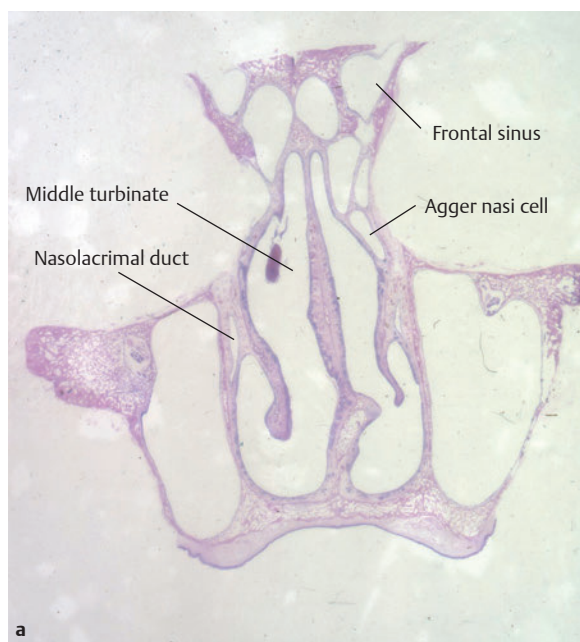
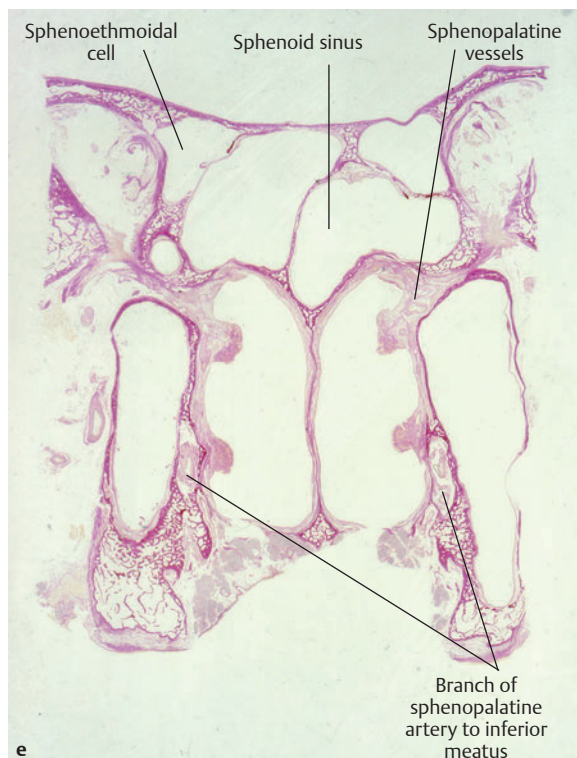
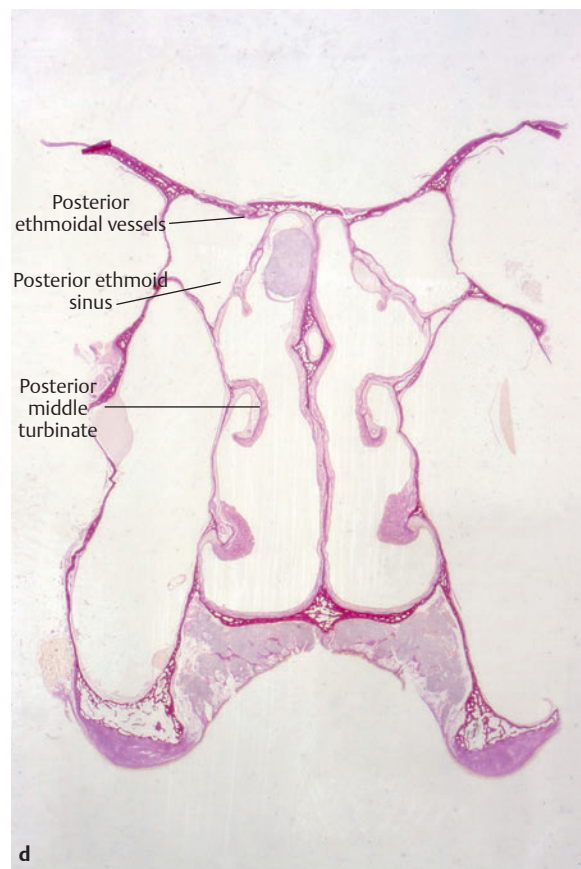
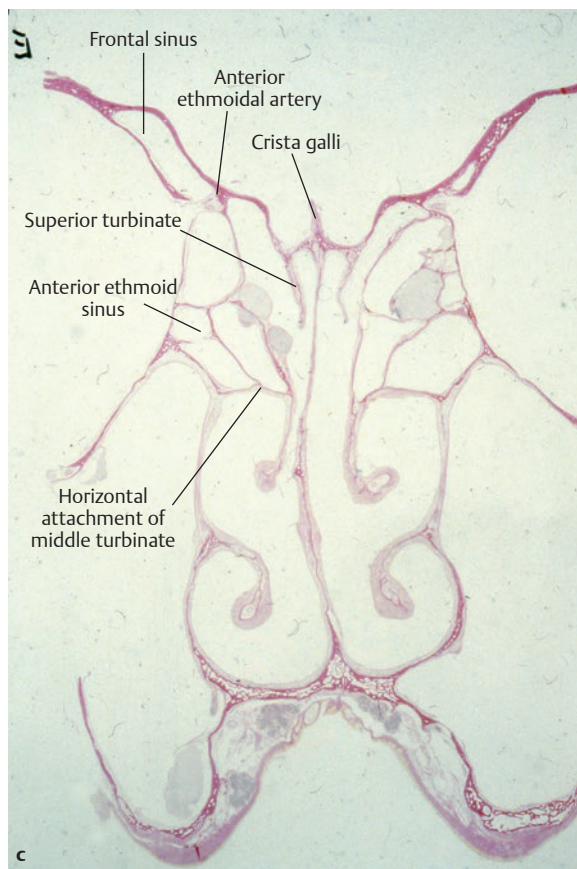


Fig. 3.1a–e Coronal sections through a midfacial block; hematoxylin and eosin. Anterior to posterior.

Fig. 3.1c–e ▶

◀ continued



the posterior ethmoids may extend superiorly and laterally to the sphenoid, forming a sphenoethmoidal cell. The optic nerve and carotid artery are usually found in the lateral wall of these cells when present, rendering these structures at risk from disease and surgery.

The maxillary sinus also has areas of potential weakness, notably the medial wall through the maxillary hiatus, which in life is closed by the inferior turbinate, uncinate process, and bulla of the ethmoid, lacrimal, and perpendicular plate of the palatine (Figs. 3.6, 3.7, 3.8, 3.9, 3.10, 3.11). However, areas without bone such as the natural ostium, anterior and posterior fontanelles, and accessory ostia are easily breached by disease. In the sinus roof, in the infraorbital canal and foramen, and in the floor the dental roots provide access to the orbit, cheek, and oral cavity.

Attached to the posterior wall of the maxilla are the pterygoid plates, which are part of the sphenoid. The space between the plates and the sinus is the pterygo-maxillary fissure, through which the maxillary artery runs. This in turn connects with the pterygopalatine fossa and the infratemporal fossa. This is an area in which angiofibromas typically arise. The pterygopalatine fossa is divided into a neural component composed of pterygopalatine ganglion and maxillary nerve and a vascular

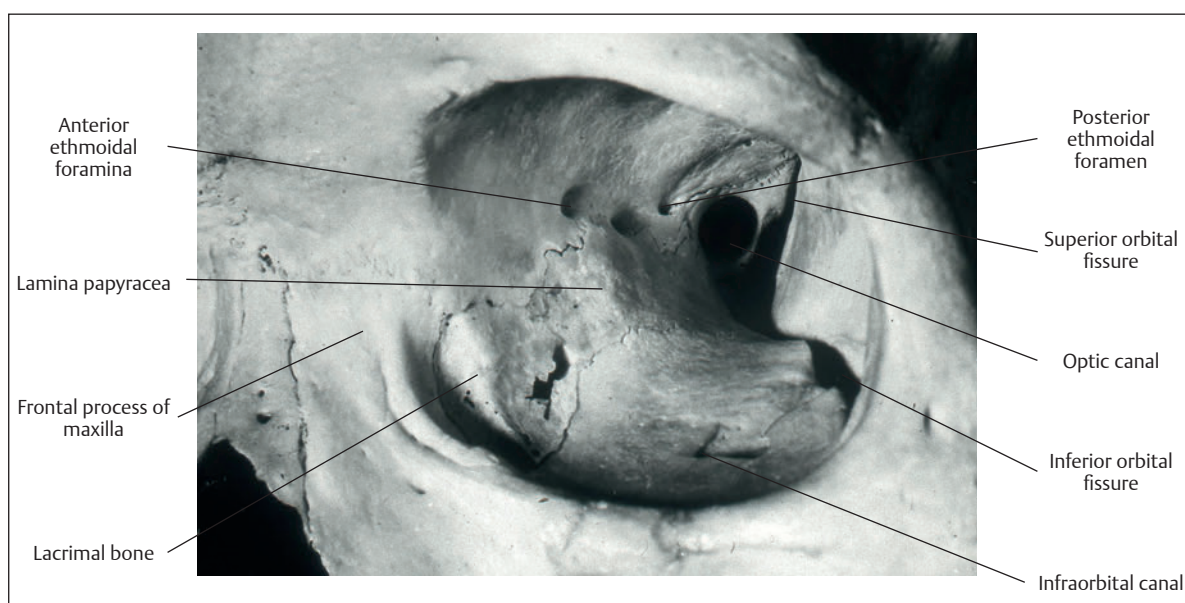


Fig. 3.2 Photograph of a left orbit.

component containing the terminal part of the maxillary artery and its branches. The infratemporal fossa lies beneath the skull base between the side wall of the pharynx and ascending ramus of the mandible. It contains the pterygoid muscles, branches of the mandibular nerve, the maxillary artery, and the pterygoid venous plexus in the lateral pterygoid muscle. While the bone of the posterior wall of the maxillary sinus is strong, once it invaded by a malignant tumor such as a squamous cell carcinoma, the excellent blood supply of these areas rapidly facilitates tumor spread and therefore has a significant detrimental effect on prognosis.

