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The term fast-flow or high-flow congenital vascular malformations originated by John Mulliken [1, 2] is based on the Hamburg classification and lesion flow characteristics as in the International Society for the Study of Vascular Anomalies (ISSVA) classification of 1996 [3, 4]. The details are:

- (a) Truncular arterial malformations (AM): aneurysms, coarctation, ectasias, stenosis, and arteriovenous fistulas (AVF)
- (b) Extratruncular arteriovenous malformations (AVM): complex combined regional syndromes

Truncular arterial malformations develop in the late phase of embryonic development, and they have a much better prognosis in terms of recurrence than the extratruncular arteriovenous malformations.

These arterial malformations occur as aneurysms, coarctations and stenosis, or ectasias. The surgical repair applies regular techniques of reconstructive vascular surgery for tangential resection of an aneurysm [5], resection of

the aneurysm and replacement by autologous interposition graft (Fig. 37.1), autologous venous patch graft plastic to treat an arterial stenosis, a coarctation (Fig. 37.2b) or an alloplastic bypass graft to treat several stenoses, or a long-distance stenosis in a coarctation (Fig. 37.2a) [6].

Congenital AVFs of major named vessels are also treated by standard vascular surgery techniques as, e.g., interruption of the venous connection and reconstruction of the arterial defect by an alloplastic interposition graft (Fig. 37.3). AVFs of larger peripheral vessels can be treated by interventional catheter techniques as well as by vascular surgery: ligation and interruption of each AV fistula (Figs. 37.4 and 37.5) [7].

Extratruncular AVMs represent completely different challenges. They are an active lesion, and they have a high tendency to progress and to worsen and to reexpand after treatment. Truncular ones are cured after treatment. Before an indication for treatment of AVMs is worked out, it is mandatory to classify the clinical findings following the Schobinger classification of AVM (Table 37.1). This classification was worked out to assess AVM lesions in different clinical stages and clinical conditions objectively based on the patient's clinical status to select the best-suited time for treatment as a practical guideline [8]. Most (77 %) stage I lesions progress to a higher stage by adulthood. It exists as an increased risk

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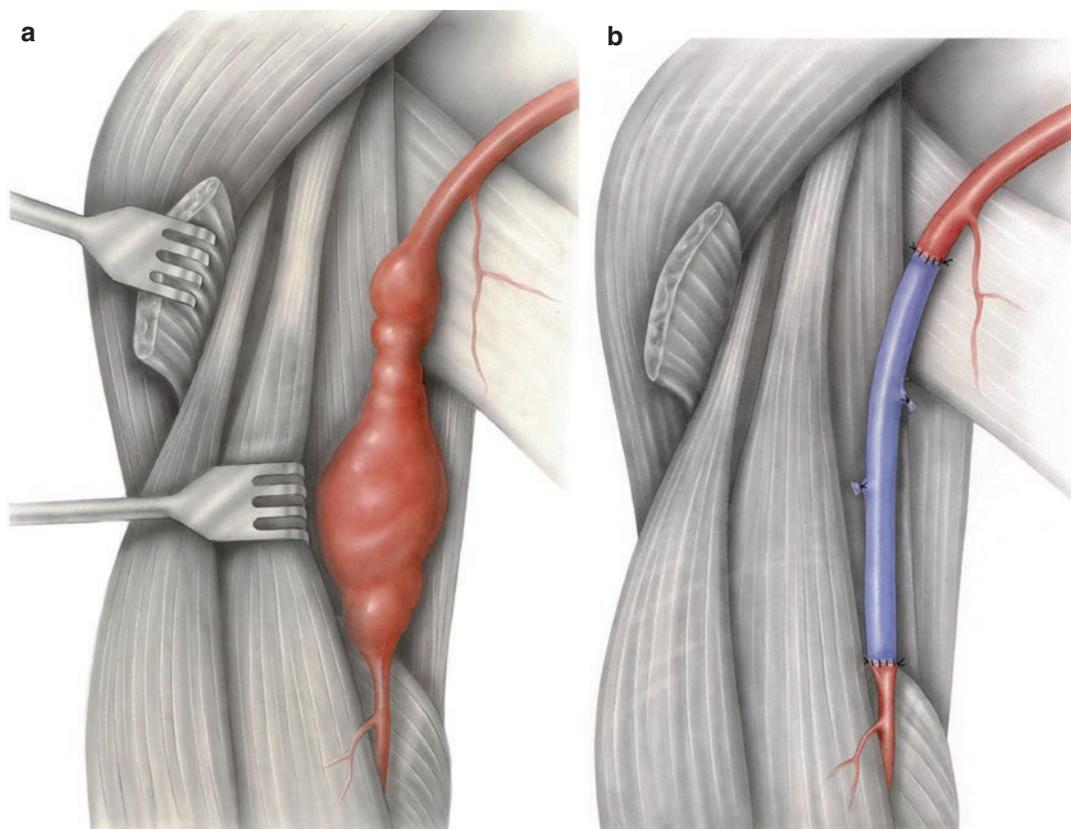


