Localized intravascular coagulopathy in venous malformations

Prof. I. Baumgartner
Localized intravascular coagulopathy (LIC)

- Subset VM exhibit localized LIC
  ➔ pain, thrombosis & excessive bleeding during surgical procedures

- LIC can progress to disseminated intravascular coagulopathy (DIC) and life-threatening hemorrhage even when coagulation parameters (PT, INR, PTT) are normal
Coagulation disorders in venous malformation

**LIC**
- D-dimer >1,000 ng/mL a/o
- fibrinogen <200 mg/dL
- incidence between 42% and 88%
- lesion size ($P<0.001$), presence of phleboliths ($P=0.005$)

**DIC**
- conversion of LIC to DIC
- consumption of platelets and coagulation factors
- increase in PT & decrease in FV earliest signs
- **bleeding**

Arch Dermatol. 2008 Jul;144(7):861-7; Pediatric (AVWS) Radiology 2015, 45 (11), 1690-1695
Localized intravascular coagulopathy (LIC) and lesion size

- Patients with LIC
- Patients without LIC

Small: < 250 cc (approximately 8 cm in diameter)
Medium: 250 - 500 cc (approximately 10 cm in diameter)
Large: > 500 cc in volume

Pediatric Radiology 2015, 45 (11), 1690-1695
Association of LIC with venous malformations

- Prospective, consecutive series
  140 patients with VM
    59 (42%) high D-dimer levels
    6 (4.3%) low fibrinogen (85-176 mg/dL), 1 (0.7%) < 100 mg/dL

large size (>10 cm²) sig. associated with positive D-dimer test, independently of localization
  59% large VMs (>10 cm²) vs 23% small VMs with high D-dimer (p<.001)

none exhibited PE

Arch Dermatol. 2008 Jul;144(7):873-77
Management algorithm

- ASS low efficacy (vs. Kasabach-Merritt phenomenon, platelets not involved in LIC)
- OAC decrease coagulation factors, not sufficient to prevent thrombin formation in LIC
# Sclerotherapy complications

N=127, sclerotherapy for venous malformation

4 (3.1%) severe complications, all related to coagulopathy

<table>
<thead>
<tr>
<th>Complication</th>
<th>Management</th>
<th>N</th>
<th>Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Skin damage</td>
<td>Not necessitated</td>
<td>8</td>
<td>I</td>
</tr>
<tr>
<td>Pain or swelling or both for &gt; 2 weeks</td>
<td>Analgesics and/or compression garment</td>
<td>6</td>
<td>I</td>
</tr>
<tr>
<td>Thrombophlebitis outside the malformation</td>
<td>Not necessitated</td>
<td>3</td>
<td>I</td>
</tr>
<tr>
<td>Unusual swelling and loss of range of joint motion</td>
<td>Analgesics</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pain, swelling, skin damage, and loss of range of motion</td>
<td>Physiotherapy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Digital loss of range of joint motion</td>
<td>Analgesics, physiotherapy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Skin damage, infected</td>
<td>Physiotherapy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intra-abdominal bleeding, mild</td>
<td>Antibiotics, per oral</td>
<td>4</td>
<td>II</td>
</tr>
<tr>
<td>Deep vein thrombosis at ankle level</td>
<td>Corticosteroids, anticoagulants</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Skin damage</td>
<td>Blood transfusion (one unit)</td>
<td>1</td>
<td>II</td>
</tr>
<tr>
<td>Bleeding at injection site, skin damage and sepsis</td>
<td>LMWH medication</td>
<td>1</td>
<td>II</td>
</tr>
<tr>
<td>Intra-abdominal bleeding (6 liters)</td>
<td>Skin graft under local anesthesia</td>
<td>2</td>
<td>IIIa</td>
</tr>
<tr>
<td>Bleeding at injection site, sepsis, abscess, worsening of DIC</td>
<td>Transfusions of blood and coagulation factors,</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intra-cerebral hemorrhage and exitus</td>
<td>i.v. antibiotics, abscess drainage, ICU treatment</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bleeding at injection site, skin damage and sepsis</td>
<td>and prolonged hospitalization</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intra-abdominal bleeding (6 liters)</td>
<td>Intermediate care-unit treatment, i.v. antibiotics,</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>long hospitalization</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Blood transfusion, intra-arterial embolisation,</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>ICU treatment</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Phlebology 2015, October, 1-15**
Rivaroxaban for treatment of LIC in venous malformation (case report)

LMWH recommended in LIC: invasive procedures, active bleeding, very low fibrinogen levels (\(0.5–1.0\) g/L) associated with a bleeding diathesis

Evolution of markers of intravascular coagulation

J Thromb Thrombolysis (2014) 38:121–123; Blood Coagulation and Fibrinolysis 2015, 26:00–00
Conclusion

Higher VM severity scores associated with more severe LIC

Sclerotherapy, surgery or pregnancy can trigger conversion to DIC, with bleeding related to factor consumption

Prophylactic dose of LMWH (100 IE/kg) recommended in LIC with recurrent pain and before invasive treatment

Therapeutic dose of LMWH is recommended in LIC with DVT

NOACs seem to have comparable efficacy and might become an ideal oral treatment alternative to LMWH
Localized intravascular coagulopathy in venous malformations

Prof. I. Baumgartner

Clinical & Interventional Angiology, Swiss Cardiovascular Center, University Hospital Bern