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We hope that making available the relevant information on Pachyonychia Congenita will be a means of furthering research to find effective therapies and a cure for PC.

hernia is described. It presented in the midline between vagina and rectum and consisted of pelvic colon with omentum and to some extent a sliding hernia of rectum. It is surmised that sepsis had been the chief cause of failure in previous surgery.

Success in the present instance depended on adequate reduction of the hernia, careful ligation of the sac, together with a meticulous fascial repair, and, most important of all, repair of the muscular deficiency.

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NAIL-BED ABLATION

HISTOLOGICAL GROUNDS FOR RADICAL OPERATION

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A COMPLETELY satisfactory method of ablating the nail bed has yet to be described. The results of nail-bed ablation for ingrowing toenails performed in St. Thomas's Hospital, between 1948 and 1956, amply demonstrate the deficiencies of methods now in general use.

In 1950 Zadik described a method by which the nail-forming matrix is removed as far distal as the

circumstances nail formation is not localized to the region of the matrix (*Fig. 60*), as commonly described in anatomical and histological text-books (*Gray's Anatomy*, 1958).

Samman postulates that nail production from sites other than the matrix is pathological—probably inflammatory or traumatic in origin. No confirmation of an inflammatory stimulus could be found in

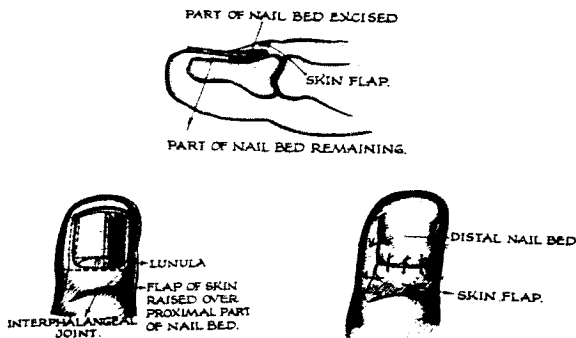


FIG. 59.—Zadik's operation. (By kind permission of the 'Journal of Bone and Joint Surgery'.)

lunula, and primary closure effected by a skin-flap (*Fig. 59*). This method offered several advantages in that it satisfied the anatomical criteria as to removal of all the nail-forming matrix and left a cosmetically acceptable toe, only a short period of hospitalization being required. Zadik claimed excellent results in 16 cases, but no details of diagnoses or periods of follow-up were quoted.

A series of 31 ablations were therefore carried out by this method, carefully following the technique as described. The results of this series were not very successful, and further inquiry showed that recent work (Samman, 1959) indicates that under certain

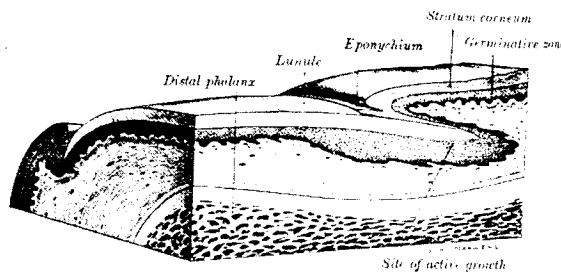


FIG. 60.—A longitudinal section through the root of a nail. (From 'Gray's Anatomy', 32nd ed., by kind permission of Longmans, Green & Co.)

19 sections of nail beds from pathological specimens in this series, and it is suggested that trauma may initiate this process.

Many methods of removing the nail-forming matrix have been in vogue, and Keyes (1934) described four types of operation which have remained the standard methods of ablation.

1. The Standard or Von Bergman operation, in which a wedge resection of the nail and bed about 0.5 cm. wide, including the wall at the affected side, is removed, better known in this country as the Watson-Cheyne operation.

2. Winograd's operation, which involves exposure of the nail-forming matrix by a small incision in the eponychium (*Fig. 60*). Following removal of a

1-in.-1-in. strip of ingrow bed are curetted, leaving intact.

3. Complete excision. Nowadays partial amputation or a skin-graft after



FIG. 61.—Normal nail

combined with this describes a method of

4. Plastic operation of the nail and matrix, consisting of a wedge of normal nail ulcerated by the aff

The St. Thomas's 50 patients operated on by the hospital examined. Of these ablations, who had

Table I.—RESULTS.

	ST.
	0
	20
Nail re-growth	
Failure rate	3
Satisfactory results	

* Indicates failure of recurrence of symptoms.

clinic for several years. In a letter to a questioner, it was stated that 26 were selected for re-operations, and that 26 ablation had been

1. Removal of nail bed: 26 ablations.
2. Removal of nail bed with shortening of toe: 10 ablations.
3. Wedge resection of nail and matrix: 7 ablations.

Personal Series. Operated on by the author: Fifteen only, with

$\frac{1}{4}$ -in.- $\frac{1}{4}$ -in. strip of ingrowing nail, the matrix and nail bed are curetted, leaving the soft tissues otherwise intact.

3. Complete excision of the nail and matrix. Nowadays partial amputation of the distal phalanx or a skin-graft after excision of the matrix may be



FIG. 61.—Normal nail bed. Region of matrix. ($\times 50$.)

in this series as having adequate follow-up. Of these, 11 suffered from ingrowing toenail, 3 from onychogryphosis, and 1 from lichen planus.

As often more than one nail was involved, the numbers of ablations were 14, 4, and 8 respectively. The period of follow-up of the personal series has



FIG. 62.—Nail bed ablated for ingrowing toenail. ($\times 50$.)

combined with this procedure, and Nuttall (1942) describes a method of total closure by flaps.

4. Plastic operations with superficial excision of the nail and matrix, sometimes combined with removal of a wedge of normal tissue below the area of wall ulcerated by the affected nail.

