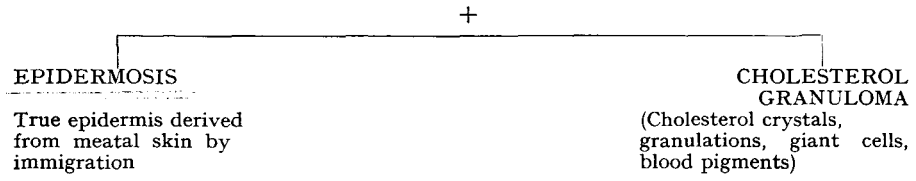
ATTIC CHOLESTEATOSIS

By A. TUMARKIN (Liverpool) *

I AM very honoured to be taking part in this symposium and I feel it is a particular privilege to follow Professor Rüedi who has taught us all so much about the pathology of the mastoid bone. I have been an earnest student of his work ever since 1937 when he published his famous monograph on the development of the middle-ear spaces (Rüedi, 1937).

I have to admit that my own views on the nature of cholesteatosis have altered very considerably during the past few years, I have been greatly influenced by Professor Rüedi and even more so by Dr. Friedmann (Friedmann, 1955, 1957), who has converted me to the immigration theory (Fig. 1). Unfortunately the problem is complicated by the fact that the term cholesteatosis, as used to-day, covers two entirely different entities.

FIG. 1.
Cholesteatosis includes two different entities



In the first the attic is filled with epidermis derived from the skin of the meatus by a process of immigration. For clarity I shall refer to this as epidermosis. Secondly there is cholesterol granuloma characterized by cholesterol crystals, granulations, giant cells and blood pigments. I have been studying this condition for the past 10 years. Fig. 2, is the slide I showed on this screen eight years ago. At that time I said "This picture differs markedly from cholesteatosis" (Tumarkin, 1950). Unfortunately many writers, being impressed by the cholesterin crystals do regard it as a variety of cholesteatosis, and this leads to ambiguity. To avoid confusion, therefore, I shall avoid the word "cholesteatosis" wherever possible. I shall speak instead of "epidermosis" or of "cholesterol granuloma" as the case may be (Fig. 3). Any theory to be complete must explain them both; it must explain why epidermosis is so insidious and why it is usually, although not invariably, associated with a hypocellular mastoid.

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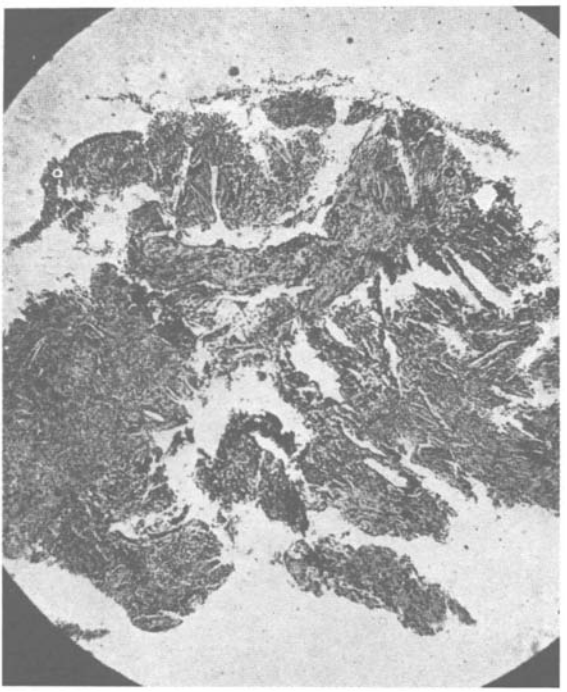


FIG. 2.

Micro-photograph of granulations removed from a case of chronic enclosed mastoiditis. The disease had been present for over a year and yet the drum was intact. Diagnosis was based on the lack-lustre gun metal appearance of the drum plus typical clouding of a pneumatized mastoid.

