

# Plastic Surgery Considered

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“I had always said that plastic surgery was something I would never have done. Never! But now I really love the results,” said Isobel of Pennsylvania, USA. “However, there was a short time immediately after the surgery, when I would have said that I shouldn’t have had it done.”

Isobel, who has PXE, had plastic surgery done on her neck in December 1996. Though her primary care physician and the plastic surgeon had both tried to prepare her for the immediate after-effects of this major surgical procedure, she was nevertheless aghast. After the 3 1/2-hour procedure, she felt terrible, and the swelling and bruising seemed worse than she expected. Besides, the hospital discharged her after only one night. That initial recovery period was rough. However, by the time she took a vacation trip six weeks later, Isobel was completely recovered and delighted with the outcome of the surgery. For Isobel, authorities approved insurance coverage without undue difficulty.

Plastic surgeons have used certain procedures effectively with individuals who have PXE. In a session at the International Centennial Meeting on Pseudoxanthoma Elasticum in November 1997, workshop presenter Dr. Hubert Weinberg of Mt. Sinai Hospital, Division of Plastic Surgery, discussed

plastic surgery for individuals who have PXE. He explained indications for surgery, pre-operative considerations, and the risks and uncertainties associated with plastic surgery, and touched on the issue of insurance coverage. (*An audiotape of this workshop is available from PXE International.*)

The wrinkles, folds and looseness of the skin that may result from PXE, what Dr. Weinberg called “laxity and skin redundancy”, can be removed from the neck, armpits, thighs, breasts and abdomen by means of plastic surgery. However, while plastic surgery is effective in reducing excess skin, it has not been helpful in changing the “cobblestone” appearance of the skin. When PXE causes the mineralization of elastin in the skin, the skin loses its ability to spring back. The skin sags and stretches. Elasticity cannot be restored to the skin, not even by means of plastic surgery. Therefore, the individual affected by PXE may want to make use of a procedure called “direct excision”. The folds and loose skin can be, essentially, smoothed out by pulling it all in one direction, somewhat as one might do when making a bed. After smoothing out the folds, extra skin remains. In bed-making, the extra sheet is turned down over the blankets. In direct excision, the extra skin is cut away and the incision is closed.

One problem with direct excision is that the incision is not always easy to hide, and it may leave an obvious surgical scar. When done to reduce the loose skin around the armpit, the surgeon can hide the scar under the arm. However, if scarring is obvious, even this scar might be visible if one were to wear a sleeveless shirt. Direct excision is the only option for some procedures. In these cases, individuals should weigh the possibility of scarring and the location of the scar against the expected results.

Dr. Weinberg emphasized that someone who has a history of scarring, especially with obvious or thickened scars, would want to be particularly aware of where the surgical scar would be located, and whether the scar would be preferable to the loose skin. He explained that, in the general population, some people tend to scar readily while others do not. A “good healer” is more likely to have good results from direct excision. The surgeon can assess this for the potential patient. Moreover, a surgeon can plan to locate the scar in a less conspicuous spot.

Procedures other than direct excision are more commonly done for people with PXE. A “face lift”, “neck lift” or tightening of the skin of the breasts or abdomen is more like a “redraping” of the skin, according to Dr. Weinberg. Many times the surgeon tightens underlying muscle and removes fat. He or she makes incisions at strategic and less conspicuous spots. For example, in a face-lift or a combination face lift and neck lift, incisions may be located at the scalp line or behind the ears. For these types of surgery, the term “redraping” may be as clear an analogy of what is done as is possible.

In abdominoplasty, plastic surgery to the abdomen, the surgeon rearranges the skin in several directions, then replaces it in a slightly different position. He or she makes an incision, with the possibility of scarring, around the navel so that it ends up in its original position. For the breasts, during a mastoplasty, skin may be re-shaped around the nipples, with the possibility of incisions around the nipples. Anyone considering plastic surgery should find out exactly where the incisions will be and understand that permanent scarring is a possibility.

In plastic surgery, as in every kind of major surgery, there are always risks and the possibility of complications. Before deciding upon surgery, an affected individual should have a very thorough pre-operative cardiac workup, as well as an evaluation of all other systems. Since many procedures require general anesthesia, Dr. Weinberg particularly emphasized the need for a good, extensive pre-operative evaluation. He recommended that the cardiac workup should include, at the very least, a cardiogram, chest x-ray and ultrasound of the heart. Affected individuals should not smoke for several weeks before the surgery. Smoking is not advisable for anyone.