Fig. 37.1 (a) Truncular arterial malformation (AM): aneurysm of the brachial/axillary artery. (b) Resection of the aneurysm and reconstruction by autologous venous interposition graft

58 during puberty. Pregnancy does not increase risk
59 of progression for stage I AVMs.

60 The primary option for treatment of extratruncular
61 AVMs are interventional catheter techniques. These
62 follow best the recommendations given by the
63 arteriographic classification [8] proposed for
64 extratruncular AVM lesions based on the
65 arteriographic findings of the “nidus.” Such
66 better management and predicting of the
67 outcome of endovascular treatment is possible.

68 However, in several cases sole vascular surgery
69 is indicated or more often in combination
70 with interventional treatment [7–9]. Resection
71 plus embolization results in a better control
72 compared to embolization alone. Early intervention
73 for stage I lesions may give improved long-term
74 control.

For example, when localized infiltrating
75 AVMs cannot be treated by direct puncture
76 sclerotherapy or by catheter embolization (Fig.
77 37.6), the en bloc resection after precise interrup-
78 tion of the afferent and efferent vessels is one
79 option. In cases with secondarily dilated venous
80 plexus by AVM, again the principal vessels
81 (artery and vein) have to be surgically liberated
82 from the fistulous communications precisely
83 (Fig. 37.7). In addition the secondarily dilated
84 veins have to be reduced and/or resected [10, 11].
85

86 A combined treatment means first interven-
87 tional embolization followed by surgical excision
88 of the localized AVM, sometimes together with
89 adjacent tissues (Fig. 37.8) [12].

