Personal experience with reconstruction of congenital pigmented nevi
Experiência pessoal com reconstrução de nevos congênitos pigmentados

ALEX MARGULIS1, ALLAN BILLIG2

ABSTRACT

Introduction: Surgical excision is the treatment of choice for congenital pigmented nevi. When confronted with lesions comprised of large surface areas that preclude primary approximation of margins, subcutaneous expansion of adjacent tissue and subsequent expanded reconstruction of the area excised is the method of choice. Methods: In this article, the authors provide an overview of the literature and present selected cases of their experience with tissue expansion in the pediatric population, covering different anatomical locations and their respective challenges. Conclusion: If performed with careful attention to several key points, tissue expansion can provide a safe and effective method by which one can satisfactorily reconstruct exposed areas remaining after large nevi excision.

Keywords: Nevi and Melanomas. Nevus. Melanoma. Melanosis.

RESUMO

Introdução: A excisão cirúrgica é o tratamento de escolha para os nevos pigmentados congênitos. Quando confrontado com lesões constituídas de grandes áreas de superfície que impedem aproximação primária das margens, expansão subcutânea do tecido adjacente e subsequente reconstrução expandida da área excisada é o método de escolha. Método: Neste artigo, os autores fazem uma revisão da literatura e apresentam casos selecionados de sua experiência com a expansão do tecido na população pediátrica, cobrindo localizações anatômicas diferentes e seus respectivos desafios. Conclusão: Se realizada com muita atenção para vários pontos-chave, a expansão do tecido pode fornecer um método seguro e eficaz para que se possa reconstruir de forma satisfatória as áreas expostas após a excisão nevos grandes.


1. MD, Plastic Surgeon, Head, Center for Pediatric Plastic and Craniofacial Surgery Hadassah Medical Center, Senior Lecturer, Hebrew University School of Medicine, Jerusalem, Israel.
2. MD, Plastic Surgeon, Center for Pediatric Plastic and Craniofacial Surgery Hadassah Medical Center, Hebrew University School of Medicine, Jerusalem, Israel.

Correspondence: Alexander Margulis MD
Center for Pediatric Plastic and Craniofacial Surgery, Hadassah Medical Center
PO.Box 12000, Jerusalem, Israel, 91200.
E-mail: ralexm@hadassah.org.il
INTRODUCTION

Congenital Melanocytic Nevi

Congenital pigmented nevi (CMN) are classically defined as melanocytic nevi that are present at birth. In one recent large series study, at least one pigmented lesion with clinical and dermatoscopic features of a nevus has been observed in 1-6% of neonates. When compared to acquired melanocytic nevi, the histopathology of CMN tend to extend deeper into the dermis and subcutaneous tissue, are arranged in single file between the collagen bundles, and track along skin appendages and neurovascular structures. CMN’s are roughly categorized by size group, the small one being less than 1.5 cm in diameter, medium size from 1.5 to 20 cm in diameter, and the large and giant exceeding 20 and 50cm in diameter, respectively. In general, authors sometime reserve the term “giant” for nevi larger than 40 cm in diameter. Estimating the final size in adulthood involves calculating the amount by which the particular anatomic site enlarges into adulthood. Although small and medium sized nevi are relatively common, large or giant nevi occur only in approximately 1 in 20,000 births. The colors of CMN range from tan to black, with various shades in between. The borders are often irregular. In reflection to their distribution, large CMN’s are sometimes referred to as “bathing Suit” or “garment” nevi. Approximately 75% of those with large CMN’s also have multiple and smaller satellite nevi disseminated throughout the body. Many large CMN’s present with an increased density of coarse and dark terminal hairs. In terms of size and consistency, CMN may initially appear flat and even, but may become more elevated and lighter, darker, or mottled with time, as well as acquire a “pebbly” cerebriform surface. The risk of developing melanoma with small and medium sized CMN is controversial and thought to be less than 1% over a lifetime. If melanoma does indeed occur, it appears at or after puberty and arises at the dermal-epidermal junction. As for large CMN, several prospective and retrospective cohort studies show the risk of developing cutaneous or extracutaneous melanoma is now approximately 5% over a lifetime. When melanoma does appear in conjunction with large CMN, it usually presents in half of the patients within the first 5 years of life (median age of 3.5 years). The primary sites for large CMN are, in order of frequency, trunk (69%), head/neck (8%), CNS (8%), retroperitoneum (5%), extremity (2%), and others (8%). In contrast to melanoma developing from small and medium sized CMN, large CMN melanoma tends to develop deep within the dermis and subcutaneous tissue, and this can make detection difficult. Other malignancies that may occur in association with large CMN are rhabdomyosarcoma, liposarcoma, and malignant peripheral nerve sheath tumors. Melanoma is less likely to occur in patients with CMN restricted to the head/neck or extremity. Another condition is neurocutaneous melanocytosis, which is the development of malignant melanocytes within the CNS in addition to the skin. Approximately 2/3rds of these patients present with large CMN accompanied by numerous satellite nevi (more than 20 come with a 5 fold increase for NCM). Such CNS melanocytosis can be detected by gadolinium-enhanced-MRI.

