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REVIEW

New Approaches to the Treatment of Congenital Vascular Malformations (CVMs)—A Single Centre Experience

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Objective. *A retrospective review of the results of management of congenital vascular malformation (CVM) patients was made to assess the efficacy of newly introduced approaches.*
Methods. *CVMs were categorised according to the Hamburg classification on the basis of minimally invasive tests. Invasive studies such as angiography are used to provide a road map for treatment. A new multidisciplinary approach was adopted, which accepts the integration of embolisation and sclerotherapy with traditional surgical therapy. Embolo-sclerotherapy was used as an independent therapy and as an adjunctive therapy to surgery.*
Results. *Ninety-nine out of a total of 294 venous malformation patients underwent ethanol sclerotherapy with an immediate success rate of 98.8%, requiring 419 sessions of treatment. Interim results were excellent with no evidence of recurrence (mean follow-up 18.2 months). Most of the 25 patients treated surgically received pre-operative embolo-sclerotherapy, each with excellent interim results and minimum morbidity (mean follow-up 21.2 months). Forty-eight patients among 76 arteriovenous malformation patients underwent embolo-sclerotherapy independently (32/48) or adjunctively (16/48). Independent therapy on 32 produced excellent interim results (25/32) requiring a total of 171 sessions (mean follow-up 19.2 months).*
Eighty-nine extratruncal (ET) forms of lymphatic malformations received multiple sessions of sclerotherapy with OK-432 (108/120 sessions) or ethanol (12/20 sessions). OK-432 was used in 51 paediatric patients with the ET form and produced an excellent response in cystic type lesions (40/45) requiring 61 sessions with no evidence of recurrence (mean follow-up 24.2 months), whereas a mixed result was obtained in the cavernous type (3/6). OK-432 sclerotherapy was used as a pre-operative adjunctive therapy in 7 patients requiring 21 sessions with 17 cavernous type of the ET form, and produced good to excellent results after surgical excision of 14 lesions.
Conclusion. *New approaches to the treatment of CVMs based on a multidisciplinary approach can improve results by fully combined surgical treatment with embolo-sclerotherapy.*

Keywords: Congenital vascular malformations; Hamburg classification; Surgical therapy; Embolo/sclerotherapy; Multi-disciplinary team approach; Venous malformation; Arteriovenous malformation; Lymphatic malformation.

Introduction

Congenital vascular malformations (CVMs) remain difficult diagnostically and therapeutically despite continued efforts over the decades. CVMs have a notorious reputation due to their variety, with a wide range of clinical presentations from a simple birthmark to a life-threatening condition containing embryonic remnants of a developmental defect. The condition has been further complicated by an unpredictable clinical course, confusing nomenclature, erratic response to treatment, frequent recurrence, and high morbidity following conventional treatment.[1–4]

The poor results of treatment over the last decade, are partly the result of ill-planned treatment strategies and a cavalier approach based on limited knowledge of CVMs.[5,6] Recurrence also plays a part as embryonic remnants of CVMs retain developmental potential.[7] Disastrous surgical experiences have allowed ill

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founded prejudice to develop concerning the treatment of CVMs.[5] The Hamburg classification[4,6,8] was introduced to provide proper information about the aetiology, anatomy, and pathophysiology of CVMs, and has became the basis for contemporary diagnosis and management.[4,6] The new concept of a multidisciplinary approach[9–12] integrates surgical and non-surgical treatments.[1,13–15] We organised the CVM (congenital vascular malformation) clinic at Samsung Medical Center, Seoul, Korea as a referral centre, based on a multidisciplinary approach in 1994. A retrospective review of interim results of the diagnosis and management of CVM patients (January 1995–December 2001), was made to assess the various issues raised by this new approach.

Diagnosis

Definition (Classification)

The classification of CVMs is difficult, since CVMs are a group of birth defects that occur as a consequence of developmental arrest at various stages of embryonic life. This affects the whole peripheral vascular (arterial, venous, lymphatic, and capillary) system. Traditional nomenclature of these various CVMs, based mainly on of name-based eponyms (e.g. Klippel–Trenaunay Syndrome, Parkes–Weber Syndrome) add further confusion, and therefore a proper classification of CVMs has been developed to provide proper aetiological, anatomical, pathophysiology, histological, and embryologic information in addition to haemodynamic information.[3,4] Various classifications have been proposed, including the (high and low) flow-based classification, but have failed to fulfil this requirement.[6]

A classification was published based on a consensus workshop for CVMs held in Hamburg, Germany, in 1988 by the ISSVA (International Society of Study of Vascular Anomaly).[4,6] It classified CVMs according to the predominant vascular defect: arterial, venous, AV (arteriovenous) shunting, lymphatic and combined. Each vascular defect was further classified into truncal (T) and extratruncal (ET) forms, based on the embryonic stage of developmental arrest.[7–9,16]

ET forms, develop from the earlier stages of embryonic life at the reticular stage. They possess embryonic characteristics of developmental potential originating from mesenchymal cells (angioblasts). This feature allows them to grow whenever stimulation (e.g. trauma, surgery, hormonal therapy, pregnancy, and menarche) is given. The ET form is further classified by its clinical presentation as diffuse and infiltrating or as limited and localised. T forms develop from a later stage of embryonic life along axial vessels and lack embryonic characteristics. The haemodynamic impact of the T forms is more severe in general than that of the ET form. A lesion is further classified by its clinically presentation as either an aplasia or as an obstruction type (e.g. hypoplasia, aplasia, hyperplasia, stenosis, membrane, and congenital spur), or as a dilatation type (e.g. localised-aneurysm and diffuse-ectasia).[4,6] This concept fulfils most of the requirements to provide aetiological, anatomical and pathophysiological information required for clinical management of CVMs. The Hamburg classification further elucidated the unique relationship between vascular malformations and vascular tumours (infantile/neonatal haemangiomas) to remove confusion concerning these two different entities which produce similar vascular anomalies.[2,17–19] We adopted this modified Hamburg classification[4,6] as the basis of a new structure of contemporary CVM management at our clinic.

CVMs and haemangiomas

CVMs and haemangioma are both categorised as 'vascular anomalies', but the conditions are entirely different, so a precise understanding of their differences is essential for the proper diagnosis and management of these conditions.

Haemangioma (infantile/neonatal) is the most common tumour of infancy. It is a rapidly growing, but self-limiting vascular tumour that usually appears during the first 4 weeks of life. Explosive growth is common, and is generally triggered postnatally, but almost invariably undergoes spontaneous regression before the age 5–7 years. The rapid growth of haemangiomas is the result of endothelial proliferation with increased mitotic activity. In contrast CVMs are not hypercellular and have a 'mature' endothelium with normal endothelial mitotic activity, and grow *pari passu* with the child.[2,17–19] The differentiation of haemangioma and CVM is generally possible given the clinical history and the physical examination alone. However, a deeply seated haemangioma in subcutaneous tissue or muscle without involvement of the papillary dermis can be confusing, and occasionally needs histological confirmation. In addition to their differentiation from haemangiomas, CVMs[2,17–19] need to be differentiated from other vascular/non-vascular conditions involving soft tissue (e.g. haematoma, neurofibroma, and sarcoma). The proper differentiation of CVMs from various malignant