90 Infiltrating AVM very often cannot be treated
91 by direct puncture or by catheter embolization

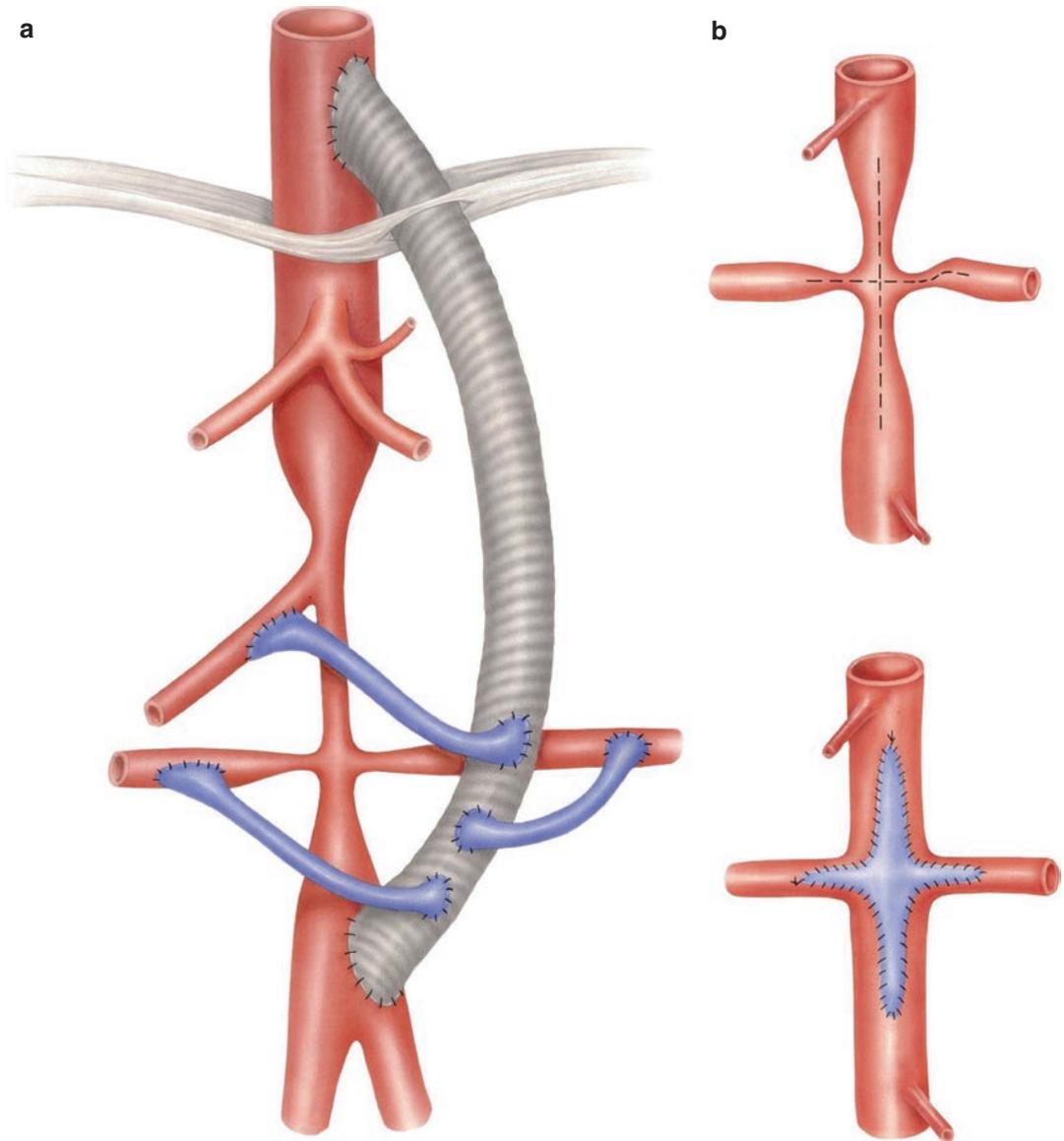


Fig. 37.2 (a, b) Truncular arterial malformation: stenosis or coarctation of the abdominal aorta. Treatment by (a) vascular bypass surgery: allopastic or autologous venous grafts, (b) patch plastic autologous, or allopastic

92 techniques [13, 14]. In this situation, the surgical
 93 technique of Belov [7] can be adopted: clamping
 94 of the infiltrated part of the tissue followed by a
 95 continuous Blalock suture and resection of the
 96 overcoming part of the AVM (Fig. 37.9). The
 97 advantage of this technique is that a cutdown of
 98 the infiltrated tissues with dramatic blood loss
 99 can be avoided.

A further sophisticated technique can be rec- 100
 101 ommended in cases where the possibilities of
 102 interventional treatment have come to an end
 103 but leftovers of the nidus or of the infiltrating
 104 AVM have to be treated in order to avoid an
 105 early recurrence. That is the technique
 106 according to Loose [15] (Fig. 37.10): (1) iden-
 107 tification of tiny AVMs by Doppler ultrasound,

Fig. 37.3 (a) Truncal arteriovenous fistula (AVF) of major named vessel; treatment by interruption of the fistulas and (b) reconstruction of the arterial side by alloplastic interposition graft