The presence of CMN often has psychosocial ramifications, especially in patients with larger nevi located in visible areas such as the face. Such children are more likely to suffer from anxiety and depression, exhibit aggressive behavior, and have social/developmental problems. Furthermore, the psychological burden imposed on the family (siblings and parents), as well as potential bonding difficulties with affected neonates and accepting the child’s condition, can and should not be underestimated.

Management

Small and medium sized CMN can be managed on an individual basis and depending on several factors, namely worrisome changes to the CMN, cosmetic appearance, functional impairment, and anxiety of parents and patient. Among the accepted modalities of treatment of CMN of small and medium size is simple observation and periodic evaluation, curettage and dermatabrasion, ablative therapy with laser, and surgical excision with primary closure.

In large CMN, surgical removal is highly recommended mainly as prophylaxis against the potential development of melanoma. Furthermore, surgical scars, even if extensive, are at times more preferable to patients and parents than the unsightly presence of a large nevus. The specific method of surgical removal of medium or large CMN is serial excision or prior tissue expansion of adjacent (or distant) and uninvolved skin, and subsequent flap reconstruction of the remaining area after excision.

Tissue Expansion

When faced with the task of reconstructing congenital and acquired deformities, tissue deficiency is the foremost underlying problem faced by surgeons. Experience has demonstrated that tissue expansion (TE) provides a reliable mechanism for replacing missing soft tissue. The skin and underlying tissue available from tissue expansion provides for like-quality and like-texture to replace the remaining open wound after lesion excision. In addition to providing additional bulk in cases of skeletal reconstruction where deficient volume must be addressed, tissue expansion allows for moving composite tissue from distant sites to provide skin coverage following soft tissue tumor resection.

The tissue expansion technique takes its roots from lessons learned in distraction osteogenesis. Bone traction led to the concept that mechanical stress could yield lengthened tissue. It was Putti who extrapolated from this that an increased area of soft tissue could be obtained in the same manner. In the 1950’s, Neuman was the first surgeon who experimented with tissue expansion for soft tissue. The idea was reintroduced in the 1970’s by Radovan, this time incorporating a contemporary device with an internally placed port. And in 1982, the Plastic Surgery Educational Foundation recognized tissue expansion as a new advance in reconstructive surgery. Today, the many and varied TE designs have increased its worth as a reconstructive option.

Physiology

When constant mechanical stress is applied to skin over time, two phenomena are observed: mechanical and biological creep, the former based on morphological changes that occur on a cellular level (stretching), and the latter based on gap-junction disruption and induction of cell proliferation. On a molecular level, many factors such as cytokines, growth factors, hormones, adhesion molecules, cytoskeletal elements, and signal transduction proteins are induced as a result of tissue expansion. The basis behind the reliability of expanded flaps is the vast vascularization promoted when the tissue is expanded. Both number and caliber...
of vessels within an expanded flap are superior to those in a non-expanded yet identical anatomic area. VEGF, amongst other angiogenic factors, are expressed in higher quantity on the surface of expanded flaps compared to non-expanded controls.

