Congenital Vascular Malformations: Surgical Strategies and Techniques

Vascular malformations, angiodysplasias represent a group of diseases, which is characterized by congenital vascular defects encountered in enormous complexity and variety. In earlier times, these diseases could only be described (Klippel and Trenaunay 1900) while their pathomorphology could not be clarified.

Today, vascular malformations are recognized as a clinical entity and are no longer a source of confusion in terms of diagnosis, classification and treatment. Furthermore, it has become possible to precisely categorize every lesion clinically encountered by taking the actual classification from 2014 as a basis:

<table>
<thead>
<tr>
<th>Vascular Tumors</th>
<th>Vascular Malformation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign</td>
<td>Morphological/embryological subclassification</td>
</tr>
<tr>
<td>Locally aggressive or borderline</td>
<td>extratruncular forms</td>
</tr>
<tr>
<td>Malignant</td>
<td>truncular forms</td>
</tr>
<tr>
<td>Arterial</td>
<td>Venous</td>
</tr>
<tr>
<td>Venous</td>
<td>Arteriovenous</td>
</tr>
<tr>
<td>Arterial</td>
<td>Lymphatic</td>
</tr>
<tr>
<td>Venous</td>
<td>Microvascular</td>
</tr>
<tr>
<td>Arteriovenous</td>
<td>Combined</td>
</tr>
</tbody>
</table>

Since then, various non-invasive and invasive diagnostic tools have become available. It is mandatory that initially, special diagnostic procedures have to be performed so that the different forms and types can be associated. Only after this identification can a treatment strategy be developed.

Diagnostic

Three different diagnostic pillars have been proven of the years:

- clinical findings,
- functional examinations,
- radiological examinations.

If we follow these principles, we will be able to achieve the main basic diagnostic goals, which are: Definition of the predominantly involved vascular system.

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Determination of the extent of the malformation and the involvement of adjacent structures

Definition of the disturbed haemodynamic: local, regional or systemic.

There are some guidelines in diagnostics of congenital vascular defects which include the diagnostics stages (Lee et al. 2013):

- radiological tests without contrast agent,
- functional tests, not invasive,
- functional tests, invasive,
- radiological test with contrast agent: venous circulation,
- radiological test with contrast agent: arterial circulation.

Through these diagnostic procedures in stages, a clear identification of the findings becomes possible. Only after clarification of the previously mentioned diagnostic goals, a reliable association and classification of vascular anomalies will be possible.

Therapeutic strategies

A vascular surgical treatment of congenital vascular defects can only be successful,

A) if a strict indication for surgery is performed

B) if the basic principles of the therapeutic strategy are followed, which are:

- to start in early childhood (at the ages of 3 – 7)
- to influence the pathophysiological process and abolish the haemodynamic dysfunction,
- to perform harmonized individual therapy,
- to perform surgery radically without loss of function,
- to perform stepwise surgical treatment,
- to perform a multidisciplinary therapy.

Surgical and non-surgical treatment can be performed within these therapeutic strategies. Non-surgical methods have proven to be true when surgery is impossible to perform or when an operation has not yet been performed. Non-surgical methods of treatment are:

- Compression bandages
- Sclerotherapy
- Laser therapy
- Percutaneous interventional catheter embolization treatment.

These therapeutic methods can be adopted individually or in combination with surgery. The indication for a sole treatment by compression bandages as a result of a therapeutic frustration as recommended by Baskerville
et al. (1985) cannot be accepted in light of up-to-date
knowledge and experience.

The onset of surgery has a special importance in chil-
dren with a vascular bone syndrome and a limb-length
discrepancy. Here, the optimal occasion for surgery is
the range between three and seven years of age because
full compensation or partial compensation of the length
discrepancy of the limbs can be expected during this
period. The consequence is that in such cases, vascula-
lar surgery must be considered to achieve a reduction of
a length discrepancy (see Fig. 1). If an additional, for
example orthopaedic treatment of the correction of a
length discrepancy has to be performed, the following
indication should be strictly followed:

Surgery must only be performed in the affected limb,
when there is no indication for vascular surgery,
when the correction of the length is not sufficient in a
severe length discrepancy after vascular surgery,
after the end of growth (here, the temporary epiphys-
iodesis is excluded).

