Features compression after open and endovascular operation in vascular malformation

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AV-malformations: hemodynamic phenomenons

- Primarily development of collateral circulation
- The blood shunted through the AV circuit fails to reach the periphery causing peripheral ischemia
- Turbulence near AV-shunts. Trauma of vascular wall
- Reverse blood flow in distal segments of arteries
- Dilatation of efferent and afferent veins, connected with the fistulae area (secondary varicosity)
- The venous pressure increases markedly also in the veins running back to the heart
AVM – the basic positions of medical tactic

- Hemodynamic validity of any intervention
- Angiography – only superselective
- Superselective embolization directed to the nidus of malformation
- Endovascular embolization - sometimes only part of complex treatment; optimal – further operative treatment (if surgical intervention is possibly…)

*Without intervention in AV-shunting area efficacy of operation is problematically*
• Sustained compression pressure should never exceed the intraarterial pressure (ankle pressure)

• Stiff, middle pressure bandages (30 - 40 mmHg) reduce venous oedema, improve nutritional flow and venous pumping function

• Intermittent pressure waves (pumps, stiff bandages + movement) enhance arterial inflow

• “Modified inelastic bandages“ are the basic treatment in AVM
## Compression and arterial circulation

<table>
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<tr>
<th>Sustained Compression</th>
<th>Intermittent Compression</th>
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<tr>
<td>Light pressure enhances arterial flow</td>
<td>Enhances arterial flow</td>
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<tr>
<td>Strong pressure reduces arterial flow</td>
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</table>

- Light pressure enhances arterial flow
- Strong pressure reduces arterial flow
Leg ulcer due to multiple arteriovenous malformations in the lower extremity of an elderly patient.

Ueda I¹, Ihanobe K², Morita M³, Nakahara C², Katsuoka K¹

Abstract
A 66-year-old woman with a history of deep vein thrombosis (DVT) presented with an irregularly shaped leg ulcer surrounded by pigmentation on the left lower limb. In addition, the circumference of her left thigh had gradually increased. The ulcer did not respond to topical treatment and enlarged, therefore, she visited our hospital. Arteriography of the left lower limb showed multiple arteriovenous malformations (AVMs), based on which we made a diagnosis of a leg ulcer due to multiple AVMs. Transcatheter arterial embolisation with a mixture of N-butyl-2-cyanoacrylate and lipiodol was performed six times in the period of about a year for treating the AVMs. The ulcer was managed with bed rest, surgical debridement, continuous pressure support with elastic wrap and topical treatment. After 15 months, the ulcer healed, leaving pigmentation and scarring. It is quite rare for AVMs to progress in the elderly. We speculate that the DVT had caused occult AVMs to become symptomatic following an increase in size.

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Foot ulcer due to arteriovenous malformation: report of a case.

Lee SG¹, Ohtoshi E, Matsuyoshi N, Ohta K, Horiguchi Y, Imamura S

Abstract
A 41-year-old woman had erosive eruptions surrounded by irregularly shaped pigmentation on the lateral aspect of her right foot, where she had noted gradually increasing warmth and pain for 10 years. The eruptions waxed and waned without complete healing, and an ulcer which had formed one year previously did not respond to topical treatments. Arteriography performed on the right lower extremity disclosed multiple diffuse arteriovenous malformations in the right lower leg and foot. The ulcer was treated by bed rest, surgical debridement, and topical application of bucladesine sodium ointment. After three months, the ulcer healed, leaving a shallow scar and pigmentation.

Ulcers associated with arteriovenous fistula within a Stewart-Bluefarb syndrome: arterial and/or venous therapy?]

