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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.
Reconstruction of a Functional and Esthetic Nail

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The most common cause of nail bed deformity is trauma, but may also be caused by infection, tumor, ischemia, or congenital anomalies. The methods of correction of many deformities are included in articles by Morrison, Shepard, Fleege, Beasley, and the discussion sections.

Not all problems will be discussed here since new problems are encountered daily and other deformities have no method of correction reported in the literature. For some of these we will postulate a method of repair, which from the knowledge of anatomy and physiology of the nail may possibly work, but may not and we will so state.

NONADHERENCE

Nonadherence of the nail after trauma is the most common nail deformity. If the nonadherence is distal it may not cause problems unless the buildup of dirt beneath the nail causes infection or unsightly appearance. The more proximal nonadherence is a problem in that the nail is frequently unstable when trying to pick up small objects and repeatedly tears loose where it is adhered, thus causing pain.

As you read in the first article in this issue, new cells push the previously formed cells upwardly and distally. The nail bed and the nail in all its stages of development are a continuum. If an injury occurs to the nail bed, the continuum is interrupted by the formation of scar. If the wound is closed carefully and it heals with minimum scar, the scar does not usually interfere with the continuing process of nail formation. However, if the scar is wide and the progress of nail cells streaming into the nail is interrupted for a significant distance, they will not restream into the nail distal to the scar. The nail then does not attach to the nail bed at or distal to the scar. Treatment of nonadherence in the past has consisted of removing the scar and attempting to close the nail bed to narrow the scar. Unfortunately, there is more tension on the nail bed sutures with a second closure than there was with the primary. It has been our experience that nail bed closure under tension leaves significantly more scar than closure without tension. The best method of treating a nonadherent nail is resection of the scar and, if there is evidence of any tension on closure, the scar should be replaced with a split-thickness nail bed graft from another area of the same nail or toenail. In Figure 1 an actress who had injured her thumb in a car door several years previously had approximately 50 per cent nonadherence of her nail. She intermittently caught the nail, partially avulsing it with pain, bleeding, and replacement by a new nail. Removal of the nail and exploration of the nail bed revealed a significant area of scarring near the central portion of the nail. This scar was outlined through the operating microscope and a

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Figure 1. A 50-year-old woman after car-door injury to the thumb nail has less than full adherence of the nail to the nail bed and has repeated episodes of tearing it free. B, The nail removed. The area of scarring is seen. C, The area of scarring is marked for excision with the operating microscope. D, A template of rubber glove is made, and a split thickness nail bed graft is taken from the large toe in the same shape. E, A piece of 0.020th fiber reinforced silastic sheeting is placed into the nail fold over the nail bed graft and sutured with a horizontal mattress suture. F, The patient one year later has almost complete adherence and no further symptoms. (Copyright, Southern Illinois University School of Medicine, Division of Plastic Surgery, Springfield, Illinois; with permission.)

template was made with a section of rubber glove. The large toenail was then removed and the template placed on the toenail bed being careful not to reverse the template. A split-thickness nail bed graft of approximately 0.008 inch was removed from the toenail and sutured into the defect where the scar had been excised. A nail-sized section of 0.020 fiber-reinforced silicone sheet was sutured with horizontal mattresses to hold it in the nail fold. At 1 year the patient has almost full adhesion of the thumb nail and has been asymptomatic for 3 years. This technique is especially effective for longitudinal scars where there is no “jumping over” of the scar but a continuous scar the length of the nail. In this situation the nail is nonadherent over the scar and then secondarily splits. The best treatment is excision of the scar and rather than closing the defect under tension to place a longitudinal split-thickness nail graft into the defect.

Other authors have advocated the use of dermal grafts to correct nonadherence, but in our hands it has not been successful.

SPLIT NAIL

The split nail is frequently caused by a longitudinal scar in the germinal matrix where nail formation as a single unit is prevented by the scar interspersed between the two germinal matrix segments. It may also occur after an injury involving the sterile matrix where the scar creates a longitudinal nonadherence with secondary splitting of the nail caused by lack of addition of sterile matrix cells to the nail with resultant weakness of the nail. The increased stress placed on this portion of the nail due to nonadherence to the nail bed causes cracks and splits. Various techniques for treatment of the split nail have been advocated. Carter's, sutured the free edges of the cracked nail together with suture for several months and
reported elimination of the split. Johnson advocated resection of the scar, lateral mobilization of the germinal and sterile matrix from the paronychium, and approximation of the matrix. His paper shows three nails with fair to good results, but we have had little success with this technique. Seckel uses a silicone sheet sutured between the nail and the nail bed after repair of the nail bed beneath the split.