The frontal sinus is unique in size and shape to each individual once developed and may be absent in ~1% of a white population. As it arises embryologically from the anterior ethmoids, (Fig. 3.12) it has a complex anatomy with asymmetric septations and a range of cellular pneumatization that has been the subject of many classifications (Kuhn). The drainage into the middle meatus is also variable, more like an hourglass than a “duct” and is referred to as the frontonasal recess. However, this is influenced by the configuration of the agger nasi and suprabullar cells. Given the complexity of these clefts and the capacity for retrograde mucociliary flow back into the sinus, it is perhaps surprising that the frontal sinus is so rarely a primary site of neoplasia.

Nasal Septum

The nasal septum is composed of a small membranous area, the quadrilateral and vomeronasal cartilages with a contributions from the upper and lower lateral cartilages and bone, including the perpendicular plate of the

ethmoid, vomer, and crests of the maxilla and palatine (Figs. 3.3, 3.6, 3.9, 3.13). Primary tumors of the septum are rare, but it is one of the commoner sites of origin for chondrosarcoma, which can spread covertly into the skull base and/or palate. Anteroinferiorly the upper lip and gingivobuccal sulcus can also be affected.

External Nose

Tumors arising within the nasal cavity and vestibule or on the columella may readily spread into the bone and cartilaginous superstructure and can involve the subcutaneous tissues and skin. The nasal bones are united as a pair in the midline (Fig. 3.14) and are supported by the nasal spine of the frontal and perpendicular plate of the ethmoid. They have attachments to the frontal bone superiorly at the nasofrontal suture and with the frontal process of the maxillary bone on either side at the nasolacrimal suture. The bones themselves may be eroded, with a mass appearing at the glabella as can occur with adenocarcinoma. Inferiorly they form the pyriform aperture together with the maxillary bones, intersected by the quadrilateral septal cartilage.

In addition, upper and lower nasal cartilages form the malleable part of the lower external nose. The upper lateral cartilages lie inferior to and are overlapped by the nasal bones, the frontal processes of the maxillae, and the lower lateral cartilages in 72% of cases.¹ They are continuous medially with the septal cartilage. The lower lateral cartilages are composed of a medial and lateral crus. The medial crura contribute to the columella which lies in front of the septal cartilage. These cartilages and the

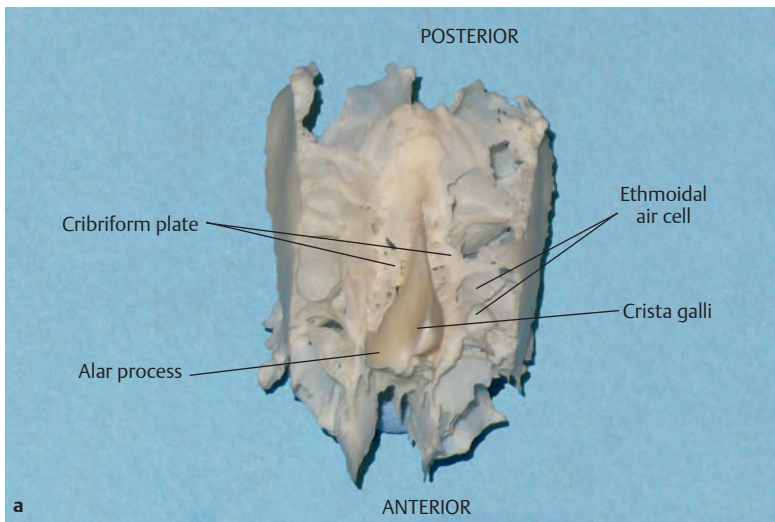
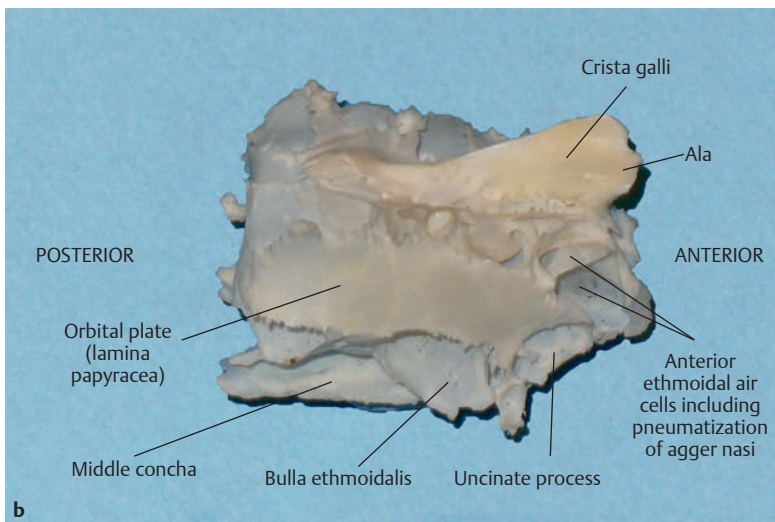
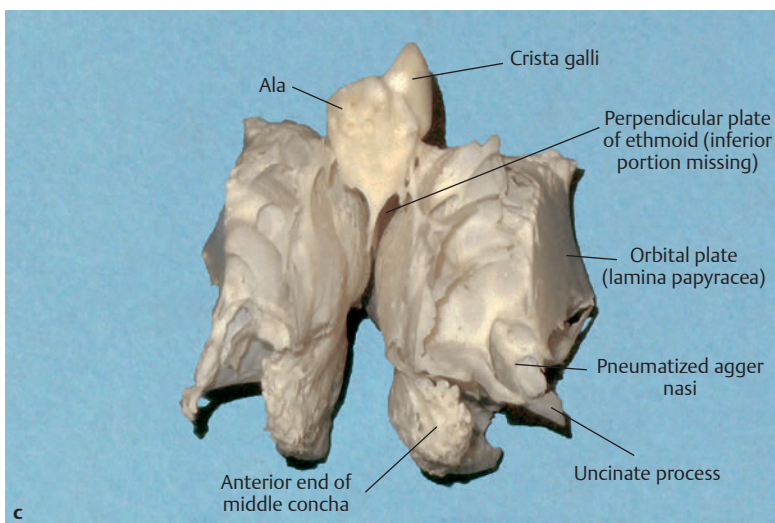


Fig. 3.3a–e Photographs of a disarticulated ethmoid bone.

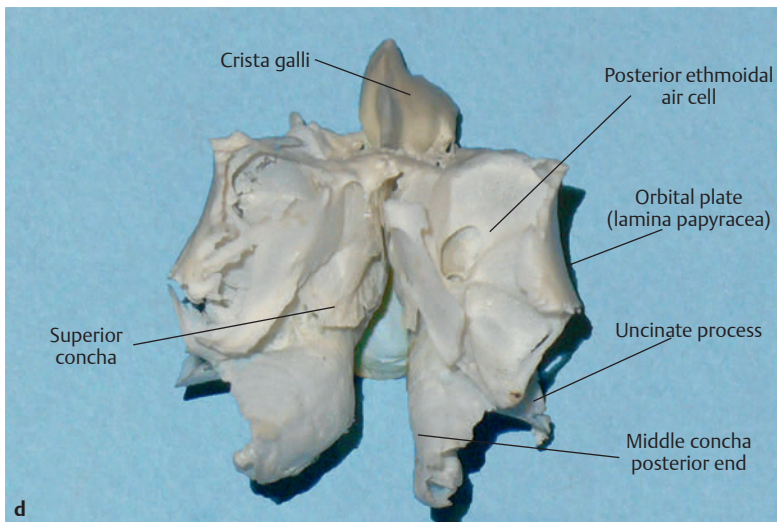
a Superior view.



b Lateral view.

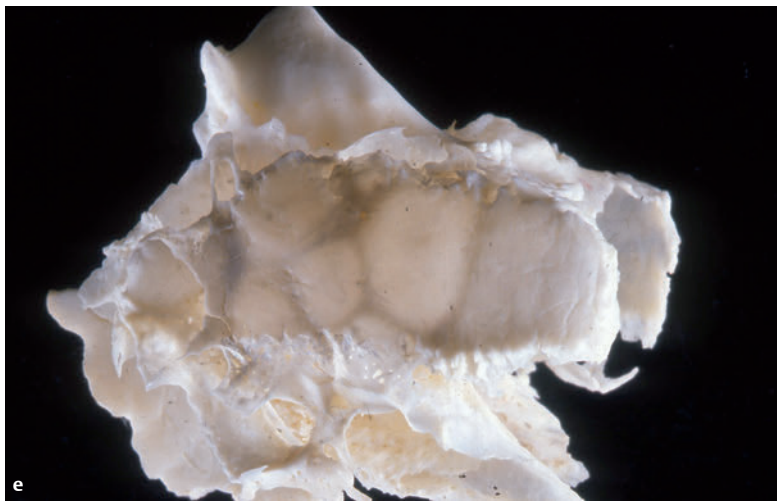


c Anterior view.



