Chapter 12: Complications of suppurative otitis media

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Complications of suppurative otitis media arise when infection spreads from the middle ear cleft to structures from which it is normally separated by bone. Before antibiotics were regularly used, these complications arose more frequently from acute middle ear suppuration than from chronic middle ear disease. Nowadays, most otological experience indicates that chronic middle ear infection is the greater hazard, although some writers (Gower and McGuirt, 1983) have still found a higher incidence of intracranial complications from acute infections. This probably reflects the high proportion of young patients in their series, and illustrates the fact that complications of acute otitis media are commoner in the young. Although the overall incidence of complications has fallen greatly with antibiotic treatment, it cannot be overstressed that the mortality from intracranial complications is as high now as it was 20 years ago (Editorial, 1982).

Whether the preceding disease has been acute or chronic, spread of infection can follow a number of possible routes.

1. By extension through bone that has been demineralized during acute infection, or suffered resorption by cholesteatoma or osteitis in chronic destructive disease.

2. By the spreading of infected clot within small veins through bone and dura to venous sinuses - the lateral and the superior petrosal - and so to intracranial structures. Apparently intact bone may be transgressed by thrombophlebitis within its Haversian vascular system. Thrombophlebitic spread from the lateral sinus to the cerebellum and from the superior petrosal sinus to the temporal lobe explains the frequent association between these complications.

3. Through normal anatomical pathways - the oval or round windows into the internal auditory meatus, the cochlear and vestibular aqueducts, dehiscence of the thin bony covering of the jugular bulb, dehiscence of the tegmen tympani, and dehiscent suture lines of the temporal bone.

4. Through non-anatomical bony defects caused by trauma - accidental or surgical - or by neoplastic erosion.

5. Through other surgical defects; in particular, the vestibular opening deliberately created at a stapedectomy operation, and possibly through the fenestration opening into the lateral semicircular canal.

6. Into brain tissue along the periarteriolar spaces of Virchow-Robin. This spread does not affect the cortical arterioles themselves and explains abscess development in the white matter with no apparent continuity of infection to the brain surface.

Chronic middle ear disease extends slowly and many of its complications are caused by the progressive and relentless erosion of bone, thus exposing the structures at risk to
damage - the facial nerve, the labyrinth, the dura. Acute infections cause complications earlier, through the thrombophlebitic mechanisms and the anatomically available pathways.

Despite the apparent 'skipping' of a brain abscess into the white matter, the general pattern of infective spread, through the mechanisms described, is progressive from one structure and tissue plane to the next. Progress is from the middle ear cleft to extradural spaces and venous sinuses; through dura to the cerebrospinal fluid spaces; and into brain tissue. It should not then be surprising that multiple complications are common, arising in one-third of cases, and that certain associations, such as those between lateral sinus thrombosis and cerebellar abscess, are frequent.

The propensity for spread of infection, and the development of complications depends on:

(1) patient attributes - age, immune state, intercurrent chronic disease such as diabetes mellitus or leukaemia

(2) bacterial attributes - virulence, susceptibility to chemotherapeutic elimination. For example in acute suppurative infection, *Streptococcus pneumoniae* type III and *Haemophilus influenzae* type B have sinister reputations

(3) efficacy of treatment of the underlying middle ear disease.

Infecting bacteria predominantly responsible for infections have varied in accounts over the years, and still do from one report to another. One explanation is the difference in the nature of bacteria associated with acute and with chronic, cholesteatomatous, disease. Some variations may represent genuine differences in the pattern of microbial infections in different parts of the world, while others indicate changes that have taken place in patterns of infection over the years. Lastly, previously unidentified bacteria such as anaerobes (Ingham, Selkon and Roxby, 1977) have been recognized by new bacteriological techniques. These explanations for apparent inconsistencies should be remembered when reading the sections below on the individual complications.

Although cholesteatoma, with posterior marginal or attic disease, is considered to be the hallmark of an ear with a poor prognosis, Browning (1984), in a study of patients in the West of Scotland, has shown that a brain abscess may arise from ears with mucosal disease, and from ears previously treated by modified radical mastoidectomy. He has also indicated that the risk of any patient with chronic otitis media developing a brain abscess is as low as 1 in 3500.

The complications to be discussed fall into two main categories:

(1) those within the cranial cavity:
   - extradural abscess
   - subdural abscess (empyema)
   - sigmoid sinus thrombophlebitis
   - meningitis
   - brain abscess
   - otitic hydrocephalus
(2) those within the temporal bone:
   facial paralysis
   labyrinthine infections.

Rarer complications, for example subclavian vein thrombosis, and internal carotid aneurysm (Kimmelman and Grossman, 1983), have also been reported.

Petrositis, which may be considered an unusual extension of mastoiditis, is described in Chapter 9.

**Intracranial infections**

**General principles**

Certain aspects of the presentation, diagnosis and management of intracranial complications are common to all; so it is pertinent to make some general comments concerned with principle here, before considering individual features under separate heading below. This plan may help to avoid unnecessary repetition, and should emphasize that these complications have to be considered as a group, since they are multiple in about one-third of instances.

The symptoms of intracranial spread of infection are those of infection, and compression of brain tissue. Headache, malaise, fever, drowsiness are all suspicious symptoms. Otalgia is not a feature of uncomplicated cholesteatoma. Any of these should alert the otologist to the possibility of a complication, and provoke initiation of appropriate investigation and treatment. More specific features of individual complications will be discussed under the appropriate headings. At any time one complication may be clinically dominant with others emerging from the investigative findings. Investigation and treatment must run concurrently. To delay treatment until investigation is complete may allow disease to proceed beyond the chance of recovery. The principles of treatment, common to all intracranial complications include:

1. systemic antibiotic therapy
2. local neurosurgical attention to the complication(s) identified
3. treatment of the ear lesion.

Antibiotics have radically reduced the incidence of complications, but have had much less effect on the mortality of established complications. Of equal importance, they have altered the clinical pattern of presentation, introducing an element of ‘masking’. Specific examples will be discussed under individual complication headings, but it is indicative of the problem that Pfaltz and Griesemer (1984) found normal otoscopic appearances in 10% of children with mastoiditis.

It is unwise to offer rigid recommendations for antibiotic therapy, for several reasons. First, the armamentarium of available drugs is changing rapidly, so suggestions at the time of writing could be obsolete within a short period. Second, the pattern of infecting organisms changes, as has been discussed above. Lastly, bacterial resistance varies with time, and from one place to another. Microbiologists are usually familiar with the drugs locally available, and often have developed policies for treating particular types of infection. Advice from the
The pathologist in the hospital where treatment is undertaken should inform the otologist's selection of antibiotics.

Certain general principles can however be addressed. Antibiotics must be used in large doses, and preferably should be administered intravenously. Drugs should be chosen on probabilities, without waiting for culture and sensitivity reports. Changes should be based on clinical responses and determined by bacteriological reports. When the ear infection is acute, a drug competent to kill *Haemophilus influenzae* must be used. Since this organism is becoming increasingly resistant to ampicillin, chloramphenicol is usually the drug of choice. The recommended dose is generally 100 mg/kg per day (Brand, Caparosa and Lubic, 1984). The risks of agranulocytosis and aplastic anaemia demand that its administration be repeatedly monitored by blood counts (every 2 days). It is usual to combine chloramphenicol with a penicillin. Since many staphylococci produce beta-lactamase, methicillin or flucloxacillin may be chosen. The possibility of Gram-negative infections, and particularly with *Pseudomonas aeruginosa*, in chronic ear disease requires consideration of antibiotics such as azlocillin, ticarcillin - these have replaced carbenicillin, since they are more effective. Aminoglycosides, of which gentamicin is at the moment the typical example, are effective against Gram-negative aerobes. Gentamicin is generally given in a dose of 4.5 mg/kg per day, after a loading dose of about 1.5 mg/kg - the precise quantity depending on serum creatinine, and lean body mass. The risks of otoxicity and nephrotoxicity must be minimized by measuring peak (20 minutes after intravenous administration) and trough (predose) serum levels. The cephalosporins also offer members that are bactericidal for beta-lactamase-producing cocci, and for Gram-negative rods. Metronidazole, in a dose of 400-600 mg 8 hourly remains the drug of choice for Gram-negative anaerobic organisms such as *Bacteroides fragilis*. This drug has been shown to penetrate well into the pus within brain abscesses (Ingham, Selkon and Roxby, 1977). At one time sulphadiazine, in combination with a penicillin, was favoured because of its ability to pass the blood-brain barrier. It is well recognized now that penicillin, and many other drugs enter the cerebrospinal fluid readily in high doses when the meninges are inflamed. Chloramphenicol, penicillin and metronidazole all penetrate abscess capsules in effective doses (Maurice-Williams, 1983). The numerous different combinations described in 'case report' literature should be considered against this background of principles.

**Treatment of the ear**

Acute otitis media will usually be cured by the antibiotic selected for the treatment of its complications, but occasionally, a myringotomy will be needed. If cortical mastoidectomy becomes necessary, it is customary to advise removal of the bony covering of the sigmoid sinus, and also of the middle fossa dura, since extradural pus or granulation tissue can exist deep to intact bone.

Chronic middle ear diseases poses the eventual need for some form of radical or modified radical mastoidectomy. Timing is important. Generally, with exceptions that will be described, it is advisable to wait until the intracranial complications have been controlled before operating on the ear. Deterioration of the state of a complication, despite appropriate treatment could, however, impose the need for earlier intervention.
Investigations

The advent of computerized tomographic (CT) scanning and, in particular, the introduction of the latest generation of high resolution scanners with the ability to reconstruct images in different planes, has revolutionized the investigation of intracranial complications to the same dramatic extent that antibiotics have affected the incidence and prognosis. By the use of CT scanning, intracranial masses - extradural abscesses - can be identified, localized and monitored during treatment. Before the advent of these techniques, masses could be recognized by straight X-rays, which might show shift of a calcified pineal gland, by EEG, by arteriography, which offered the best way to demonstrate supratentorial masses, by distortion of the vascular pattern, or by air encephalography and ventriculography, with certain attendant risks. In the near future magnetic resonance (MR) imaging may prove to be better than CT scanning. Evaluations are at present in progress.

Lumbar puncture, to provide cerebrospinal fluid for examination, still has an essential role in diagnosing meningitis, but its risks in the presence of raised intracranial pressure must be recognized. Its contribution to the diagnosis of lateral sinus thrombosis is discussed under that heading. Specialized forms of angiography for example digital subtraction venography, are also mentioned below.

Extradural abscess

Involvement of the dura mater by spreading disease constitutes pachymeningitis. Most often such spread is preceded by bone loss, through demineralization in acute infection, or erosion by cholesteatoma in chronic disease. Non-infected cholesteatoma may expose and coat the dura with matrix, without inflammatory reaction, but more often an inflammatory response produces granulation tissue on the surface of the dura. Fortunately, the dura is tough and resistant to invasion and destruction. Commonly, the result of infection reaching its outer surface is the development of a collection of pus between it and the more superficial bone. This constitutes an extradural abscess, and this is the commonest of all intracranial complications arising from middle ear infections.

