



# Pachyonychia Congenita Project

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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.

## Surgical Pearl: Mini-incisions for the extraction of steatocystoma multiplex

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**S**teatocystoma multiplex is an autosomal-dominant disorder that causes multiple smooth papules and nodules on the trunk and proximal extremities and less commonly on other sites, such as the axillae, neck, face, or the scrotum.<sup>1,2</sup> Sebaceous glands are regularly found in the epithelium of the cyst. On histologic examination the cysts show a thin wall containing sterile sebum and cellular debris. Molecular genetic mapping studies have shown that mutations in the gene encoding for keratin 17 (K17) can cause either steatocystoma multiplex, pachyonychia congenita, or a combination of both.<sup>3,4</sup> Eruptive vellus hair cysts are considered to be a variant of steatocystoma multiplex. The lesions vary in size from a few millimeters to several centimeters. The condition usually appears during adolescence and, despite its benign character, many patients understandably ask for removal of the most apparent lesions.<sup>5</sup>

Various surgical and medical therapies have been used for the treatment of steatocystoma multiplex, but some have been proven to be inefficient and others have resulted in unsightly scarring. Previous therapies have included radical excisions,<sup>2,6</sup> carbon dioxide laser therapy,<sup>7</sup> cryotherapy,<sup>8</sup> needle aspiration,<sup>9</sup> or oral retinoids.<sup>10</sup>

From 1997 through 1999 we have treated 5 cases with steatocystoma multiplex. The cysts were opened by mini-incisions of 1 mm with a No. 11 surgical blade. The creamy content was squeezed out; the wall was excochleated with a very small (1-mm) curet and completely removed with a forceps. The incisions were closed with adhesive strips. This tech-



**Fig 1.** Steatocystoma multiplex on the chest of a 30-year-old man.



**Fig 2.** Squeezing of a cyst after mini-incision.

nique resulted in virtually unnoticeable scarring (Figs 1-4) and recurrences were not observed at the treated sites. Recently, Adams, Mutasim, and Nordlund<sup>1</sup> have found that the same technique produces excellent aesthetic results, and it is likely that others have also successfully applied this simple procedure. In patients with Fitzpatrick skin types IV-VI or in patients with a history of hypertrophic scar-

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**Fig 3.** Extraction of the cyst wall.



**Fig 4.** Postoperative result. Scars are virtually unnoticeable.

ring or keloid formation, it may be prudent to start with the treatment of one single lesion to observe wound healing. With this precaution kept in mind, extraction through mini-incisions appears to represent the best currently available modality for the treatment of steatocystoma multiplex.

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