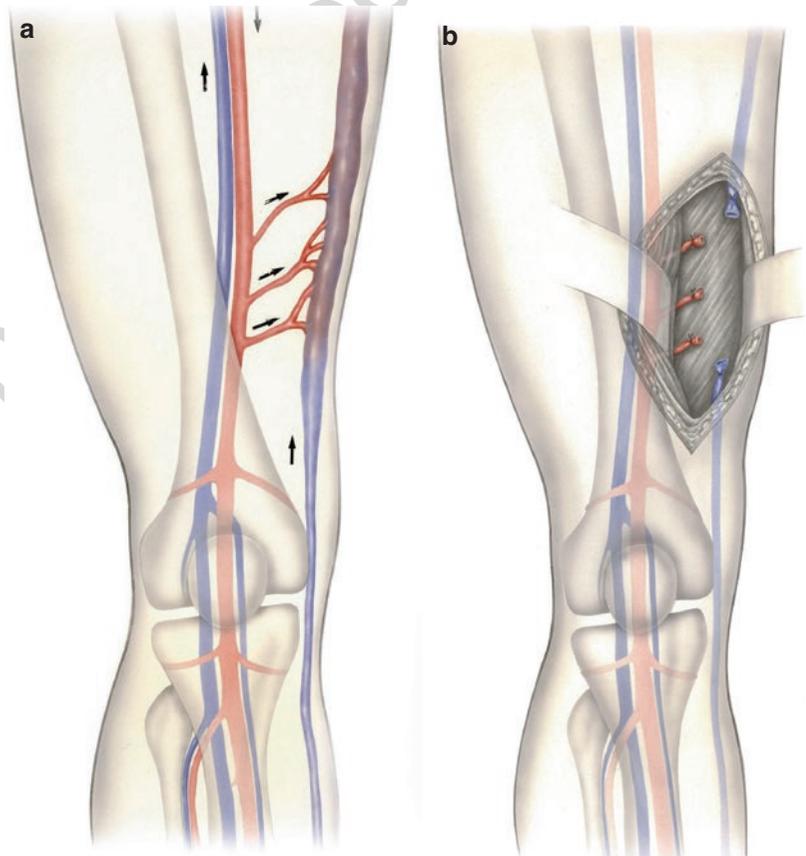
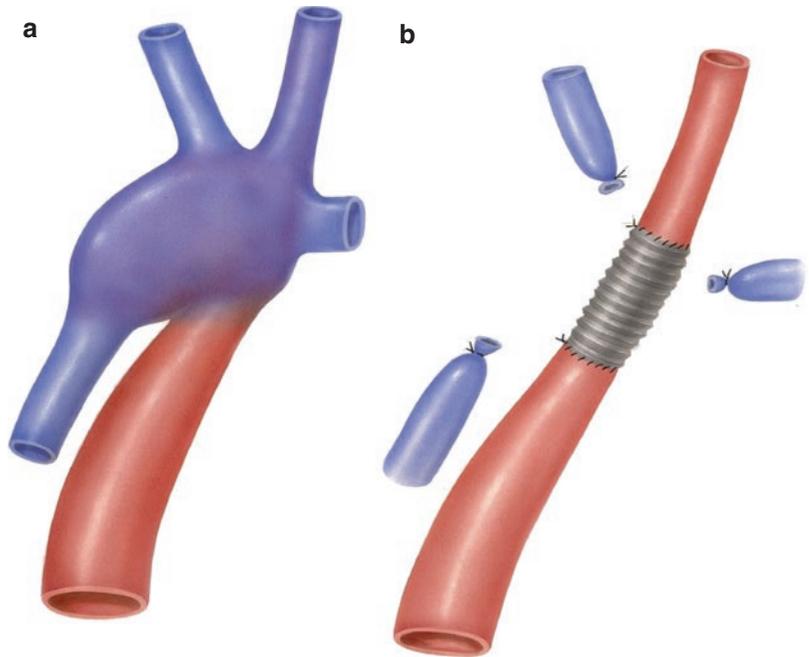


Fig. 37.4 (a) Truncal arteriovenous fistulas (AVF) of larger arterial vessels and branches which could not be occluded by interventional catheter techniques sufficiently. (b) These residual AVFs can be treated by vascular surgery: ligation and interruption of each arteriovenous fistula