Excision of the scar with primary approximation is not often successful because of the tension required for closure. As mentioned previously we have had little success with the technique employed by Johnson following trauma, but have had success with lateral nail bed mobilization following tumor removal. This is because there is less scarring with tumor to interfere with flap rotation.

If the scar or lesion causing the split is narrow and not in the germinal matrix, or if only the distal portion of the germinal matrix is involved, a split-thickness graft of sterile matrix can be placed into the excised defects with usually good results (Fig. 2). It must be remembered that replacement of germinal matrix with sterile matrix will not be successful, whether it is a full-thickness or a split-thickness graft. Lost germinal matrix

Figure 2. A, A split nail following a 3-mm punch biopsy for the diagnosis of ganglion. B, The defect at the base of the nail measures exactly 3 mm in diameter. C, After removal of the nail, the scar at the distal border of the germinal matrix is seen. D, A split thickness nail bed graft is removed from the dorsum of the nail bed. E, The split thickness nail bed graft is sutured in place with 7-0 chromic sutures. F, The patient's nail 1 year later. (Copyright, Southern Illinois University School of Medicine, Division of Plastic Surgery, Springfield, Illinois; with permission.)
must be replaced by germinal matrix from another finger or toe to achieve any chance of nail growth.

If a germinal matrix graft from the toe is used to replace a defect in the finger germinal matrix, one wonders what happens to the four-to-one growth differential. Although the edge of the nail-generating toenail graft appears to be confluent with the nail produced by the finger germinal matrix (Fig. 3), we do not know whether the germinal matrix of the toe speeds its growth or if that of the fingers slows.

RECONSTRUCTION OF THE EPONYCHIUM

The most common cause of loss of the eponychium is a burn. Many techniques of rotation flaps have been described to correct this deformity. Unfortunately the dorsum of a burned finger is, in most cases, so scarred that the flaps do not rotate well or rarely form a cosmetic eponychium. Shepard, elsewhere in this issue, discusses improvement from lining the undersurface of these rotated flaps with split-thickness nail bed grafts.

Rose used a free composite graft from the helical rim of the ear to recreate the eponychium with good results. Our best success has been with resection of tumors of the eponychium or removal of the scarred eponychium and its replacement with either the dorsal roof of the nail fold from the large or second toe depending on the amount needed. This graft is essentially a composite graft of nail bed and skin (Fig. 4).

![Figure 3. A, A split nail with a pterygium due to injury to the germinal matrix. B, An area of scar is removed from the sterile matrix, and a defect is left in the germinal matrix. The tissue is seen over the eponychium is a full thickness germinal matrix graft from a second toe. C, The germinal matrix graft sutured in place. D, At 3 months, the germinal matrix graft can be seen to be producing nail. E, At 6 months, the sterile matrix graft produced nail is growing distally. (Copyright, Southern Illinois University School of Medicine, Division of Plastic Surgery, Springfield, Illinois; with permission.)](image-url)
CROOKED NAIL

It appears simple to correct a nail that grows at an undesired angle by moving the germinal matrix so that the nail grows in the desired direction. Unfortunately, this is not as simple as it appears and Shepard points out that it is more successful to align the entire nail bed including the sterile matrix. It is frequently necessary to use full-thickness skin grafts or flaps to maintain the sterile matrix in alignment after position change or split-thickness nail bed grafts as advocated by Shepard. If a split-thickness skin graft is used or the wound is allowed to heal secondarily, the nail is again pulled out of alignment with healing and wound contracture.

HOOKED NAIL

A nail is usually hooked longitudinally but may be hooked horizontally. Lateral hooking, if severe and progressive, is referred to as a pinscher nail. Although there are no causes for this horizontal curvature such as a midline ganglion, in the majority of patients the cause is unknown. It is a relatively rapid occurrence in which the patient notes that the nail starts to curve volarly and cut into the paronychium. It may progress to actually pinching off the nail bed as the nail edges approximate. When the pinching becomes sufficient to squeeze the nail bed the patient has discomfort and is seen for care. Various techniques for treatment have been advocated including wedge resection of the midportion of the phalanx to flatten the edges of the nail. We insert strips of dermal graft beneath the paronychium to elevate and flatten the lateral portions of the nail bed (Fig. 5). If all else fails, surgical extirpation of the nail and nail bed may be necessary to achieve relief.

The hooked nail is usually the result of fingertip amputation. They also result from congenital absence of soft tissue of the tip of the finger resulting in tension on the nail bed pulling it over the tip of the finger (Fig. 6). Release of the contracted soft tissue of the tip to allow the nail bed to return to the top of the bone will improve the defect significantly. The shape of the nail follows that of the nail bed; therefore, if the nail bed is curved the nail will follow the curve and may grow into the volar surface soft tissue of the fingertip.