◀ continued

d Posterior view.



e Lateral view backlit to show lamina papyracea.

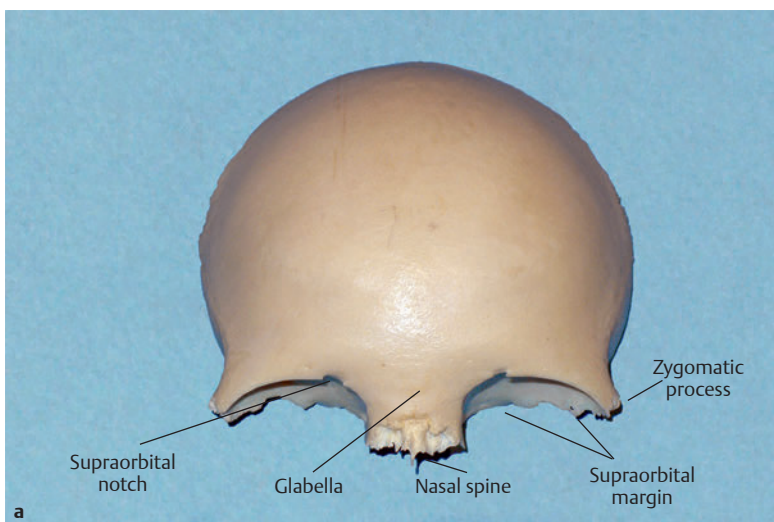
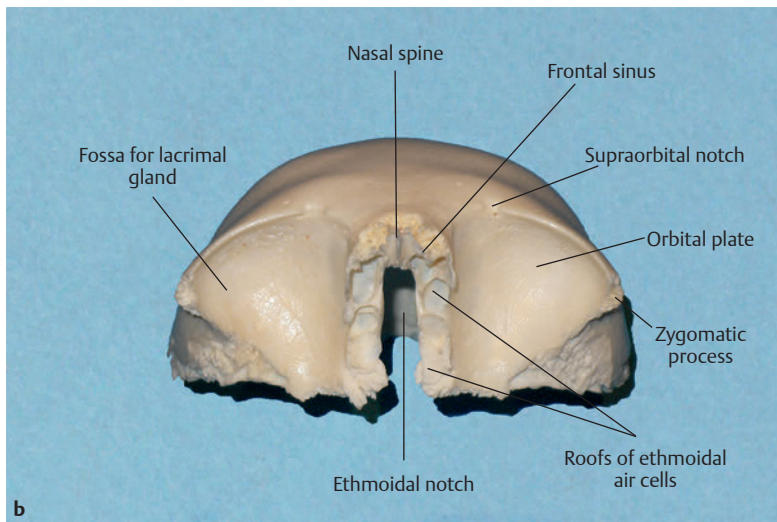


Fig. 3.4a, b Photographs of a disarticulated frontal bone.

a Anterior view.

Fig. 3.4b ▶



◀ continued

b Inferior view.

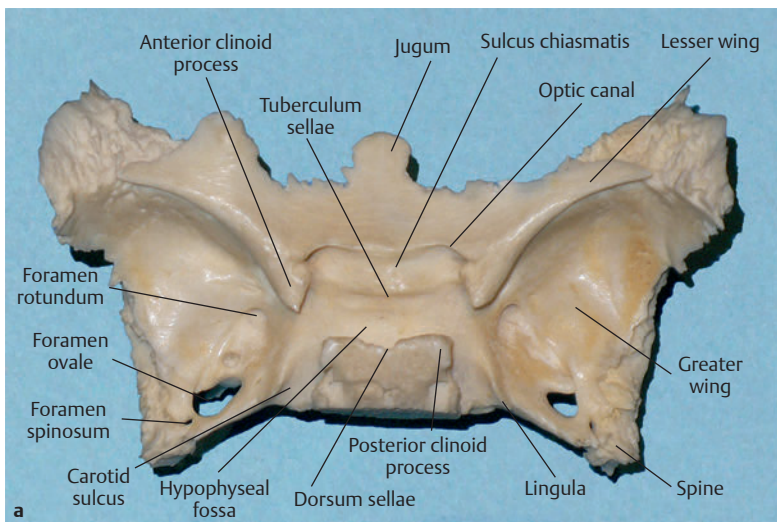
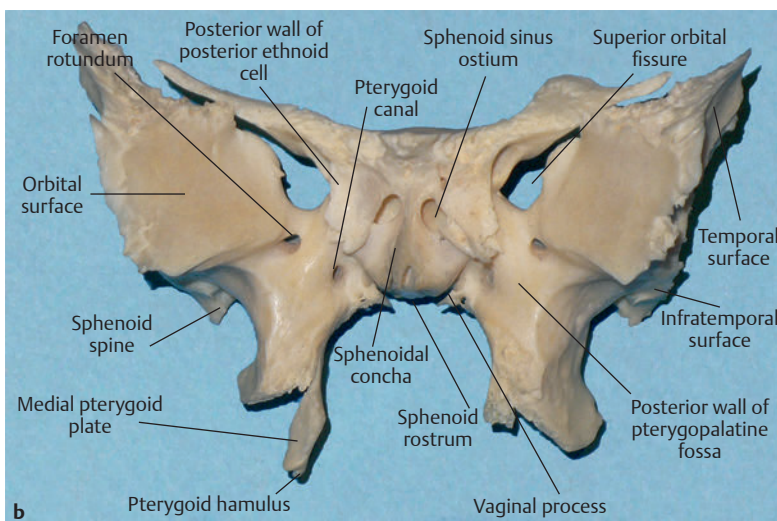
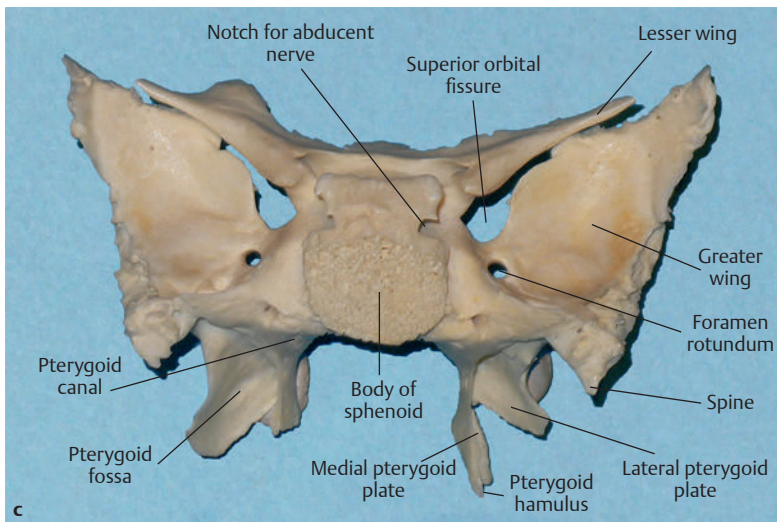


Fig. 3.5a–c Photographs of a disarticulated sphenoid bone.

a Superior view.



b Anterior view.



◀ continued

c Posterior view.

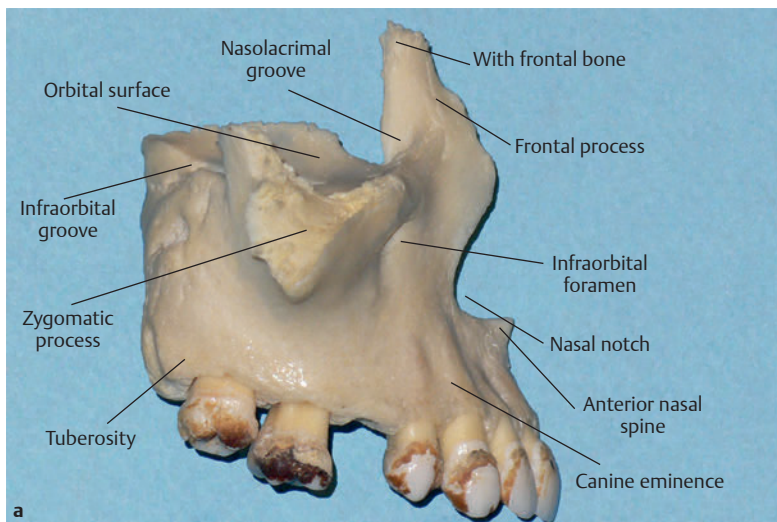
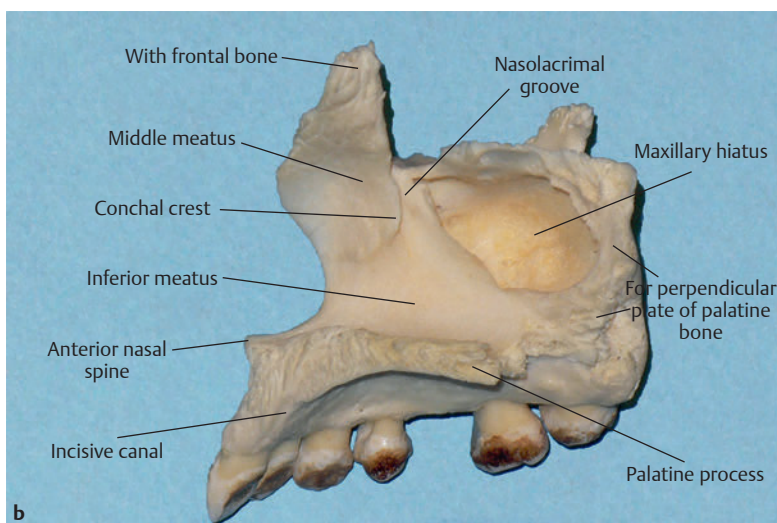