A middle fossa extradural abscess may strip dura from bone extensively on the inner surface of the squamous temporal bone, even to the extent of producing a sizeable intracranial mass, which, by raising intracranial pressure can, albeit rarely, cause focal neurological signs and papilloedema. Erosion through the skull to the exterior from there would produce a subperiosteal abscess, the classical but rare 'Pott's puffy tumour'. Most middle fossa extradural abscesses are confined to the upper surface of the tegmen tympani, with much less dramatic results, since firm attachment of the dura to the arcuate eminence prevents separation of a large area from the bone, and impedes the development of a large volume of pus. More rarely, an extradural abscess may develop medially to the arcuate eminence, over the petrous apex. Irritative involvement there of the Gasserian ganglion of the trigeminal nerve, and of the sixth cranial nerve, produces the characteristic features of Gradenigo's syndrome - facial pain, diplopia, and aural discharge. Posterior fossa extradural abscesses are limited anatomically by the attachments of the dura laterally to the groove for the sigmoid sinus, and medially to the region of the internal auditory meatus and the subarcuate fossa. Posterior extension around the sigmoid sinus produces a sigmoid sinus-perisinus abscess. This may contribute to the development of, and may be associated with, thrombophlebitis developing.
within the sigmoid and transverse sinuses. Very rarely such a perisinus abscess may extend through the jugular foramen into the neck.

Clinical features

The clinical pattern depends on the site of the abscess, its size, duration and the rate of its development. The discussion above touched on some of the features associated with the more unusual patterns of extradural pus but, in most instances, the features are vague and rather non-specific. Indeed, many times an extradural abscess is an incidental finding uncovered during mastoid surgery. With chronic ear disease, a complaint of headache, broadly spread on the side of the affected ear, especially when accompanied by malaise, is a suspicious symptom. If the abscess communicates freely with the middle ear, there is, characteristically, intermittent relief from pain during episodes of aural discharge.

Diagnosis

Ultimately, diagnosis depends on operative findings. Suspicion may be confirmed by the appearances of a CT scan, and this investigation is essential to exclude a possible brain abscess in those unusual patients presenting with raised intracranial pressure.

Management

Suspicion of dural inflammation or an extradural abscess is an indication for surgical exploration. Released pus is evacuated, and enough bone should be removed for an area of healthy dura to be exposed all the way round the diseased portion. Granulation tissue attached to the dura should not be disturbed, for fear of breaching the dura and infecting the subdural spaces. The possibility of other coexisting complications must be considered and excluded or treated. Appropriate antibiotic treatment will be needed, especially when the extradural abscess complicates acute otitis media. In the absence of other complications, recovery should be as rapid and as complete as after uncomplicated mastoid surgery.

Subdural abscess (empyema)

Spread of infection through the dura exposes the subdural space to the hazards of infection, which become manifest as widespread leptomeningitis or, if the accumulating fluid is contained, as subdural effusion or abscesses (subdural empyemas). The rate of spread probably determines the clinical and pathological pattern, and the type of organisms may also be important. Dawes (1979) described the predominance of non-haemolytic streptococci in subdural abscess. As is the case with other intracranial complications, the condition is frequently, if not usually, associated with other complications, and that should be expected from an understanding of the way in which it develops.

As has been noted, the dura is very resistant to destruction, and granulation tissue developing on its inner surface as an inflammatory reaction tends to obliterate the adjacent space. This granulation tissue may eventually be converted to fibrous tissue. Eventual necrosis of the dura may lead to infection of the subdural compartment. At first, a seropurulent effusion collects. Gower and McGuirt (1983) identified sterile effusions in the subdural space in a small proportion of patients with subdural fluid collections, and observed that none had
been recorded before the advent of CT scanning. Eventually, a seropurulent effusion becomes frankly purulent, and extends over the surface of the cerebral hemisphere to an extent limited by the granulation tissue obliteration of the space. Continuing granulation tissue invasion loculates the developing abscess.

The abscess may remain small near the site of dural penetration, or it may extend widely with the production of a volume of pus large enough to act as a space-occupying lesion. Adjacent cortical veins may become involved with thrombophlebitis, which is responsible for some of the clinical features. This process may produce multiple small abscesses within the brain adjacent to the preceding subdural infection. The subdural pus tends to accumulate near the falx cerebri, and particularly where that structure joins the tentorium cerebelli. Healing may be associated with fibrosis in the limiting granulation tissue, with obliteration of the subdural space. The established pathological pattern is commonly one of numerous multiloculated abscesses over the convex surface of the cerebral hemisphere, and between the hemispheres along the falx. Although non-haemolytic streptococci are often the infecting organisms, Gower and McGuirt (1983) reported the finding of *Haemophilus influenzae* in all of eight non-sterile effusions, although only one of these was of the sinister type B.

**Clinical features**

The development of a subdural empyema is heralded by the development of severe headache and drowsiness, which is followed by the onset of focal neurological symptoms, both irritative as fits, and paralytic.

Drowsiness may develop over a few hours and proceed quickly to coma. Paralysis of one upper or lower limb may rapidly extend to hemiplegia. Hemianopia and hemianaesthesia occur, and if the lesion is on the dominant side, aphasia develops. Epileptic fits of Jacksonian type, starting locally and spreading to affect one side of the body, may precede the weakness. These fits sometimes become increasingly frequent, and are probably the result of the cortical thrombophlebitis. Papilloedema is not common, nor are cranial nerve palsies, although they have been described in the fully developed picture. The site of the fits, and the pattern of weakness should suggest the position of the empyema.

**Diagnosis**

Meningism may accompany the headache. Despite meningism, the clinical picture can be distinguished from that of meningitis by virtue of the characteristic neurological localizing features. The rate of development, over hours rather than days, is much faster than would be expected from a typical brain abscess. In children suspected of having meningitis, subdural empyema should be seriously considered if there is no response to treatment, or if motor seizures occur (Gower and McGuirt, 1983). Nowadays, a definitive diagnosis depends on CT scanning. Lumbar puncture is helpful. The cerebrospinal fluid pressure is raised, but the sugar content is normal, and cultures are sterile. The fluid may occasionally be turbid if there is marked pleocytosis. If CT scanning is not available, angiography and exploratory burr holes may be needed to clinch the diagnosis.
Management

This complication must be managed in close cooperation with a neurosurgeon. Treatment comprises the administration of massive doses of systemic antibiotics, drainage of the subdural fluid with irrigation of the subdural space if the collection is purulent, and treatment of the ear disease. The choice of antibiotic will probably now include intravenous penicillin and chloramphenicol, because of the increasing presence of Haemophilus influenzae in acute infections. The possibility of Gram-negative organisms from chronic ear disease may impose the need to use aminoglycosides. Acute ear infection will almost always require myringotomy and sometimes cortical mastoidectomy, while appropriate surgical treatment to the mastoid will be demanded by chronic infection, usually after the patient’s general state has been stabilized by neurosurgical treatment. The neurosurgical management involves at least one burr hole to sample the fluid, and several may be made in order to establish an irrigation system. Previously, when disease was often more advanced before diagnosis, it was common practice to advise burr holes on both sides of the skull, since the abscess can track under the falx to the opposite hemisphere.

Antiepileptic medication must be prescribed to suppress fits, and may have to be continued for many months after recovery.

Lateral sinus thrombophlebitis

Thrombophlebitis may develop in any of the veins adjacent to the middle ear cleft. Of these, the lateral sinus, comprising the sigmoid and transverse sinuses, is the largest, threatening the greatest risks when it is filled with suppurating blood clot. The process is usually, but not invariably, preceded by the development of an extradural perisinus abscess. Mural thrombus then partly fills the sinus. Progressive expansion of this clot eventually occludes its lumen. The clot may become partly organized, and may be partly broken down and softened by suppuration. From this stage on, the release of infecting organisms and infected material into the systemic venous circulation causes bacteremia, septicaemia, and septic embolization. Extension or propagation of the thrombus upwards may extend to the confluence of the sinuses - the torcular Herophili - and beyond there to the superior sagittal sinus. Invasion of the superior or inferior petrosal sinuses may extend the disease to the cavernous sinus. Venous thrombophlebitis extending into brain substance accounts for the very high association of this complication with brain abscesses. Downward propagation of thrombus into and through the internal jugular vein can reach the subclavian vein (Surkin et al, 1983; Albert and Williams, 1986).

The harmful effects then derive from the release of infective emboli into the circulation, and from the haemodynamic disturbances caused to venous drainage from inside the cranial cavity.

The use of antibiotics has greatly reduced the incidence of lateral sinus thrombosis. Formerly, most instances were associated with acute otitis media in childhood; now the incidence is much higher in chronic ear disease (Teichgraber, Per-Lee and Turner, 1982), although one series, reported by Gower and McGuirt (1983), was dominated by acute infections. The mortality from lateral sinus thrombosis before the days of operative treatment was 100%. This fell to 50% in the early 1900s (Gower and McGuirt, 1983), while reports
since the advent of antibiotic treatment still describe death rates over 20% (Seid and Sellars, 1973).

Before the use of antibiotics, the commonest infecting organism was the beta-haemolytic streptococcus, and this could reliably be cultured from the blood. Its propensity for red cell destruction caused progressive anaemia, which was a characteristic feature of the disease in the pre-antibiotic era. Nowadays, a large variety of mixed flora may be found. Aural cultures by Venezio, Naidich and Shulman (1982) grew *Proteus mirabilis*, staphylococci, *Streptococcus pneumoniae*, and *Bacteroides oralis*, and in only one of 14 cases were blood cultures positive. Similar floral diversity, with cultures producing *Bacteroides*, streptococci, enterobacteriaceae and other Gram-negative rods was reported by Teichgraeber, Per-Lee and Turner (1982). The latter account describes many negative cultures.

**Clinical features**

The classical pattern, before antibiotic modification, was of a severe pyrexial wasting illness in a patient with middle ear infection. Usually this would develop over several weeks, but occasionally a fulminating infection by virulent organisms would arise soon after the onset of an acute otitis media. Fever was high and swinging, following a so-called 'picket fence' pattern. Rigors, with profuse sweating, occurred as the temperature rose rapidly to 39-40°C and then fell. The shivering during these rigors has been described as so violent as to shake the bed, but few otologists practising today will have seen this phenomenon. Headache and neck pain were the rule. Emaciation was accompanied by progressive anaemia.

Many clinical features depended on the gradual extension of thrombus, effectively limiting the systemic dissemination of infection. As clot extended down the internal jugular vein, it would be accompanied by perivenous inflammation, with tenderness along its course. This tenderness descended the neck with the clot, and might be accompanied by perivenous oedema, or even suppuration in jugular lymph nodes. Perivenous inflammation around the jugular foramen occasionally caused paralysis of the lower three cranial nerves. Raised intracranial pressure produced papilloedema and visual loss. Hydrocephalus could develop if the larger or only lateral sinus was occluded, or if the clot reached the superior sagittal sinus. Extension to the cavernous sinus, along the superior petrosal sinus, presented with chemosis and proptosis of one eye. This could spread to the other eye if the circular sinus became involved.