If the skin of the tip of the finger is amputated, healing by secondary intention will draw normal skin inward and pull the nail bed over the tip, resulting in curving of the nail to varying degrees even though the fingertip may look relatively normal. The most severe hooking results when fingertip and bone are amputated. This is because the nail bed needs support of the underlying closely adherent distal phalanx to remain flat. If the amputation is volarly angulated and there is extra nail bed on the dorsum, the nail bed will drop, contract, or be pulled with sutures over the tip of the finger causing hooking.
Figure 5. A, A 47-year-old woman who, over a period of several months, had developed hooking of the radial side of the nail. B, Looking end on the pinching and digging in of the nail can be seen. The patient was bothered by pain. C, An incision was made at the tip and the radial paronychial fold elevated from the periostium with an elevator. D, A strip of dermal graft taken from the volar aspect of the wrist was pulled into place beneath the perionychium with a suture to build up and flatten the perionychium. E, The AP appearance of the nail 6 months later with the patient asymptomatic. F, The distal view of the nail at six months. (Copyright, Southern Illinois University School of Medicine, Division of Plastic Surgery, Springfield, Illinois; with permission.)

Figure 6. A, A 7-year-old girl who was born with congenital absence of pulp creating a hooked nail. B, A lateral view of the same patient. C, A lateral view following buildup of the tip soft tissue with a thenar MP crease flap. D, Volar view of the same patient at 1 year. (Copyright, Southern Illinois University School of Medicine, Division of Plastic Surgery, Springfield, Illinois; with permission.)
The best method of treatment is prevention; avoidance using the nail bed to cover the amputated tip. Replacement of the bony support and flap coverage initially may give good results.

Correction of a hooked nail requires recreation of the defect with subsequent reconstructing normal anatomy of the fingertip to as near normal as possible. This may require release of scar on the fingertip and replacement with full-thickness skin graft, cross-finger flap, V-Y flap, or thenar metacarpophalangeal crease flap to allow the nail bed to return to the dorsum of the distal phalanx (Fig. 7). Another technique is to shorten the nail bed to the length of the bone so that a shorter, straighter nail is present.

Bone grafts may be used to replace amputated portions of the distal phalanx and give support to the nail bed; however, there is only bone approximation at one end and chances of resorption of the graft is high. Soft tissue coverage with a local flap or a cross-finger flap is essential. A proposed solution when there is enough distal phalanx is to place four pins through and use a distraction device to separate the fragments after dividing the bone and using subsequent bone grafts. If there is not enough bone for the pins to be placed through the distal phalanx, two can be placed in the middle phalanx and the distal phalanx pins distal to the joint and site of bone division. After adequate or as much as possible distraction has occurred, a bone graft is placed between the two fragments of bone encouraging a lower incidence of graft resorption because there is approximation to bone at both ends.

A free vascularized composite graft of nail bed tip and bone from a toe may be used and is described by Morrison elsewhere in the issue.

Composite grafts from the toe pad may be used after release of the tip contracture, but it is rare that one graft will provide enough soft tissue volume to solve the tissue deficit and multiple stages must be used.

Unfortunately, there is no simple answer to the hooked nail. Many are improved after any of the before-mentioned surgical procedures, but it is rare that the result is outstanding.

BONY IRREGULARITY

Approximately half of the nail bed injuries are associated with fractures of the distal phalanx. There is a higher incidence of post-healing surface deformities of the nail if there was a distal phalanx fracture present. All patients considered for correction of nail bed deformities should undergo radiographic examination of the distal phalanx. If inclusion cyst exists in bone, it needs to be removed because it may cause or contribute to the deformity. If there is a bony exostosis, it must be removed to flatten the bone and nail bed so that the nail can grow on a straight flat surface, as was aptly described by Ashbell et al.

Malalignment of fractures is a common cause of nonadherence of the nail bed and must be corrected as the first step to correct the nail deformity. It is imperative with associated nail bed injuries and tuft fractures that the fracture be accurately aligned. If the nail is replaced on the nail bed after fracture reduction, it will usually maintain the bone reduction since it was grown to fit. On occa-
ension a displaced fracture needs to be reduced and the nail replaced to hold reduction while a small pin is secured down the distal phalanx. This is most common when the fracture is unstable enough that the nail alone will not maintain reduction.

If bony angulation or offset is present causing nonadherence or other nail deformity, the bone must be osteotomized and the deformity corrected. Also, nonunion of the distal phalanx will frequently cause deformity of the nail bed due to lack of support and abnormal motion at the nail bed. Bone grafting of the distal phalanx is usually necessary to obtain stable healing.

Care must be taken when placing pins longitudinally in the distal phalanx that they are not passed through the length of the sterile matrix or longitudinally between the sterile matrix and the periosteum. This will create a scar elevating the nail bed and can also create a ridge on the nail.