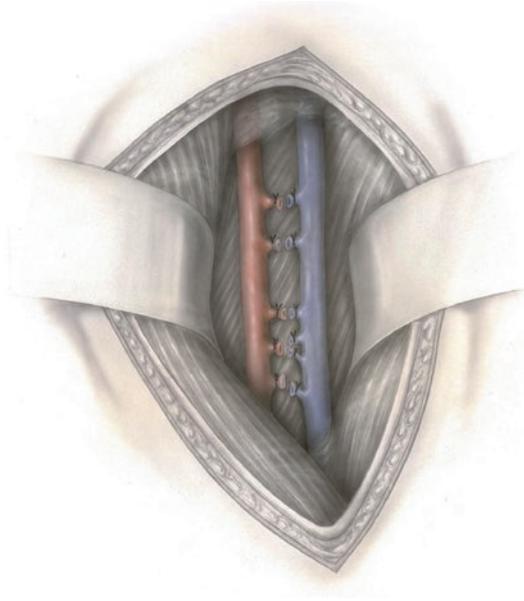
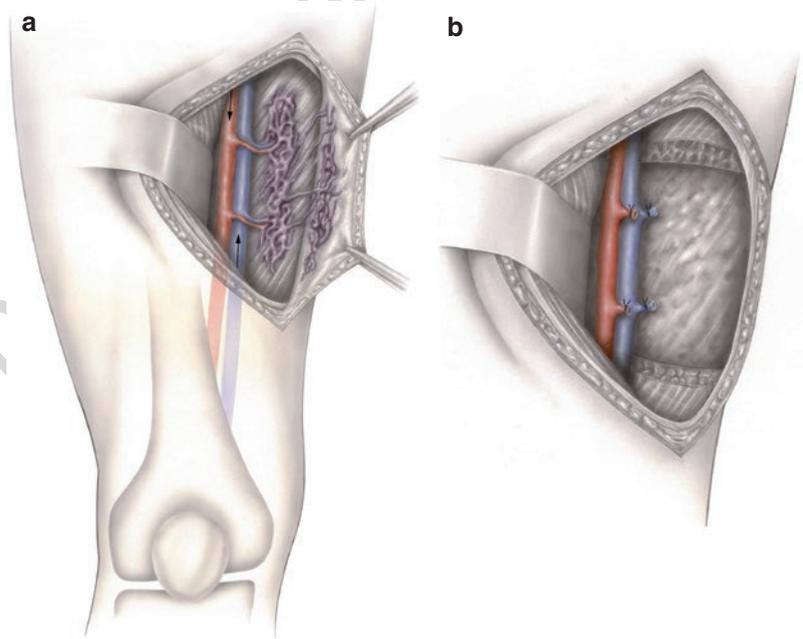


Fig. 37.5 Multiple truncal arteriovenous fistulas (AVF) can be treated by interventional catheter occlusion and/or by ligation by vascular surgery

Fig. 37.6 (a) Extratruncular infiltrating arteriovenous malformation (AVM). (b) Treatment by interventional catheter occlusion and/or by vascular surgical ligation of residual AV fistulas and en bloc resection of infiltrated muscle together with superficial veins



| Table 37.1 Schobinger classification of AVM | t1.1 |
|---|------------------------------|
| Stage I (Quiescence): Pink-bluish stain, warmth, and arteriovenous shunting are revealed by Doppler scanning. The arteriovenous malformation mimics a capillary malformation or involuting hemangioma | t1.2 t1.3 t1.4 t1.5 |
| Stage II (Expansion): Stage I plus enlargement, pulsations, thrill, bruit, and tortuous/tense veins | t1.6 t1.7 |
| Stage III (Destruction): Stage II plus dystrophic skin changes, ulceration, bleeding, tissue necrosis. Bony lytic lesions may occur | t1.8 t1.9 t1.10 |
| Stage IV (Decompensation): Stage III plus congestive cardiac failure with increased cardiac output and left ventricle hypertrophy | t1.11 t1.12 t1.13 |

(2) clamping of the AVMs under sonographic supervision, (3) surgical closure of tiny AVMs by over-and-over sutures, and (4) Doppler ultrasound control to ensure the complete closure of the specific AVM. These procedures have to be continued until no more AVM is to be detected by ultrasound [10, 15–20].

