

in treating VLS [3].

In VLS, histologic findings are typically characterised by orthokeratotic hyperkeratosis, hydropic degeneration of the basal cells, oedema of the upper dermis, and homogenization of collagen associated with a predominantly lymphocytic inflammatory infiltrate [3]. Since these pathological changes predominantly involve the superficial dermal layer, we believe that nanofat grafting is the best procedure for improving the symptoms and signs of VLS.

Nanofat grafting is a simple procedure that allows the administration of adipose stem cells and growth factors in a very superficial layer to treat several pathological conditions that do not show spontaneous healing, such as lichen sclerosus. The standard lipotransfer technique is much more suitable for treating volume deficits.

This was a preliminary study for a longer series of VLS cases showing that nanofat grafting leads to satisfying results due to the regenerative capacities of ADAS cells.

References

1. Zuk PA, Zhu M, Mizuno H, et al. Multilineage cells from human adipose tissue: implications for cell-based therapies. *Tissue Eng* 2001;7:211-28.
2. Tonnard P, Verpaele A, Peeters G, et al. Nanofat grafting: basic research and clinical applications. *Plast Reconstr Surg* 2013;132:1017-26.
3. Gutierrez-Pascual M, Vicente-Martin FJ, Lopez-Esteban JL. Lichen sclerosus and squamous cell carcinoma. *Actas Dermosifiliogr* 2012;103:21-8.
4. Gunthert AR, Duclos K, Jahns BG, et al. Clinical scoring system for vulvar lichen sclerosus. *J Sex Med* 2012;9:2342-50.
5. Casabona F, Priano V, Vallerino V, et al. New surgical approach to lichen sclerosus of the vulva: the role of adipose-derived mesenchymal cells and platelet-rich plasma in tissue regeneration. *Plast Reconstr Surg* 2010;126:210e-211e.

Congenital Insensitivity to Pain and Anhidrosis

Jin Yong Shin, Sun-Woo Kim, Si-Gyun Roh, Nae-Ho Lee, Kyung-Moo Yang

Department of Plastic and Reconstructive Surgery, Chonbuk National University Medical School, Jeonju, Korea

Correspondence: Si-Gyun Roh
Department of Plastic and Reconstructive Surgery, Chonbuk National University Medical School, 20 Geonji-ro, Deokjin-gu, Jeonju 54907, Korea
Tel: +82-63-250-1860, Fax: +82-63-250-1866
E-mail: pssroh@jbnu.ac.kr

No potential conflict of interest relevant to this article was reported.

Received: 10 Apr 2015 • Revised: 1 May 2015 • Accepted: 6 May 2015
pISSN: 2234-6163 • eISSN: 2234-6171
<http://dx.doi.org/10.5999/aps.2016.43.1.95>
Arch Plast Surg 2016;43:95-97

Copyright © 2016 The Korean Society of Plastic and Reconstructive Surgeons
This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/3.0/>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

Hereditary sensory and autonomic neuropathy (HSAN) or hereditary sensory neuropathy has five different clinical subtypes. Congenital insensitivity to pain and anhidrosis (CIPA) is HSAN type IV. CIPA is a rare disease with an autosomal recessive inheritance. Recurrent episodes of fever, no sweating, insensitivity to pain, and self-injury are symptoms of CIPA. In addition, most patients suffering from CIPA experience mental retardation [1]. CIPA has been reported to occur with a genetic mutation of the



Fig. 1.

(A) Right hand. On the right index finger, an erythematous swelling was observed. There was yellowish discharge and a heating sensation. Further, multiple crusts and scars were observed. (B) Left hand. On the left hand, multiple crusts and scars were observed.



Fig. 2.
X-ray of the right-hand oblique view. Distal phalangeal bone of the right index finger had disappeared.

neurotrophic tyrosine kinase 1 (NTRK1) gene [2]. This gene is encoded in the tyrosine kinase receptor, which is responsible for nerve growth factor. Thus, the signals for pain, heat, and cold cannot be transmitted to the brain. A previous study reported that approximately 20% of patients with CIPA die due to hyperpraxia before the age of 3 years [3]. Here, we report on a 33-month-old female with CIPA who presented with a pathognomonic clinical feature.

This 33-month-old female child visited our department. Both her hands were crusted, with yellowish discharge, swelling, redness, and heating sensation on her right index finger (Fig. 1). On the X-ray, the distal phalanx of the right index finger had disappeared and the distal phalanx of the contralateral index finger was destructed (Fig. 2). The patient was diagnosed with cellulitis and osteomyelitis. She sucked and bit her fingers even though her fingers had a large number of open wounds. Therefore, we applied an elastic bandage on both of her hands in order to stop the sucking. Antibiotic treatment was also administered. During follow-up, swelling was observed on the left dorsum of the foot with no pain. She was diagnosed with a fracture of the first metatarsal bone (Fig. 3). She visited the orthopedic surgery department, and a splint was applied. Two days later, she was transferred to our department because of a splint sore. At this time, we realized that she could not feel the pain. She did not sweat easily and therefore had recurrent fever and did not wear clothes well for fever control. She had repeated oral ulcers and injuries to the hands and feet, such as contact burns. The Sequenced Language Scale for



Fig. 3.
X-ray of left foot. Fracture of the first metatarsal bone was observed.