Klose J¹, Kröger K, Grabbe S, Dissemond J

Abstract
We report on a 46-year-old female patient with a 2-year history of ulceration over the dorsum of her right foot associated with a congenital arteriovenous fistula. About 12 years ago she had an ulcer at the same site. Despite an insufficient occlusion of the arteriovenous fistula after coil-embolization complete healing of the ulcer was achieved for a period of 10 years. At present hyperpigmentation could be seen surrounding the ulcer as a clinical sign for a venous insufficiency. The ulcer healed completely under a conservative therapy of the venous component of the arteriovenous fistula. The pathogenesis and therapy of ulcers associated with arteriovenous fistula within a Stewart-Bluefarb syndrome are discussed in this case report.
Stewart-Bluefarb syndrome: Report of five cases and a review of literature.

Paras K, O'Connor AG, Bester L.

Abstract
Stewart-Bluefarb syndrome is a rare angioproliferative disorder characterised by acroangiodermatitis associated with an underlying arteriovenous shunt. This condition should be differentiated from acroangiodermatitis of Mali classically described in association with chronic venous insufficiency. Patients with Stewart-Bluefarb syndrome typically present with lower leg pigmented macules, papules and plaques that can coalesce to form larger confluent patches of pigmentation. Recognition of Stewart-Bluefarb syndrome may be difficult or delayed as the cutaneous manifestations may resemble a variety of other dermatological conditions. Most commonly, acroangiodermatitis may be confused with Kaposi's sarcoma and the condition is often referred to as 'Pseudo-Kaposi's sarcoma'. Acroangiodermatitis may also resemble or coexist with pigmentation of chronic venous insufficiency. As seen in this report, acroangiodermatitis may also be clinically confused with the 'cavernous' form of a capillary malformation. Here, we describe five patients with Stewart-Bluefarb syndrome. In one female and two male patients the diagnosis was delayed as the acroangiodermatitis closely resembled other conditions. All underlying arteriovenous communications were initially diagnosed on duplex ultrasound and confirmed with magnetic resonance angiography. Four patients were found to have a congenital arterio-venous malformation while one was diagnosed with a post-thrombotic arterio-venous fistula. Management included observation and intervention using a variety of techniques including percutaneous or trans-catheter embolisation, endovenous laser, radiofrequency ablation and foam ultrasound guided sclerosis therapy. This case series highlights the challenges involved in the diagnosis and management of Stewart-Bluefarb syndrome. Given the local and systemic sequelae of high flow shunts, correct diagnosis and early detection of the underlying arteriovenous abnormality is crucial in the long-term management of these patients and in preventing the associated complications.

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Venous status ulcers due to congenital agenesis of the inferior vena cava in a 16-year-old male.

Phair J, Trestman E, Stableford J.

Abstract
We report a case of agenesis of the infrarenal inferior vena cava in a 16-year-old boy with venous ulceration in the gaiter region bilaterally. Duplex imaging was performed revealing an iliofemoral venous system. Magnetic resonance venography then confirmed this and pelvic collateralization. This patient’s condition has been successfully managed with wound care. This case is a rare example of a congenital malformation of the inferior vena cava with presenting symptoms of venous stasis ulceration in a pediatric patient.

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Conservative Therapy for Surgically Untreatable Extensive Arteriovenous Malformation from the Lower Extremity to the Pelvis with Secondary Consumptive Coagulopathy

Yoshiko Watanabe, MD, PhD, Toru Iwashashi, MD, PhD, Naozumi Saiki, MD, Nobusato Koizumi, MD, PhD, Toshiya Nishibe, MD, PhD, and Hiitoshi Ogino, MD, PhD

We present a woman with surgically untreatable extended arteriovenous malformations (AVM) and consumptive coagulopathy, which had been controlled by conservative compression and anticoagulation therapies for 17 years. At age 13, she was diagnosed with extended AVM in the entire leg and pelvis. At age 16, limited surgical resection of the enlarged superficial vein in the left calf was performed for persistent leg pain. One year later, anticoagulation therapy was performed for massive bleeding from hemorrhoids due to AVM and coagulopathy. Despite its intractability, her condition has been favorably controlled with conservative methods, including compression and anticoagulation therapies.
AVM and Compression: our experience

Compression hosiery
- 20-40 mm Hg – preferable and very safe
- Two component stocking system !!!