Embolic propagation of infected clot and organisms produced infiltrates in the lung fields and septic spread to large joints and subcutaneous tissues. Other viscera and the pleuroperitoneal cavity were also targets for embolization. Although these distant effects usually developed late in the course of the disease, they could be presenting features if the insidious nature of the sinus disease had prevented its earlier recognition. Even today there are reports of chest disease caused by septic pulmonary emboli, from a completely inapparent lateral sinus thrombosis (Hawkins, 1985). This is even more likely to occur now, perhaps because of the 'masking' effects of antibiotics on the primary ear disease.

This masking by antibiotic treatment has muted the more dramatic and often diagnostic clinical character of the disease (Teichgraeber, Per-Lee and Turner, 1982). The clinical picture
has so far been described in the past tense, to indicate that the disease has changed, but it is important to remember these dramatic features as they may still be encountered occasionally. What follows is an attempt to describe the clinical pattern more likely to be met today, in those patients who have been treated with antibiotics before coming to the attention of the otologist.

Patients always feel ill and persisting fever is still usual, but often without the violent swings and rigors of earlier times. Earache and neck pain with mastoid tenderness and stiffness along the sternomastoid muscle are universal features. These, together with fever, should be recognized nowadays as the most consistent clinical features of lateral sinus thrombosis. Anaemia is now rare. Cases have even been described with no evidence of ear infection (Hawkins, 1985). Papilloedema is still a common finding - described in 50% of instances by Wolfowitz (1972). The state of mental awareness may be impaired, with drowsiness, lethargy and coma. Other intracranial complications should be expected in nearly 50% of patients with lateral sinus thrombosis. Of these, meningitis and brain abscess are the most frequent, and their symptoms can so dominate the illness that the lateral sinus thrombosis may be inadvertently overlooked.

Extension of infected clot down the internal jugular vein is always accompanied by tenderness extending along its course down the neck, and localized oedema over the thrombosing internal jugular vein may still be seen.

Very rarely, thrombosis may extend to the subclavian vein (Surkin et al, 1983; Albert and Williams, 1986). In the latter report, engorged collateral veins developed over the shoulder, and intravascular clotting was so extensive that it mopped up platelets and caused thrombocytopenia.

The clinical examination will usually, but not always, indicate middle ear infection. Tenderness over the mastoid process and along the sternomastoid muscle is almost always apparent, and must be regarded seriously as an important sign exciting suspicion of this complication. Examination of the fundi may show papilloedema. A rare finding is pitting oedema over the occipital region, well behind the mastoid process, caused by clotting within a large mastoid emissary vein; this constitutes Griesinger's sign. Physical signs of other associated complications are often present.

There is no single pathognomonic sign of lateral sinus thrombophlebitis. Vigilance, a high level of suspicion, and investigation along the lines suggested below should secure recognition of this dangerous complication.

Investigation

A full blood count may show anaemia with a raised white cell count and raised erythrocyte sedimentation rate, but none of these possible abnormalities is sufficiently specific to be helpful in making or excluding the diagnosis.

Blood cultures, with specimens taken as the temperature rose to its swinging peak, used to be considered a most important diagnostic step, but their value nowadays has become
greatly diminished, since bacteraemia, with rigors, is so much less common, and because of the development of more reliable diagnostic tests.

A lumbar puncture should be performed, if papilloedema does not suggest that raised intracranial pressure might precipitate coning. Examination of the cerebrospinal fluid is the most important way of identifying meningitis. In uncomplicated lateral sinus thrombosis, the white blood count in the cerebrospinal fluid will be low when the cause is chronic middle ear disease, and somewhat raised in acute otitis media (Gagnon, Sierra-Dupont and Huot, 1976). The cerebrospinal fluid pressure is usually normal. Variations in protein and sugar levels in the cerebrospinal fluid are not sufficiently consistent to be useful.

The Queckenstedt, or Tobey-Ayer test is traditionally recommended whenever a lumbar puncture for possible intracranial infection is indicated. Queckenstedt (1916) had described the manoeuvre as a means of recognizing spinal cerebrospinal fluid obstruction, but Tobey and Ayer (1925) declared the test diagnostic of lateral sinus thrombosis. The test involves measurement of the cerebrospinal fluid pressure and observing its changes on compression of one or both internal jugular veins by fingers on the neck. In the normal subject, compression of each internal jugular vein in turn is followed by a rapid rise of cerebrospinal fluid pressure of 50-100 mmHg, above the normal level. There is an equally rapid fall on release of pressure. It is normal for there to be a difference in rise on the two sides, but unusual for it to exceed 50 mmHg. In a typical case of lateral sinus thrombosis, pressure over the vein draining the occluded sinus causes either no rise, or a very slow one of 10-20 mmHg. Compression of the normal internal jugular vein, on the other hand, produces a rapid pressure rise to two or three times the normal level. Unfortunately, there are instances in which the Tobey-Ayer test may suggest lateral sinus thrombosis, when there is none and also false negative results, with a normal finding in the presence of lateral sinus thrombosis. As Albert and Williams (1986) have fully discussed, the false negative results stem from collateral channels draining the dural venous sinuses. False positive results appear if a normal lateral sinus is very small or absent, creating an erroneous impression of occlusion by disease.

Before the advent of CT scanning and angiography, the Tobey-Ayer test, combined with blood cultures was considered of high diagnostic importance. Now the emphasis has changed.

CT scanning

A CT scan is an essential investigation for any patient with suspected intracranial complications. It may show the increased density of fresh clot (Venezio, Naidich and Shulman, 1982). Filling defects within the sinus can often be shown with iothalamate (Conray) enhancement, and failure of opacification may be evident. Septic thrombosis shows as intense inflammatory enhancement of the sinus walls and of the adjacent dura. Other findings, which are non-specific, include cerebral oedema, reduced ventricular size because of oedema, parasagittal haemorrhages, and tentorial enhancement from collateral venous flow. Computerized tomographic scanning is essential also to identify or exclude accompanying complications, for example brain abscess and subdural empyema.
Angiography

Despite the help available from CT scanning, and its literally vital role in exposing other complications, the definitive investigations for lateral sinus thrombosis (before operative exposure) involve angiography, to demonstrate the obstruction and its site and extent, and the anatomical arrangement of the individual's venous drainage. There is a possible risk of displacing loose infected thrombus, but the consensus view is that vascular studies must be undertaken whenever a lateral sinus thrombophlebitis is suspected.

Arteriography, with radiopaque dye in the carotid artery can show the venous outflow during the venous phase. The demonstration is made clearer by subtraction angiography. This technique involves precisely registered superimposition of a negative arteriogram on a positive film of the bone structures. The effective cancellation of the skeletal image leaves the vascular pattern clearly exposed.

Digital subtraction venography is the preferred vascular imaging technique available at present. The contrast material is administered intravenously, and so without anaesthesia, and without any of the risks of arteriography. The imaging is produced by digital computer techniques. These allow the much diluted agent to be traced even after passing through the heart and onwards into the systemic circulation for a second systemic venous transit.

For completeness, radioisotope scanning with gallium should be mentioned. This is a technique that can show the 'hot spots' of sepsis.

Treatment

Treatment consists of the administration of antibiotics together with exposure of the lateral sinus and incision and removal of its contents.

The principles involved in the choice of an antibiotic for treating any intracranial infection have been discussed in the introductory comments to this chapter, and those guiding rules should govern the selection of the most appropriate agents for managing a patient with lateral sinus thrombophlebitis. Intravenous administration will generally be recommended. As Teichgraeber, Per-Lee and Turner (1982) advise, most patients should receive a combination of two or more of the following: ampicillin, chloramphenicol, a cephalosporin and an aminoglycoside.

In the past, anticoagulants were recommended. Most writers now agree that there is no regular place for their use except in those very rare instances where spreading thrombus has reached the cavernous sinus (Hawkins, 1985).

Surgical

Of all intracranial complications, lateral sinus thrombophlebitis is the most important for which operation should be undertaken early, in order to expose and treat the infected lesion. The same could be said for extradural abscess, but this is a much less serious condition, and often discovered incidentally at the time of operative exploration. This early intervention contrasts with the principles governing the otological surgical management of
other intracranial complications, where it is almost always advisable for the complication to be treated, medically or with neurosurgical attention, first and for the infecting ear to receive operative attention later, when the patient's condition has greatly improved. Exceptions to this principle, when treating the other complications, usually arise if the ear continues to infect the intracranial contents, preventing the expected improvement in the brain abscess or meningitis. Lateral sinus thrombophlebitis, however, like an extradural abscess, is a localized, often purulent lesion to which easy access is available only through the mastoid region. Drainage of the infected site then requires a mastoidectomy operation.

Before operation intensive medical treatment must be started, and the timing of mastoidectomy should depend to some extent on the response. During this early period, under otological care, the temperature chart should be watched every 4 hours, and the central nervous system should be examined once or preferably twice daily. Unless there is very rapid improvement, and certainly if these is deterioration, mastoid exploration should be carried out within the first 2 days.

If the lateral sinus thrombophlebitis follows acute otitis media and coalescent mastoiditis, cortical mastoidectomy is needed. In chronic otitis media, a radical mastoidectomy is undertaken through a postaural incision. With the temporal bone drill, the mastoid is opened and the region of the sinus plate approached. A perisinus abscess may declare itself with an outflow of pus through a tract in necrotic bone over the sinus. That necrotic plate can be separated from the underlying sinus with probes and curettes.

Often, there is no bone necrosis and the sinus plate is firm, healthy and intact. The sinus must then be deliberately uncovered, by drilling the plate, at first with a fast cutting burr, and later as the soft tissue becomes visible through the thinning bone with a diamond paste burr. When the bone is tissue paper thin, it can be lifted off the underlying sinus with flat blunt dissectors. At this stage, the sinus should be fairly widely exposed upwards towards the jugular bulb. Its appearance determines further action.

The normal healthy sinus is soft, bluish in colour and compressible with a blunt probe. If this is the case a small needle should be inserted through the wall to seek a free flow of venous blood. Such a flow would indicate that the diagnosis of lateral sinus thrombophlebitis was incorrect and, apart from stopping the bleeding by placing a small piece of free muscle tissue on the puncture, no further action is needed. The sinus might feel firm, and appear white and opaque. This would suggest that its lumen was occluded with fibrosing clot or fibrous tissue. In these circumstances it should be opened with a sharp instrument, and the absence of blood or necrotic debris confirmed by inspection. Lund (1978) pointed out that an obliterated cord of scar tissue is sometimes found as testimony to a 'silent' lateral sinus thrombophlebitis. If the sinus wall is covered with granulation tissue or if it is necrotic, the sinus must also be opened and the abscess and necrotic tissue within it removed.