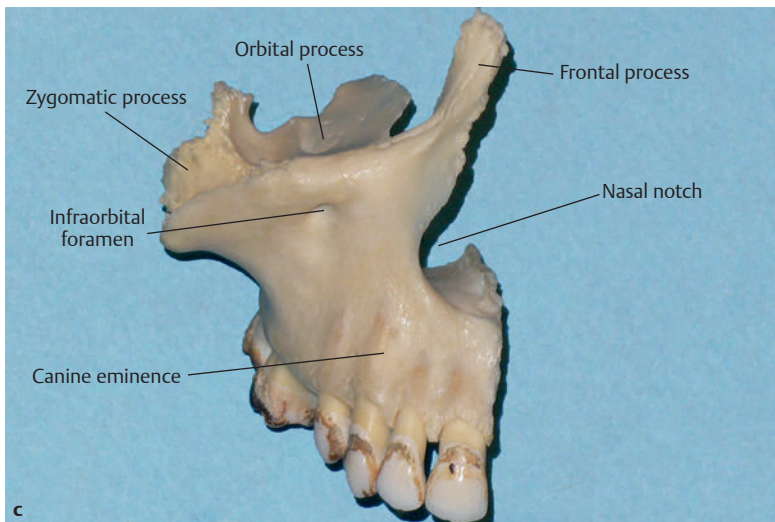
Fig. 3.6a–c Photographs of a disarticulated right maxillary bone.

a Lateral view.



b Medial view.

Fig. 3.6c ▶



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c Anterior view.

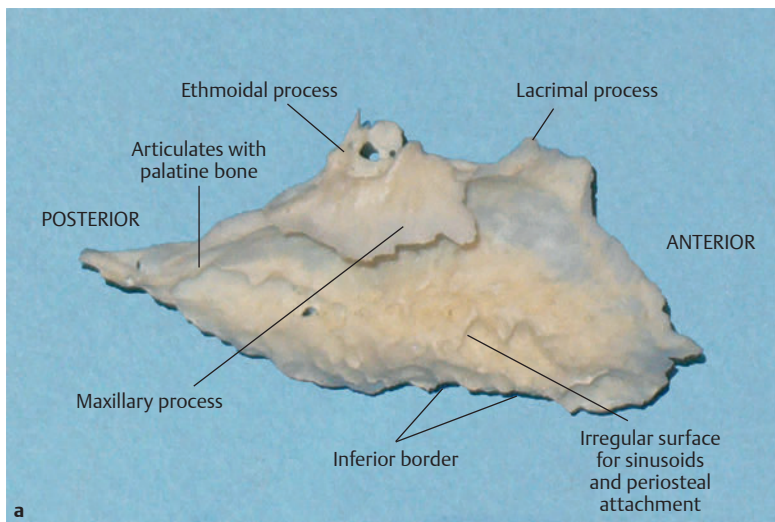
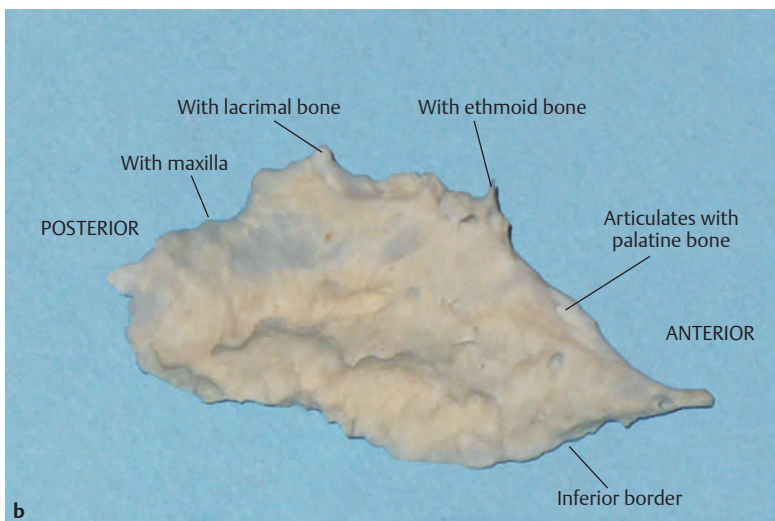


Fig. 3.7a, b Photographs of a disarticulated right turbinate bone.

a Lateral view.



b Medial view.

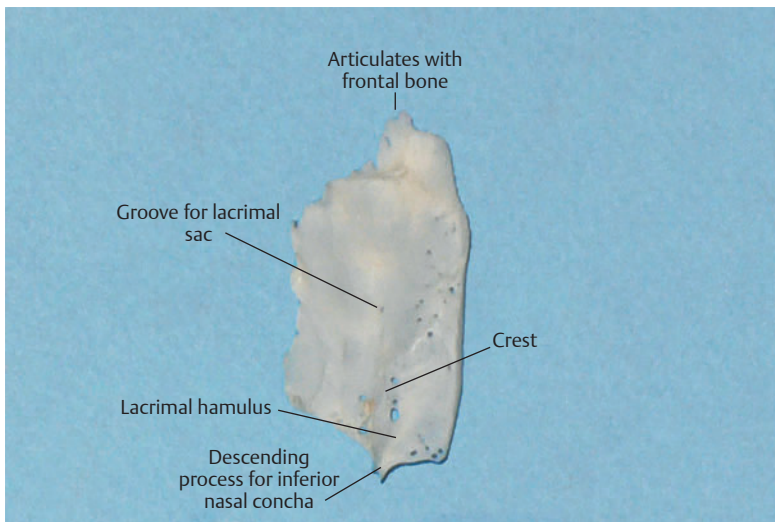


Fig. 3.8 Photograph of a disarticulated left lacrimal bone.

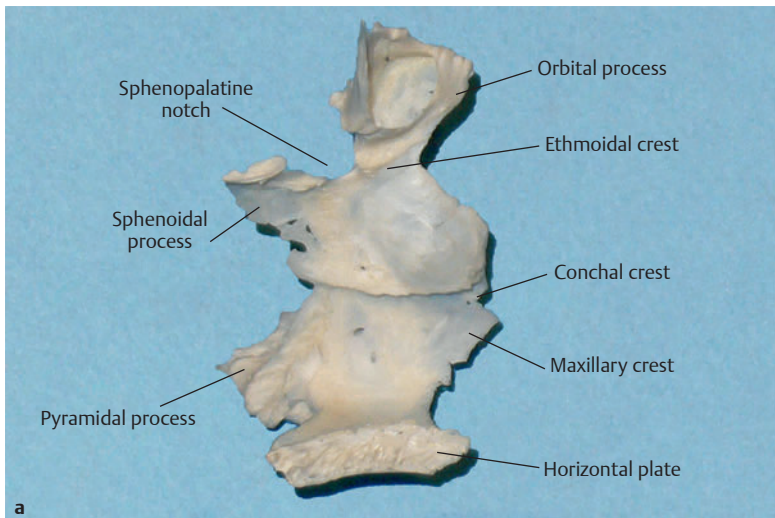
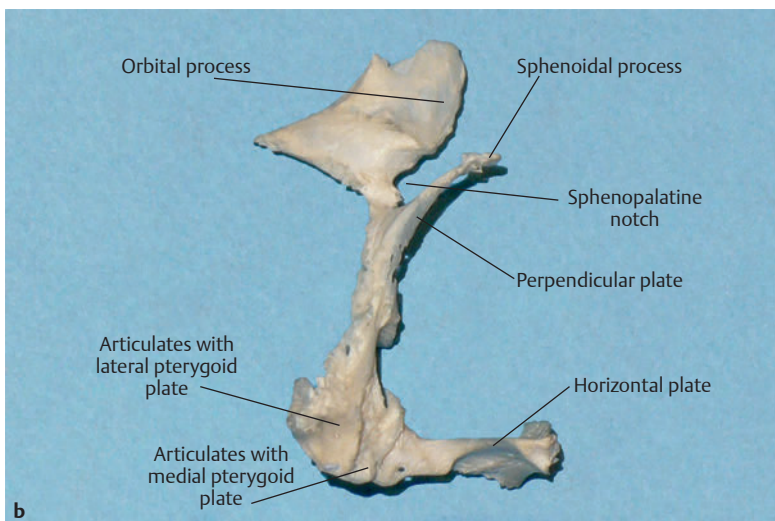


Fig. 3.9a, b Photographs of a disarticulated left palatine bone.

a Medial view.



b Posterior view.