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Fig. 37.7 Extratruncular arteriovenous fistulas (AVM) with secondarily dilated venous plexus. Treatment by interventional catheter occlusion of the arteriovenous fistulas and/or vascular surgery and ligation of the residual AV fistulas and resection or extirpation of the secondarily dilated veins

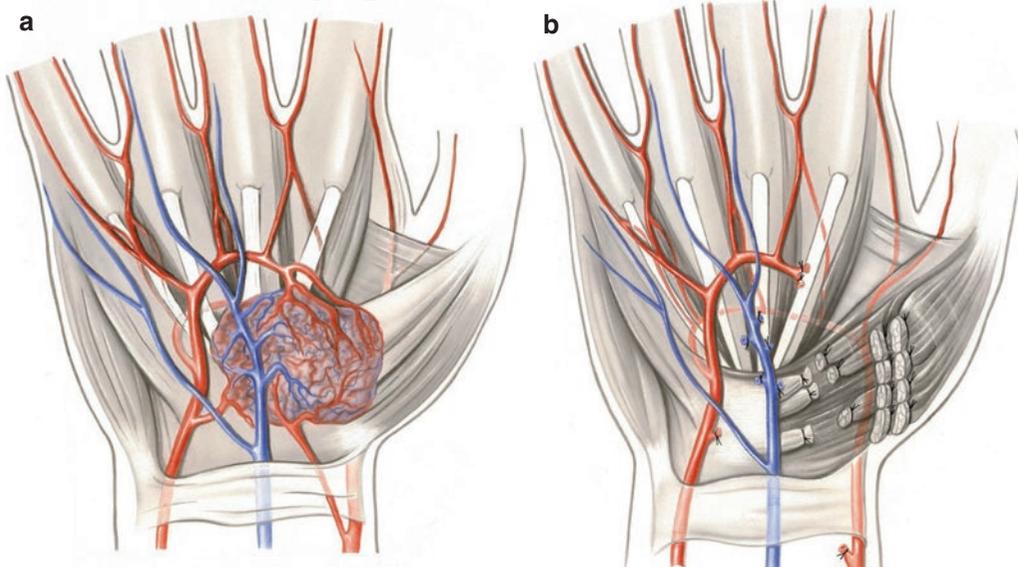
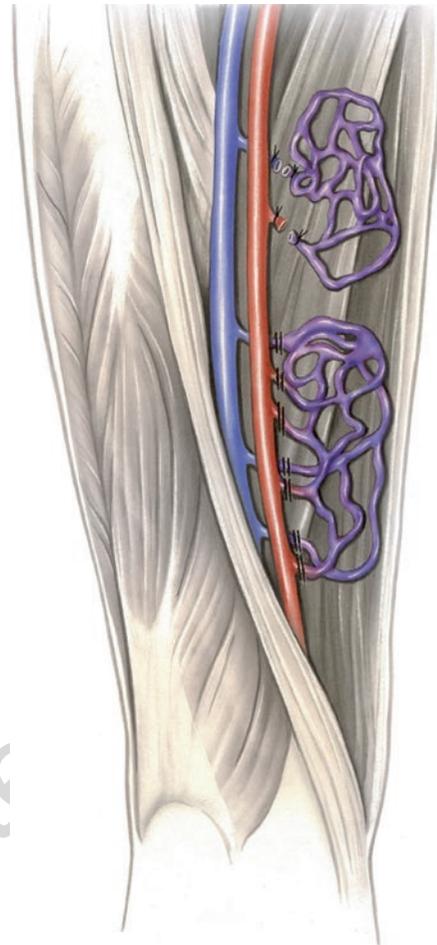


