

To Look for the Last Piece of Puzzle of Congenital Vascular Malformation Hemodynamic Issues

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The congenital vascular malformation (CVM) is a group of various birth defects developed in various peripheral vascular systems during the various stages of embryonal life to make the condition more complicated. It is therefore, unique vascular disorder of extreme variety with stigma of totally unpredictable behavior and complicated characteristics.

Depending upon the stage of embryonal life when the developmental arrest occurs, it maintains embryologic characteristics originated from the mesenchymal cells with potent evolutibility to grow again whenever the condition is suitable, like cancer-cell.

In addition to this embryologic characteristics, the hemodynamic characteristics are also different and complicated, depending upon the stage of embryogenesis when the defect(developmental arrest) developed either in reticular stage with reticular network of vascular system or after the reticular stage to develop to the trunk of vessels. And, it often affects more than one vascular system with subsequent impact to the entire circulations system directly and/or indirectly to make hemodynamic status more complicated.

The CVM, therefore started to be known as an enigma among vascular disorders along the turn of the last century with notorious reputation for its totally unpredictable behavior mostly due to embryological characteristics and further became the curse to the vascular surgeon who led most of challenge due to its hemodynamic complexity.

It has remained with much confusion and con-

trovery through the century from the terminology (nomenclature) to the treatment strategy due to this complex nature.

In spite of much advances we have made in its concept and management through the last two decades, the CVM is still ultimate challenge to the surgeons often as symbol of humiliation. Though most of etiological, embryological, anatomical, and pathophysiological aspect of CVMs are now well investigated with reasonable answers, the complex nature of the CVM has not been fully understood yet, mostly due to this newly recognized hemodynamic complexity.

The hemodynamic aspect of CVMs in particular still leads significant confusion since the CVM affects more than one vascular system in most of occasions and remains for the further clarification on various hemodynamic impacts as well as its consequences by each vascular system affected by the CVM.

Comparing to the vascular malformation primarily involving only one vascular system, that is arterial, venous or lymphatic system, the malformation involving two(or more) vascular systems(e.g. arteriovenous or veno-lymphatic system) results in much more complicated hemodynamic consequences and its clinical impact is often quadrupled, to increase the risk of complication and morbidity through the management.

The arteriovenous(AV) shunting malformation in particular has been known to be the most complexed form of CVM hemodynamically with enormous impact to the entire cardiovascular system, proximally and distally through the arteriovenous communication(e.g. AV fistula). Eventually entire

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arterial and venous as well as lymphatic systems are involved together with the heart to result in utmost hemodynamic complexity.

Therefore, the AVM remains ultimate challenge among various CVMs due to this hemodynamic complexity, and the proper interpretation of hemodynamic consequences of dynamic change of AV shunting status will be the final key to manage the AVM safely. Hemodynamic problem of the AVM is not limited to the lesion itself but extended more to its secondary phenomenon (e.g. arterial and/or venous aneurysm or ectasia). It is extremely difficult and delicate to manage the primary hemodynamic problem as well as its hemodynamic consequences to the secondary phenomenon safely with reduced morbidity and complication through proper prediction and prevention of its natural outcome (e.g. rupture and/or thrombosis).

Our experiences of various treatments to the CVMs, mostly based on ethanol-oriented embolo/

sclerotherapy with/without combination of surgical therapy, have shown extremely high acute complications as well as morbidity: 112 patients among total 362 patients (34%) through 212 sessions out of total 951 sessions (22%), though following remarkable success of the therapy. And most of these complications (e.g. tissue damage) and/or morbidity (e.g. transient pulmonary hypertension) were found to have close relationship to the inadequate prediction and/or assessment of this hemodynamic aspect of complicated natures of the CVMs.

With presently available limited knowledges in hemodynamic aspect of various CVMs, proper prediction and prevention of various hemodynamic complications and morbidity following the treatment and/or its natural course is substantially curtailed. Active incorporation of advanced biomechanical engineer knowledges with clinical knowledge only can bring the last piece of puzzle to break this stalemate in current management of CVMs.