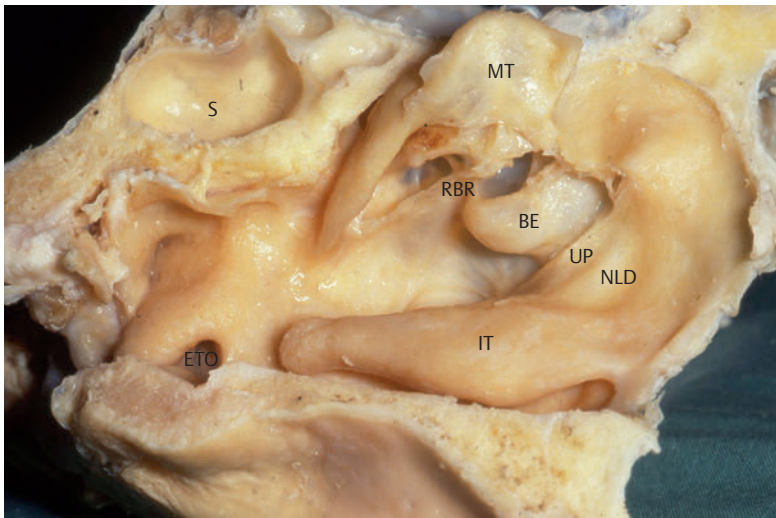


Fig. 3.10 Lateral wall of nose: cadaver specimen with middle turbinate lifted to show middle meatus. BE, bulla ethmoidalis; ETO, eustachian tube orifice; IT, inferior turbinate; MT, middle turbinate; NLD, nasolacrimal duct; RBR, retrobulbar recess; S, sphenoid; UP, uncinate process.

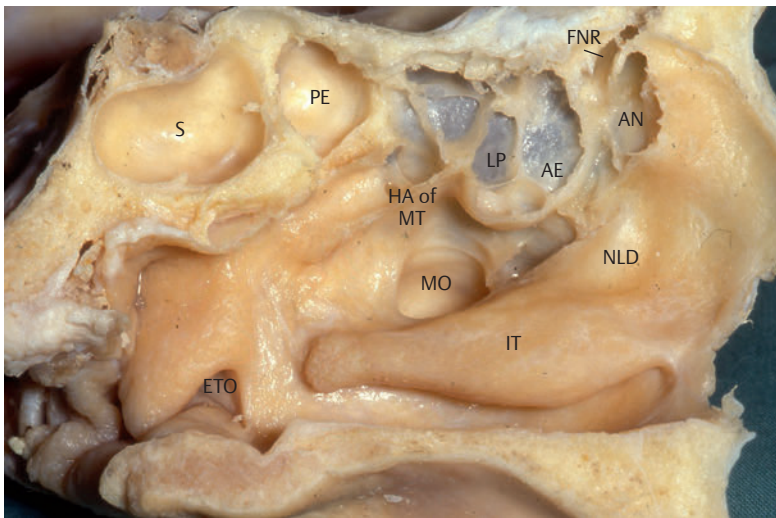


Fig. 3.11 Lateral wall of nose: cadaver specimen with dissection to show structures of lateral wall: AE, anterior ethmoid; AN, aggr nasi; ETO, eustachian tube orifice; FNR, frontonasal recess; HA of MT, horizontal attachment of middle turbinate duct; IT, inferior turbinate; LP, lamina papyracea; MO, maxillary ostium; NLD, nasolacrimal; PE, posterior ethmoids; S, sphenoid.

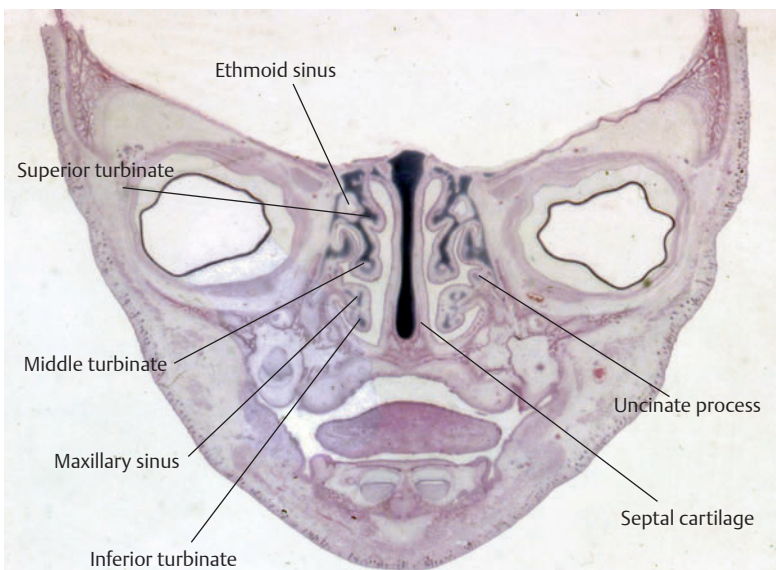


Fig. 3.12 Coronal section through a 24-week fetus (hematoxylin and eosin).

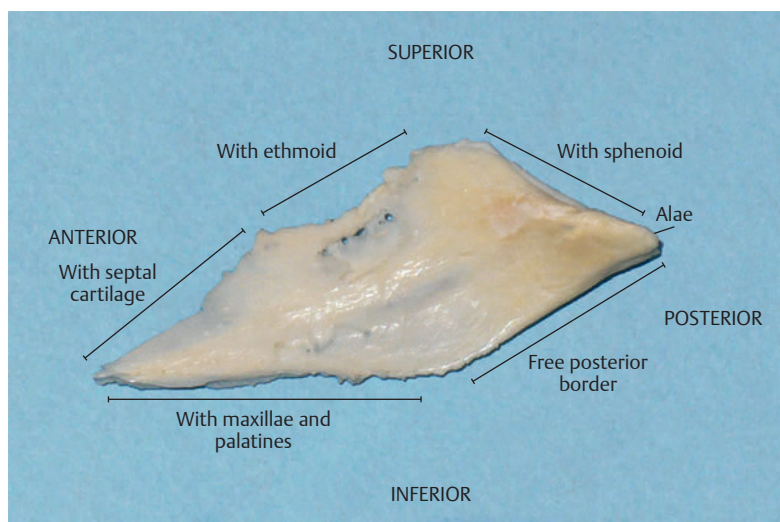


Fig. 3.13 Photograph of a disarticulated vomer.

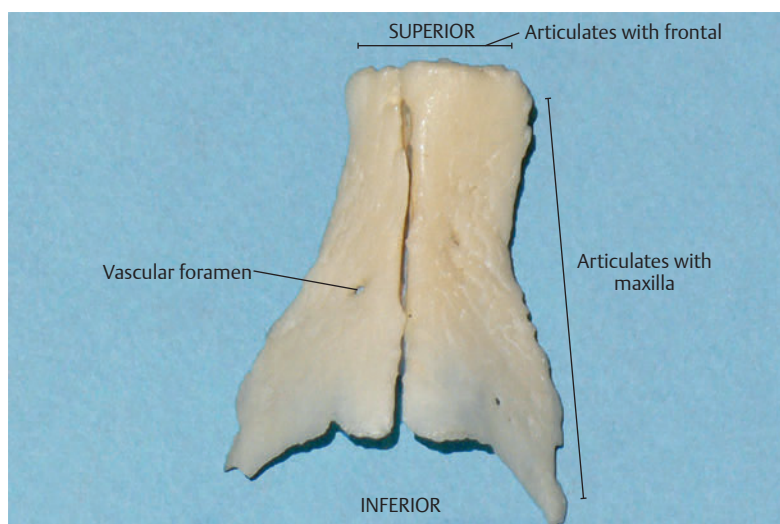


Fig. 3.14 Photograph of disarticulated nasal bones.

fibrous tissue that connects them present little resistance to tumor spread, though this is fortunately rare in the anterior nasal cavity. However, tumors that arise on the anterior septum and columella are able to spread bilaterally to the cervical lymphatics and therefore have a particularly poor prognosis.

Paranasal Sinuses

Histology

The lining of the nose and paranasal sinuses may be grossly divided into skin anteriorly, olfactory epithelium superiorly, and ciliated columnar respiratory epithelium elsewhere. In addition there may be areas of squamous metaplasia on the anterior ends of the inferior and middle turbinate, reflecting areas of aerodynamic “trauma.”

