Scope of the problem: Varicose veins

<table>
<thead>
<tr>
<th>Year</th>
<th>Population</th>
<th>N</th>
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Varicose veins
## Scope of the problem: Venous ulcers

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<th>Male %</th>
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Venous ulcers
Venous ulcers
Combination venous disease
Overview

- **Anatomy**
- Physiology and pathophysicsiology
- Diagnosis
  - Ultrasound
  - Functional studies
  - Venography
- Therapies
  - Deep reflux
  - Perforator reflux
  - Superficial reflux
## Terminology

<table>
<thead>
<tr>
<th>Older Term</th>
<th>New Term</th>
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<tr>
<td>Greater Saphenous or Long saphenous vein (LSV)</td>
<td>Great Saphenous Vein (GSV)</td>
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<tr>
<td>Lesser Saphenous Vein (LSV)</td>
<td>Small Saphenous Vein (SSV)</td>
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<tr>
<td>Giacomini Vein</td>
<td>Cranial Extension of the SSV</td>
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<tr>
<td>SFJ or Crosse</td>
<td>Confluence of the superficial inguinal veins</td>
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<tr>
<td>Dodd or Hunter Perforators</td>
<td>Femoral Canal Perforators</td>
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<tr>
<td>Sherman and Boyd Perforators</td>
<td>Paratibial Perforators</td>
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<tr>
<td>Posterior Arch Vein</td>
<td>Posterior Accessory GSV</td>
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<tr>
<td>Cockett Perforators</td>
<td>Posterior Tibial Perforators</td>
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<tr>
<td>Communicating veins</td>
<td>intersaphenous veins</td>
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</table>

* Nomenclature of the veins of the lower limbs: An international interdisciplinary consensus statement, Alberto Caggiati, MD, John Bergan, MD, Peter Gloviczki, MD, Georges Janetet, MD, Colin P. Wendell-Smith, MD, and Hugo Partsch, MD. *JVS* Aug 2002
Great Saphenous Vein

Confluence of superficial inguinal veins

Anterior thigh circumflex vein

Posterior thigh circumflex vein

Anterior accessory great Saphenous vein

Great Saphenous Vein

Posterior accessory great Saphenous vein
Anatomy of the SSV

Figure 3.5  The small saphenous vein. (a) Normal termination; (b) low termination; (c) high termination.
Anterior Thigh PV’s

Medial Ankle PV’s

PV’s of the Femoral Canal

Paratibial PV’s

Posterior Tibial PV’s

Posteromedial Thigh PV’s

Multi-level investigation
Perforators by region

- **Area 1**: Connect GSV in thigh to femoral vein (Hunter’s, Dodd’s, Boyd’s) – usually feed varicosities
- **Area 2**: Connect GSV and branches in calf to deep system (usually PTV) in multiple planes, usually feed ulcers
- **Area 3**: Connect SSV gastrocnemius and soleus veins – usually feed varicosities
Tibial Variation 1

Diagram showing:
- GSV or Posterior Arch Vein
- Perforating Vein
- Fascia or Gastrocnemius Aponeurosis
- Superficial Posterior Compartment
- Intermuscular Septum or Paratibial Fascia
- Tibia
- Posterior Tibial Vein & Artery
- Deep Posterior Compartment
Deep venous anatomy

- Directly parallels arterial anatomy above the knee
- Paired venous channels for every arterial channel below the knee (venae comitantes)
Overview of Venous Disorders

- Anatomy
- Physiology and pathophysiology
- Diagnosis
  - Ultrasound
  - Functional studies
  - Venography
- Therapies
  - Deep reflux
  - Perforator reflux
  - Superficial reflux
Venous valvular function

• In the normal vein, valves prevent the reflux of blood following muscle pump contraction

• Abnormal valve function results in reflux
  – Primary valvular insufficiency (pregnancy, obesity, heredity)
  – Secondary valvular insufficiency (post-phlebitic syndrome)
Venous function: Muscle pumps

- Veins contain two thirds of all circulating blood
- In the lower extremities, the muscle pumps are the “peripheral heart” of the venous system, returning blood to the right atrium against gravity
- 4 Muscle pumps: foot, calf, popliteal, thigh
Veins and How They Work
Venous function: Muscle pumps

Figure 4.1  Pressure profiles in the veins of the foot, calf, popliteal fossa and upper thigh at rest and on walking. Foot venous pressure is progressively reduced by contractions of the calf muscle pump whose excursions of pressure are significantly greater than those of the popliteal or ‘groin pumps’. The heavy black arrows indicate intramuscular pressures.
Venous function: Muscle pumps

- Calf muscle pump is most important, with greatest capacity and pressure profile.