FIG. 3.
A Complete theory of Cholesteatosis must explain.

(1) Epidermosis
(2) Cholesterol granuloma
(3) Insidious onset
(4) Hypocellularity
(5) Marginal perforation
(6) Drum retraction

It must explain why the perforation is characteristically marginal and why the drum is so frequently retracted.

Finally it must take note of the fact that dry retraction of the drum head—including, of course, dry perforation of Shrapnell's membrane—is by far the commonest pathological condition encountered within the middle ear. For example, in unilateral epidermosis we usually find that the opposite ear shows a retracted drum.

I suggest that dry collapse is the essential primary pathology and epidermosis only occurs when some extra secondary factor is superadded.

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Our first task, therefore, is to explain "dry collapse" and then to consider what converts this into active epidermosis.

I take my stand on the Bezold Habermann theory which postulates obstruction in the eustachian tube with secondary intratympanic vacuum, a condition which is extensively common in children, especially slum children who suffer from nasopharyngitis and adenoid hyperplasia. When the eustachian tube is chronically obstructed the drum tends to cave in,

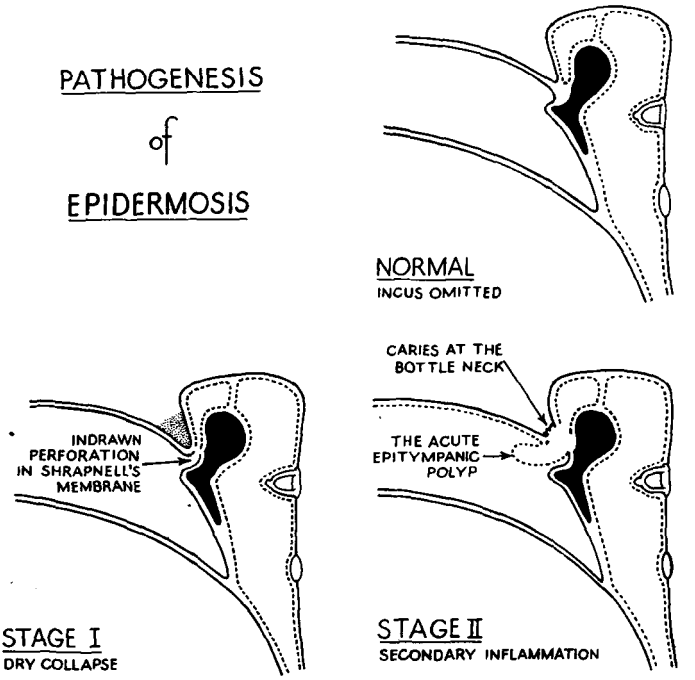


FIG. 4.

either at the pars tensa or at the pars flaccida. At the same time the process of pneumatization falters and may entirely cease.

The idea that a vacuum—by its simple mechanical effect could inhibit pneumatization is resisted by many authorities. I myself find it entirely plausible and recently Ojala (1957) has proved conclusively that pneumatization in the humerus of a growing chicken can be inhibited by simply blocking the air channel to the developing air cell system. He concluded that exactly the same thing can occur in the mastoid of a growing child.

In many cases nothing more happens and the individual may go through life unconscious that his mastoid is hypocellular, his drum indrawn and his hearing slightly impaired (Fig. 4).

