Endovascular Management of the Congenital Vascular Malformation is Not a Panacea: Editorial

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Congenital vascular malformation (CVM) has maintained its notorious reputation as an enigma in modern medicine throughout the century; due to its embryological characteristics, the CVM is known as a unique vascular disorder of extreme variety with stigma of totally unpredictable behavior.¹

Before a modern diagnostic technology became available through the last century, the knowledge on this unique embryonic tissue remnants was so limited that a cavalier (and overaggressive as well) approach to the CVMs, mostly led by surgeons alone based on limited knowledge and experiences resulted in often disastrous outcomes with high recurrence and high complication/morbidity. Naturally such poor outcome added further confusion on its management with erroneous prejudice to the CVMs for many decades.

However through last three decades, the nature of the CVMs affecting the entire circulation systems, artery, vein, and/or lymphatics in various extents and severities, has been correctly understood, and a new concept on its management has emerged based on a new classification-Hamburg Classification to provide an improved strategy.²

The old concept based on name-based eponyms was timely replaced by Hamburg classification which in turn provided a proper ground to adopt various new technologies for accurate diagnosis and subsequent management of the CVMs.

Such new approach based on a new concept opened the door for the endovascular therapy as a new additional armamentarium to the traditional open surgical therapy, to establish a multidisciplinary (team) concept.³

New endovascular approach with various embolo-sclerotherapies was adopted as a possible solution to compensate earlier mistakes solely based on the open surgical treatment; it is now fully integrated as one approach through a multidisciplinary strategy to achieve an acceptable goal for the contemporary management of the CVMs.⁴
However, through the last two decades we learned the lessons on quite a few critical risks involved to the endovascular approach through painful mistakes mostly by the ignorance; ‘indiscriminating’ enthusiasm to the endovascular therapy in its initial period gave much more harm as the price for the learning curve.

Through the experience with absolute ethanol sclerotherapy, for example, we confirmed that the endovascular therapy is NOT a panacea to all different conditions of the CVMs. Its benefit can be maximized only by careful use with appropriate precautions to limit to ‘surgically inaccessible’ lesions. And ‘surgically accessible’ lesions would be better off with a more effective management, with open surgical/excisional therapy, with less morbidity providing a potential chance of ‘cure’.

The primary aim of the endovascular therapy in the CVM management should remain for ‘surgically inaccessible’ lesions as the treatment of choice, within the boundary of a multidisciplinary team approach to improve the management outcome with reduced morbidity and minimum recurrence.

The secondary aim/role of endovascular therapy could be expanded to the ‘surgically accessible’ lesions as a supplement to improve subsequent surgical/excisional therapy through preoperative as well as postoperative embolo-sclerotherapy.

Such peri-operative, especially preoperative, therapy could reduce surgical morbidity as well as complications drastically so that such benefit can exceed beyond the boundaries of the traditional surgical approach. Hence, an appropriate embolo-sclerotherapy, before, during, and/or after the surgical excision is highly desirable to achieve a far superior outcome than the conventional excisional surgery alone even for a simple surgically accessible lesion.

Regardless of the type of the CVM lesions and modalities of the therapy, ultimate goal of the therapy is the precise control of the “nidus” of the lesion itself to prevent recurrence and/or fast deterioration of the lesions with full consideration of its embryological characteristics in addition to its hemodynamic consequences.

All ‘extratruncular’ type CVM lesions as the result of a defective development in the ‘early’ stage of embryogenesis keep its unique embryological characteristics to progress, originated from its mesenchymal cells/angioblasts. These lesions will grow and recur by various endogenous and exogenous stimulations (e.g. female hormone, menarche, pregnancy, trauma, surgery).

Previous approach to the AVM, for example only with the strategy to shut off the feeding artery with ligation or embolization, leaving the nidus of the lesion intact, is totally wrong; such a mistake would provoke a more aggressive neovascular recruitment by this primitive tissue only to make the condition worse with no exception.

In addition, such endovascular therapy based on various modalities of the embolo-sclerotherapy can provide only ‘effective control’ at best; it should not be abused aiming for a ‘cure’ overzealously. Even the absolute ethanol cannot provide a chance of cure without enormous collateral damage, for example. Therefore, in view of unique embryological characteristics of (extratruncular) CVM lesions, the endovascular therapy is mandated for ‘controlled’ aggressiveness within the accepted boundary of the “palliative” concept only when the benefit d exceeds the morbidity following the treatment.

Besides, proper selection of the embolo-scleroagent should be made to fit best to each lesion with different location and severity. Most of presently available embolosclero-agents (e.g. absolute ethanol, Onyx, N-butyl cyanoacrylate, coil and/or contour particles) are far from the perfect accompanying significantly high risk of complications and/or morbidities; a careful plan, from the diagnosis and treatment to the long term follow up assessment, is mandated for each different agent with different limitation, risk, and morbidities (e.g. bleomycin).

Therefore, proper assessment on the response should be included upon the decision on the subsequent session of the endovascular treatment for its agent to be used, the amount, and the delivery route, etc.
Preferred modality of embolosclerotherapy should be selected based on positive balance of benefit by the treatment over the morbidity accompanied by the treatment, unless the treatment is indicated regardless of its price for a life- or limb-threatening condition (e.g. hemorrhage of venous malformation; high cardiac output failure by AV malformation).

Critical role of careful assessment on benefit versus risk of morbidity before the commitment cannot be overemphasized, and the ultimate goal of the treatment has to be clearly defined with a realistic expectation.

By the same token, the same principle of the endovascular therapy cannot be implied to all the CVM lesions as ‘one size fits to all’. Because, not all the CVMs are the same in their clinical significance, natural progress, response to the therapy, etc. and they are all different from each other. For example, early aggressive approach to all the AVM lesions, either macro- or micro-AV shunting, is generally warranted whenever and wherever possible to reduce the serious impact as well as the consequence of the AVM lesion. However, the venous malformation or lymphatic malformation seldom becomes a life- or limb-threatening condition on the contrary to the AVM.

Therefore, a ‘discriminating’ use of same embolo-sclerotherapy is warranted depending upon the type of the CVMs.

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