This evacuation must extend in both directions - upwards towards the confluence of the sinuses, and downwards if necessary as far as the jugular bulb. Pus and any unorganized thrombus should be removed. In the past, it was advised that clot removal should be extended in each direction until blood flowed freely from either end of the opened sinus. It is now generally agreed (Teichgraeber, Per-Lee and Turner, 1982; Hawkins, 1985) that it is unnecessary to remove organized thrombus, and that it is no longer desirable to follow clot...
centrally until free blood flow is established. If profuse venous bleeding is encountered, the lumen of the sinus should be obliterated with a ribbon gauze pack, impregnated with an antibacterial agent, inserted between the bone and sinus wall; bismuth iodoform paraffin paste (BIPP) should be avoided since it is radiopaque.

Whenever cortical mastoidectomy is performed for coalescent mastoiditis, even if intracranial complications are not suspected, the sigmoid sinus should be exposed and needled. This is a routine measure to avoid missing an unsuspected lateral sinus thrombophlebitis. The same investigation of the sinus is not recommended during radical mastoidectomy for chronic ear disease, unless the operation is being conducted during the management of an intracranial complication.

Internal jugular vein ligation used to be considered important to prevent dissemination of infected clot. The current consensus view, as summarized by Teichgraeber, Per-Lee and Turner (1982) is that ligation should be reserved for the very rare cases in which septicaemia does not respond to initial antibiotic treatment and surgery and that it should be considered for children showing signs of embolization.

**Meningitis (leptomeningitis)**

This is a major and serious complication of middle ear infection, and probably still the commonest intracranial complication (Gower and McGuirt, 1983). Before the days of antibiotics most sufferers died. Nowadays, recovery is usual provided that recognition is prompt and treatment expeditious. The patients at greatest risk are those with multiple complications, which may be overlooked because of the severe symptom of the meningitis. As with all other otogenic complications, the incidence, particularly that from acute otitis media, has fallen greatly with the use of antibiotics. Although most reports indicate that it is more frequently now a complication of chronic ear disease, childhood otogenic meningitis is seen most often as a complication of acute middle ear infection (Gower and McGuirt, 1983). It is probably fair to say that, in adults, it is now more commonly a complication of chronic disease.

Although spread may be through any of the channels previously described, such as preformed pathways, otogenic meningitis usually arises by direct spread through necrosing bone from the middle ear cleft. The rate of development depends on factors discussed in the introduction to this chapter, and particularly on the virulence of the organism, the resistance of the host, and the development of preformed access by bone erosion. Suppurative labyrinthitis, described later in this chapter, offers access to the cerebrospinal spaces through the internal auditory meatus and through the vestibular and cochlear aqueducts. Rarely, rupture of an established brain abscess into the subarachnoid space may lead to meningitis. Under the least favourable circumstances meningitis can develop within hours of onset of acute suppurative otitis media.

The organisms usually responsible for acute infection are *Haemophilus influenzae*, especially type B, and *Streptococcus pneumoniae*, of which type III has a sinister reputation for causing rapid complications. Infection from chronic ear disease may be caused by any of the organisms normally found in these conditions (Lampe and Edwards, 1984). Gram-negative
enteric organisms, *Proteus*, and *Pseudomonas*. Anaerobes such as *Bacteroides* species have also been reported (Siegler, Faiers and Willis, 1982).

The initial inflammatory response of the pia-arachnoid to infection is an outpouring of fluid into the subarachnoid space, with a rise in cerebrospinal fluid pressure. This fluid soon becomes permeated with white blood cells, and then with rapidly multiplying bacteria. The organisms feed on glucose, and thereby reduce its level in the cerebrospinal fluid, producing the characteristic biochemical feature of pyogenic meningitis. Once purulent, a sticky exudate is formed. This accumulates at first in the basal cisterns, and more rarely at the vertex. Free flow of cerebrospinal fluid is impeded by exudate obstructing the ventricular foramina to cause a non-communicating hydrocephalus. Obstruction to cerebrospinal fluid flow in the subarachnoid spaces may produce communicating hydrocephalus. Irritation of upper cervical nerve roots by inflammatory exudate is the basis for the classical features of this condition - neck pain and neck stiffness. Exudate collecting around exit foramina of cranial nerves can cause palsies in the late stages of the disease. Spread of infection along the Virchow-Robin spaces into the brain substance sometimes leads to brain abscess, while accumulations of the exudate in loculated masses on the cerebral surface are no different from those found in subdural empyema, which has been discussed under that heading.

**Clinical features**

The two most constant and reliable early clinical features are headache and neck stiffness. At first, the headache is often localized to the side of the head of the infected ear, but soon becomes generalized and 'bursting'. There is malaise and pyrexia, often to 39°C. Initial neck stiffness shows as resistance to flexion; later, rigidity or retraction develop. Mental hyperactivity usually colours this early stage, with restlessness and fretfulness in children. Anxiety, punctuated by periods of drowsiness is usual in adults. At this stage, the tendon reflexes may be exaggerated. Photophobia is a constant characteristic symptom and, before neck stiffness is marked, the patient may lie curled up away from the light. Vomiting, caused by raised intracranial pressure is also a feature.

As the condition progresses, all these symptoms become more severe. The headache may be excruciating, and neck rigidity is marked, with a positive Kernig's sign, retraction and later on opisthotonus. The temperature remains uniformly raised, with none of the swinging pattern, which used to characterize lateral sinus thrombosis. Gradually, the tendon reflexes become less marked, and the abdominal reflexes may be lost.

Deterioration is marked by alternating delirium and stupor, passing finally into coma. The tendon reflexes disappear, and cranial nerve palsies develop. Eventually Cheyne-Stokes respiration follows, with fixed dilated pupils, then coma and death.

Any focal neurological signs, especially in the early stages, should arouse suspicion of a subdural or cerebral abscess. Similarly epileptic fits do not occur with otherwise uncomplicated meningitis. The neck stiffness, which is so typical of the disease may be delayed for several days from the onset, especially if the first accumulation of exudate is vertical, rather than in the basal cisterns.
Diagnosis

The diagnosis is made by examination of cerebrospinal fluid. Any patient with middle ear infection, headache, and neck stiffness must undergo a lumbar puncture. At the same time, suspicion of other complications must always be entertained, and possible brain abscesses and subdural empyemas need to be excluded, preferably by CT scanning. In the earliest stages of otogenic meningitis the only abnormality on lumbar puncture is a rise in fluid pressure above the normal 100-150 mmHg. As the infection proceeds white cells accumulate in the cerebrospinal fluid, and the fluid becomes cloudy and then turbid in appearance. On histological inspection, most of these cells will be found to be polymorphonuclear leucocytes, which are not present in normal cerebrospinal fluid. They increase in number to reach the range of 0.1-10 x 10^9/L (100-1000.000/mm³), although with Staphylococcus epidermidis, counts below 0.1 x 10^9/L may be met. As the inflamed blood-brain barrier allows free passage into the cerebrospinal fluid, its constitution approximates more closely to that of serum, and this can be demonstrated by biochemical tests. Thus the protein content may rise from a normal 150-400 mg/L to a raised level of 2-3 g/L. The chloride content may fall from the normal 120 mmol/L to 80 mmol/L. The appearance of bacteria in the cerebrospinal fluid is accompanied by a fall of cerebrospinal fluid glucose levels from the normal value of 1.7-3.0 mmol/L to zero. Bacteriological examination of cerebrospinal fluid is first undertaken by direct examination after Gram staining, and then by culture of the fluid. Despite positive diagnostic findings on cellular and biochemical testing, positive bacteriological diagnosis is by no means the rule, and so treatment cannot wait for, nor depend upon, it. The lumbar puncture findings are decisive, when no mass shows on CT scanning, even without bacteriological recognition. There is no other complication in which the cerebrospinal fluid sugar level is lowered, and few in which the white cell count is so high. A brain abscess, if leaking into the subarachnoid space may cause a huge rise of cerebrospinal fluid white cell count even to more than 50 x 10^9/L (50.000/mm³), and a subdural abscess to counts over 0.1 x 10^9/L. In both of these the cerebrospinal fluid pressure may be raised, but in neither is the cerebrospinal fluid sugar level reduced. In the presence of cerebrospinal fluid pleocytosis, CT scanning should exclude either form of abscess, leaving a diagnosis of meningitis unchallenged.

Treatment

Surgical

As with most other complications, treatment of the intracranial sepsis should take precedence over management of the otitis media. Medical treatment of the meningitis is of paramount importance, and any operation for the ear condition should, if possible, be deferred for several days until the patient's general condition has improved. Years ago, before antibiotics offered hope for cure, appropriate ear surgery was undertaken as soon as the diagnosis had been made. Nowadays, urgent surgical intervention should be advised only if the expected response to treatment does not appear. Certainly deterioration or failure of response over 48 hours implies loculated infection in the mastoid, needing surgical drainage. In acute otitis media, Gower and McGuirt (1983) would advise early middle ear drainage, with myringotomy, either repeated, or possibly assisted in drainage by insertion of a ventilation tube. Coalescent mastoiditis is an indication for cortical mastoidectomy but, in many, if not most instances, cure of the meningitis by antibiotics cures the preceding acute
infection. As with other complications, chronic middle ear disease needs eradication by some form of radical mastoidectomy, but again that should be deferred if possible until the dangerous meningitis is under control.

**Medical**

The lumbar puncture used for diagnosis may be repeated several times to reduce intracranial pressure, possibly a second time in the first 24 hours, and then daily until improvement is assured. Subsequent punctures are needed to check the state of the patient, and discharge from hospital must not be considered until the cerebrospinal fluid characteristics have returned to normal. Differential white cell counts are needed, since improvement may be indicated by a change from polymorphs to macrophages, even though the total count persists at, say, the 0.2 x 10^9/L level. In earlier days, the initial and subsequent lumbar punctures were used to instil intrathecal antibiotics, and in particular pure crystalline penicillin in a dose not exceeding 5000-10,000 units in 5 mL of normal saline. Intrathecal antibiotics pose a risk of epilepsy, and their use now has largely been abandoned.

The mainstay of medical treatment rests with large doses of systemic antibiotics. A few years ago a standard choice was intramuscular penicillin, intrathecal penicillin, and sulphadiazine (which, of all sulphonamides, most readily crosses the blood-brain barrier). Streptomycin might have been recommended as an occasional adjunct because of its efficacy against *Haemophilus influenzae*. Today, as has been discussed in the introduction to this chapter, it is more difficult to be dogmatic. Because of its frequent role as a causative agent, *Haemophilus influenzae* must be a target of any regimen, and since more and more strains are becoming resistant to ampicillin, chloramphenicol is considered the first choice, combined with ampicillin or penicillin. The risk of toxic effects from chloramphenicol must be minimized by regular blood cell counts.