- Components:
  1. Dilated valveless sinusoids w/in gastrocnemius and soleus
  2. Direct perforator veins
  3. Valves directing blood (superficial to deep; distal to proximal)
  4. Fascia surrounding muscles transmits high IM pressures to venous system
What’s This?

No calf pump activation!
Classification of venous disease severity: CEAP

Box 6.1  CEAP classification of lower limb chronic venous disease

<table>
<thead>
<tr>
<th>C</th>
<th>for Clinical signs (grade 0–6), supplemented by (A) for asymptomatic and (S) for symptomatic presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>E</td>
<td>for Etiologic classification – congenital (E_C), primary (E_P), secondary (E_S)</td>
</tr>
<tr>
<td>A</td>
<td>for Anatomic distribution – superficial (A_S), deep (A_D), or perforator (A_P), alone or in combination</td>
</tr>
<tr>
<td>P</td>
<td>for Pathophysiolgic dysfunction – reflux (P_R) or obstruction (P_O), alone or in combination</td>
</tr>
</tbody>
</table>

**Clinical classification**
- Class 0: No visible or palpable signs of venous disease
- Class 1: Telangiectases or reticular veins
- Class 2: Varicose veins
- Class 3: Edema
- Class 4: Skin changes ascribed to venous disease (e.g., pigmentation, lipodermatosclerosis, venous eczema)
- Class 5: Skin changes as in class 4 with healed ulceration
- Class 6: Skin changes as in class 4 with active ulceration

**Etiologic classification**
- Congenital (E_C)
- Primary (E_P) – with undetermined cause
- Secondary (E_S) – with known cause
  - post-thrombotic
  - post-traumatic
  - other

**Anatomic classification**
- *Superficial veins (A_S, D):*
  - Segment 1: Telangiectases/reticular veins
  - Great saphenous vein
- *Deep veins (A_D, S):*
  - Segment 6: Inferior vena cava iliac
  - Segment 7: Common
  - Segment 8: Internal
  - Segment 9: External
  - Segment 10: Pelvic – gonadal, broad ligament, other
  - Femoral
  - Segment 11: Common
  - Segment 12: Deep
  - Segment 13: Superficial
  - Segment 14: Popliteal
  - Segment 15: Crural – anterior tibial, posterior tibial, peroneal (all paired)
  - Segment 16: Muscular – gastrocnemial, soleal, other

**Perforating veins (A_P, D, S):**
- Segment 17: Thigh
- Segment 18: Calf

**Pathophysiolgic classification**
- Reflux (P_R)
- Obstruction (P_O)
- Reflux and obstruction (P_R, O)
Clinical classifications of CEAP

0. No venous disease
1. Spider veins
2. Varicose veins
3. Edema
4. Lipodermatosclerosis
5. Healed ulcers
6. Active ulcers
C1: Telangectasia
C2: Varicose veins
C4: Skin changes

- Telangiectasias
- Atrophie blanche
- Pigmentation
C5, C6 Venous ulceration

C5: Healed ulcer

C6: Active ulcer
Overview of Venous Disorders

• Anatomy
• Physiology and pathophysiology
• **Diagnosis**
  – Ultrasound
  – Functional studies
  – Venography
• Therapies
  – Deep reflux
  – Perforator reflux
  – Superficial reflux
Anatomic venous diagnosis: Duplex ultrasound

1. Scan **entire venous system** from groin to ankle

2. Exam should be focused on identifying thrombus, **reflux**, or both

3. Thrombus: hyperechoic, **incompressible**

4. Reflux: identified with **patient standing**, and/or **valsalva**
Venous Insufficiency, Reflux

- **Reflux** = > 0.5 seconds
- **Positioning**: hydrostatic pressure is the key: the patient should be standing (90 mmHg for 6’ pt) with weight on contra-lateral leg, slant table is also adequate – 66% false negative if done supine
Venous Insufficiency, Reflux

- **Color Flow:**
  - normal antegrade flow is **BLUE**
  - abnormal retrograde flow is **RED = REFLUX**
  - normal antegrade flow is toward the heart: from right side of screen to the left side of the screen