Blood Supply

The blood supply of the nose and sinuses is grossly derived from the external and internal carotid with considerable overlap and crossover between the right and left sides. The sphenopalatine, greater palatine, facial, and superior labial branches supply the majority of the nasal cavity with a contribution superiorly from internal carotid via the anterior and posterior ethmoidal arteries. Sinusoids on the inferior turbinate and adjacent anterior septum (anterior septal tubercle [Fig. 3.15]), under autonomic control, regulate airflow. The blood supply of the septum has become more appreciated of late due to the frequent use of the Haddad nasoseptal flap based on the branches of the sphenopalatine artery in reconstruction of the larger posterior skull base defects.⁹

The cavernous venous system drains into the facial veins anteriorly, into the pterygoid plexus posteriorly,

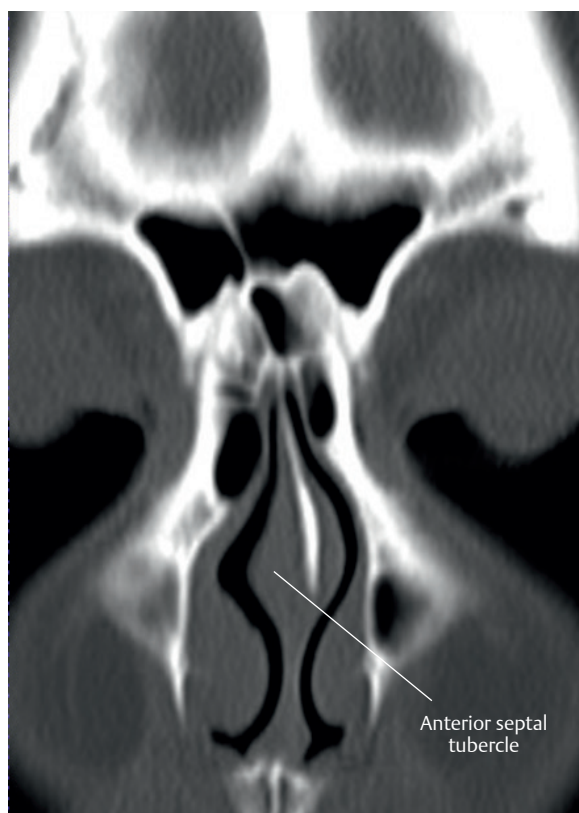


Fig. 3.15 Coronal CT scan through the anterior nasal septum, showing the anterior septal tubercle.

and superiorly has connections with the superior ophthalmic veins and superior sagittal sinus. These pathways may in part be responsible for the pattern of local spread for tumors such as olfactory neuroblastoma, which may appear in the contralateral eye or anywhere on the dura.

The maxillary sinus receives its blood supply via branches of the facial, maxillary, infraorbital, and greater palatine arteries with venous drainage to the anterior facial vein and pterygoid plexus. Once this area is infiltrated by malignancy, such as a squamous cell carcinoma, complete surgical excision is difficult. The frontal sinuses are supplied by the supraorbital and anterior ethmoidal arteries with venous connections through the diploic veins into the sagittal and sphenoparietal sinuses and also with the superior ophthalmic veins. The sphenoid sinuses are supplied by the posterior ethmoidal vessels and nerves.

Nerve Supply

The nerves may also provide routes of tumor spread, as graphically demonstrated by adenoid cystic carcinoma, whose capacity for perineural spread either directly or embolically, severely compromises attempts at curative resection. The maxillary division of the trigeminal nerve

supplies the majority of the septum via the nasopalatine nerves with a contribution from the anterior superior alveolar and anterior ethmoidal branch of the nasociliary nerve anteriorly. The posteroinferior septum also receives a small supply from the nerve of the pterygoid canal and the posterior inferior nasal branch of the anterior palatine nerve. On the lateral wall again the majority is supplied by branches of the maxillary and pterygopalatine ganglion, with anterior contributions from the anterior superior alveolar, anterior ethmoidal, and infraorbital nerves. Superiorly, olfactory epithelium extends down from the olfactory niche onto the superior turbinate and septum. The extent varies between individuals and diminishes with age. Some evidence of olfactory epithelium has been reported on the medial middle turbinate, but this does not appear to be functional.

The sinuses receive their nerve supply via the supra-orbital nerve (frontal), posterior ethmoidal and pterygopalatine ganglion (sphenoid), and maxillary division of the trigeminal, which supplies sensation to the maxillary sinus via the infraorbital, superior alveolar, and greater palatine nerves, all of which may act as conduits for tumor spread.

Lymphatic Drainage

Fortunately the lymphatic drainage from the sinuses is relatively poor to the retropharyngeal and jugulodigastric nodes and to the pterygopalatine fossa, but this is not true of the nasal vestibule, anterior septum, and columella, from where bilateral cervical spread can occur to the submandibular region.

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4 Diagnostic Strategy

Presentation

While the clinical features of each specific pathology are covered in their respective chapters, a few general observations may be made. The principal problem in the diagnosis of sinonasal tumors, both benign and malignant, has been related to late presentation. Patients develop relatively innocuous symptoms, often unilateral, that are ignored by both patient and primary care physician. Unlike hoarseness or dysphagia, which are recognized as of potential significance, nasal obstruction, discharge, even serosanguinous discharge, and reduced sense of smell are common complaints more often associated with allergy, inflammation, or infection than with a tumor. With hindsight the unilateral nature and usually spontaneous onset should elicit concern, but in the case of malignancy it is more often the spread beyond the sinonasal confines to the cheek, eye, or upper alveolus that leads to diagnosis. Fortunately, with malignancy neck disease remains relatively rare, as does systemic disease with a few notable exceptions (Table 4.1).

Local Disease

A mass in the nasal cavity will produce unilateral nasal obstruction, discharge that is often bloodstained, a reduction in the sense of smell, and occasionally facial discomfort.¹ Frank epistaxis is less common than might be supposed, although it typically occurs with vascular lesions such as juvenile angiofibroma, olfactory neuroblastoma, and malignant melanoma.

Tumors that arise in or involve the ethmoidal labyrinths obviously affect the nasal cavity at an early stage and may spread across the midline, for example, adenocarcinomas, but more often present with orbital symptoms due to early erosion of the lamina papyracea. These include proptosis, a sign that is almost always due to a mass either within or outside the orbit. The direction and degree of displacement will depend on the position of the mass and its speed of growth. This in turn may be associated with diplopia if displacement occurs quickly but may be absent if the change in position takes place slowly. Similarly, the optic nerve and its blood supply can tolerate impressive stretching if it occurs slowly, but vision may be lost rapidly when additional inflammation or infiltration is present. Diplopia may also occur as a consequence of direct muscle infiltration or involvement of the neural supply, especially in the cavernous sinus. Once at the orbital apex, whether extra- or intraperiosteal, the disease can spread into the middle cranial fossa. Tumors that invade or arise within the skull base may directly affect the optic nerve(s) and chiasm. This can occur with meningioma and chondrosarcoma and may tragically be

bilateral. Visual loss can also result from chemosis due to corneal exposure, keratosis, and ulceration.

The incidence of orbital invasion by sinonasal malignancies will vary with the histology, but overall visual symptoms occurred in 50% of a cohort of 220 cases.¹ This rose to 62% if the tumor arose in the ethmoid, compared with 46% with malignant tumors of the nasal cavity. However, it should be noted that the orbital periosteum, unlike the lamina papyracea, is relatively resistant to penetration by malignant tumors, so the presence of orbital symptoms does not indicate intraperiosteal spread per se, nor does the lack of orbital symptoms and signs necessarily mean that the eye has not been infiltrated. A recognition of this led to a change in strategy for the management of the orbit in the 1980s.² It has been estimated that between 66% and 82% of patients with ethmoidal malignancy have erosion of the lamina, whereas the periosteum is involved in 30 to 50%.³⁻⁶

Epiphora due to nasolacrimal involvement is quite often encountered with anterior ethmoidal and maxillary lesions as a result of secondary compression or distortion of the system.

Superior extension of the tumor into the anterior cranial cavity can occur through the lateral lamella of the cribriform plate, along the anterior and posterior neurovascular bundles, or directly through the fovea, but this is generally asymptomatic. Classically this occurs with olfactory neuroblastoma. CSF leakage and meningitis are exceptionally rare and even when the dura has been breached and extensive frontal lobe infiltration occurs, any personality changes are usually too subtle to be noticed. Occasionally patients with direct involvement of the olfactory bulbs and tracts may develop reduction or distortion of the sense of smell, for example, in frontal meningioma, although this rarely attracts much attention.

Tumors arising within the anterior ethmoids/middle meatus can spread into the maxillary sinus and/or into the frontal sinus via the frontal recess, occasionally producing a mucocele, although this is a rare phenomenon in the presence of a malignant tumor. We have had three patients with sinonasal malignancies who have presented in this way (0.3%). It is not known why the frontal and sphenoid sinuses are rarely the site of primary malignant tumors and are more often involved by local spread or involvement from the surrounding bone. A frontal sinus tumor is most likely to present with swelling of the forehead, whereas sphenoid tumors produce orbital symptoms, in particular visual loss.

Malignancy of the maxillary sinus will spread medially into the nasal cavity, producing obstruction and serosanguinous discharge, as before, but may also spread superiorly, particularly into the infraorbital canal,⁷

Table 4.1 Clinical features arising from sinonasal tumors

Primary symptoms	
Nasal cavity	• Nasal blockage • Serosanguinous discharge • Hyposmia
• Inferiorly into palate	• Mass
• Anterosuperiorly into the nasal bone	• Glabellar mass
• Externally into skin	• Mass/ulceration
• Superiorly into anterior cranial fossa	• Minimal—subtle personality change • Headache
• Posteriorly into nasopharynx and eustachian orifice	• Middle ear effusion
Ethmoid sinuses	
• Medially into nasal cavity	• As above, can cross to contralateral side to produce bilateral symptoms
• Inferolaterally into maxilla	• Facial pressure due to mucus retention
• Medially into orbit	• Proptosis • Diplopia • Visual loss • Chemosis • Epiphora
• Superiorly into the anterior cranial fossa	• Minimal—subtle personality change
Maxillary sinus	
• Medially into nasal cavity	• As above
• Anteriorly into cheek directly or via infraorbital canal	• Mass or ulceration of skin • Paresthesia
• Posteriorly into pterygoid region and infratemporal fossae	• Trismus and pain
• Inferiorly into the palate or alveolar ridge	• Mass • Loosening of the teeth • Malignant oroantral fistula
• Superiorly into orbit	• As above
Secondary symptoms	
Lymphatic	• Cervical lymphadenopathy in levels I & II and facial
Systemic	• Bone pain • Dyspnea • Liver pain • Skin nodules • Localizing neurologic symptoms & signs • General malaise • Confusion

producing paresthesia of the cheek as well as orbital symptoms. Direct anterior spread through bone or via the infraorbital foramen may produce a mass in the cheek, which in turn may ulcerate. Inferior spread from the maxilla produces a mass in the oral cavity, loosening of teeth, and/or a malignant oroantral fistula. Posterior spread into the pterygoid region and infratemporal fossa is associated with trismus and pain.