MATERIAL

The St. Thomas's Series.—The notes from 50 patients operated on for ingrowing toenails on the files of the hospital during the period 1948-56 were examined. Of these, 29 patients, with a total of 43 ablations, who had either been attending a follow-up

Table I.—RESULTS WITH STANDARD OPERATIONS—ST. THOMAS'S SERIES

	OPERATION 1 26 Ablations	OPERATION 2 10 Ablations	OPERATION 3 7 Ablations
Nail re-growth	8	3	6*
Failure rate	31 per cent	30 per cent	85 per cent
Satisfactory results	18	7	1

* Indicates failure of operation—re-growth of nail with associated recurrence of symptoms.

clinic for several years or responded satisfactorily by letter to a questionnaire submitted by the author, were selected for review. The following methods of ablation had been used:—

1. Removal of nail and radical ablation of nail bed: 26 ablations.
2. Removal of nail and radical ablation of nail bed with shortening of the terminal phalanx and primary closure: 10 ablations.
3. Wedge resection of the nail, nail fold, granulations, and matrix: 7 ablations.

Personal Series.—Eighteen patients were operated on by the author using the method of Zadik. Fifteen only, with a total of 26 ablations, are included



FIG. 63.—High-power view showing minimal inflammatory reaction. Nail bed ablated for ingrowing toenail. ($\times 320$.)

varied from 3 to 18 months. In addition, 24 nail matrices were examined histologically. These consisted of 5 normal controls, 16 with ingrowing toenails, and 3 with onychogryphosis. The sections were cut at 5μ , stained with hæmatoxylin and eosin, and examined with a light microscope.

RESULTS

The results for the St. Thomas's series are given in Table I. It will be seen that relief of symptoms and no recurrent nail growth occurred with 70 per cent of patients for both Operations 1 and 2. It will also be observed that failure, as indicated by the need of further operation, was greatest with Operation 3 (85 per cent).

The results in the personal series are given in Table II. It will be seen that with Zadik's operation a completely successful result was obtained in only 27 per cent of ablations, some degree of nail re-growth appearing in 73 per cent.

The results of the histological survey show that no significant abnormality could be detected in the region of the nail matrix (Figs. 61, 62). The high-power section from a case of ingrowing toenail (Fig. 63) shows a minimal inflammatory reaction, but

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DISCUSSION

It has always been thought that nail production occurred in the region of the lunula. The nail-forming matrix is stated to lie in the apex of the epithelial invagination near the proximal end of the terminal phalanx and extends to the distal margin of the lunula. In 1927 Pinkus suggested that a small part of the roof of the invagination might also form

Table II.—ABLATIONS BY ZADIK'S METHOD—
PERSONAL SERIES

NUMBER OF ABLATIONS PERFORMED	PERFECT RESULTS	MINIMAL NAIL RE-GROWTH	RE-GROWTH WITH SYMPTOMS	NOT TRACED
Ablations for Ingrowing Toenails: 14 Patients				
19 Big toes only	2	4	8	5
Ablations for Onychogryphosis: 3 Patients				
4 Big toes only	3	1	0	0
Ablations for Lichen planus: 1 Patient				
8	2	3	3	0

Average period of follow-up 9 months; variation of 3-18 months.

part of the matrix, and Lewis (1953, 1954) further suggested that the nail consisted of three parts: the dorsal nail growing from the posterior half of the roof of the nail fold; the intermediate from the region known as the matrix; and the ventral part growing from the distal half or two-thirds of the nail bed immediately below the visible nail, though both dorsal and ventral components may not be present.

Samman (1959) confirmed from studies, both from the histochemistry of this region and its vascular pattern, that Lewis's findings were correct and suggested that nail production from the dorsal and ventral components was probably pathological, following either a traumatic or inflammatory stimulus.

From the studies of nail-bed sections no definite difference could be found between the control and abnormal specimens. No significant inflammatory change was found in any of the sections, and although the tissues examined did not extend much farther than the lunula, it is unlikely that the inflammatory changes suggested by Samman would be limited to the distal two-thirds of the nail bed. However, trauma is known to play an important part in the aetiology of ingrowing toenails (Lloyd-Davies and Brill, 1961). The fact that re-growth of the nail occurred in only 1 out of 4 cases of onychogryphosis but in 12 out of 14 cases of ingrowing toenail suggests that, in the absence of any stimulus to aberrant nail formation, only the intermediate component is active. The work of Lewis and of Samman suggests that this ventral area of nail production may be responsible for the re-growth of nails after ablation by many methods. The experience gained in this study would seem to confirm this, and be a strong argument against the continued use of the Zadik type of operation in which the area of ventral nail production is

not excised. It seems clear (Table III) that those operations involving more radical ablation carry a greater chance of success, although sufficient numbers are not available to allow a valid statistical assessment.

Table III.—RECURRENCE RATES

	I ABLATION OF NAIL BED	II ABLATION OF NAIL BED WITH SHORTENING OF PHALANX	III WATSON- CHEYNE OPERATION	ZADIK'S OPERATION
Recurrence rate	31 per cent	30 per cent	85 per cent	73 per cent
Total ablations	26	10	7	26

Zadik (1961) has modified his original operation so that a greater amount of tissue is removed from the area of the nail folds, and the proximal extensions of the exposure have been discarded. Nevertheless, it is evident that radical ablation of the nail bed is still the operation of choice, particularly for long-standing ingrowing toenails.

CONCLUSIONS

1. Radical methods of nail-bed ablation carry the highest chance of success as the distal part of the nail bed must be removed to prevent re-growth of the 'ventral' nail.

2. The poor results in this series may be due to this factor, and it is doubtful whether sufficient tissue is removed in Zadik's operation.

3. The stimulus to aberrant 'ventral' nail formation is probably traumatic, associated with repeated minor trauma of the ingrowing toenail.

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ART

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