Compression bandages with high stiffness (inelastic bandages)
- > 20 mm Hg – hemodynamically more effective, than stockings
- Save, but they are frequently blamed for losing effectiveness over time

Elastic bandages
- After surgical interventions
- BUT be carefully!

Adjustable devices (CircAid, Velcro band, …)
- We haven’t experience. Maybe – in combined AVL malformations..
Features of secondary varicose veins in case of AVM

- Increased pressure in the superficial veins
- The AVM area is always marked congested superficial veins
- Sharp thinning of the venous walls, frequent formation of venous aneurysms
- Trophic skin changes
- *All this confirms the need for eccentric compression*
Must we perform phlebectomy in AVM? No!
Endovascular embolization of AVM

- Often embolization – multisession procedure
- Using Hystacril and 90 alcohol - a strong pain effect
- Skin reactions after embolization (important accents for compression !)
- Eccentricity of compression is the most important property
Free tissue reconstruction following right gluteal, lumbar region and upper third of the right hip malformation tissues excision

- Post-operative compression:
  standard elastic bandages & compression garment
AVM, trophic ulcer and Compression

Two component stocking system
mediven ulcer kit ®

mediven ulcer kit ® = Compression & Stiffness
Venous malformations (VM)

- Pain, oedema, reduction of QoL
- Functional impairment
- Bleeding
- Trophic disorders and venous ulcers
- Thrombosis

# VM: Treatment Options

<table>
<thead>
<tr>
<th>Truncular (T)</th>
<th>Surgery &amp; Thermo-(sclero) obliteration</th>
<th>Conservative options</th>
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<tbody>
<tr>
<td>Obstruction</td>
<td>Resection with prosthesis placement</td>
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<tr>
<td>Dilatation</td>
<td>Avulsion (phlebectomy)</td>
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<td></td>
<td>Perforant ligation</td>
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<tr>
<td></td>
<td>Sclerotherapy</td>
<td></td>
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<tr>
<td>Exatruncular (ET)</td>
<td></td>
<td><strong>Compression</strong> &amp; Vasoactive Drugs (VAD) ?</td>
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<tr>
<td>Infiltrating</td>
<td>Radical resection (?)</td>
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<td></td>
<td>Partial resection</td>
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<td></td>
<td>Sclerotherapy</td>
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<td>Thermocoagulation</td>
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<tr>
<td>Limited</td>
<td>Resection</td>
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</tbody>
</table>
Venous malformations

- Accuracy diagnosis
- No necessity for endovascular embolization
- Operations in CVI principals (reflux abolition, pathological venous volume reduction, step-by-step surgery)
- Constant compression (CCI 3)
Adequate compression in vascular malformation – hosiery CCI III

- Hypoplasia of femoral vein and superf. iliac vein
Compression & MRI findings

- Pressure 20 mm Hg - deep veins volume reduction 60 - 80 %
- Increase of pressure to 50 mm Hg - same results
- Compression of superficial veins – only 70 mm Hg

Uhl J.-F., 2011
Popliteal vein aneurism (PVA) & CT scan with compression (CCI-2)

- **30 mm Hg**
- AVP wasn’t visible
- Superficial veins were without changes !!!
## Compression & VM

<table>
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<tr>
<th></th>
<th>Pressure 20-30 mm Hg (mean)</th>
<th>Pressure &gt; 30 mm Hg</th>
</tr>
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<tbody>
<tr>
<td>Stiffness &lt; 10 mm Hg</td>
<td>P S</td>
<td>P S</td>
</tr>
<tr>
<td>Stiffness &gt; 10 mm Hg</td>
<td>p S</td>
<td>P S</td>
</tr>
</tbody>
</table>