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He is, however, vulnerable—especially if the collapse has gone on to actual perforation of the drum head. Suppose he succumbs to infection at a later date. Inflammatory exudate forms within the middle-ear cleft and leaks away through the preformed perforation. Thus there is no distension of the drum, no otalgia. Instead there is the typical insidious otorrhea followed by the sprouting of granulations from the carious bone surrounding the bottle neck of the perforation. But the bottle neck is already lined by the epidermis which has been sucked in by the preceding vacuum. Inevitably that epidermis responds in accordance with its nature—it tries to grow over and protect raw granulations—it enters the attic and epidermosis begins. Thus the pathogenesis of epidermosis requires a preliminary stage of dry collapse and later an inflammatory process to set it alight. Some authorities maintain that acute inflammation can produce epidermosis de novo—that is without any such preliminary dry collapse. Dr. Friedmann has produced typical epidermosis in the auditory bullae of healthy guinea-pigs by the artificial introduction of pyogenic organisms and Professor Rüedi believes that a violent necrotizing tympano mastoiditis can destroy the membrane right down to the annulus and so permit meatal skin to invade the middle ear. I think this must be uncommon. Acute suppurative otitis media tends to burst the pars tensa at the centre. Epidermosis cannot occur unless the resulting central perforation spreads right out to the annulus. Such a disastrous spread is unlikely in this era of antibiotics, and any epidermosis which did occur would bear little resemblance to the familiar attic epidermosis which we are now discussing.

Apart from acute suppuration, however, there are other exudative reactions that can contribute to the development of cholesteatosis. For example in response to intratympanic vacuum the mucous membrane must become engorged and in some cases petechial hæmorrhages will occur. Small granulations will spring up and the whole cleft may become filled with sanious exudate. This possibly explains why blood pigments figure so prominently in cholesterol granuloma. It may explain the curious condition of the blue drum in which the mastoid is more or less filled with an indolent sanious exudate. Fig. 2 was obtained from such a case. The patient had been under observation for over a year during which time there has never been any otorrhœa whatsoever. When we opened the mastoid we found typical cholesterol granuloma. This sort of sterile mechanical process is comparatively rare. More commonly, possibly as a response to virus infection, the middle ear becomes filled with a glue-like exudate. Within recent years we have been studying this condition of exudative otitis media amongst the children of the Liverpool slums and have been appalled at its frequency. Treatment is difficult and protracted, nevertheless it can be extremely rewarding. Unfortunately in some cases the exudate recurs obstinately and finally symptoms of a more violent nature appear. The child begins to suffer with recurring bouts of otalgia

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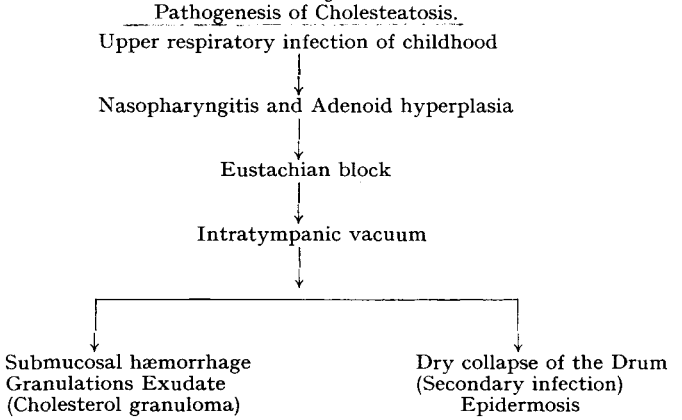
and finally otorrhœa. We have explored the mastoid in some cases and have found all types of pathology from the acute purulent granulations to the characteristic granuloma. We have, therefore, come to regard cholesterol granuloma as merely one aspect of a complex pathological process. We believe, however, that it occurs in its purest form when inflammation is minimal, more especially when a cellular mastoid bleeds under the mechanical effect of a vacuum.

Lastly this vexed question of pneumatization. I believe the hypocellular mastoid is always abnormal. It results either from inhibition of pneumatization, as I have just described, or from condensing osteitis following inflammation. And so it is not surprising that cholesteatosis is commonly associated with a hypocellular mastoid. Nevertheless that association is not invariable. Why should it be? The pneumatic system develops quite rapidly and if a child reaches the age of 5 or 6 before developing eustachian obstruction it will have acquired a comparatively large pneumatic system.

Thus we need not be surprised to find epidermosis within a fully cellular mastoid and, since the delicate trabeculae of the pneumatized bone can offer so little resistance to the spread of disease, it is in these very cases that we find the most widespread invasion of the whole petromastoid structure.

