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The NLN Mission Statement

The mission of the NLN is to create awareness of lymphedema through education and to promote and support the availability of quality medical treatment for all individuals at risk for or affected by lymphedema.

The NLN is dedicated to:

- promoting research into the causes, prevention and treatment of lymphedema;
- securing adequate insurance coverage for medically necessary, safe and effective treatment;
- expanding the number and geographical distribution of lymphedema treatment facilities and certified therapists

To achieve these goals, the NLN disseminates information about lymphedema to health care professionals so they can appropriately counsel their patients on its avoidance, and prescribe safe, effective treatment for those affected by this condition. The NLN also provides this information to the general public.

Congenital Vascular Malformation: What You Always Wanted To Know, But Were Afraid To Ask

By *ByungBoong Lee, MD, PhD, FACS*

HISTORICAL BACKGROUND

Congenital vascular malformation (CVM) remains one of the most difficult and challenging diagnostic and therapeutic enigmas in the modern practice of medicine. Its clinical presentations are known to be extremely variable, ranging from an asymptomatic birthmark to a life threatening condition. These extremely variable findings of CVM have proven to be a major challenge, even for the most experienced clinicians. To compound these challenging conditions, the extremely rare incidence of CVM has made it quite difficult to improve the learning curve to the optimum degree for proper diagnosis and subsequent treatment. Hence, patients with vascular malformations are usually bounced from one clinician to another, only to increase the disappointing outcomes, often with further complications and recurrence, making the condition worse in general.

Even though CVM has been known for centuries as an enigma in medicine and many attempts have been made to control this ever-challenging problem, the work has been performed primarily by surgeons, with disastrous results after surgical intervention, often compounded by a poor understanding of the nature of the disease as a whole and a subsequently ill-planned and over-aggressive approach by the surgeon.

Recently, however, a better understanding of the anatomy and pathophysiology of the disease nature and advancements in medical technology have provided a more accurate and safe diagnosis for the improved management of this disease, in general. And the new concept of total care management through a multidisciplinary approach slowly has replaced the decades-old, over-conservative attitude to CVM, wrongly based on prejudice following the disastrous experiences

by an over-aggressive surgical approach in earlier decades.

Since 1980, surgeons and physicians of this specialty on both sides of the Atlantic looked into possible collaboration for a better strategy with a new attitude through two globally recognized CVM workshops, one in Hamburg in 1988 and another in Denver in 1992. The urgent necessity to accept this new approach through multidisciplinary coordination was fully confirmed, and a new classification of the wide spectrum of this disease, which had defied proper classification and compounded therapy for decades, was correctly introduced.

Though significant differences in the concept of CVM remained between two major streams across the Atlantic, a few fundamental issues were properly assessed together through the Seoul consensus workshop of 1996. The importance of proper sharing of knowledge through a common language, that is, using a common classification of CVM throughout the world, was upheld once again for optimum communication. We also concluded that a multidisciplinary approach is required for the advanced care of CVM to provide better handling of this complex

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vascular problem beyond the limits of conventional management, with a well-orchestrated effort for maximum coordination among various related specialists.

The Congenital Vascular Malformation (CVM) Clinic—opened as one of the vascular specialty clinics of the Vascular Center at Samsung Medical Center and Sungkyunkwan University, Seoul, Korea, in 1996—remains a model for the extensive multidisciplinary approach to this old problem with a new concept for advanced care through the collaboration of 15 different specialty departments. Of the 15 specialties involved, vascular surgery, orthopedic surgery, plastic and reconstructive surgery, pediatric surgery and interventional radiology are the leading core members of our team approach in this new method of multidisciplinary management.

NEW & OLD TERMINOLOGY

CVM is a relatively new terminology representing one distinctive vascular disease entity that has been well documented since the 16th Century. Traditional name-based terminology for CVM, such as Klippel-Trenaunay Syndrome (*see photo*) and Parkes-Weber Syndrome, for example, have been used extensively and have become a symbol of the complex nature of the disease. However, this name-based nosology only added more confusion and failed to represent accurate information on pathophysiology, anatomy, and embryology of various CVMs.