**Expander**

Although the initial intension was for advancement flaps, tissue expansion experience has demonstrated that expanded transpositional and rotation flaps may be the preferred choice for reconstruction. Transposition, although requiring a considerable investment of forethought and planning, provides for greater versatility in flap design and range. Keeping this in mind, choice of expander, anatomical placement, incision for introduction, and expansion routine vary depending on the site to be reconstructed and adjacent tissue. Several types of TE’s exist, differing in size, shape, and type of filling valve. They follow three basic patterns: round, rectangular, and crescent shape (Figure 1). Expander volumes range from 100cc to 2000cc, and saline solution as the medium for expansion is delivered in a controlled fashion through a valve port—either integrated into the prosthesis itself, or attached to a silicone tubing allowing distance from the prosthesis. The latter, which is more common usage, necessitates placing the valve port over a firm surface of bone or tissue and, if necessary, sutured into place to avoid undesirable movement and flipping. Expanders currently used possess slightly firm backing that allows the redundant envelope to be folded underneath the base when first placed. This avoids future folds and firm points that may lead to overlying skin distress and eventual compromise. As the expander increases in size when inflated, the redundant envelope underneath gradually unfolds beneath the enlarging flap.

Preoperative planning is necessary in order to assess the best possible option for site of incision, TE placement, flap movement in relation to defect, and postoperative scarring. Color, texture, and contour of the recipient site must correspond to the donor site as best as possible in order to maximize the final aesthetic and functional outcome. In order to prevent, or minimize, TE failure or extrusion, the donor site must be free of infection, scars, or trauma. In cases of nevus repair, the incision location is chosen within the border of the lesion to be excised.

**Surgical Technique**

An adequate pocket is dissected to allow placement of the chosen expander, the dimensions of which extending approximately 1 cm more in all directions than the size of the TE to be placed. When a remote port is used, it should be placed over a region of firm skeletal support in order to ease outpatient filling. At the end of the same procedure of TE placement, approximately 10-20% of the total expansion volume is filled in order to assess and possibly revise TE characteristics, namely location and presence of firm surface folds.

Draining tubes are placed in order to minimize potential dead-space. The TE incision site is usually sutured in a watertight fashion with 4-0 Nylon string (buried) followed by nylon running sutures. The area is bandaged softly with Bacitracin ointment or Xeroform gauze.

Serial injections are initiated approximately 7-10 days post-operative, and the draining tubes are removed after the first few post-op visits. After thorough education by the hospital nursing staff, the pediatric patients’ family or caregiver is encouraged to begin a home expansion protocol. Expansion should render the skin tight and tense, but without any severe pain or compromise to the skin. In such cases of excessive filling, the extra volume must be removed. The expansion protocol extends between 8-12 weeks, after which the TE is removed and reconstruction commences. Parents are encouraged to record the scheduled amount injected each time throughout the expansion process.

The amount injected each time is approximately 20-50 cc for the scalp, 5-10 cc for the cheek, 10-20 cc for the forehead and neck, and 50-100 cc for the trunk. The frequency of injections is approximately once per week. Broad spectrum antibiotics are recommended from the time of surgery until approximately 7-10 days post-op (or when the drains are removed). A low threshold for returning the patient on antibiotics is maintained, especially when a concurrent infection is apparent elsewhere in the body. Early treatment with antibiotics allows for minimization of TE infection and possible loss.

**REGIONAL CONSIDERATIONS**

**Scalp**

Despite existing concerns that tissue expansion in the scalp may affect cranial vault morphology, research has shown no distortion or other untoward effects on cranial sutures by CT analysis. Even though cranial molding may appear, any deformity usually corrects itself within 3-4 months after TE removal. Typically, TE’s for nevi repair in the scalp are placed at 6 months of age. For correction of CMN in the scalp, TE’s can be placed serially, with larger expanders placed after each stage in order to distribute the expansible forces evenly over the hair follicles. Previous studies have shown that tissue expansion in hair-baring areas does not induce proliferation of hair follicles. For TE placement in the scalp, a pocket is dissected subgaleally but above the periosiutum. The overlying flap is designed such that consideration is given to the major blood supplies to the scalp (superficial temporal, postauricular, occipital, and supraorbital vessels). Port placement is variable, but the pre-auricular area has shown least port migration. In hemi-scalp reconstruction for both nevi and scar alopecia, temporal hairline reconstruction is best accomplished using a large transpositional flap from the occipital region.
Forehead
Expanded flap reconstruction of nevi in the forehead poses the greatest challenge for the plastic surgeon, mainly because of the potential morbidity involved and possible disfigurement of integral facial structures such as the eyebrow. Past research has revealed a 24% aesthetic complication rate when reconstructing the forehead with tissue expansion. To minimize these complications, certain guidelines have been adopted over the years, among them are bilateral expansion for reconstruction of midforehead lesions; Serial expansion; Supraorbital and temporal lesions best managed by transposition of the expanded skin medial to the lesion; Preference for parietal region expansion rather than temporal region expansion for reconstruction of hairline; If brow is elevated, abnormality should be corrected by interposing non-hair-bearing forehead skin; Maximal avoidance of overextension of the flap. If necessary, continue with TE expansion in order to provide adequate coverage for defect.

