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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.
The Vein Hook Successfully Used for Eradication of Steatocystoma Multiplex

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BACKGROUND Steatocystoma multiplex is characterized by the formation of the numerous cutaneous cysts in the exposed area leaving some cosmetic problems for the patients. Only surgical excision has been effective, and its several variations were done with limited success. Because the patients usually have many cysts, excision of cysts was tedious for the doctors and left scars on the patients.

METHOD Five patients agreeing to participate in this experiments were selected. The vein hook used for ambulatory phlebectomy was employed to eradicate the cysts. The skin was incised approximately 2 to 3 mm in length. Then the mosquito forceps removed the cysts by gently squeezing or hooking the inner or outer cyst wall. By completely removing tissue around the cyst, recurrence was able to be prevented.

RESULT It took approximately 1 minute to excise one cyst completely, it left no hypertrophic scars except for transient postinflammatory hyperpigmentation, and it had no recurrences for 14 to 30 months on five patients.

CONCLUSION The use of this instrument is very simple and time-saving, providing excellent success rate with favorable cosmetic results. It can be a good alternative for eradication of the cysts in steatocystoma multiplex.

The authors have indicated no significant interest with commercial supporters.

Steatocystoma multiplex is a disorder clinically characterized by numerous cutaneous cysts with nonkeratinizing walls. The walls are lined by thin epidermis and sebaceous glands within or adjacent to the cyst walls histopathologically. There is no known medical treatment for this disease, and surgical excision is the only effective treatment. Patients usually have so many cysts, however, that it takes a long time for surgeons to excise them all, which can leave hypertrophic scars in spite of high success rate.

Numerous surgical procedures have been tried to overcome these limitations; for example, 18-gauge needle aspiration, 3-mm punch excision, laser therapy, cryotherapy, incision, and artery forceps extraction have not been satisfactory considering time consumed, scar formation, and more importantly, success rate.

We employed a vein hook used for ambulatory phlebectomy for patients with varicose vein of the lower extremities. Its tip is properly bent twice, so after being inserted through the narrow incision and twisted, it can hook the outer or inner surface of the cyst or its capsule (Figure 1). When the cyst is pulled, the hook easily separates the cyst from the surrounding soft tissue. The procedure with the vein hook was anticipated to decrease the size of the incision, reducing the time needed for removing cysts from surrounding soft tissues and for the excision of the cysts. This also diminishes the possibility of scars for good cosmetic results to the patients.

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The practical technique is to mark each cyst with pen and to inject 1:100,000 epinephrine-premixed lidocaine under each cyst. By injecting under the cysts, the cysts then are pushed up just under the epidermis for easy separation from the surrounding tissue in addition to patients' comfort. Next, the skin is incised about 2 to 3 mm using a No. 11 blade at a right angle (Figure 2A), and the cysts are removed by vein hook without puncture of the walls if possible (Figure 2B). In contrast, if the wall is punctured, the cyst should be squeezed to cause the contents to come out first. The hook then inserts to grasp either the inner or the outer wall for the cyst and pulls it out gently. When the cyst is exposed, the baby mosquito forceps grasp the portion of the cyst and pull it out gently (Figure 2C). Other grasps are frequently needed to remove the complete lining of the cysts adhering to the surrounding tissues (Figure 2D). The wounds are cleaned with gauze soaked in alcohol, and then the margins are approximated by steri-strips.

Five typical cases of steatocystoma multiplex are reported, which were treated by the above-mentioned simple surgical technique (Table 1). On these five cases, our method proved successful. They had a mild transient hyperpigmentation for about 2 to 4 weeks, and all resulted in satisfactory cosmetic work. Two were eager to remove the remainder of the cysts (Figures 3A and 3B).

We expect “simple incision or puncture for hooking out the cyst by phlebectomy hook” to be very successful in steatocystoma multiplex patients. It is possible to work by making small incisions,
approximately 2 to 3 mm, to minimize the risk of tissue injury, infection, the resultant hyperpigmentation, scarring, and operation time. Accordingly, this method might be the first line of treatment of the steatocystoma multiplex instead of conventional excisional surgery.

### References


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