Image courtesy of Olivier Pichot, MD
Duplex – Mickey Mouse View of SFJ
The “most” significant landmark: The Saphenous Compartment

Hyper-echoic saphenous fascia

“Egyptian eye”

Contains: saphenous veins, and nerves

NOTE: Saphenous tributaries, collateral and communicating veins lie external to this compartment

The superficial fascia that covers the GSV has been named “saphenous fascia,” and the compartment in which it runs, “saphenous compartment.”
Saph Compartment “sheath”

Multi-level investigation
Great Saphenous Variations – Saphenous Sheath and Tributaries

Ricci and Georgiev - *Journal of Vascular Technology*
Overview of Venous Disorders

• Anatomy
• Physiology and pathophysiology
• **Diagnosis**
  – Ultrasound
  – **Functional studies**
  – Venography
• **Therapies**
  – Deep reflux
  – Perforator reflux
  – Superficial reflux
Functional venous diagnosis: Ambulatory venous pressures

1. Measure baseline VP in foot or ankle in standing position. Reflects distance from RA to foot (avg. 80-90mmHG)

2. Exercise (toe raises)

3. Measure VP with exercise (nl 25 mmHG)

4. Measure VP recovery time (nl 25-30 sec, severely abnl <5 sec)
The common denominator in the pathophysiology of venous disease.

Instead of dropping, the intravenous pressure rises during exercise.

May be due to reflux, obstruction, pump failure or a combination of the above.
Functional venous diagnosis: Ambulatory venous pressures

Figure 4.2  Normal and abnormal foot venous pressure profiles. The lower trace, deep venous incompetence results in pressure swings with very little fall in mean pressure and a rapid return to high resting pressure.
Ambulatory Venous Hypertension: Causes

Muscle pump failure
  primary
  secondary

Venous obstruction
  thrombotic
  non thrombotic

Venous valvular incompetence
  primary / segmental
  secondary / entire vein
Overview of Venous Disorders

• Anatomy
• Physiology and pathophysiology

• **Diagnosis**
  – Ultrasound
  – Functional studies
  – Venography

• Therapies
  – Deep reflux
  – Perforator reflux
  – Superficial reflux
Anatomic venous diagnosis: Venography

- **Ascending venography**
  - Used primarily to evaluate for clot
  - Needle placed in foot and dye followed from distal to proximal
Anatomic venous diagnosis: Venography

- **Descending venography**
  - Used primarily to evaluate for reflux
  - Sheath placed in CFV and dye followed from proximal to distal to evaluate valve location and function
Overview of Venous Disorders

• Anatomy
• Physiology and pathophysiology
• Diagnosis
  – Ultrasound
  – Functional studies
  – Venography
• **Therapies**
  – Deep reflux
  – Perforator reflux
  – Superficial reflux
Deep vein reconstruction for reflux

• **Workup and evaluation:**
  1. Clinical indications: C4-C6 in CEAP
  2. Noninvasive lab: identify site and severity of reflux, including PPG or AVP to determine relative importance
  3. Ascending and descending venography for anatomic delineation
Deep vein reconstruction for reflux

1° Reflux:
- Valvuloplasty or valve repair
- Identify site of most severe reflux
- Treat one site only
Deep vein reconstruction for reflux

- **20 Reflux (PPS):**
  - Vein-Valve transplant
  - Venous transposition
Overview of Venous Disorders

• Anatomy
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• Diagnosis
  – Ultrasound
  – Functional studies
  – Venography
• **Therapies**
  – Deep reflux
  – **Perforator reflux**
  – Superficial reflux
Perforator interruption procedures

• **Indications:**
  – Advanced CVI (CEAP C4 – C6: skin changes to active ulcers)

• **Contraindications:**
  – PVD, DM, morbid obesity, connective tissue disorder, severe deep venous reflux
Perforator interruption procedures: Linton procedure

- Linton procedure
  - Incision(s) along medial calf with identification and ligation of perforators
  - Fraught with complications, mostly wound related
  - Cure is worse than the disease
Perforator interruption procedures: 
SEPS

- **SEPS (Subfascial endoscopic perforator surgery)**
  - Use of endoscopic technique avoids problem of long incision through most damaged skin and soft tissue
  - Limited ability to treat important Cockett I perforators
  - Results not as good as hoped
Perforator interruption procedures: Minimally invasive techniques