Certain risks and possibilities of complications relate particularly to those with PXE, while others apply equally to the general population. There is no good evidence as to whether affected individuals may heal more slowly than the general population. Another possible complication in individuals with PXE is the slight possibility of “calcium extrusion” in which a small bit of calcified matter may emerge through an incision. This may be associated with

delayed healing or more scarring. Certain individuals may develop more extensive scarring. Cheloids or hypertrophic scars occur in the general population and can cause difficulty for affected individuals as well.

Among the complications that infrequently occur, and occur in the general population, Weinberg listed the risk of infections, reaction to anesthesia, and the risk of hematoma (bleeding under the skin). Certain complications are specific to the procedure to be done, or to the part of the body that is being treated. An infrequent but possible risk associated with a face-lift is injury to nerves of the face, more likely the loss of sensory nerves than those that affect movement. Mastoplasty, the procedure done to reshape the breast, carries a risk of asymmetry or decreased sensitivity of the nipples, and difficulties in breast-feeding. Women would do best to wait until their childbearing is finished. Plastic surgery does not halt the process of aging, loose skin or folded skin due to PXE, nor do its results last if a patient's skin stretches due to childbearing or weight gain. Dr. Weinberg suggested that it is important to be realistic about what plastic surgery can accomplish and to be aware of possible complications.

One of the concurrent frustrations that affected individuals deal with in deciding whether to undergo plastic surgery is that, in more than a few cases, insurance coverage is initially denied. However, potential patients who are willing to appeal the decision and to fight for coverage have been successful, while others have had no trouble in their insurance coverage. The key to the decision often rests in the definition of plastic surgery as either reconstructive surgery or cosmetic surgery. According

to the definition of The American Society of Plastic and Reconstructive Surgeons ([www.plasticsurgery.org](http://www.plasticsurgery.org)), reconstructive surgery corrects abnormal structures caused by congenital defects, or by trauma or disease. Reconstructive surgery is commonly indicated if the surgery corrects the function of the body. On the other hand, cosmetic surgery restores normal structures of the body to improve appearance and self-esteem. Many insurance carriers approve payment for reconstructive surgery and reject it for cosmetic surgery. The same procedure might be considered cosmetic by one insurer and reconstructive by another. PXE International can provide a letter for you to accompany your request for coverage. In this letter, PXE International clearly states that only surgery can mitigate the skin effects of this congenital disorder. People seeking plastic surgery for PXE are seeking to correct an abnormal structure, affected by disease.

Because PXE is a congenital condition, corrective procedures should be defined as reconstructive surgery. However, authorities sometimes reject claims at first because the loose skin of a person with PXE does continue to carry out its function. Careful appeals and, when necessary, involvement of one's state insurance commissioner, have worked to gain insurance coverage for those with PXE who have been willing to keep fighting.

Mary, of Kansas, USA, worked through the process for insurance approval prior to plastic surgery, and her insurance company eventually approved the claim. She suggests being entirely clear about what one's policy does provide for, contacting others with PXE who have secured approval for insurance

coverage, and assembling a complete set of documents: letters from the personal physician, the plastic surgeon, and those knowledgeable about PXE. She recommends that the applicant send pictures. The authorities denied her first application, as she expected. She remained optimistic that the process of appeal would eventually work for her. She is very happy with the result of her surgery.

The bottom line, like so many things for PXE, is that people considering plastic surgery are entering somewhat

uncharted waters. As we begin to collect information from individuals and plastic surgeons alike, we ask anyone with any experience in this matter to contact us. We can help each other if we share experiences. In addition, our sidebar offers language you or your plastic surgeon can use with your insurance company to educate it about this issue.

*Mary and Isobel are willing to answer questions from others who have PXE and are considering plastic surgery. Contact PXE International to reach them.*

### **Language to Use in Communicating with Insurance Company**

PXE causes a number of systemic manifestations. One of the most common, and most severe, is mineralization of the mid-dermis in the flexor areas of the body. The neck, mouth, chin and sometimes other areas on the face are affected. Additionally, PXE affects the antecubital fossae, axillae, groin, and other areas, as well.

The effects of this mineralization include a pebbly plaque that coalesces to result in lax, redundant folds of skin. In severe cases, these lax folds lead to clinically significant manifestations, including infection and trauma. PXE causes these effects and thus these effects are a result of this congenital disorder.

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