Proper consensus for better understanding of its nature was drawn, for the first time in history, with new nomenclature/terminology and classification of the CVM through the Hamburg consensus meeting in 1988. Based on this new terminology/classification, new knowledge of its pathophysiology, anatomy and embryology has been rapidly accumulated over the last two decades. Subsequently, new concepts for the contemporary diagnosis and management of the various CVMs have emerged to clarify many old and confusing issues, including the rela-

tionship of primary lymphedema which once was abandoned, but belatedly confirmed as a lymphatic malformation with other CVMs.

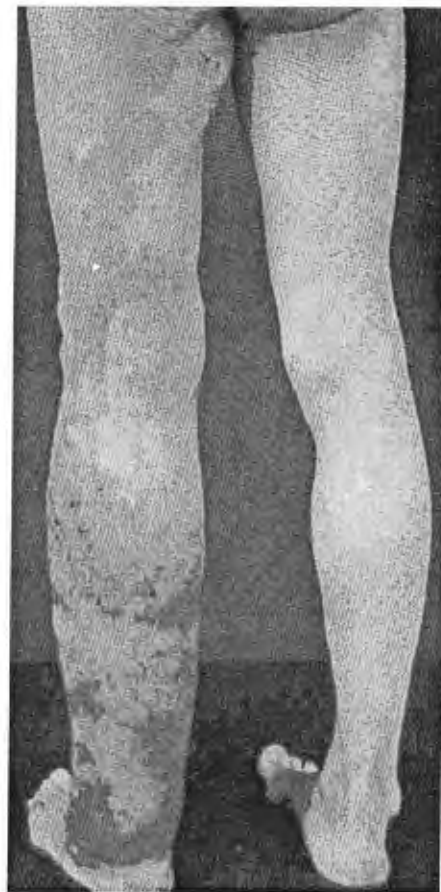
The new terminology, based on the Hamburg Classification, has at last replaced the old name-based terminology, which had been almost a trademark of all the different varieties of CVM for decades. The Hamburg Classification of CVM now is accepted as the formal nomenclature/terminology to classify various CVMs by most of leading members of the International Society for the Study of the Vascular Anomaly (ISSVA). Other terminologies like angiodysplasia or cavernous/capillary hemangioma, initiated by the specialists (e.g. pathologists) in this field in earlier decades, also added to the confusion and, therefore, have been encouraged to be abandoned as well (although it is still popular not only in the medical community, but also among laymen).

DEFINITION-CLASSIFICATION

CVM is one of two different components of the "vascular anomaly," together with "neonatal/infantile hemangioma" which represents only vascular tumors. Proper differentiation between these two entities, therefore, is extremely important since both have entirely different backgrounds; embryology, histopathology, physiology, and anatomy, will require different clinical management.

While vascular malformation represents a pure birth defect originated from mesenchymal cells before the birth following developmental arrest in the peripheral vascular system during the embryonal life, hemangioma in the new definition is a pure vascular tumor with self-limited growth developed mostly after birth. The CVM, therefore, represents entire varieties of birth defects developed along the peripheral vascular (arterial, venous, and lymphatic) systems during the embryonal life with various etiologies, mostly unknown. What its clinical significance will be totally depends on the components of vascular systems involved in this birth defect. When the arterial system alone is

Thirty-year-old Korean male with Klippel Trenaunay Syndrome (KTS), chronic venous hypertension and increased length of leg.



involved (e.g. carotid coiling), the defect is named an arterial malformation (AM); venous malformation (VM) when venous system only is involved (e.g. venous aneurysm); lymphatic malformation (LM) when lymphatic system only is involved; arteriovenous malformation (AVM) when arterial and venous systems both are connected abnormally, bypassing the normal capillary system and establishing direct/indirect arteriovenous (AV) shunting status (e.g. AV fistula); combined defects when various kinds of CVMs are existing in a mixed status (e.g. hemo-lymphatic malformation-HLM as mixed form of VM and LM); capillary malformation (CM) when capillary system only is involved (e.g. portwine stains).