• The application of techniques such as RF and foam, previously successfully applied in the treatment of superficial reflux, will likely revolutionize the therapy of perforator reflux
  • No incisions or dissection
  • Able to reach even inframalleolar perfs
## Ulcer healing with foam perforator ablation

<table>
<thead>
<tr>
<th></th>
<th>N</th>
<th>2 wks</th>
<th>4 wks</th>
<th>6 wks</th>
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<td>9</td>
<td>1 (11%)</td>
<td>1 (11%)</td>
<td>8 (100%)</td>
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<td><strong>Foam</strong></td>
<td>16</td>
<td>12 (75%)</td>
<td>16 (100%)</td>
<td>16 (100%)</td>
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</table>

*Bergan et al, Ann Vasc Surg Jan 06*
Overview of Venous Disorders

- **Anatomy**
- **Physiology and pathophysiology**
- **Diagnosis**
  - Ultrasound
  - Functional studies
  - Venography
- **Therapies**
  - Deep reflux
  - Perforator reflux
  - **Superficial reflux**
Varicose Veins - Cause

REFLUX

• Normal = no “backflow” through one way valves
• Vessel bulging = valves can’t close
• Valve damage = valves don’t close
Varicose Veins – Greater Saphenous Distribution

- Most common finding
- Varicosities along the medial thigh and calf
- 10% of patients are at risk of ulceration
- Skin changes seen along the medial ankle
Varicose Veins – Lesser Saphenous Distribution

- Less frequent than Greater Saphenous
- Varicosities may be seen on the posterior calf and lateral ankle
- Skin changes seen along the lateral ankle
Varicosities due to pelvic or Giacomini reflux

- Begin during pregnancy
- Increased symptoms during pre-menstrual period and after intercourse
- May be associated with pelvic congestion syndrome
Treatment Options

- Compression therapy
- Surgery
- Endovenous occlusion
- Sclerotherapy
Elastic compression stockings

- Must be graduated
- Replace q 6 months
- Calf high generally sufficient to improve venous outflow
- Use custom model for atypical leg shapes
Compression therapy

- Reduces symptoms of aching, fatigue, pain, and swelling
- Increases fibrinolytic activity
- Increases TCpO2
- Mainstay of treatment for venous ulcers
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<th>Compression Strength</th>
<th>Indications</th>
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<td>8-15mm</td>
<td>Leg fatigue, mild swelling, stylish</td>
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<tr>
<td>15-20mm</td>
<td>Mild aching, swelling, stylish</td>
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<tr>
<td>20-30mm</td>
<td>Aching, pain, swelling, mild varicose veins</td>
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<tr>
<td>30-40mm</td>
<td>Aching, pain, swelling, varicose veins, post-ulcer</td>
</tr>
<tr>
<td>40-50, 50-60mm</td>
<td>Recurrent ulceration, lymphedema</td>
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</table>
Surgery: High ligation and stripping

- For decades, this surgical technique was the standard of care in the treatment of symptomatic superficial venous reflux
- Used in conjunction with surgical removal of varicosities
- Long term results were acceptable, with approximately 20% recurrence at 5 years
GSV High L & S: Indications

• Symptomatic reflux C 2- C6 refractory to conservative measures
  • Pain, swelling, fatigue
  • Phlebitis
  • Ulceration
GSV High L & S: Contraindications

- Deep venous obstruction
- GSV thrombosis
- Severe deep system reflux
- PAD
ESCHAR trial: Role of Surgery in patients with venous ulcers

- 500 patients with active or recent ulceration randomized to GSV or SSV L & S vs. compression alone
- No difference in ulcer healing between the 2 groups
  - 65% @ 24 weeks
- Ulcer recurrence was twice as high in the compression alone group

Ligation & stripping technique
Why endovenous therapy for superficial reflux?