Fig. 37.8 (a) Extratruncular infiltrating arteriovenous malformation (AVM) with AV nidus. (b) Treatment by interventional catheter embolization and afterward

vascular surgery by en bloc resection of the infiltrated tissues/nidus with partial resection of the adjacent tissues

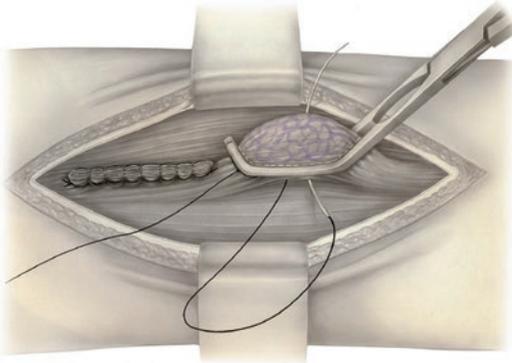


Fig. 37.9 Extratruncular localized infiltrating arteriovenous malformation (AVM): when sclerotherapy or embolization treatment are not possible or not successful, the surgical technique according to Belov [7] is an option: clamping of the infiltrated part of the tissue followed by a continuous Blalock suture and resection of the overlying part of the AVM

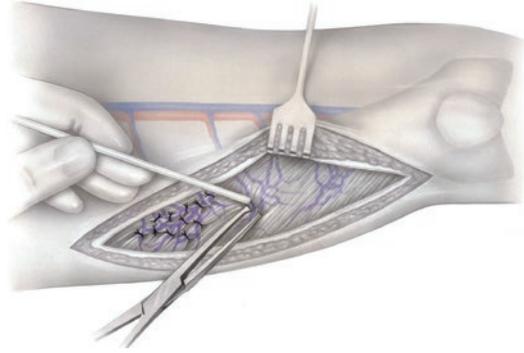


Fig. 37.10 Extratruncular diffuse infiltrating AVM not to be sufficiently treatable by interventional techniques can be treated by the surgical technique according to Loose: before surgery the AVMs are identified by color. Doppler imaging and precisely marked on the overlying skin. During surgery ultrasonic Doppler mapping of AV fistulas, clamping, and over-and-over sutures of the AV fistulas and directly afterward ultrasonic control of the complete closure of the AV fistulas. This has to be continued until every AV fistula of the specific region is closed up