The nature/characteristics of each CVM is further complicated by the embryonal stage when its developmental arrest has occurred resulting in vascular defects. Its behavior is quite different depending on the embryonal stage during which the defect developed. When it occurs in an

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CVM...

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earlier stage of embryogenesis, the defect, designated as the extratruncular (ET) form remains, possessing unique characteristics of mesenchymal cells, that is, evolutionary potentials that are extremely dangerous for the clinician to encounter. When this ET form gets adequate conditioning/stimulation (e.g. hormonal change, pregnancy, trauma, surgery), it grows/recurs, often explosively, making the clinical management quite complicated. The truncular (T) form, meanwhile, as the result of developmental arrest in the latter stage of embryogenesis, lacks this crucial characteristic of mesenchymal cells and maintains only hemodynamic consequences (e.g. venous aneurysm).

Therefore, in addition to proper identification of each CVM involved, further precise identification of whether T form and/or ET form of CVM should be involved is extremely important to the clinician for safe management. For example, all the cardiac (septal and valvular) malformations and most neurovascular malformations (e.g. cerebral aneurysms) belong to this T form, which means they can be handled without risk of recurrence, while CVMs developed in the peripheral vascular system should be handled with extreme care since most of them belong to the ET form with high potential risk of recurrence when stimulated.

Due to the complicated conditions of CVM, in addition to its rarity, CVMs often mislead proper diagnosis and, subsequently, result in mismanagement, worsening the condition and contributing to the many inaccurate prejudices regarding CVMs. Therefore, this critical information of the embryologic status of various CVMs based on the new classification must become the major guideline, not only for the diagnosis, but also for the treatment strategy point of view.

DIAGNOSIS AND MANAGEMENT

The diagnosis of CVMs is complicated due to their complex nature and various

embryologic characteristics, but precise assessment/diagnosis is extremely crucial for deciding the proper treatment approach.

Proper diagnosis of such complicated CVMs finally has become feasible only in the last decade, with various new diagnostic technologies for the arterial, venous and lymphatic system based on the most recent information available on distinctively different arteriodynamics, venodynamics and lymphodynamics. New information on the lymphodynamics, in particular, recently uncovered regarding autoregulatory peristaltic function, in contrast to the venodynamics, finally corrected much of the misconception about the lymphatic system and provided proper guidelines for the diagnosis and treatment not only for the LM but also for the combined form of HLM.

With precise diagnosis of the T and/or ET form of various CVMs, a proper decision regarding indications for treatment of each component of the CVM have become feasible, as well as for the selection of the right modality of treatment.

Since not every CVM can be or should be treated, depending upon its kind (e.g. CM), type (T or ET) and/or location (e.g. proximity to airway) and severity (e.g. expanding, bleeding), etc.—except in the case of a life or limb threatening one—the decision for treatment as well as selection of the treatment approach now can be made safely, preferably by a multidisciplinary team approach.

Therefore, many believe CVMs, in general, can no longer be neglected by the vascular specialists in medicine/surgery, who will encounter this problem anyway as frontline clinicians, and a basic knowledge and understanding of CVMs is mandated, despite the fact that CVMs are relatively rare—though no more so than cardiac anomalies—when compared to other vascular problems.

CLINICAL IMPLEMENTATION

In order to adopt the new concept of contemporary management of CVMs, we initiated the CVM Clinic in 1995, based on

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the multidisciplinary team approach, together with a separate Lymphedema Clinic, which also handles a substantial portion of the T-form of LM known as primary lymphedema, a type of chronic lymphedema.

The CVM Clinic was organized as one of the vascular specialty clinics of the newly established Vascular Center at the Samsung Medical Center (SMC), the flagship hospital of Sungkyunkwan University, Seoul, Korea, as a groundbreaking symbol of the multidisciplinary approach to various vascular specialty problems. The CVM Clinic is based on a fully integrated multidisciplinary team consisting of 15 specialty departments working cooperatively to achieve an orchestrated effort among related clinicians for maximum benefit.