• The use of endovenous therapy eliminates the shortcomings of high L & S while providing excellent therapy

• Compared to high L & S:
  – Fewer local complications
  – No scarring
  – Dramatically reduced recovery time
  – Local anesthesia
  – Equivalent short and long term results
Types of endovenous therapy: RFA and EVLT

• Similarities
  – Generator + catheter set up
  – Percutaneous access
  – Local anesthesia + tumescence (outpatient)
  – Recovery less than 24 hours
  – Staged treatment of varicosities as needed
Types of endovenous therapy: RFA and EVLT

- **Radiofrequency ablation**
  - Uses radiofrequency energy to heat vein walls, coagulating protein and collapsing/sealing vein
  - Single manufacturer (VNUS Medical Technologies, Inc.)
  - Uniform treatment and research parameters
  - Has been assessed in two RCTs comparing RFA to high ligation and stripping
Types of endovenous therapy: RFA and EVLT

- EVLT (endovenous laser therapy)
  - Mechanism of action more complex: laser energy heats blood, with release of steam from bubbles coagulating protein and collapsing/sealing vein
  - Multiple manufacturers and types of laser (*including laser type, wavelength, and continuous vs. pulse mode*)
  - Diverse treatment and research parameters
Indications for endovenous ablation

- **Superficial system reflux causing**
  - Pain with or w/o varicosities (heaviness, fatigues, aching throbbing, etc.)
  - Edema
  - Ulceration
  - Superficial phlebitis

- **Perforator reflux causing**
  - Ulcers
  - Contributing to painful varicosities and edema persisting AFTER GSV ablated
Contraindications to EVA

- Obstructive component to venous hypertension (post DVT)
- Tortuosity
- Deep reflux is NO LONGER a contraindication
“Egyptian Eye” = treatment target

- Use u/s to cannulate in long or short view: KNOW BOTH
- Identify tip of catheter/fiber 1cm below SFJ
- Under u/s guidance provide tumescent anesthesia
  - 1cm deep, “halo” w/in sheath
  - Reconfirm cath/fiber tip
Post-treatment evaluation

- **Immediate and @ 72 hrs:**
  - Check for clot at SFJ
  - Check for closure of target vein

- **Long term follow up:**
  - Evaluate for continued closure (6 mos and yearly)
  - Regression (early)/progression (late) of reflux in other venous segments (SSV and SSV CE, deep system)
GSV Closure by Radiofrequency Ablation
GSV Ablation by Laser

Image courtesy of Nick Morrison, MD and Diana Neuhardt, RVT
Clinical results after RFA

Merchant 2005 multicenter trial

Clinical benefits persist @ 4 Y despite duplex findings
## Endovenous Ablation: Comparison of short term success

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<td>Hingorani 2004</td>
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<td>73</td>
<td>96%</td>
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<td>Shortell 2005</td>
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<tr>
<td>Min 2003</td>
<td>EVLT</td>
<td>499</td>
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<td>0.7%/2%</td>
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Endovenous ablation: Comparison of intermediate and long term success

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<tr>
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<td>2 ry</td>
<td>EVLT</td>
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## Complications: Comparison of short term outcomes†

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<th>One year</th>
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<td>EVLT</td>
<td>RFA</td>
<td>EVLT</td>
<td>RFA</td>
<td>EVLT</td>
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<td>15%</td>
<td>0.4%</td>
<td>0</td>
<td>0</td>
<td>0</td>
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<tr>
<td>Induration</td>
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<td>25%</td>
<td>0</td>
<td>2%</td>
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<td>0</td>
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<tr>
<td>Dysesthesia</td>
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<td>20%</td>
<td>9%</td>
<td>3%</td>
<td>4%</td>
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<td>2%*</td>
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<td>0</td>
<td>0</td>
<td>0</td>
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<tr>
<td>Edema</td>
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<td>18%</td>
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*only w/early experience

†composite data from multiple studies
## Complications: Comparison of venous thromboembolic events

<table>
<thead>
<tr>
<th>Study</th>
<th>Type</th>
<th>N</th>
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<th>CFV CE</th>
<th>PE</th>
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<td>1 (0.3%)</td>
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<td>Shortell 2005</td>
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<td>4 (2%)</td>
<td>0</td>
</tr>
<tr>
<td>Min 2001</td>
<td>EVLT</td>
<td>499</td>
<td>0</td>
<td></td>
<td>0</td>
</tr>
<tr>
<td>Anastasie 2003</td>
<td>EVLT</td>
<td>232</td>
<td>2 (0.9%)</td>
<td></td>
<td>0</td>
</tr>
<tr>
<td>Proebstle 2003</td>
<td>EVLT</td>
<td>37*</td>
<td>1 (2.7%)</td>
<td></td>
<td>0</td>
</tr>
</tbody>
</table>