References

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| <p>117 118 119 120 121 122 123 124 125 126 127 128 129 130 131 132 133 134 135 136 137 138 139 140 141 142 143 144 145 146 147 148 149</p> | <p>1. Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. <i>Plast Reconstr Surg.</i> 1982;69:412–22.</p> <p>2. Enjolras O, Mulliken JB. Vascular tumors and vascular malformations (new issues). <i>Adv Dermatol.</i> 1997;13:375–423.</p> <p>3. Mulliken JB. Classification of vascular birthmarks. In: Mulliken JB, AE Y, editors. <i>Vascular birthmarks: hemangiomas and malformations.</i> Philadelphia: WB Saunders; 1988. p. 24–37.</p> <p>4. Lee BB, Baumgartner I, Berlien P, Bianchini G, Burrow P, Glovicki P, Huang Y, Laredo J, Loose DA, Markovic J, Mattassi R, Parsi K, Rabe E, Rosenblatt M, Shortell C, Stillo F, Villavicencio L, Zamboni P. Diagnosis and treatment of venous malformations Consensus Document of the International Union of Phlebology (IUP) updated 2013. <i>Int Angiol.</i> 2014. PMID:24566499.</p> <p>5. Mattassi R, Loose DA. Treatment of arterial malformations. In: Mattassi R, Loose DA, Vaghi M, editors. <i>Hemangiomas and vascular malformations.</i> Milan: Springer-Italia; 2009. p. 209–13.</p> <p>6. Belov S, Loose DA. Surgical treatment of congenital vascular defects. <i>Int Angiol.</i> 1990;9:175–82.</p> <p>7. Belov S. Operative- technical peculiarities in operations of congenital vascular defects. In: Balas P, editor. <i>Progress in angiology.</i> Turin: Edizioni Minerva Medica; 1991. p. 379–82.</p> <p>8. Lee BB, Baumgartner I, Berlien HP, Bianchini G, Burrows P, Do YS, Ivancev K, Kool LS, Laredo J, Loose DA, Lopez-Gutierrez JC, Mattassi R, Parsi K, Rimón U, Rosenblatt M, Shortell C, Simkin R, Stillo</p> | <p>F, Villavicencio L, Yakes W. Consensus document of the International Union of Angiology (IUA)-2013, Current concepts on the management of arteriovenous malformations. <i>Int Angiol.</i> 2013;32(1):9–36.</p> <p>9. Loose DA. Combined treatment of congenital vascular defects: indications and tactics. <i>Semin Vasc Surg.</i> 1993;4:260–5.</p> <p>10. Loose DA. The combined treatment of arteriovenous malformations. In: Mattassi R, Loose DA, Vaghi M, editors. <i>Hemangiomas and vascular malformations.</i> Milan: Springer-Italia; 2009. p. 195–204.</p> <p>11. Loose DA. Treatment of arteriovenous malformations. In: Mattassi R, Loose DA, Vaghi M, editors. <i>Hemangiomas and vascular malformations.</i> Milan: Springer-Italia; 2009. p. 215–21.</p> <p>12. Loose DA, Weber J. Indications and tactics for a combined treatment of congenital vascular defects. In: Balas P, editor. <i>Progress in angiology.</i> Turin: Edizioni Minerva Medica; 1991. p. 373–8.</p> <p>13. Riles TS, Rosen RJ. Peripheral arteriovenous fistulae. In: Rutherford RB, editor. <i>Vascular Surgery.</i> 4th ed. Philadelphia: WB Saunders Company; 1995. p. 1211–7.</p> <p>14. Lee BB, Bergan JJ. Advanced management of congenital vascular malformations: a multidisciplinary approach. <i>Cardiovasc Surg.</i> 2002;10:523–33.</p> <p>15. Loose DA. Systematik, radiologische Diagnostik und Therapie vaskulärer Fehlbildungen. In: Hohenleutner U, Landthaler M (Hrsg.) <i>Operative Dermatologie im Kindes- und Jugendalter. Diagnostik und Therapie von Fehl- und Neubildungen.</i> Berlin/Wien: Blackwell; 1997.</p> <p>16. Jackson JE, Mansfield AO, Allison DJ. Treatment of high-flow vascular malformations aided by flow</p> | <p>150 151 152 153 154 155 156 157 158 159 160 161 162 163 164 165 166 167 168 169 170 171 172 173 174 175 176 177 178 179 180 181 182 183</p> |
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- 184 occlusion techniques. *Cardiovasc Intervent Radiol.* 1996;19:323–8. 193
- 185 194
- 186 17. Mattassi R, Loose DA, Vaghi M. Surgical techniques 195
- 187 in vascular malformations. In: Mattassi R, Loose DA, 196
- 188 Vaghi M, editors. *Hemangiomas and Vascular malfor-* 197
- 189 *mations. An atlas of diagnosis and treatment.* 2nd ed. 198
- 190 Milan: Springer Italia; 2015. p. 249–54. 199
- 191 18. Mattassi R, Loose DA, Vaghi M. Principles of treat- 200
- 192 ment of vascular malformations. In: Mattassi R, 201
- Loose DA, Vaghi M, editors. *Hemangiomas and vas-*
- cular malformations. Milan: Springer Italia; 2009. 193
- p. 145–51. 194
19. Weber JH. Interventional therapy in arteriovenous 196
- congenital malformations. In: Mattassi R, Loose DA, 197
- Vaghi M, editors. *Hemangiomas and vascular malfor-* 198
- mations.* Milan: Springer Italia; 2009. p. 153–62. 199
20. Mattassi R. Surgical treatment of congenital arterio- 200
- venous defects. *Int Angiol.* 1990;9(3):196–202. 201

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