Metastatic Disease

Although it is mandatory to examine the neck, fewer than 10% of epithelial malignancies present with cervical disease, reflecting the paucity of lymphatic drainage from the sinuses. This is more often a concern with tumors of the nose and some of the lymphomas and sarcomas. The submandibular, jugulodigastric, prefacial and postfacial nodes are most commonly involved by tumors from the septum and in particular the columellar region; these

sometimes spread bilaterally, which is invariably associated with a poor prognosis. However, careful examination may reveal more cervical disease than had hitherto been suspected, as with olfactory neuroblastoma, and the possibility of locoregional spread should always be considered during follow-up even in unusual locations such as the cheek.⁸

Systemic metastases are generally uncommon but may occur with longer follow-up and in the presence of uncontrolled local disease. Again this is particularly true of olfactory neuroblastoma and also of malignant melanoma. Adenoid cystic carcinoma is also known to spread along perineural lymphatics, either directly or by embolization, often presenting at some distance from the original tumor, although patients can survive for some time with disseminated disease. Patients may be unaware of systemic metastases for some time and it is a matter of debate how aggressively one should seek them out in the case of tumors where the therapeutic options may be limited. Notwithstanding this, particularly at presentation, complaints of an unresolving nonproductive cough, bone pain, or significant fatigue should prompt further investigation as this can have a direct bearing on the management of the primary lesion.

Examination

In addition to a general ENT examination, including careful inspection and palpation of the oral cavity, midface, and neck, the mainstay of diagnosis remains endoscopy of the nasal cavity and nasopharynx. An initial inspection with a 4 mm 0° or 30° scope using the traditional “three passes” technique to adequately consider the inferior, middle, and superior segments of the nose may immediately determine the problem, but specific attention should be paid to the olfactory niche and middle and superior meatuses. Ideally the nasopharynx should also be examined using either a rigid or a flexible endoscope. However, in many cases the associated edema and nasal secretions obscure a good view of the tumor itself. The nose should then be anesthetized/decongested using whatever combination of topical solution is available. In our own service, a preparation containing phenylephrine hydrochloride and lidocaine hydrochloride is applied topically on a short length of ribbon gauze into either side of the nose for 5 minutes, which shrinks the lining and reduces sensation, but even this may not give adequate exposure.

Where there is any element of suspicion, imaging is undertaken (see later) followed by a formal examination and biopsy. In our own practice this is conducted as a day case under general anesthetic combined with local anesthesia/decongestion (usually using Moffatt’s solution [1 mL 2% sodium bicarbonate, 2 mL 10% cocaine, 2 mL 1:1000 epinephrine⁹]), which ensures that adequate

representative tissue is taken. Of course, this may be done under local anesthesia alone, but one should not succumb to the temptation of a quick “smash and grab” in a busy outpatient clinic. Under endoscopic control it is rarely necessary to use any external open approach that risks transgression of normal tissue planes. Exceptions to this would be a rare lesion in the frontal sinus lateral to the midpoint of the orbit.

Tissue must be subjected to expert histopathology. Diagnosis can usually be reached using formal saline fixation but occasionally fresh tissue may still be required in some lymphoreticular disease.

Histopathology

The sinonasal region is the region with the greatest histological diversity in the body and this is reflected in the WHO classification,¹⁰ which has been largely used in this document (Table 4.2) and our own series (Table 4.3). However, there have been several changes to this classification over the years and sometimes the histopathological classification is at odds with clinical findings. The best example of this is the erroneous inclusion of angiofibroma in the nasopharynx, where it merely presents in common with several other areas but does not actually arise. Our own clinical and imaging studies show that angiofibroma actually originates in the pterygopalatine fossa at the anterior aspect of the pterygoid (vidian) canal and initially erodes the sphenopalatine foramen.

Immunohistochemistry is frequently required, particularly for any small cell or undifferentiated carcinoma. Due to the histological diversity, frozen section can pose some difficulties on initial biopsy, although it has an important role during subsequent resection once the diagnosis has been established.

Other Investigations

Imaging

(See also Chapter 5.)

Fine-detail three-plane computed tomography (coronal, axial and sagittal plane) combined with magnetic resonance imaging provides an accurate demonstration of tumor extent and can sometimes indicate the type of histology.¹¹ Although together these modalities produced an accuracy of 98% in predicting extent of tumor, the assessment of spread through the orbital periosteum and dura still requires microscopic confirmation. MRI alone is not sufficient as early erosion of the cribriform plate is still best shown on coronal CT.^{12,13}

The extent to which imaging beyond the midface and brain is undertaken will be determined by the histology and patient symptoms. It is not routinely undertaken

Table 4.2 Histopathology and ICD-O codes^a according to WHO classifications of tumors

A Nasal cavity and paranasal sinuses
(1) Malignant epithelial tumors
Squamous cell carcinoma • Keratinizing squamous cell carcinoma ICD-O 8070/3 • Nonkeratinizing (cylindrical cell, transitional) carcinoma; currently no separate ICD-O code • Verrucous carcinoma ICD-O 8051/3 • Papillary squamous cell carcinoma ICD-O 8052/3 • Basaloid squamous cell carcinoma ICD-O 8083/3 • Spindle cell carcinoma ICD-O 8074/3 • Adenosquamous carcinoma ICD-O 8560/3 • Acantholytic squamous cell carcinoma ICD-O 8075/3
Lymphoepithelial carcinoma ICD-O 8082/3
Sinonasal undifferentiated carcinoma ICD-O 8020/3
Adenocarcinoma • Intestinal-type adenocarcinomas ICD-O 8144/3 • Sinonasal nonintestinal-type adenocarcinomas ICD-O 8140/3
Salivary gland-type carcinoma • Adenoid cystic carcinoma ICD-O 8200/3 • Acinic cell carcinoma ICD-O 8550/3 • Mucoepidermoid carcinoma ICD-O 8430/3 • Epithelial-myoepithelial carcinoma ICD-O 8562/3 • Clear cell carcinoma ICD-O 8310/3 • Myoepithelial carcinoma ICD-O 8982/3 • Carcinoma ex pleomorphic adenoma ICD-O 8941/3
Neuroendocrine tumors • Typical carcinoid ICD-O 8240/3 • Atypical carcinoid ICD-O 8249/3 • Small cell carcinoma, neuroendocrine type ICD-O 8041/3
(2) Benign epithelial tumors
Sinonasal papillomas • Inverted papilloma (Schneiderian papilloma, inverted type) ICD-O 8121/1 • Oncocytic papilloma (Schneiderian papilloma, oncocytic type) ICD-O 8121/1 • Exophytic papilloma (Schneiderian papilloma, exophytic type, everted type) ICD-O 8121/1
Respiratory epithelial adenomatoid hamartoma; no ICD-O code
Salivary gland-type adenomas • Pleomorphic adenoma ICD-O 8940/0 • Myoepithelioma ICD-O 8982/0 • Oncocytoma ICD-O 8290/0
(3) Malignant soft tissue tumors
Fibrosarcoma ICD-O 8810/3
Undifferentiated high grade pleomorphic sarcoma ("MFH") ICD-O 8830/3
Leiomyosarcoma ICD-O 8890/3
Rhabdomyosarcoma ICD-O 8900/3 • Embryonal ICD-O 8910/3 • Alveolar ICD-O 8920/3
Angiosarcoma ICD-O 9120/3
Kaposi's sarcoma ICD-O 9140/3
Malignant peripheral nerve sheath tumor ICD-O 9540/3
Liposarcoma ICD-O 8850/3