These, in my opinion, are the basic factors responsible for cholesteatosis and they enable us to answer the question as to the relation between

FIG. 5.
Pathogenesis of Cholesteatosis.



epidermosis and cholesterol granuloma. We see that from one point of view they are different insofar as epidermosis arises by immigration of meatal epithelium, following dry collapse, whereas cholesterol granuloma is a variety of exudative mastoiditis (Fig. 5). On the other hand they are closely related since both are produced in the growing child's mastoid by

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the same basic disease. Thus in listing the aural complications of the upper respiratory disease of childhood, although we demarcate many different entities from dry collapse to acute suppuration we should not regard our demarcations as water tight. We are dealing with a continuous spectrum of disease and we must not be surprised to find elements of more than one type present in any given case.

MY POINT IS THAT EVERY ONE OF THESE CONDITIONS IS A RESULT—WHETHER IMMEDIATE OR REMOTE—OF THE DIFFUSE UPPER RESPIRATORY DISEASE OF CHILDHOOD.

I now turn to the question of management.

Thirty years ago this discussion would have been dominated by the intra-cranial complications. That I make no further reference to them is a measure of the great progress that has been made in Otology (Fig. 6).

FIG. 6.
Types of Attic Suppuration.

(6) Advanced disease with intracranial complications
(5) Advanced disease without intracranial complication, but with necrotic ossicles, ulceration of pars tensa as well as pars flaccida
(4) Advanced disease with necrotic ossicles, perforation pars flaccida, but intact pars tensa
(3) Early disease with viable ossicles and intact pars tensa
(2) Combined attic epidermosis and mastoid Cholesterol Granuloma
(1) Acute Epitympanic polyp

Nevertheless we still encounter far too many cases in Group V, i.e. advanced pathology with disorganized ossicles and perforations in the pars tensa and the pars flaccida. In the past we have been happy enough to secure a dry ear in such cases, believing that sacrifice of the ossicular chain inevitably meant a poor functional result. Nowadays, however, hopes have been raised high by the surgical triumphs of our Continental friends. It seems, however, that the astounding successes that can undoubtedly be achieved with other types of pathology are not so easily obtained in epidermosis. The basic difficulty in reconstructing a middle ear invaded by epidermosis is the need to eradicate completely all traces of the invading epidermis before applying a skin graft. Zöllner himself spoke very feelingly on this point at the recent International Congress:

> "Wherever . . . Cholesteatoma . . . can be removed radically . . . primary plastic surgery is promising—but if the hypotympanum must be curetted radically we often encountered recurrences of Cholesteatoma underneath the plastic flap, despite the most careful curettage. We, therefore, avoid any inclusion of the skin within the area of plastic surgery.
>
> "Our procedure has become more and more conservative as the years have gone by, but this consideration must not prevail at the expense of the dependable healing of the disease process. That is why we do not cover a recessus epitympanicus, affected with Cholesteatoma, with a plastic flap . . ." F. Zöllner . . . Int. Cong., 1957.

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Recently Richtner (1957) published a small series of cases dividing them into seven groups. He used mainly Type IV Tympanoplasty of which he speaks very highly. Nevertheless he admits that his worst results were achieved in cholesteatosis, only one case in seven improved.

My own experience of these operations is negligible and I look forward to hearing from others who can speak with authority on this important problem.

Suppose now we are dealing with stage IV in which the pars tensa is intact, but the ossicles are not viable. The incus and head of malleus must be removed and the question arises—shall we carry out Type III Tympanoplasty? Here again the problem of completely eradicating the epidermis confronts us—but in addition the use of a free graft under these circumstances is open to criticism.

This graft serves three purposes:

(1) It replaces defects in the pars tensa.
(2) It epithelializes the mastoid cavity.
(3) It provides a mechanical bridge between the reconstituted pars tensa and stapes.