Face and Neck, and Pan-Facial
In this region, it is imperative that the surgeon adhere to the subunit principle. The principle dictates that incisions be placed such that the succeeding scars be camouflaged within the natural lines of the face such as the nasolabial fold. Similar to the forehead, undue tension in the face region can disfigure critical structures. Such disfigurement can lead to lower positioning of the lid and lid drooping, and oral incompetence. Neale et al report a 10% rate of lower eyelid ectropion and a less than 10% rate of lower lip deformity when cervical skin flaps are advanced cephalad to the cervicomandibular region. Expanded transpositional or rotational flaps from the lateral cheek and/or neck and postauricular region minimizes the risk of these problems. Needless to say, tissue expansion also allows for full-thickness skin-graft coverage of large surface areas. Pre-expansion in distant areas allows for large skin-grafts to be harvested and used for resurfacing the peri-orbital, cheek, and forehead aesthetic units, especially when local tissue is not available for flap expansion. When the skin is expanded, the total area available is meant to be adequate for coverage of both area to be grafted and donor site. Above the clavicle is the ideal place for TE placement for grafts meant for the face, due particularly to the excellent color and texture match.

Trunk
The lower abdomen is perhaps the most easily expanded site and ideal for TE placement and subsequent expanded flap for local or full thickness graft for distant, coverage of excised giant nevi. In children, is it important to avoid tissue expansion in areas, such as the upper abdomen or chest itself, which can lead to breast tissue distortion. Port placement is frequently chosen in the anterior thigh or, less frequently, over anterior ribs. When treating giant nevi on the back or buttock region, TE on the posterior trunk has become the modality of choice. Serial expansion is frequently performed in order to achieve enough surface area to cover particularly large nevi excisions. For giant nevi treatment, expansion in the trunk can be performed as early as 6 months of age. The lower back is most suitable for larger transposition flaps to cover areas of the buttocks and, similarly, the abdomen for areas of the anterior thigh. As in other areas, transposition flaps in the posterior trunk region allow for better aesthetic and functional positioning of the remaining scars.

Extremities
With regard to tissue expansion, the extremities have traditionally been viewed as areas of limited value and with a higher complication rate (47% vs. 23% in non-limb areas). In a ten-year retrospective review of more than 200 cases, Casanova found a 19.4% complication rate (15.5% major, and 4.9% failure). Despite this, expansion in the limbs is possible. The surgeon must understand the limitations inherent in the basic geometry of the limbs, and that flap coverage is better attainable circumferentially rather than axially. Careful planning, choice of TE size, and a possibly slower rate of expansion can all lead to a lower risk of infection, extrusion, and flap failure. Unstable infected wounds in the extremity are a contraindication for local TE placement. Keeping all limitations in mind, one may consider other options for attaining adequate tissue for coverage after giant nevi excision, such as expanded pedicled flaps or expanded free flaps.