Synovial cell sarcoma ICD-O 9040/3
Alveolar soft part sarcoma ICD-O 9581/3
Malignant fibrous histiocytoma ICD-O 8830/3
(4) Borderline and low malignant potential tumors of soft tissue
Desmoid-type fibromatosis ICD-O 8821/1
Inflammatory myofibroblastic tumor ICD-O 8825/1
Glomangiopericytoma (sinonasal-type hemangiopericytoma) ICD-O 9150/1
Extrapleural solitary fibrous tumor ICD-O 8815/1
(5) Benign soft tissue tumors
Myxoma ICD-O 8840/0
Leiomyoma ICD-O 8890/0
Rhabdomyoma ICD-O 8900/0
Hemangioma ICD-O 9120/0
Schwannoma ICD-O 9560/0
Neurofibroma ICD-O 9540/0
Meningioma ICD-O 9530/0
(6) Malignant tumors of bone and cartilage
Chondrosarcoma ICD-O 9220/3
Mesenchymal chondrosarcoma ICD-O 9240/3
Osteosarcoma ICD-O 9180/3
Chordoma ICD-O 9370/3
(7) Benign tumors of bone and cartilage
Fibrous dysplasia; no ICD-O code
Osteoma ICD-O 9180
Osteoid osteoma ICD-O 9191/0
Osteoblastoma ICD-O 9200/0
Osteochondroma (exostosis) ICD-O 9210/0
Chondroma ICD-O 9220/0
Chondroblastoma ICD-O 9230/0
Chondromyxoid fibroma ICD-O 9241/0
Giant cell lesion; no ICD-O code
Giant cell tumor of bone ICD-O 9250/1
Ameloblastoma ICD-O 9310/0
Nasal chondromesenchymal hamartoma; no ICD-O code
(8) Hematolymphoid tumors
Extranodal NK/T cell lymphoma ICD-O 9719/3
Diffuse large B cell lymphoma ICD-O 9680/3
Extramedullary plasmacytoma ICD-O 9734/3
Extramedullary myeloid sarcoma ICD-O 9930/3
Histiocytic sarcoma ICD-O 9755/3
Langerhans cell histiocytosis ICD-O 9751/1
Juvenile xanthogranuloma; no ICD-O code
Rosai–Dorfman disease (sinus histiocytosis with massive lymphadenopathy); no ICD-O code

continued ►

Table 4.2 Histopathology and ICD-O codes^a according to WHO classifications of tumors (continued)

(9) Neuroectodermal tumors
Ewing's sarcoma ICD-O 9260/3
Primitive neuroectodermal tumor (PNET) ICD-O 9364/3
Olfactory neuroblastoma (esthesioneuroblastoma) ICD-O 9522/3
Melanotic neuroectodermal tumor of infancy ICD-O 9363/0
Mucosal malignant melanoma ICD-O 8720/3
Heterotopic central nervous system tissue (nasal glioma); no ICD-O code
(10) Germ cell tumors
Immature teratoma ICD-O 9080/3
Teratoma with malignant transformation ICD-O 9084/3
Sinonasal yolk sac tumor (endodermal sinus tumor) ICD-O 9071/3
Sinonasal teratocarcinosarcoma; no ICD-O code
Mature teratoma ICD-O 9080/0
Dermoid cyst ICD-O 9084/0
(11) Secondary tumors
B Nasopharynx
(1) Malignant epithelial tumors
(2) Benign epithelial tumors
(3) Soft tissue neoplasms
Nasopharyngeal angiofibroma ^b ICD-O 9160/0
(4) Hematolymphoid tumors
(5) Tumors of bone and cartilage
(6) Secondary tumors
^a Whenever available.
^b Although juvenile angiofibroma is known to arise from the posterior nasal cavity and not the nasopharynx, the WHO classification includes this lesion here.

for all sinonasal malignancy, but poorly differentiated tumors such as sinonasal undifferentiated carcinoma (SNUC), neuroendocrine carcinoma and lymphoreticular lesions require more extensive staging. Similarly adenoid cystic carcinoma, which has a tendency to spread to the lung, requires chest CT.

Ultrasound and Fine Needle Aspiration

Ultrasound of the neck should be offered at presentation and during follow-up to selected patients, if available, in combination with fine needle aspiration, for example, for olfactory neuroblastoma.^{14,15} This is widely available but requires specialist expertise in head and neck/sinonasal pathology.^{16,17}

Additional Tests

The accuracy and utility of positron emission tomography (PET) remains to be established in the nose and sinuses but may prove of value in staging and revealing recurrence, particularly as it becomes more readily available.^{18,19}

A radionuclide bone scan should be considered in individuals where bone metastases are suspected and hematological investigations including bone marrow aspirate may be appropriate in cases of chloroma (leukemic deposits), lymphoma and individuals where bone and liver secondaries are suspected.

Table 4.3 Types of histopathology in our personal cohort of sinonasal neoplasia (*n* = 1506)

Classification	Type of histopathology	<i>n</i>
Epidermoid	• Squamous	320
	• Inverted papilloma	114
	• Other	46
Nonepidermoid	• Adenocarcinoma	117
	• Adenoid cystic	54
	• Other glandular	15
Mesenchymal	• Fibrosarcoma	23
	• Malignant fibrous histiocytoma	6
	• Alveolar soft part sarcoma	5
	• Other	6
Vasoform	• Angiofibroma	155
	• Hemangiopericytoma/glomangiopericytoma	13
	• Vascular malformations/hemangioma	21
	• Angiosarcoma	1
Muscular	• Rhabdomyosarcoma	26
	• Leiomyoma	3
	• Leiomyosarcoma	5
Cartilage	• Chondrosarcoma	42
Bone	• Osteosarcoma	12
	• Osteoma	55
	• Ossifying fibroma	31
	• Fibrous dysplasia	18
	• Other	8
Lymphoreticular	• B cell lymphoma	60
	• NK/T cell lymphoma	33
	• Extramedullary plasmacytoma	13
	• Other	6
Neuroectodermal	• Olfactory neuroblastoma	80
	• Malignant melanoma	115
	• SNUC	24
	• Schwannomas	17
	• Meningioma	11
	• Carcinoid	7
	• PNET/Ewing's sarcoma	6
	• Other	6
Germ cell tumors	• Mature teratoma	1
	• (Dermoids)	7
Odontogenic	• Ameloblastoma	6
	• Ameloblastic fibroma	5
	• Odontogenic keratocyst	3
Metastases		10
Abbreviations: PNET, primitive neuroectodermal tumor; SNUC, sinonasal undifferentiated carcinoma.		

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5 Imaging of the Paranasal Sinuses and Nasopharynx

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Introduction

Imaging of the paranasal sinuses and nasopharynx is essential in the diagnosis of patients with pathology in these regions. In the last three decades we have witnessed a tremendous advance in the therapy of patients with inflammatory, infectious, and neoplastic diseases of the head and neck. During the same period we have also seen revolutionary changes in imaging technology that have greatly influenced the evolving surgical and medical therapies.

The use of computed tomography (CT) imaging technology is well known for the evaluation of inflammatory disease affecting the paranasal sinuses. A combination of CT and magnetic resonance imaging (MRI), and at times positron emission tomography (PET) imaging, is obtained in the work-up of patients with suspected sinonasal and nasopharyngeal malignancy. There are many confounders in imaging, which occasionally make it difficult to distinguish inflammatory and infectious diseases from neoplasms. However, these three main imaging modalities can be used in a complementary fashion to help reduce this uncertainty and to assist with staging, biopsy, and treatment planning for tumors.

The objective of this chapter is to highlight the application of imaging information with regard to the identification of the typical findings in inflammatory, infectious, and neoplastic disorders of the paranasal sinuses and nasopharynx. Additionally, we will describe some of the pitfalls of diagnosis encountered with imaging. We will also review radiological staging of sinonasal (SN) and nasopharyngeal (NP) malignancies as well as the imaging of postsurgical and post-radiation therapy patients.

The efficacy and quality of information provided by radiological assessment primarily depends on the way in which the images are acquired. For example, depending on the plane of acquisition, the slice thickness, and the presence or absence of contrast, a CT image can vary greatly in its utility in assessing a potential sinonasal or nasopharyngeal tumor patient.¹ Described in each section are the current standards for imaging of these regions. Occasionally these vary depending on the clinical requisite (i.e., contrast medium is rarely administered in patients with neutropenic fever being assessed for acute sinusitis), but for the most part they are fairly consistent and reliable.²

Paranasal Sinuses

Imaging Protocols

Computed Tomography

Given its superior bony resolution, CT is typically the best imaging modality for the display of the delicate regional bony anatomy as well as of the mucosal changes in the presence of inflammatory disease. It is also superior in the detection of bony involvement by pathology, in particular periosteal reaction and bony erosion. Particularly when intravenous (IV) contrast is used, CT can also provide excellent soft tissue information, although it is inferior to the soft tissue contrast resolution provided by MRI.

Images are acquired in the axial plane, preferably with 0.5 mm slice thickness. The imaging plane should start above the skull base structures and include the entirety of the paranasal sinuses and nasal cavity, the orbits, and the middle and anterior cranial fossae. Coronal and sagittal reformatted images are reconstructed from the axial imaging acquisition. If derived from the thin-section source images, these reformatted images should demonstrate excellent spatial resolution, essentially indistinguishable from the axial source images.

Coronal images represent the optimal plane for endoscopic correlation. Sagittal planes are very helpful in improving the 3D conceptualization of the regional morphology.³ However, when employing these images for ensuring the accuracy of the spatial orientation in one's mind, the sagittal images should always be correlated with an additional orthogonal plane, that is, a coronal or axial plane image. The application of multiplanar reconstruction and the use of cross-hairs for localization purposes is especially helpful (Fig. 5.1).

If contrast administration is contemplated, one should first consider doing an MRI examination. Intravenous contrast is typically administered for improved soft tissue resolution. Information provided by MRI is superior for this purpose and avoids the radiation dose received with CT.

Magnetic Resonance Imaging

MRI provides excellent soft tissue contrast resolution. It offers multiplanar capabilities, and does not involve ionizing radiation, which is of particular advantage when imaging children and women of childbearing age.