Rifampicin has recently been shown to be an effective agent against *Haemophilus influenzae* type B that has previously not responded to chloramphenicol (Lewis and Priestley, 1986). This drug has the unique ability to penetrate pus and kill phagocytosed organisms - even after oral administration.

Agents likely to be effective against Gram-negative organisms must also be considered when the infection is secondary to chronic middle ear disease. In these categories are azlocillin, ticarcillin, and some newer cephalosporins such as ceftazidime. All are less toxic than aminoglycosides like gentamicin. Whichever combination is selected, the preferred route is intravenous. Systemic therapy must be continued for at least 10 days after apparent clinical recovery. If *Bacteroides* are found on anaerobic culture, metronidazole should be administered in a dose of 400 mg 8-hourly (Siegler, Faiers and Willis, 1982).

Failure of an adequate response may be the result of:

(1) organisms resistant to the chosen antibiotics: a change should be planned, guided as far as possible by bacteriological data

(2) persisting leakage of infected material into the cerebrospinal fluid; urgent surgical treatment of the ear must then be considered
(3) presence of a previously unidentified other complication; CT scanning will be needed urgently for its recognition

(4) leakage into the cerebrospinal fluid from an unrecognized brain abscess.

**Brain abscess**

Otogenic brain abscesses almost always develop in the temporal lobe or the cerebellum of the same side as the infected ear from which they emanate. They are found in the temporal lobe approximately twice as frequently as in the cerebellum. In children, 25% of all brain abscesses are otogenic, while in the adult, with a greater predominance of chronic ear disease, the proportion of brain abscesses caused by ear infection is nearer 50%. Of the various routes of spread previously described, the commonest responsible for brain abscess is by direct extension of infection through an osteitic tegmen tympani, with formation of a middle fossa extradural abscess. Although the dura is very resistant to infective invasion, a local pachymeningitis may be followed by thrombophlebitis penetrating the cerebral cortex of the temporal lobe, or by extension of infection along periarteriolar Virchow-Robin spaces into the cerebral white matter. Cerebellar abscesses are frequently preceded by lateral sinus thrombophlebitis. They usually lie within the lateral lobe of the cerebellum, which may be adherent to the lateral sinus or to a patch of dura underneath Trautmann's triangle. The frequently repeated observation that intracranial complications are often multiple can be understood by this pattern of progressive involvement.

Formation of an abscess starts with an area of cerebral oedema and encephalitis. Rarely, this oedema is poorly contained and proceeds to massive cerebral oedema with spreading encephalitis. More often, the development and extension of the abscess is contained by the formation of a capsule. This fibrous tissue restriction depends on a microglial and blood vessel mesodermal response to the inflammatory process, and is variable in its rate of development. In general, capsular formation takes 2-3 weeks, and while it proceeds, the central part of the affected brain liquefies. After these stages of initial encephalitis and abscess localization, there may be a period of abscess enlargement, as renewed or continuing infection increases the volume of contained pus. Now, the features of a space-occupying lesion dominate, with rising intracranial pressure and focal neurological damage. Abscesses in the posterior fossa - in the cerebellum - cause raised intracranial pressure earlier than those above the tentorium, and rapidly rising intracranial pressure may cause 'coning' or impaction of the flocculus and brainstem into the foramen magnum, followed by fatal disruption of vital centres in the brainstem. If capsular development is slow, softening of brain around the developing abscess may allow further spread of infection into the relatively avascular white matter, with the formation of secondary abscesses, separate from the original or connected by a narrow stalk. In this way a multilocular abscess is formed. Eventually, an abscess may rupture into the ventricular system or subarachnoid space, with overwhelming meningitis and death.

Like other complications the incidence of otogenic brain abscess has fallen, but the mortality rate, even in the past decade was still 40% (Editorial, 1977). A report by Fischer, McLennan and Suzuki (1981) of their experiences during the 1970s described a mortality of 14%, so this is still a life-threatening complication, although there has been an improvement
in outlook as a result of earlier recognition by CT scanning, better understanding of bacteriology, and the addition of steroids to the treatment regimen to control cerebral oedema.

**Bacteriology**

The bacterial flora is usually a complex mixture of aerobes and obligate anaerobes. Anaerobic streptococci are the commonest organisms (Maurice-Williams, 1983). Pyogenic staphylococci are also common, especially in children, and *Streptococcus pneumoniae* and *Strep haemolyticus* are often found. Gram-negative bacilli - *Proteus mirabilis, Escherichia coli,* and *Pseudomonas aeruginosa* - are cultured with increasing frequency. This may reflect the higher incidence of otogenic abscesses from chronic ear disease, or the fact that the Gram-positive organisms are so often sensitive to, and therefore eradicated by, the most commonly prescribed system antibiotics. The recognition of obligate anaerobes, first suggested as infecting agents by McFarlan (1943) has shown that many supposedly sterile abscesses are infected by organisms of the *Bacteroides* genus, especially *B. fragilis,* and organism that produces a highly active beta-lactamase (Editorial, 1977; Ingham, Selkon and Roxby, 1977).

**Clinical features**

The earliest stage of 'encephalitis', when brain tissue is invaded, causes headache, fever, malaise and vomiting, followed by drowsiness. The symptoms may be slight, and are easily masked by those of an acute otitis media, but drowsiness should always arouse suspicion. These early features may be hidden completely by a dramatic complication such as meningitis, or even by lateral sinus thrombophlebitis. The cerebral disturbance is sometimes so slight as to be ignored, and in chronic otitis media, headache must be considered the most important symptom. Persistent headache with chronic middle ear infection is always suggestive of intracranial spread and, although it may be caused by a much less sinister cause such as an extradural abscess, the possibility of local encephalitis cannot be excluded.

If this early phase of localized encephalitis progresses rapidly to a generalized form before containment by encapsulation, drowsiness may progress to stupor and then coma and death from tentorial herniation. More often, the period of local encephalitis is followed by a 'latent period' during which the pus becomes contained within the developing fibrous capsule. Throughout this stage, which may last from 10 days to several weeks, there are no symptoms, and the preceding encephalitis illness may be forgotten, if indeed it ever attracted the patient's attention.

The next stage of an enlarging abscess causes first some clinical features, as a result of the alteration in cerebrospinal fluid dynamics, which are common to abscesses in any part of the brain, and second site-specific features caused by focal neurological impairment. Naturally, the neurological effects of damage in the temporal lobe differ from those in the cerebellum.

Abscesses are surrounded by an area of cerebral oedema and low grade encephalitis, which fluctuates in size causing variation in the severity of the symptoms. There is malaise and anorexia, weakness and lethargy. With rising intracranial pressure the pulse rate slows, and the temperature may fall to subnormal levels. Constant headache is usual, and vomiting
of cerebral type occurs in many patients. The drowsiness varies and may alternate with irritability. Thought processes may become slow. Papilloedema is often found, but only if cerebrospinal fluid pressure has remained raised for 2 or 3 weeks. As explained before, papilloedema appears earlier with an abscess in the cerebellum than in the temporal lobe. A long-standing abscess may be associated with emaciation. If intracranial pressure continues to rise, the patient eventually lapses into coma, the ipsilateral pupil dilates and finally both become fixed and dilated. Death eventually supervenes, either from the effects of raised intracranial pressure, or from overwhelming meningitis following intraventricular rupture of the abscess contents.

These are features common to all abscesses, and during their development specific neurological signs must be sought to try to localize the site of the space-occupying lesion.

**Cerebral-temporosphenoidal abscesses**

A cerebral abscess in the dominant (usually left) hemisphere often causes so-called nominal aphasia. The patient cannot name a common everyday object such as a key, a pen or a screwdriver, but he or she is able to demonstrate its use correctly. Visual field defects arise from involvement of the optic radiations. Most commonly there is a quadrantic homonymous hemianopia, affecting the upper part of the temporal visual fields, or much more rarely the lower quadrants. The fields lost are on the side opposite to that of the lesion, because of damage to fibres arising in the two retinae of the same side. Although best examined by formal perimetry, the patient's state of consciousness may prevent adequate cooperation. Simple clinical testing by confrontation can then be useful. Motor paralysis develops as the abscess enlarges. Upward development affects facial movement on the opposite side, and then, progressively, paralysis of the upper and lower limb. Inward expansion affects first the leg, then the arm, and finally the face.

**Cerebellar abscesses**

The focal features of cerebellar involvement include weakness and muscle incoordination on the same side as the lesion. Ataxia causes a tendency to fall to the side of the lesion. Clinical tests for cerebellar faults, show that the ipsilateral side is ataxic on specific testing, with a tendency to past-pointing when attempting to touch a target with a finger. The finger-nose test exposes intention tremor and past pointing, and dysdiadochokinesis is demonstrated by attempts at rapid alternating supination and pronation of the forearm, and by difficulty in rapidly touching each finger to the thumb tip. Spontaneous nystagmus may be present; this is coarse and irregular, but very variable in its appearance. Generally, it beats to the side of the lesion, and this contrasts with the paralytic jerk nystagmus of suppurative labyrinthitis. As mentioned previously, intracranial pressure rises early and rapidly with a cerebellar abscess, and the effects of this rising pressure may dominate the clinical pattern before focal signs can be recognized clinically.

**Investigations**

The investigation and management of brain abscess require a team approach, and at the earliest stage of clinical suspicion, advice should be sought from neurosurgical colleagues.
in order to plan the most expedient way of investigating the possible complication, and

treating it, and the underlying ear disease.

Radiological

Computerized tomographic scanning with and without intravenous iothalamate
(Conray) enhancement is without question the most important investigation in the diagnosis
of brain abscess. Not only can the position and size of the abscess be identified, but the
appearance of localized encephalitis can be distinguished from those of an encapsulated
abscess. The CT scan will also help to identify associated complications such as subdural
abscesses, and lateral sinus thrombophlebitis. Scanning is also the most valuable method for
observing the progress of an abscess during treatment. In the absence of CT scanning, and
before this technique became available, supratentorial masses were best demonstrated by
carotid arteriography. Upward and medial displacement of the middle cerebral artery would
indicate a temporal lobe mass. Plain X-rays of the skull are of limited value, they may show
displacement of a calcified pineal gland. Ventriculography used to have a role for
demonstrating posterior fossa masses.

Nowadays, there is no useful place for electroencephalography. Before the availability
of CT scanning, it was useful for showing abnormal delta-wave activity, which was said to
allow accurate localization in about 50% of abscesses.

Lumbar puncture

Great care is needed when cerebrospinal fluid is removed in the presence of raised
intracranial pressure, because of the risk of coning. If the patient is stuporose, has violent
headache, or papilloedema, sampling should be performed only in a neurosurgical unit with
immediate availability for intervention. Earlier in the development of brain abscess, lumbar
puncture is particularly valuable, especially to exclude meningitis, for which it is the
definitive investigation. Usually with brain abscess, a lumbar puncture will show some rise
of cerebrospinal fluid pressure, with raised protein content. Any rise in white cell count is
much less than that encountered in meningitis. If an abscess is leaking into the cerebrospinal
fluid, very high cell counts may be found. The glucose content of the cerebrospinal fluid
remains unaffected - at a normal level.