In collaboration with other plastic surgeons, I’ve devised a useful algorithm for repairing complex defects in the upper extremity. We believe that in order to achieve the best possible results in terms of color and contour match, it is best to evaluate the limb in thirds (arm, forearm, and hand) and to note whether or not the lesion in circumferential. For non-circumferential lesions in the arm, expanded transpositional flaps from the shoulder or upper back serve well. If a lesion in the arm is large and circumferential, an expanded free flap is the optimal choice. For large lesions located from mid to lower forearm, tissue from the flank region is expanded and made to serve as a large pedicled flap through which the forearm, after excision of the lesion, can be placed for vascularization from the recipient forearm bed. After a delay period of approximately 3 weeks, the pedicle can be divided. Placement of “crimping sutures” during this delay period allows for a more accurate flap and donor site coverage area at time of division. It is important that the location of the flap be chosen such that the extremity is in a comfortable position of rest during attachment. A comfortable rest position of the extremity minimizes the need for complex immobilization.

For the complex lesions on the hand region, expanded full thickness skin grafts from the abdomen or groin are the modalities of choice. Regional expansion is more suitable for smaller, select lesions on the dorsum of the hand, given adequate donor area for TE placement is available.

In the lower extremities, tissue expansion is even more limited than in their upper counterparts. However, with experience, the surgeon can achieve successful expansion results with limited risk for infection or extrusion of the expander. In general, tissue expansion and flap transposition proximal to the knee is easier than distal to it. As in the upper extremity, manipulation of an expanded flap in the lower extremity is easier circumferentially rather than axially. Below the knee tissue expansion is, for the most part, limited. In select cases, tissue expansion for pedicled flaps from the thigh can be used to cover deficient areas below the knee after giant nevi excision.

Complications
The main complications encountered with tissue expansion can be divided into major and minor categories. The major include infection, expander exposure, and flap ischemia. Even though early signs of infection following surgery are best treated with antibiotics and expander removal, it is possible
to circumvent expander loss by maintaining a low threshold for intervention. Furthermore, even if the expander is indeed removed and the source of infection eliminated, the tissue gained from the already expanded implant can be used to cover deficient areas. Thorough wound irrigation with antibiotics can further provide a margin of safety. As for expander extrusion, if a small area of the incision site is exposed yet the surrounding tissue is unaffected, one can still avoid total loss of the implant by reducing the amount and frequency of expansion. In our experience, the likelihood of implant salvage is more promising if the expander is in a dependent position relative to the open incision site. Conservative treatment with antibiotics and local wound care can further delay implant loss. The open wound, if kept clean, will remain stable or eventually close by secondary intention, thus allowing completion of the intended expansion.

Minor complications, on the other hand, include transient pain during expansion, seroma formation, appearance of dogears at the donor site, and subsequent widening of any scarring. In one study, such minor complications were observed in as much as 17% of the cases followed.

**METHODS**

Between the years 2003 and 2009 we performed 133 expanded flap reconstructions in 61 patients. The youngest patient age was 7 months, with the mean age of patient being 3 years and 7 months. In each case, between 1 and 4 tissue expanders were placed during surgery. In the majority of cases, the tissue expander chosen was Softspan, Bauer design, SSP-rectangular in shape with soft bottom. The range of TE volume was between 70 and 750cc, the specific size chosen based on the characteristics of the nevus and the needs of the patient. Every expander was connected to a remote injection port, and weekly injections were carried out in the clinic. Overall, the overall time need for complete tissue expansion was approximately 10-12 weeks, with a 4-month hiatus in between rounds.

Complications were seen in 12 patients (19.6%) and included infection (49%, Figure 2), extrusion (25%, Figure 3), hematoma (16%), and device failure (9%).

**RESULTS**

Here we present pre and post-operative photographs of selected cases, categorized into various anatomical regions. Patient consent was provided in all cases. Wherever possible, photographs of the patient during the expansion period are also provided (Figures 4 to 11).
Figure 5 – Tissue expansion of forehead. A: pre operative aspect. B: during expansion. C: post-operative aspect.

Figure 6 – Tissue expansion of orbital region. A: pre operative aspect. B: during expansion. C: post-operative aspect.

Figure 7 – Tissue expansion of cheek. A: pre operative aspect. B: during expansion. C: 1-year post-operative aspect.

DISCUSSION

Reconstruction of congenital nevi, especially if large, is an extremely daunting task for the plastic surgeon. Although a relatively straightforward procedure, the ability to anticipate expanded implant characteristics and subsequent flap area coverage is vital for maximal results. In my experience from over 100 pediatric expanded flap reconstructions, several key considerations for different anatomical locations are noteworthy.