But in these cases the pars tensa is intact. There can surely be no gain in first denuding the drum of its epithelium and then skin grafting it?

Again, the survival of the free graft in the mastoid cavity is, to say the least, precarious. I believe that the tympanomeatal flap—as in fenestration—has a much better chance of survival, and the same argument applies to the third function of supplying a mechanical bridge. Thus, from every point of view, a pedicle flap should be superior to a free graft, whenever the pars tensa is intact.

The fashioning of such a flap is complicated by the presence of a perforation in Shrapnell's membrane and the tissues are more friable than normal. Nevertheless the flap can be made and in more successful cases the perforation should heal over. This is the sort of tympanoplasty I am trying at present and I am sure that many others are doing likewise.

Next, suppose we are dealing with Stage III in which the pars tensa is intact and the ossicles are viable. In the past I have aimed at exposing the invading epidermis wherever it ramifies. My idea was simply to evacuate all debris and to provide access to the skin lined cavity, so that suitable toilet could prevent the reaccumulation of squamae and ultimately the ear could heal completely. It was with this idea that I proposed the conservative permeatal atticotomy (Tumarkin, 1947). There was nothing new in that idea of exposure and conservation. It has been pursued by many surgeons both before and since my time. Stacke himself attempted something of the sort. Dundas Grant alway advised preservation of the matrix and indeed that idea is implicit in the various conservative radical operations that have been proposed from time to time. Dingley (1955) recently

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claimed excellent results from this sort of conservative surgery and Baron (1949) of San Francisco ardently supports this view. This sort of operation can frequently give highly satisfactory results. Unfortunately the exposed ossicles are vulnerable and demand periodic toilet. I have been saddened to see, on more than one occasion, an apparently brilliant result return years later with a neglected cavity full of stinking debris. I have learnt to choose my cases for this sort of operation, not only on their pathology, but also on their social status and personal hygiene (Fig. 7). I have tried to improve matters by covering the ossicles with a tympano-meatal flap. I call this segregation of the ossicles and I described it to the

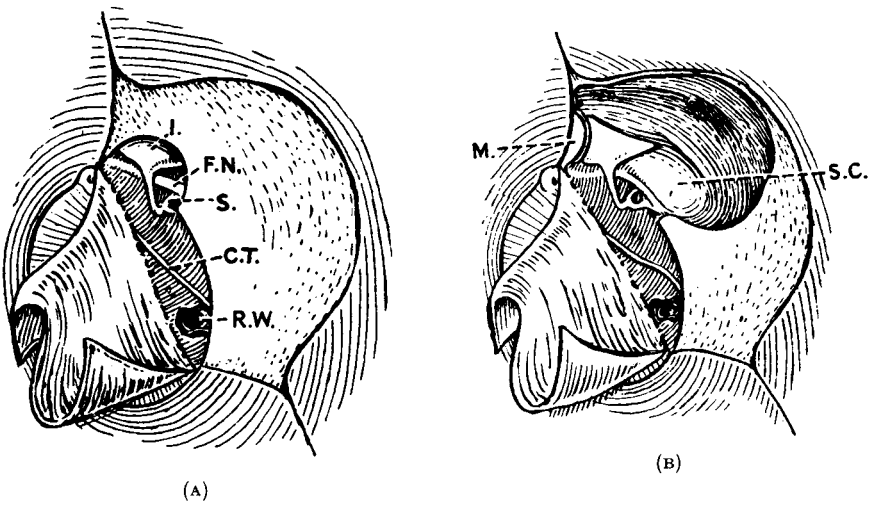


FIG. 7.

(A) The flap has been elevated down to the annulus and turned forward across the drum bringing into view the posterior third of the tympanic cavity. The first semilune has been removed bringing into view the long process of the incus I and the facial nerve F.N. R.W.—Round Window. C.T.—Chorda Tympani. S.—Stapes.