*SSV
RFA vs EVLA: RECOVERY Trial

- Multi center RCT single blinded RFA vs. EVLA (980nm), 69 pts, 87 limbs
- F/U 2, 7, 14, 30 days p-op
- Findings
  - Success rate 100% both groups
  - RF associated with reduced pain, bruising, tenderness
RFA vs EVLA: RECOVERY Trial

– Study Limitations

• Single blind – investigators knew which therapy patients received
• Only one laser tested
• Industry sponsored by VNUS, manufacturer of RF catheter
Additional issues: *Options for managing associated varicosities*

- Treat **concomitantly** with GSV/SSV (few)
  - General/regional anesthesia for ablation
  - Very large, numerous vv
  - Pt lives far away

- Treat in **staged** fashion (most)
  - Local anesthesia for ablation
  - Average size and # of vv
  - Follow up easy
Natural history of varicosities post-ablation

- Most patients have involution of largest varicosities
- Many have complete involution of all varicosities
- Some areas respond better than others

<table>
<thead>
<tr>
<th>Location/type of vv</th>
<th>Pts with resolution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Largest varicosities</td>
<td>28%</td>
</tr>
<tr>
<td>All veins</td>
<td>13%</td>
</tr>
<tr>
<td>Medial thigh</td>
<td>47%</td>
</tr>
<tr>
<td>Below knee</td>
<td>30%</td>
</tr>
<tr>
<td>Posterior</td>
<td>4%</td>
</tr>
</tbody>
</table>

Monahan et al, JVS 2005;42
Additional issues: Options for managing associated varicosities

Staged treatment options

- Stab avulsion
- Sclerotherapy
  - Saline
  - Chemical (sotradecol, polidecol)
  - Foam (sotradecol, polidecol)
- No treatment
Surgical Treatment of Varicose Veins: Phlebectomy

- Very esthetic method of removing varicose veins
- Usually requires only local anesthetic
- Especially useful for branches of GSV, LSV
Ultrasound-guided sclerotherapy

- Highly technical procedure
- Requires advanced ultrasound skill
- Extremely versatile – may be used for nearly all veins
- Efficacy enhanced with foamed solutions: Sotradecol
Ultrasound-guided Sclerotherapy

- Nearly any size vein can be treated
- Needle location inside vein, as well as movement of sclerosant and response of vein (spasm) visible
- Efficacy enhanced with foamed sclerosant
Sclerotherapy of Telangiectasias: Technique

Injection of colorless solution creates illusion of vein disappearance; damage to endothelium leads to fibrosis of vein
Summary of venous disorders

- Disorders of the venous system range from cosmetic to limb-threatening in nature.
- Understanding of the unique hemodynamics and pathophysiology of the venous system are key to developing strategies for therapy.
- As with arterial disease, the use of endovenous techniques is increasing our ability to treat patients effectively.
Low Flow Venous Malformations, Angiomas, and Klippel-Trenaunay Syndrome
Background

• Vascular malformations
  – spectrum of disorders ranging from minimal to significantly disabling conditions impacting patient’s anatomic, functional and emotional integrity

• Incidence: VMs = 1.2-1.5%
  – 2/3 are low flow

• Etiology: unknown, but genetic predilection

• Classification and diagnosis controversial

• Treatment:
  – Small lesions may be cured
  – Extensive lesions therapy palliative/goal oriented
Classification Challenges

- Inconsistent, archaic and often contradictory nomenclature for low flow venous malformations:
  - hemangioma
  - cavernous hemangioma\(^1\)
  - birthmarks (naevi)
  - port-wine stains\(^2\)
  - angiomas

1) Archaic term cavernous hemangioma is still used rather than venous malformations (VM) (Rutherford, Vascular Surgery.2000;1636)
2) The 19th century expression for capillary malformations (CM) (Rutherford, Vascular Surgery.2000;1633)
All Vascular Anomalies

TUMORS

- Infantile HEMANGIOMA most common type

VASCULAR MALFORMATIONS (VM)

- HIGH FLOW (arterial component)
- LOW FLOW (venous and/or lymphatic components)
LFVMs: Clinical Presentation and Dx

- LFVMs are developmental errors in vasculogenesis leading to structural and functional anomalies
- Present at birth but often not clinically apparent until later in life
- Symptoms: skin discoloration, varicosities, pain, decreased mobility, swelling, bleeding, osteomuscular hypertrophy
LFVMs: Clinical Presentation and Dx