For the scalp, it is best to wait until the child is at least 6 months old before a tissue expander is introduced. For this region, flap design must take into consideration the major blood supplies of the region, such as the superficial temporal, post-auricular, occipital, and supra-orbital vessels. Furthermore, the remote tissue expander port has shown least migration when situated in the pre-auricular region.

The forehead region is particularly challenging due to the potential distortion of vital landmarks such as the eyebrow. Here, the surgeon should employ bilateral expansion of the forehead when treating mid-forhead lesions, strive for serial expansion, design a flap such that its transposition is medial to...
the lesion, prefer parietal region expansion over temporal region expansion when addressing reconstruction of the hairline, and strictly avoid flap overextension.

When dealing with lesions on the face, the author recommends adherence to the “subunit principle”, with strategic placement of incisions such that the remaining scars be camouflaged within the natural lines of the face. Expanded transpositional or rotational flaps from the lateral cheek and/or neck and postauricular region, as opposed to cervical advancement flaps, minimizes the risk of lower eyelid ectropion and lower lip deformity. Distant expansion for the purpose of full-thickness-skin-grafts for the peri-orbital, cheek, and forehead regions is highly acceptable, especially when the circumstances for local expansion with flap is not favorable. For such grafts, the ideal location for expansion is immediately above the clavicle.

For lesions in the trunk region, it is important to avoid tissue expansion in areas such as the upper abdomen or chest itself, which can lead to breast tissue distortion. Port placement is recommended in the anterior thigh region. The lower back is most suitable for large transposition flaps to cover areas of the back and buttocks, and similarly the abdomen for areas of the anterior thigh. Expansion can be initiated as early as 6 months of age.

For the extremities, perhaps the most important point to remember is that flap coverage is more attainable when designed circumferentially rather than axially. Also, the higher complication rate observed in the extremities (infection, extrusion, and flap failure) can be minimized by careful planning, appropriate choice of TE size, and possibly slowing the rate of tissue expander inflation. For successful repair of complex nevi in the upper extremity, the author employs a useful algorithm that addresses the extremity in thirds (arm, forearm, and hand). For non-circumferential lesions in the arm, expanded transpositional flaps from the shoulder or upper back serve well. If a lesion in the arm is large and circumferential, an expanded free flap is the optimal choice. For large lesions located from mid to lower forearm, tissue from the flank region is expanded and made to serve as a large pedicle flap through which the forearm, after excision of the lesion, can be placed for vascularization from the recipient forearm bed. For complex lesions on the hand, expanded full thickness skin grafts from the abdomen or groin are acceptable.

For the most part, tissue expansion in the lower extremity is limited, especially distal to the knee. In select cases, tissue expansion for pedicled flaps from the thigh can be used to cover deficient areas below the knee after giant nevi excision.

Needless to say, careful patient selection is imperative from the beginning. After considering and eliminating other suitable yet less risky treatment options, the patient (when old enough) and parents must both be receptive to the idea of tissue expansion as the best option and choice. Among them, there must be a thorough understanding of the expansion process and its potential complications. As for technique, the proficient surgeon should be able to understand potential problems and know how to avoid them. The type of expander, location and length of incision, and expansion routine should all be tailored to the patient and lesion in question. A low threshold should be maintained for prophylactic antibiotics, especially when infection is anticipated or likely. Careful wound management cannot be overemphasized. Lastly, and perhaps most importantly, overall heightened attention to all the details throughout the expansion process will ensure maximal consideration, management, and treatment of the patient.

CONCLUSION

Surgical excision is the treatment of choice for congenital melanocytic nevi. In cases of large lesions, staged tissue expansion and subsequent flap reconstruction offers the advantage of supplying tissue with sufficient surface area for coverage and with similar color and texture of the area involved. With increased vigilance by the surgeon throughout the process, the various stages involved in tissue expansion and subsequent expanded flap reconstruction are relatively safe, with high final-outcome success and minimal complication rates. In my experience, addressing key focus points, both in the patient and surgeon, along with maximal attentiveness for complications and their prompt and effective treatment, assures for satisfactory results that are both medically effective and aesthetically pleasing.

REFERENCES

34. Marulis’ textbook for pediatric tissue expansion.