Burr hole needling

The definitive diagnosis was formerly established by needling the brain through a burr
hole to seek pus. This is a neurosurgical manoeuvre, to be conducted by neurosurgical
colleagues. With the advent of modern CT scanning, it should no longer be necessary.

Differential diagnosis

A brain abscess should be suspected during the course of any intracranial
complication. The conditions to be differentiated include meningitis, subdural abscess, lateral
sinus thrombophlebitis, otitic hydrocephalus, and brain tumour.
In meningitis, there is high sustained fever and neck stiffness, and the cerebrospinal fluid findings are abnormal, and diagnostically typical. A subdural abscess is suggested by much more rapid evolution of focal neurological signs. Lateral sinus thrombophlebitis is often a precursor of cerebellar abscess, and an abscess should be suspected if any features not typical of a thrombosed sinus develop. Otitis hydrocephalus should easily be distinguished by the absence of focal neurological signs, the CT scan findings, and the cerebrospinal fluid findings on lumbar puncture. Finally, if evidence of a space-occupying lesion is found in a patient with middle ear disease, even though a brain abscess must be presumed, a brain tumour, of coincidental origin, requires exclusion. The CT scan appearance, especially with reconstruction in different planes, which is available on the most advanced scanners, usually makes the distinction. If not, neurosurgical exploration could be necessary.

**Treatment**

Historically several phases have been recognized in the management of otogenic brain abscesses (Brand, Caparosa and Lubic, 1984). Until this century, most patients died when all that was available was craniotomy. During the next phase, in the early part of the century, intracranial pus was evacuated at the time of mastoidectomy, by opening the dura and needling the brain along the tract established by the inward extension of the infection. There must now rarely, if ever, be a place for that form of management. Today, the treatment of the abscess involves the use of large doses of systemic antibiotics, combined with a surgical approach through a clean field. Treatment of the ear disease must take second place, in accordance with the principles discussed previously. Ideally the abscess should be completely controlled neurosurgically and with antibiotics, and predisposing chronic ear disease dealt with by radical mastoidectomy 10-14 days later. As is the case with most of the other intracranial complications discussed, acute ear infections may well resolve with the antibiotics used for the abscess treatment, and only rarely will myringotomy or cortical mastoidectomy be needed.

The first step must be urgent consultation with neurosurgical colleagues. If raised intracranial pressure has caused coma or rapid deterioration, it may be lowered temporarily by the administration of dexamethasone, 4 mg intravenously every 6 hours, or intravenous 20% mannitol, in a dose of 0.5 g/kg. This may be a life-saving measure until neurosurgical help can be obtained. Corticosteroids may also be needed after surgical treatment of an abscess if cerebral oedema persists.

Antibiotics must be started in large doses and administered intravenously. For reasons already discussed, the initial choice should include chloramphenicol, which is effective against *Haemophilus* and many enterococci, and which passes readily into the cerebrospinal fluid in high doses (Brand, Caparosa and Lubic, 1984). This should be combined with a penicillin, often chosen from those active against beta-lactamase-producing organisms. Both chloramphenicol and penicillin penetrate the capsule in effective concentrations. If *Pseudomonas* or *Proteus* species are suspected, because of the primary source in a chronically infected mastoid, an aminoglycoside may also be added. Metronidazole should also be given from the start, in view of the strong likelihood of *Bacteroides* infections (Editorial, 1977). This drug also penetrates the capsule and high concentrations have been found in the pus within abscesses after administration in doses of 400-600 mg 8 hourly (Ingham, Selkon and Roxby, 1977). Antibiotics should be administered for at least 3 or 4 weeks, with careful regard to their potential toxic effects.
Surgical

The commonest current neurosurgical options are to drain the abscess repeatedly through burr holes, or to excise it completely with its capsule. Opinion is divided on the choice of technique (Maurice-Williams, 1983). Repeated needle aspiration involves a smaller operation than excision and, since it can be performed under local anaesthetic, it is safer for very ill patients. There are, however, several disadvantages. It may do little to reduce the mass effects of the abscess, especially if the pus is thick. Forty per cent of abscesses are multilocular (Stephanor, 1978), and total removal of pus is sometimes impossible by aspiration. At best, aspiration involves repeated procedures, and as the capsule collapses, it thickens so that there is a risk of the cannula glancing off and damaging adjacent white matter. If the abscess does not collapse, excision may eventually be necessary after all. Furthermore, there is a risk of late recurrence, which may be as high as 8%.

Aspiration offers access to the pus for bacteriological examination. In order to identify obligatory anaerobes, such as Bacteroides fragilis, which has increasingly been shown to cause apparently 'sterile' abscesses, cultures must be set up rapidly (within an hour) in a low oxygen tension medium, and preferably one containing nalidixic acid to inhibit the growth of Gram-negative organisms. With each aspiration antibiotics can be instilled into the cavity of the abscess. Before the advent of CT scanning it was also usual to put 2 mL of thorotrast into the abscess on the first aspiration. This radiopaque agent is taken up by the fibrous wall of the abscess capsule, and allows subsequent changes in the lesion to be watched by straight X-ray examinations.

Primary excision is favoured by some neurosurgeons as the treatment of choice. It offers a means to decompress the brain immediately. Its main disadvantages is that the operation can cause extensive damage to cerebral tissue, especially if the abscess is multiloculated with tentacular extensions. Excision is then possibly associated with a higher incidence of residual neurological deficit. It is a more major operation than repeated aspiration, and one demanding a higher level of neurosurgical skill to minimize brain damage, and to avoid rupture into the ventricle.

Recently, open operation to remove pus from the abscess cavity under direct inspection has been advocated (Maurice-Williams, 1983). The abscess is incised widely, and all the pus removed from it and its daughter loculi. This removes the mass effect and the sepsis at one operation, as does excision, but with much less risk of further neurological damage.

Although the otologist will be interested in the possible techniques available, the choice of the procedure is a neurosurgical decision, which will be determined by the facilities and preferences of the neurosurgical colleague cooperating in the patient's care.

After successful treatment of a temporal lobe abscess, there is a high risk of epileptic seizures. If followed-up for long enough, 70% of patients will have a fit, so anticonvulsant medication is needed, and should be continued indefinitely.
Otitic hydrocephalus (benign intracranial hypertension)

This is one of the least common complications of middle ear infection. First described by Symonds (1931), this is a misnomer; it is a syndrome of raised intracranial pressure during or following middle ear infection. The most frequent victims are children and adolescents. In a review of 60 patients with benign intracranial hypertension, Foley (1955) uncovered a history or preceding acute otitis media in 13. ‘Pseudotumor cerebri’ occasionally appears in the literature as an unhelpful synonym for the condition.

Pathogenesis

The aetiology is unknown, but most accounts recognize a relationship with lateral sinus venous thrombosis. The inference is either that obstruction of the lateral sinus affects cerebral venous outflow, or that extension of thrombus to the superior sagittal sinus, impedes cerebrospinal fluid resorption by Pacchionian bodies (Pfaltz and Griesemer, 1984). However, it has been argued that superior sagittal sinus thrombosis should be associated with more neurological deficits than are found in otitic hydrocephalus, and with ventricular dilatation, which is lacking in this syndrome. Lateral sinus thrombosis usually occurs without subsequent hydrocephalus. Seid and Sellars (1973) found only one case among 13 with sinus thrombosis and, indeed, ligation of the internal jugular vein in the neck does not cause hydrocephalus; so that mechanism must also be suspect. Gower and McGuirt (1983) considered that otitic hydrocephalus should be accepted as an idiopathic benign intracranial hypertension associated with ear disease, and argued that raised intracranial pressure following lateral sinus thrombosis is a different entity. Any attempt to make this distinction is probably not helpful since most writers accept that the syndrome to which the name of otitic hydrocephalus is properly attached most often follows lateral sinus thrombosis. Thus, Foley (1955), with a large series of 44 cases, described lateral sinus disease in 27 out of 34 cases surgically explored, and Wright and Grimaldi (1973) described three cases all with lateral sinus thrombosis. Lenz and McDonald (1984) reviewed the literature to disclose 10 patients of whom only one, with bilateral ear disease, had normal sinuses. Some of the problems in explaining pathophysiology may depend on variations in venous arrangement in the skull. Clemis and Jerva (1976) described the venous patterns, showing that there is a right predominance in 35% of subjects and a left predominance in 13%; 24% showed disproportion, while there is poor cross circulation at the confluence of the sinuses in just over 10%, and an absent sinus in 4%. Foley’s (1955) series indicated more cases with right-sided ear disease, and a higher incidence of sinus thrombosis in the right-sided than in the left-sided cases. These lateralizing relationships were also evident in Lenz and McDonald's (1984) rather smaller review.

Clinical features

The leading symptoms are headache, drowsiness, blurred vision, nausea and vomiting, and sometimes diplopia. The onset may be many weeks after an acute otitis media, or many years after the start of chronic middle ear disease. Clinical examination reveals papilloedema and drowsiness. Lateral rectus palsy due to sixth nerve stretching (a false localizing sign in raised intracranial pressure) may be found on one or both sides. These signs are associated with evidence of acute or chronic middle ear infection, or with a history of a recent acute middle ear infection since recovered.
The differential diagnosis includes any other cause for raised intracranial pressure, and in particular a brain abscess. Investigations to exclude that possibility should include CT scanning. The scan will show normal ventricles.

**Treatment**

The ear disease, if acute, may have recovered. Any persisting middle ear infection has to be treated on its own merits. Management of the complication requires measures to reduce the raised intracranial pressure, in order to prevent visual impairment by papilloedema. Treatment includes the use of steroids, diuretics, and hyperosmolar dehydrating agents. Repeated lumbar puncture has been advocated, but this is not free from risk in the presence of raised intracranial pressure. Long-term thecoperitoneal shunting may occasionally be needed.

**Prognosis**

The outlook for survival is good, but treatment may be needed over many weeks or even months. Permanent deficits such as visual impairment are not common (Pennybacker, 1961), but recurrences have been reported, albeit rarely (Johnston and Paterson, 1974).

**Facial paralysis**

The management of the paralysed facial nerve is discussed fully in Chapter 24. Here, mention will be made of aspects of facial palsy as a complication of middle ear infection.

**Acute otitis media**

Facial palsy occurs in acute otitis media only in that small proportion of patients (less than 10%) with a congenital dehiscence of the thin bony wall normally separating the horizontal part of the facial nerve canal from the middle ear mucosa. Infection of the mucosa may cause an inflammatory reaction in the subjacent epineurium and perineural spaces. The diagnosis is usually straightforward, but the Ramsey Hunt syndrome (see Chapter 24) can cause confusion, since the pain and facial palsy of that condition is associated with blistering of the surface of the tympanic membrane, which may be mistaken for evidence of acute otitis media.