• Isolated or part of a syndrome (KTS, Proteus, Maffucci, Sturge-Weber etc.)
• Affects both superficial and deep underlying anatomic structures (skin, muscles, abdominal viscera, CNS)
• Management overlaps borders of different subspecialties
• **Multidisciplinary approach** is fundamental for proper diagnosis

• **Diagnostic (imaging) modalities:**
  - *Ultrasound*
  - *MRI*
  - *Arteriogram*
Treatment Options

- Surgical resection

- Sclerotherapy
  - Ethanol (USA)
  - Polidocanol (Europe, not FDA approved in the US)
  - *Sodium Tetradecyl Sulfate (STS, Sotradecol®)*
Treatment Options

• Surgical resection:
  - Most effective for encapsulated and microvascular lesions
  - Diffuse, deep, and macrovascular lesions are not amenable to surgical excision d/t risk of hemorrhage and damage to vital structures
Treatment Options

- Ethanol sclerotherapy (ES):
  - Limitations:
    - use in pediatric patients
    - general anesthesia required for all patients
  - Side effects:
    - Severe pain, EtOH toxicity, ulceration at injection site, ischemic bullae, deep vein thrombosis, tissue fibrosis, peripheral nerve palsy, pulmonary embolism and pulmonary hypertension
Treatment Options

• Sodium Tetradecyl Sulfate (STS):
  ▪ Detergent sclerosant
  ▪ FDA approved since 1946
  ▪ Brand name: Sotradecol®
  ▪ Often used for varicose veins and telangiectasias
  ▪ **Microfoam** offers best visualization under US, prevents sclerosant dilution by intrallesional blood, maximizes endothelial exposure
Treatment Options

• STS Foam Sclerotherapy (STS FS):
  - Side effects:
    ▪ Allergic reactions (from urticaria to anaphylaxis)
      Incidence 0.2%-0.3%
    ▪ Hyperpigmentation
      Incidence parallels that of other sclerosing agents
    ▪ Extravasation necrosis
Our Approach

• Multidisciplinary team
  Plastic Surgery (adult and pediatrics), ENT, Diagnostic Radiology, Interventional Radiology, Dermatology, Ophthalmology and Vascular Surgery

• All patients undergo MRI

• Arteriogram if suspicion of high flow on MRI

• Treatments in the office setting under local anesthesia

• General anesthesia for pediatric patients
Our Approach

- **Goals** are preset with each patient individually
- Successful accomplishment of these goals marks the completion of treatment
Our Approach

- Foam produced by **Tessari** method
- Procedures are performed under both **ultrasound** and direct visual guidance
- Injected areas are elevated for a minimum of 10 minutes, **compressed and wrapped** (compression remains on for 7 days)
- **General anesthesia** for pediatric pts
Clinical Cases

Case 1

• 13 year old female c/o right leg swelling, discoloration, varicosities and severe discomfort with exertion. Lesion present from birth but progressive. Referred by dermatologist.
• **Diagnosis:** Klippel-Trenaunay Syndrome (KTS)
• **Initial treatment:** Ethanol Sclerotherapy
  - complicated by popliteal DVT
• **Subsequent therapy:** STS foam sclerotherapy (*goals preset as decreased pain, swelling, increased cosmesis, mobility*)
  - No side effects
• **Outcome:** 100% goal achieved after 4 treatments, no complications
MRI before STS FS
MRI after STS FS
Clinical Cases

Case 2

- 7 yo male with right leg pain, varicosities, discoloration, decreased mobility, bleeding since birth
- **Diagnosis**: Klippel-Trenaunay Syndrome (KTS)
- **Initial treatment**: Ethanol sclerotherapy (outside facility)
- **Subsequent treatment**: STS foam sclerotherapy (*goals set at increased mobility, decreased bleeding risk @hockey*)
- General anesthesia due to age
- **Outcome**: symptoms improved after two treatments and able to play hockey, no complications
Case 3

- 53 year old male with long history of left face and left ear vascular malformation
- Diagnosis: Low flow vascular malformation
- Initial treatments: multiple surgical resections over 20 years
- Subsequent treatment: STS Foam Sclerotherapy (*goals set primarily for cosmesis, mild pain relief*)
- Outcome: symptoms improved after the first treatment, no complications