- **1\textsuperscript{st} modality** - Pressure Increase to 30-40 mm Hg
- **2\textsuperscript{nd} modality** - Pressure 20-30 mm Hg + STIFFNESS (> 10 mm Hg)

Pressure (P)    Stiffness (S)    Pressure + Stiffness (PS)
Hemodynamic goals in VM

1. Short saphenous vein
2. Giacomini vein
3. Lateral embryonic vein
4. Atypical perforants

Stages of treatment

1. RFA of SSV
2. RFA of Giacomini vein
3. Open ligation of perforant veins
4. Avulsion of dysplastic veins
5. FF-sclerotherapy of residual veins

Eccentric compression – the most important element in postoperative treatment
Thermoobliteration procedures (Laser or RF)

Diode Laser

Cool-tip RF Ablation System (Valleylab, Covidien, USA)
Foam-form sclerotherapy

- Compression: only adhesive bandages
## VM: compression modality

<table>
<thead>
<tr>
<th>VM: post-operative period</th>
<th>Medical compression stockings (MCS)</th>
<th>Bandages</th>
</tr>
</thead>
<tbody>
<tr>
<td>VM + angiomatous tissues</td>
<td>CI II, CI III, Elastic, Inelastic</td>
<td>++</td>
</tr>
<tr>
<td>VM + severe deformity of extremity</td>
<td>++</td>
<td>++</td>
</tr>
<tr>
<td>VM + lymphatic insufficiency</td>
<td>++</td>
<td>++</td>
</tr>
<tr>
<td>VM: post sclerotherapy</td>
<td>++</td>
<td>++</td>
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<tr>
<td>VM: bleeding</td>
<td>++</td>
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Potential possibility: adjustable devices, CircAid, …
Venous procedures in VM have become less traumatic, leading to less post-interventional discomfort, pain and complications.

The main benefit of post-interventional compression in superficial veins seems to benefit more from longer and eccentric compression:

- Reduction of hematoma and edema
- Reduction of pain and discomfort
- Improvement of results (occlusion of venous cavern)
- Reduction of phlebitis and DVT

Data on reduction of recurrence rate after interventions by compression is not available.

Prolonged compression – best option in different situations.
Standard CCl3 & eccentric compression in post-treatment period (AVM, VM): pain syndrome

Visual Analog Scale (VAS)

Day 2
Day 21

p=0.039

Standard C.  Eccentric C.
Capillary malformations

Before

Compression (after palliative resection) ??:
- only light !!
- only post-operative !!

After operation
CM + VM: trophic disorders of upper limbs

Arm-sleeves CCl2 in compression treatment
## Lymphatic dysplasia

<table>
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<th>Lymphatic Malformation</th>
<th>Treatment</th>
</tr>
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<tr>
<td>Microcystic lymphangioma</td>
<td>Sclerobliteration (+++)</td>
</tr>
<tr>
<td>Diffuse lymphangioma</td>
<td>Surgical treatment (++)</td>
</tr>
<tr>
<td>Cystic hygroma</td>
<td>Sclerobliteration (++++)</td>
</tr>
</tbody>
</table>

**Conservative option:** Rapamycin (Sirolimus) – VEGF synthesis reduction, TGF-β and TNF-α inhibition 2 mg/day – 6 month
Lymphatic dysplasia

- Surgical excision + Ethanol sclerotherapy
- Multisession therapy
- Padding eccentric compression
Compression therapy remains quite an important part of the conservative approach and post-operative period to the treatment of congenital vascular malformations.

No explicit directions in terms of specific applications have been mentioned in the literature.

Considering exceptionally high variety of clinically treated malformations, any compression therapy shall be assigned individually.

High stiffness materials would be prioritized in most cases in view of the pathogenic behavior of malformations (AVM & VM).

Eccentric compression very important in majority situations.

Compression therapy remains the most controversial in the treatment of patients with capillary malformations.
THANK YOU FOR ATTENTION