PACHYONYCHIA

Pachyonychia may be congenital or developmental. Its development may be secondary to chronic infection, inflammation, ischemia, or other causes. The nail may be thinned and flattened by grinding. The nail may be removed for culture and treatment with appropriate antifungal agents if fungal infection is suspected. If all else fails, extirpation of the germinall matrix, both areas of the sterile matrix, and the nail may be indicated.\(^2\)

ISCHEMIC DEFORMITIES

The most common ischemic deformity of the nail seen by the hand surgeon is that following use of the upper extremity tourniquet. Careful examination of the nail bed of a patient several weeks after use of an upper extremity tourniquet will reveal a transverse line or groove of varying degrees across the nail. Because the nail averages 3 to 4 months for a single growth, the date of the ischemic episode can be approximated. These lines may also occur after general ischemic events such as severe pneumonia or cardiac failure.

The germinall matrix is very sensitive to oxygen deprivation much like hair follicles. If the germinall matrix is used as a free graft, it may take well but will not produce nail. The amount of ischemia tolerated by germinall matrix cell without loss of growth potential varies between individuals. As a rule, the younger the individual, the more tolerant their germinall cells are to ischemia and the more likely that full-thickness germinall matrix graft will produce a nail.

A patient who has a significant ischemic episode or period of denervation of the fingertip will frequently lose volume in the pad of the finger. This may cause the hypnonychium to be exposed by skin traction which pulls the keratin plug away from the nail with resultant pain and tenderness. The distal nail is removed and a 4 to 5 mm horizontal strip of sterile matrix in the area of the hypnonychium is excised. This strip is replaced with a split-thickness skin graft.

Figure 8. A. A 65-year-old woman post-severe episode of ischemia of the hand with resultant atrophy of the pads and then pain in the hypnonychial areas with use after the sensation returned. B. The distal nail was removed and a split thickness skin graft on the side of the finger applied in its place to prevent adherence of the hypnonychium. C. The hypnonychium does not adhere to the hypnonychium and the pain is relieved. (Copyright, Southern Illinois University School of Medicine, Division of Plastic Surgery, Springfield, Illinois; with permission.)
Figure 9. A, A 5-year-old child who was growing almost no nail following being bitten by a rabbit. B, The lateral view. C, The involved small finger placed next to the second toe to show the comparative width of the nail but the decrease in the length of the toenail. D, Area for excision to create the fold and the defect of the graft is shown. E, The nail wall is raised and then the nail and perionychium taken as a unit. F, The hypoonychium is excised with the nail. Dissection carried down to the periosteum of the tip and then proximally on top of the periosteum. G, The eponychial composite graft is seen with the soft tissue contracted with the nail in its normal width. H, The composite graft ready to place in the nail fold. I, Horizontal mattress sutures holding the composite graft in place and spreading the germinal matrix. J, The new nail growing at one year. Note the sterile matrix is not long enough, and a split thickness graft of sterile matrix should have been placed so that adherence would go further distally. K, Lateral view of the growing nail. L, The donor toe. (Copyright, Southern Illinois University School of Medicine, Division of Plastic Surgery, Springfield, Illinois; with permission.)
from the side of the finger or ulnar surface of the palm. The growing nail does not adhere to the split-thickness skin graft relieving the tension of the hyponychium and relieving their pain and discomfort (Fig. 8).

ABSENCE OF THE NAIL

Absence of the nail may result from trauma or birth defect. Congenital absence frequently has significant parental sense of guilt as there are with many congenital abnormalities. Most absences of the nail involve multiple nails, many times all of either the upper or lower extremity being absent. In this situation, donor sites are limited and the best treatment remaining is excision of a nail-size section of skin from the location of the nail and placement of a split-thickness skin graft in the defect. The lunula and hyponychium may be created with full-thickness skin graft strips that are lighter in color. The surgeon must be specific with the parents and the patient in telling them that a completely normal nail cannot be created.

Absence of the nail may also be secondary to trauma. In this case, a free vascularized toenail graft gives the most reliable nail growth and most normal appearing nail. Unfortunately, it also significantly scars the finger and foot, which may not be acceptable to the patient.

Free composite nonvascularized grafts of germinal matrix may be indicated. The younger the patient, the better the chances of its success. Our success with this have been like Shepard’s in patients where a composite graft of nail fold, sterile matrix, and germinal matrix were all taken as a unit and transplanted into a pocket on the finger (Fig. 9), 4, 10, 11

Buncke’s has described the nail pouch construction where a prosthesis is wrapped in a split-thickness graft and buried beneath the skin on the dorsal tip of the finger. We have used this technique on a few occasions and it worked well initially, but as time passed the folds contracted and soon the dorsum of the finger was flat and smooth without any folds left to hold the glued nail.

REFERENCES


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