**Treatment**

The affection of the nerve is invariably a neuropraxia, and full recovery of facial muscle function is to be expected after cure of the preceding infection. Although this can usually be achieved by appropriate systemic antibiotic treatment, occasionally myringotomy or, more rarely, cortical mastoidectomy may be needed. Operative decompression of the facial nerve is unnecessary (Alford and Cohn, 1980).

**Chronic otitis media**

In chronic destructive middle ear disease, the facial nerve trunk may be exposed if its bony covering is eroded by cholesteatoma, with a subsequent inflammatory reaction to the
expanding surface of the cholesteatoma sac. Pressure of that sac may also be a factor since uninfected congenital cholesteatoma of the petrous apex invariably present with a slowly progressive facial paralysis.

**Diagnosis**

Facial paralysis of slow onset, which is insidiously progressive, should arouse suspicion of erosive disease in the temporal bone. The association with aural discharge will point attention to chronic middle ear infection, although a similar clinical pattern may be caused by neoplasms, such as carcinoma of the middle ear. In that dread disease, pain is usually a feature, and this is not a symptom of otherwise uncomplicated cholesteatoma. The absence of discharge does not exclude the possibility of cholesteatoma; meticulous examination of the tympanic membrane, preferably under the binocular operating microscope, is essential in all patients with a lower motor neuron facial palsy. If there is no suspicion of attic or posterior marginal disease, radiological examination with high resolution CT scanning may provide an explanation for the progressive facial nerve lesion. On occasion it may not be possible to examine the tympanic membrane fully without general anaesthesia. If any doubt about the state of the middle ear remains, surgical exposure of the nerve in the middle ear and mastoid region must be advised.

**Treatment**

Urgent operative exploration of the middle ear, to treat the chronic middle ear disease is needed, proceeding if necessary to a radical mastoidectomy. The facial nerve should be exposed carefully throughout its horizontal course in the middle ear and in its vertical mastoid segment, by following cholesteatoma, granulation tissue and osteitic bone. Cholesteatoma matrix may be gently removed from the surface of the 'soft' nerve, but any attached granulation tissue should be left untouched to avoid further neural injury. Healthy bony should be removed from the nerve on either side of the diseased segment, to allow space for oedema of the nerve without further compression. Naturally any packing in the cavity at the end of the operation must be inserted gently and carefully to avoid pressure on the nerve. The management of the problems caused by the facial paralysis are discussed in Chapter 24. Good recovery can be expected if no axonal degeneration had occurred before treatment.

**Labyrinthine complications**

**Pathogenesis**

**Acute middle ear suppuration**

Acute middle ear suppuration may extend to the labyrinth through the round window. The round window membrane is thinner in acute than in chronic otitis media, and its permeability may be increased. Pus cells may pass into the scala tympani perilymph by diapedesis from adjacent inflamed labyrinthine blood vessels. A fibrillary precipitate then accumulates in both perilymphatic and endolymphatic spaces, and developing endolymphatic hydrops is followed by destruction of the membranous labyrinth. Preformed fistulae into the labyrinth from the middle ear, as for example, after a stapedectomy operation offer another route for infective spread. The process may stop at any stage and, if the inflammatory changes
induced in the labyrinth by the transgression are reversible, the clinical condition is called serous labyrinthitis. Should the intralabyrinthine suppuration destroy cochlear and vestibular function in the affected ear, the complication is labelled suppurative labyrinthitis.

**Chronic destructive ear disease**

Chronic destructive ear disease can erode the bony labyrinth by cholesteatoma or osteitis, leading to similar inner ear destruction, but fully developed intralabyrinthine inflammation is preceded by thinning of the bony labyrinthine wall to produce a fistula of the labyrinth. Labyrinthine damage from slowly eroding cholesteatoma may be followed by new bone deposition. This allows destruction of one part of the labyrinth with preservation of the rest. Furthermore, bony fistulae are often closed by new bone deposition after the eroding disease has been eliminated.

Vestibular irritation caused by inflammatory disease very near to the endosteum of the bony labyrinthine lumen is sometimes termed 'paralabyrinthitis'. Very rarely, chronic osteitis around the bony labyrinth may cause necrosis of the whole otic capsule, a condition described as sequestration of the labyrinth. The term perilabyrinthitis is also encountered in writings on this topic, and its correct usage will be discussed later.

Suppurative labyrinthitis is now a rare complication of acute otitis media, because of the use of antibiotics, but the development of labyrinthine fistula remains as common, at 10% of all cases of chronic otitis media, as it was in the pre-antibiotic days (McCabe, 1984). Because of this high incidence, labyrinthine fistula should perhaps be considered the most important of the labyrinthine complications.

**Suppurative labyrinthitis and serous labyrinthitis**

As has been indicated, the distinction between the two depends on the retrospective recognition of recovery of cochlear and vestibular function; so the term serous labyrinthitis has little clinical value. The method of spread of infection has been described, but should be amplified by the observation that, on rare occasions, infection can extend from meningitis to the labyrinth through the internal auditory meatus, or through the cochlear or vestibular aqueducts. Even more rarely, the infection may be blood borne.

**Clinical features**

The patient suffering from acute or chronic middle ear infection presents with violent prostrating vertigo and vomiting. Severe hearing loss of a sensorineural type is to be expected, but will be adumbrated as a complaint by the severe disabling vertigo, especially if there has been a preceding conductive impairment from the acute or chronic middle ear disease. The patient lies still, avoiding any head movement, on the side with the infected labyrinth upwards. Examination demonstrates evidence of the preceding ear disease. The complication itself causes little systemic infective disturbance. Pyrexia or leucocytosis appear only as features of accompanying acute suppurative otitis media. At first there may be a spontaneous 'irritative' jerk nystagmus beating towards the infected ear; but this is soon replaced by a 'paralytic' jerk nystagmus, beating towards the healthy side. The direction of this nystagmus probably dictates the preference for lying on the unaffected ear; in that position, the patient's
efforts to look at a bedside visitor involve turning the eyes towards the damaged labyrinth, and in this direction gaze, the violence of the nystagmus is least. In the earliest, irritative, phase, when subsequent progress may happily confirm that serous labyrinthitis was the appropriate label, tests of cochlear function by masked bone conduction should indicate retained hearing. Loss of cochlear function as the condition is watched indicates transition to the irreversible suppurative state. The paralytic jerk nystagmus is initially third degree in its severity. Provided no additional problems develop, recovery takes place by the mechanisms common to recovery from any cause of vestibular failure, with gradual subsidence of the nystagmus through a second to a first degree state, and then to absence of spontaneous nystagmus with fixation. At this stage, after perhaps 2-3 weeks, the recovering patient will have gained fairly good balance, but will still be unsteady when trying to walk in the dark, or with the eyes closed, and will still be unhappy to make sudden head movements. Since this improvement in equilibrium depends on central compensatory changes, it may be upset later in life, long after the original infection, by other general illnesses, impaired central nervous system function, drugs or psychiatric illness. The hearing loss in the damaged ear will be total and permanent.

**Diagnosis**

The clinical pattern described above is that of sudden vestibular failure from any cause, and suspicion of suppurative labyrinthitis primarily rests with precise recognition of the underlying middle ear infection. Examination of the ears with the binocular microscope is necessary, and occasionally examination under general anaesthesia will be needed to inspect the attic fully, and to remove obscuring crusts, secretions or debris. Haemorrhagic, or bullous, myringitis can produce sudden vestibular failure with acute pain and inflammatory changes on otoscopic examination. Mastoid X-rays confirming a clear air cell system would help to support that diagnosis, but if in doubt the safest course must be to treat the illness as suppurative labyrinthitis. Once a middle ear infection has been recognized the diagnosis poses little difficulty, although traditionally a cerebellar abscess has always been considered to offer a source of confusion. In the latter complication, the patient may appear far more ill, show cerebellar signs on neurological examination, and demonstrate nystagmus persisting longer after the time of the severest vertigo. Nowadays, a CT scan should provide a definitive distinction.

**Sequelae**

During the course of the acute illness there is a continuing danger of intracranial spread of infection with the development of meningitis. In the long term, the labyrinth may remain filled with sequestered pus, and traditional teaching suggested the occasional need to drain such a labyrinth surgically. The long-term effects on balance and hearing have been mentioned above.

**Management**

As with all complications, separate attention must be given to the management of the complication itself, and to the antecedent ear disease.
Treatment of suppurative labyrinthitis requires complete bed-rest. Head movements should be avoided as much as possible. Any hearing tests must be carried out at the bedside, and not in a chair in the audiometric department. Tests of vestibular function, which inevitably excite endolymphatic movement must be eschewed. (In any case they add nothing useful to the clinical examination.) Vertigo and vomiting may be controlled by parenteral prochlorperazine or cinnarizine. If vomiting prevents hydration, intravenous fluids must be infused. It is usual to advise the administration of parenteral antibiotics, and certainly these will be needed to treat acute otitis media as the cause, but it is doubtful whether penetration into the labyrinth itself can affect the course of the labyrinthitis. The development of meningitis, however, may possibly be prevented by antibiotics. The choice of antibiotic may be decided by bacteriological examination of any available secretions. Broad-spectrum drugs such as ampicillin should be used, and if there is any anxiety about infection with *Haemophilus influenzae* type B, then intravenous chloramphenicol should be chosen. Immobility must be secured throughout treatment, and observations are planned to recognize the earliest signs of meningitis.

Treatment of an acute ear infection may demand myringotomy, and more rarely cortical mastoidectomy but, in most instances, the otitis media will recover with antibiotic therapy alone. In chronic middle ear infections, formal exploration of the mastoid will be needed to make the ear safe. Premature surgical trauma to temporal bone can promote dissemination of infection, and so mastoid exploration should be deferred until the acute symptoms of the suppurative labyrinthitis have subsided. This policy involves conservative medical treatment with continuing observation for 7-10 days before mastoid exploration is performed. Nowadays, it is not considered necessary, or indeed advisable, to drain a 'dead' labyrinth during that mastoidectomy operation.

After full recovery from the acute infective illness, vestibular head exercises (Cawthorne-Cooksey, see Appendix) may accelerate central compensation for the vestibular deficit.

**Labyrinthine fistula**

As has been explained, this is a complication of chronic otitis media. An ear, in which the endosteum of the labyrinth has been exposed by bony erosion, continuously threatens the development of suppurative labyrinthitis, and so urgent treatment is essential. The incidence of 10% of all cases of chronic mastoid disease with mastoidectomy has remained unchanged over 20 years (Sheehy, Brackmann and Graham, 1977). Fistulae occur most commonly in the dome of the lateral semicircular canal, but other parts of the bony labyrinth may be eroded, including the promontory.