Infants showed delayed mental age at 13 months and low IQ at 66 points.

Because of the elastic bandage, the patient was no longer able to suck and bite her fingers. Thereafter, the symptoms (yellowish discharge, swelling, redness, and heating sensation) were relieved. The patient was discharged after a total hospital stay of 16 days. A weak steroid ointment was applied on her fingers. This ointment has a bitter taste; thus, the patient's sucking behavior decreased. The fracture of the first metatarsal bone healed with union achieved by maintaining the splint in place. The sore also healed with the dressing. We recommended nerve biopsy to confirm the diagnosis; however, the patient's parents did not give their consent for the procedure.

CIPA is a rare disease with a short life expectancy. It is a hereditary disorder, but it could present with sporadic occurrence [4]. Specific treatment of CIPA is not known. Nevertheless, proper training is needed for prevention of injury. Patients suffering from CIPA usually visit the pediatrics department complaining of high fever. However, they may also visit a plastic

surgeon due to recurrent wounds on the hands and feet. If the plastic surgeon is aware of CIPA, proper management could be administered in order to extend the patient's life expectancy.

References

1. Swanson AG. Congenital insensitivity to pain with anhidrosis: a unique syndrome in two male siblings. *Arch Neurol* 1963;8:299-306.
2. Indo Y, Tsuruta M, Hayashida Y, et al. Mutations in the TRKA/NGF receptor gene in patients with congenital insensitivity to pain with anhidrosis. *Nat Genet* 1996;13:485-8.
3. Rosemberg S, Marie SK, Kliemann S. Congenital insensitivity to pain with anhidrosis (hereditary sensory and autonomic neuropathy type IV). *Pediatr Neurol* 1994;11:50-6.
4. Kim JS, Woo YJ, Kim GM, et al. Congenital insensitivity to pain with anhidrosis: a case report. *J Korean Med Sci* 1999;14:460-4.

Emergency Transplantation of Free Flap between Separated Thoraco-Omphalopagus Conjoined Twins

Joo Seok Park¹, Jeong Jun Park², Dae Yeon Kim³, Jin Sup Eom¹

Departments of ¹Plastic Surgery, ²Thoracic and Cardiovascular Surgery, and ³Pediatric Surgery, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea

Correspondence: Jin Sup Eom

Department of Plastic Surgery, Asan Medical Center, University of Ulsan College of Medicine, 88 Olympic-ro 43-gil, Songpa-gu, Seoul 05505, Korea
Tel: +82-2-3010-3602, Fax: +82-2-476-7471
E-mail: jinsupp@amc.seoul.kr

No potential conflict of interest relevant to this article was reported.

Received: 4 Feb 2015 • Revised: 25 Feb 2015 • Accepted: 25 Feb 2015
pISSN: 2234-6163 • eISSN: 2234-6171
<http://dx.doi.org/10.5999/aps.2016.43.1.97>
Arch Plast Surg 2016;43:97-99



Copyright © 2016 The Korean Society of Plastic and Reconstructive Surgeons
This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/3.0/>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

Conjoined twinning is one of the most uncommon congenital anomalies. Spencer has reported that while the incidence of conjoined twinning is close to



Fig. 1.
A 13-day-old thoraco-omphalopagus conjoined twins who shared the heart and the liver.

one in 200,000 live births, 1% of all conjoined twins are stillborn and 40%–60% die shortly after birth [1]. The separation of conjoined twins presents a unique challenge to many pediatric surgical specialties. A multidisciplinary approach is essential for successful twin separation. A variety of methods for providing soft-tissue coverage have been reported in the literature and include the use of skin grafts, skin substitute products, local skin flaps, and tissue expansion.

Here, we report a case of the transplantation of a latissimus dorsi musculocutaneous (LDMC) flap between separated thoraco-omphalopagus conjoined twins.

A set of male thoraco-omphalopagus conjoined twins were born with joined manubrium and upper abdomen (Fig. 1). They had a fused liver, and each had a congenital heart anomaly. Computed tomography (CT) revealed that conjoined twin 1 had supracardiac total anomalous pulmonary venous return, dextrotransposition of the great arteries, and a perimembranous ventricular septal defect, while conjoined twin 2 had supracardiac total anomalous pulmonary venous return.

Although the literature recommends that separation should be delayed until the twins gain weight, the unstable hemodynamics and deteriorating conditions of the infants prompted early surgical separation on the 14th day after birth. During the separation, an atrial connection was identified and conjoined twin 2 took most of the fused atrium. After separation of the fused heart and liver, a chest and abdominal wall defect resulted, which was larger than expected. Closure of the defect was not performed immediately because the cardiac status of the babies required further correctional surgery.

The next morning, however, the heart of conjoined twin 1 ceased to beat and resuscitation for 5 hours failed. Then, to utilize the tissue of conjoined twin 1