The CVM Clinic at the SMC presently serves over 1,200 patients with various CVMs, providing the most state-of-the-art advanced care in history, together with the Lymphedema Clinic which also serves over 1,000 lymphedema patients of various etiologies.

The CVM Clinic at Seoul is the one and only referral center specializing in CVM management in the whole of Asia, drawing worldwide attention. Recently the clinic expanded its service to include not only Asian patients, but patients from around the globe, making it one of the leading institutes of its kind in the world.

Contribution of the CVM Clinic, Samsung Medical Center, to the field of CVMs since it was established in 1995 includes:

1. Specialized only for the exclusive management of CVMs as one of world's first multidisciplinary organizations in this field.;
2. Adopted new concept of multidisciplinary approach for the clinical management of various CVMs;
3. Adopted and upheld the modified Hamburg classification as the new classification of CVMs;
4. Adopted and developed further,

various new diagnostic technologies: whole body blood pool scan (WBBPS), transarterial lung perfusion scan (TLPS);

5. Established ethanol sclerotherapy as major scleroagent to VMs;
6. Established new concept of peri (pre and post) operative embolo/sclerotherapy to various malformations (VM, AVM & HLM);
7. Fully integrated the surgical therapy with various newly introduced embolo/sclerotherapy;
8. Established discriminating implementation of OK-432 (picibanil) to the ET form of LM depending upon the anatomical characteristics (cavernous, cystic or mixed), with proper combination of surgical treatment;
9. Developed new strategy to the fistulous (high flow) type of AVMs with multidirectional approach: transvenous, transarterial and/or percutaneous direct puncture technique, utilizing various agents (e.g. contour particles, coils, balloons, N-butyl cyanoacrylate, ethanol);
10. Developing new scleroagent to the VM with (micro) foam therapy.
11. Developing new diagnostic technology for the assessment of LM: ultrasonographic and/or MR lymphangiography;
12. Major contributions made to the body of knowledge in the field by members of CVM Clinic and Lymphedema Clinic through numerous presentations and publications in the field of CVMs and lymphedema.

CLINICAL MANAGEMENT

CVM patients registered at the CVM Clinic, SamSung Medical Center (December 2003), have now reached a total number of 1,203 (Female-687 and Male-516) with various CVMs: AM-2, VM-453 AVM-145, LM-393, HLM-108, CM-7, and unclassified-95 (mostly VM, deferred for the final diagnosis due to young age),

with an age range of 14 days to 81 years (mean-23 years).

Four hundred, fifty-seven were selected for the treatment with various indications and underwent a fully integrated variety of surgical and non-surgical (embolo/sclerotherapy) management methods: 323 with sclerotherapy as independent therapy through 1165 sessions, and 134 with surgical therapy through 171 operations with/without combined embolo/sclerotherapy. Excellent results of the contemporary management of CVM by multidisciplinary team approach includes immediate failure rate of sclerotherapy in 3.9% per session (45/1165), and 11.1 % per person treated with sclerotherapy (45/405).

CONCLUSION

Although the treatment can be led primarily by the surgeon and/or interventional radiologist to perform the combination of various surgical and embolo/sclerotherapies through the multidisciplinary approach, proper referral of the patient with this ever-confusing, as well as ever-challenging, CVM to the right team that will be able to handle it properly is most crucial for the right diagnosis and disposition. The crucial role of internists, if not angiologists, who are most likely to encounter this vascular malformation first among the physicians and have to bear the most important responsibility to guide them in the right direction, cannot be overemphasized. Naturally, these angiologists, in general, ought to have a minimum basic knowledge in these CVMs, enough to recognize them properly to initiate the proper challenge to this enigma in modern medicine as the front-line encounter.

Acknowledgement: Without devotion to this ever-challenging problem by each member of the CVM Clinic, Lymphedema Clinic, together with the department manager, nurse coordinator and research coordinator, the present globally recognized achievements in the field of CVMs would have not been possible. I wish to express my deepest gratitude to each member through this presentation.

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