(B) The attico-antrotomy has been completed. The incus is fully exposed lying on the semi-circular canal S.C. The head of the malleus M. is visible. Diseased cells are removed wherever they ramify and finally the flap is turned into place over the ossicles so as to segregate them from the mastoid cavity.

Society here eight years ago. You will observe the similarity to a tympanoplasty Type II except, of course, that a sliding graft is used instead of a free graft. The results of this operation were, on the whole, disappointing. For one thing, at that time I was working by a purely permeatal approach through aural specula. Such an approach does not permit the microscopic survey which is so essential to reconstructive surgery of the ear. I have now abandoned the permeatal route and hope to do better with a wider approach of more orthodox incisions. Even so, however, I am dubious of the value of applying grafts to the ossicles under such circumstances. Not

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only is there risk of burying epidermic islands, but there is a real risk that the whole ossicular mass will become buried and bound down by adhesions. If so we are on the horns of dilemma. If we expose the ossicles they are vulnerable, if we graft them they solidify.

In the face of this dilemma it is natural to enquire whether it might not be possible in some way to retain the outer attic wall, so as to protect the ossicles from the external world. In the past I have always assumed that this was out of the question.

Consider the patient who presents with an acute epitympanic polyp, giving a history of quite recent insidious otorrhœa. I used to approach this sort of case with pessimism. Despite the short history I always felt that there was inevitably so much deep pathology within the mastoid that conservative measures must be futile. And yet everyone knows that such cases can sometimes be cured by quite simple means. Since my conversion to the immigration theory I have realized why this must be so. The mastoid may be hypocellular and it may be inflamed. Nevertheless the major pathology, i.e. the epidermic invasion, may have only just begun at the surface and, if that is the case, it can be halted and repulsed. With this altered view point I have been attacking these cases with renewed vigour. I flood them with antibiotics and then, under local anæsthesia, with adequate magnification, I remove the epithelium from the margins of the perforation, suck out the presenting polyp, and curette the bony margins. Providing the after care is intensive and meticulous this simple procedure can yield most gratifying results and I commend it to your attention.

In all these groups the dominating pathological feature is the epidermosis and I have made no reference to the state of the mastoid itself. In Group II, however, in addition to the attic epidermosis there is cholesterol granuloma in a cellular mastoid. This is particularly liable to occur in children and is one of the most interesting and important groups. Mr. Birrell has made a special survey of these children and, like everyone else, I am looking forward to hearing his account of his work and so I make no further reference to this group.

How then do we manage cholesteatosis?

In Group VI we are desperately fighting for the patient's life with little thought indeed for his hearing.

In Groups V and IV all the resources of modern microsurgical technique must be deployed and, even so, the functional results leave much to be desired.

In Group III satisfactory results can be obtained, but only at the cost of subsequent perpetual vigilance and even when the pathology is minimal, as in Group I the healed ear is still in a precarious condition. Is this the best we can hope for? Surely not—If what I have said is correct cholesteatosis takes its place along with chronic tonsillitis and chronic sinusitis and, indeed, bronchiectasis, as one of the complications of the upper respiratory

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disease of childhood. Surgery—whether it be tonsillectomy, mastoidectomy, pneumonectomy—however flamboyant is a confession of defeat. It can indeed be harmful if it diverts our energies from the humble drudgery of conservative and preventive treatment.

Our work in the Liverpool slums has convinced us that cholesteatosis is preventable. Our aim at present is to cure the underlying diffuse disease if possible without recourse to surgery. This we can often achieve. Nevertheless, there are still many cases in which, despite our best efforts, the disease progresses implacably. It is obvious that we have much more to learn about its pathogenesis. Perhaps most urgent of all is our need to elucidate the role of the viruses. Some day we shall solve these problems. Some day it will be possible to immunize children against this respiratory disease.

When that day dawns I believe that it will mean the end of chronic ear disease and that, in my opinion, is the only satisfactory way to manage cholesteatosis.

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