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

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

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

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

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

A combined treatment means first interventional embolization followed by surgical excision of the localized AVM, sometimes together with adjacent tissues (Fig. 37.8) [12].

Infiltrating AVM very often cannot be treated by direct puncture or by catheter embolization...
techniques [13, 14]. In this situation, the surgical technique of Belov [7] can be adopted: clamping of the infiltrated part of the tissue followed by a continuous Blalock suture and resection of the overcoming part of the AVM (Fig. 37.9). The advantage of this technique is that a cutdown of the infiltrated tissues with dramatic blood loss can be avoided.

A further sophisticated technique can be recommended in cases where the possibilities of interventional treatment have come to an end but leftovers of the nidus or of the infiltrating AVM have to be treated in order to avoid an early recurrence. That is the technique according to Loose [15] (Fig. 37.10): (1) identification 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.

Table 37.1  Schobinger classification of AVM

| 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 |
| Stage II (Expansion): Stage I plus enlargement, pulsations, thrill, bruit, and tortuous/tense veins |
| Stage III (Destruction): Stage II plus dystrophic skin changes, ulceration, bleeding, tissue necrosis. Bony lytic lesions may occur |
| Stage IV (Decompensation): Stage III plus congestive cardiac failure with increased cardiac output and left ventricle hypertrophy |

(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].
Fig. 37.7 Exatruncular 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) Exatruncular 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 overcom

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