A labyrinthine fistula may be silent, with no symptoms, and with its discovery at operation unexpected. Suspicion should be attached to any patient with chronic middle ear disease complaining of brief episodes of vertigo or unsteadiness. Even if longer attacks of vertigo are a complaint, and if they seem to fit the pattern of another disorder (such as Ménière's disease), an infected middle ear and possible labyrinthine fistula should remain under suspicion until operative exploration proves otherwise. It is safer to explore an ear with an intact otic capsule than to miss exploration of one with a fistula.
Assessment of the dizzy patient with chronic middle ear disease must include a careful examination of auditory function. The state of hearing in the apparently healthy ear bears heavily on treatment decisions. Although cold air caloric stimulation may be used as an indication of vestibular function, water must never be used for caloric testing in the presence of chronic ear disease and a suspected fistula.

**The fistula sign**

This is an important physical sign, which depends on transmission of air pressure changes from the external ear canal to a fistula in the labyrinth, causing endolymph movement. Raised air pressure may be produced by pressure with a finger on the tragus, but more reliably by the use of a pneumatic otoscope fitted with a speculum large enough to fit securely into the meatus, and produce an air-tight seal. Recognizable and precise effects of pressure changes arise when the fistula test is positive. The nature of these positive findings has often been incorrectly described in textbooks. The sign is not simply one of nystagmus induced by the increased pressure. As McCabe (1984) has fully explained, increased pressure causes conjugate deviation of the eyes away from the examined side. If the pressure is maintained, a jerk nystagmus develops beating towards the examined, and affected ear. As the pressure is released, the eyes return to the midline. Pulsation of pressure in the meatus causes repeated deviation of the eyes to the unaffected side with each pressure rise, and return to the primary position of gaze when the pressure falls. The patient feels dizzy during these events, and accompanying head movements away from the examiner may make continuous inspection of the eyes difficult. McCabe (1984) has also shown that the direction of deviation of the eyes on raised pressure depends on the site of the fistula. The above description of deviation towards the normal ear is the commonest finding, associated with a fistula in the most usual site in the dome of the lateral semicircular canal. A lateral canal fistula anterior to the ampulla causes deviation towards the side of the fistula. An erosion into the vestibule is indicated by rotatory horizontal deviation towards the diseased ear. Raised pressure on a fistula in the superior canal causes rotatory movement towards the normal ear. Finally, vertical deviation of the eyes suggests a fistula into the posterior canal.

It is always important to seek a positive fistula sign in any vertiginous patient, and in any patient with chronic middle ear disease. There are however both false positive and false negative results. The fistula sign may sometimes be positive after a labyrinthine membrane rupture, when there is a perilymph leak into the middle ear. In the presence of an intact tympanic membrane, a positive fistula sign has traditionally been considered to indicate syphilitic otic capsule disease (Hennebert's sign) but this has also been demonstrated in Ménière's disease. A false negative fistula sign may come about from inadequate sealing of the speculum in the meatus, or because a mass of cholesteatomatous debris protects the inner ear from the transmission of the raised pressure, or yet again if the vestibular labyrinth has previously succumbed to the disease and is unresponsive.

**Treatment of labyrinthine fistula**

Whenever chronic middle ear disease is recognized in a vertiginous patient, labyrinthine erosion must be presumed. This is so, no matter how characteristic of another disorder the pattern of vertigo may seem to be, and no matter how slender the evidence for middle ear disease available. The patient's safety demands surgical exploration of the middle
ear. Only if inspection during operation fails to reveal erosion of the labyrinth, should other possible explanations be followed.

During surgical exploration of the middle ear cleft, a labyrinthine fistula should be suspected whenever cholesteatoma is encountered, since asymptomatic fistulæ are not rare. Preoperative demonstration of a positive fistula sign will alert the surgeon to the risk, and its characteristics will indicate where the fistula may be found. Great care is needed while peeling cholesteatoma matrix off the dome of the lateral semicircular canal, and away from areas where fistula is suspected. Matrix should be removed from other sites first, and then dissection carried out slowly under high power magnification. A slight change in colour at the junction of the matrix and subjacent bone suggests a possible fistula. Eventually a small sheet of cholesteatoma matrix over the possible fistula will have been isolated. Its management requires careful consideration, for if the endosteum of the bony labyrinth is breached, a 'dead ear' with total deafness can be expected. Clearly then the state of hearing in the other ear is a factor of great importance. The options are to remove the matrix, or to leave it in place.

If an open cavity operation, such as a radical mastoidectomy, is in hand the matrix can safely be left undisturbed unless there is any suspicion of vascular infected granulation tissue deep to it. That dangerous state is unlikely if the fistula is clearly demarcated as a blush area through the matrix, but McCabe (1984) recommended biopsy and frozen section examination in all instances of doubt. If cholesteatoma and granulation tissue do extend beyond the fistulous opening, then the labyrinth must be explored as far as the limits of invasive disease. A bony fistula left under cholesteatoma matrix will close by new bone growth if the surface inflammatory condition is controlled. Whenever matrix is completely removed from a fistula, closure of the defect with connective tissue material such as temporalis fascia or perichondrium may avert the problems of perilabyrinthitis discussed later. During intact canal wall tympanoplasty, as opposed to open cavity operation, any fistula revealed will remain protected from the exterior, so vertigo caused by perilabyrinthitis will not become a problem. However, any remaining cholesteatoma matrix over a fistula, whether left deliberately or accidentally, presses the need for a second exploration after an interval of 6-12 months. By that time, the abandoned matrix will have formed a small epithelial pearl, which can easily be removed. Many surgeons would contemplate intact canal wall procedures only when the other ear has good hearing and is free from disease, and only when the patient will accept a second operation, and may be relied upon to honour that obligation.

Whatever procedure is undertaken, accurate documentation about the state of hearing and vestibular function before operation, about the findings and events during the procedure, and about any postoperative vertigo that may have been caused by vestibular damage during operation, is of paramount importance. Investigation of subsequent vestibular symptoms after mastoid surgery is very difficult when such records are not available.

**Perilabyrinthitis**

This term should be reserved for the particular condition for which it was appropriated by Cawthorne (1957). It denotes the problems caused by a fistula into the labyrinth after mastoid surgery, in the presence of retained vestibular function. The fistula may have preceded the mastoidectomy, or have been caused by it. The vertigo arises through the Tullio
phenomenon, since the stapes footplate in the affected ear is mobile. Giddiness may be provoked by pressure changes near the fistula, and cold air blown into the ear at windy street corners may cause imbalance. Sometimes the symptoms can be prevented by occluding the external meatus. Operative help consists of exploration, removal of skin from the fistula and protection by a connective tissue graft. Occasionally, deliberate labyrinthine destruction, or vestibular nerve section may be needed to procure relief.

Vertigo after mastoid surgery

Vertigo and imbalance may develop for many reasons after mastoid surgery (Ludman, 1984, 1986). Analysis of an individual problem is greatly helped by access to reliable information about the state of the ear before operation, the findings at operation, and subsequent progress. The causes include:

(1) unrelated vestibular disease
(2) persisting middle ear disease with further bone erosion
(3) perilabyrinthitis (Cawthorne, 1957)
(4) delayed endolymphatic hydrops (Nadol, Weiss and Parker, 1975; Schuknecht, 1978; Ludman, 1986)
(5) breakdown of central compensation, after loss of labyrinthine function (Ludman, 1984)
(6) cerebellar abscess
(7) vestibular nerve neuroma activity, after labyrinthectomy (Ludman, 1971, 1986).

Cochlear complications

When discussing labyrinthine complications of middle ear infections emphasis has usually been placed on vestibular symptoms. It is probable, however, as Paparella et al (1973) have explained, that sensorineural hearing loss can follow middle ear infection, without overt balance disturbance. The transmission of toxic substances through the dependent round window membrane, into the basal turn of the cochlea has been well documented. Serous labyrinthitis induced in this way may be confined to that region, causing first a temporary, and later a permanent, high frequency threshold shift. High frequency sensorineural hearing losses have been shown in chronic otitis media (for example Walby, Barrera and Schuknecht, 1983). The risk to hearing may be greater in acute than in chronic infection, because the round window membrane is demonstrably thicker in the latter condition, and pus may accumulate under pressure when the tympanic membrane is intact.
Appendix 12.1. The Cawthorne/Cookseye regime of head exercises
(From Dix (1984), with permission.)

The Cawthorne/Cookseye system of exercises is designed to restore balance and to train the eyes and muscles and joint sense by performing many exercises with the eyes closed. The movements are carried out in the following graduated stages:

**Stage 1: head kept still - in bed or sitting**

Eye movements only are practised looking up and down and from side to side and then focussing. The patient focusses on the instructor's finger held three feet away and follows the finger to one foot from the eyes.

**Stage 2: head and eye movements while sitting**

Head movements bending forwards and backwards and then from side to side are at first slow, then quick. The movements are then repeated with the eyes closed.

**Stage 3: head and body movements while still sitting**

Movements of shoulder shrugging and circling are first practised. The patient then picks up an object from the ground and looks right up with it. Bending forwards, he then passes an object (such as a ball) from hand to hand under the knees. It is important that he should relax between the various movements.

**Stage 4: standing exercises**

The following manoeuvres are carried out in turn:

1. The patient gets up and stands without support first with the eyes open and later closed;
2. the above exercise is repeated turning round while standing;
3. a large ball is thrown from hand to hand while standing.

**Stage 5: moving about**

1. Walking across the room and around a chair with the eyes open. The exercise is repeated with the eyes closed;
2. circling around a centre person who throws a large ball and to whom it will be returned;
3. standing back to back with an instructor who passes a large ball to the patient between the legs, receiving the ball back from him above the head. This manoeuvre is performed as quickly as possible;
4. walking up and down a slope with the eyes open and later closed;
5. walking up and down steps with the eyes open and later closed;
6. games involving stooping, stretching, and aiming, such as skittles, bowls or basketball.
The principles of the Cawthorne/Cookseye exercises
instructions for patients

The balance parts of the two ears complement each other, sending equal impulses to
the brain which are essential for the maintenance of equilibrium of the head and body.

If either or both balance centres are damaged, equilibrium is upset. The result of this
is vertigo or giddiness which may be accompanied by nausea and vomiting. Although this
condition may be very frightening it is not serious in that it does not, in itself, threaten life.
It can, furthermore, be overcome by carrying out special exercises.

The purpose of the exercises is to build up a tolerance mechanism in the brain which
compensates for the unequal balance of the two ears. The exercises stimulate the development
of this tolerance mechanism and the more diligently and regularly they are performed, the
sooner will vertigo disappear.

The exercises should be carried out persistently for at least 5 minutes three times daily
and for as long as vertigo persists. This may be for 1-3 months. A conscious effort should be
made to seek out the head positions and movements that cause vertigo insofar as one can be
tolerated, because the more frequently vertigo is induced the more quickly is the brain
compensation mechanism built up.

Certain medications help to control the vertigo while brain compensation is being
achieved and any such tablets should be taken regularly during the course of exercises.

As normal a life as possible is, meanwhile, to be recommended. Early return to work
and sports are helpful in rehabilitation.

Diligence and perseverance will be required but the earlier and more regularly the
balance exercise regime is carried out the faster and more complete will be recovery to
normal activity.