Recommendations for orthopaedic surgery to gain a
correction of the length discrepancy of the legs on
the not affected leg, as recommended by Servelle or
Ilizarov, must be strongly rejected.

**Surgical treatment**

Surgical treatment is indicated for the five main types of
peripheral vascular malformations which cause vascular
insufficiency, cardiac overload and limb length discrep-
ancy, disfiguration and dysfunction. The foundation of this
treatment are six different therapeutic strategies, which
are linked to special indications and special surgery

- Reconstructive vascular surgery
- Operations to reduce the haemodynamic activity of
  the vascular defects
- Operations to remove the vascular defects
- Combined treatment
- Unconventional surgical methods
- Multidisciplinary treatment

Reconstructive vascular surgery is rarely possible in pe-
ripheral vascular malformations because of the diffuse
nature of vascular defects. However, in truncular forms,
a complete elimination of the malformation is possible.
Therefore, postoperative results are usually quite good
because of the removal of the malformation and restitu-
tion of the affected vessels.

Operations to reduce the haemodynamic activity of the
vascular defects are surgical strategies which are often
indicated as well as in venous as arterial malformations.

- The removal of truncular vascular defects includes
  the following:
  - Resection of dysplastic phlebectasis
  - Resection of deep av-communication, if emboliza-
    tion is not possible
  - Resection of superficial av-communication together
    with afferent veins

Surgical techniques in resection of truncular dysplastic
phlebectasis quite often demand multiple, mostly
atypical incisions along the affected vessels and possibly radical resection of dilated veins and venous convolutes. If embolization by catheter techniques is not possible, the operative approach in truncular, deep av-communication consists of typical incisions towards the main vessels. The av-fistulas are carefully dissected and resected consecutively along the vessels.

If embolization is not possible, the superficial truncular av-communication is reached by atypical incisions along the superficial dilated afferent veins at the locations, where clinical, angiographic and haemodynamic communication has been discovered. They are resected together with segments of the afferent phlebectasias.

The removal of extratruncular vascular defects includes the following techniques:

**Extirpation of extratruncular, limited vascular malformation** (see fig. 2).

**Radical extirpation of extratruncular infiltrating vascular malformation together with tissues and organs.**

The surgical approach in extratruncular venous and av-malformations is best attained by phlebincisions. Dissection of vascular malformation is in many cases very difficult or even impossible because of infiltrative growth in encompassing tissue and fragility of the dysplastic vessels. It is also frequently accompanied by abundant haemorrhage. In such cases, a radical extirpation of the extratruncular vascular malformation together with muscles can be performed. This technique is in accordance with Malan's opinion, that the criteria of surgical treatment in vascular malformations are not different from those applied in "cancer surgery". One must be extremely radical even if this approach produces considerable mutilation (Malan 1974).

The operations to remove vascular defects are most often applied in peripheral vascular malformations. Postoperative results after early and functionally radical operations are usually good. Corrections of the lower limb length discrepancy can be achieved after vascular surgery in early childhood. Recovery of form and function is also possible after meticulously executed operation in parts of extremities where embolization is dangerous, and surgery is difficult (Mattassi, Loose, Vaghi 2015).

**Combined treatment**

The treatment of congenital, predominantly arteriovenous malformations is a task that can only be performed to the highest standard by a multidisciplinary group guided by a vascular surgeon. The treatment of congenital vascular malformations is one of the most difficult tasks in vascular surgery.

The occurrence of severe haemorrhage, endangering the patient's life during the operation, is ever present during every single one of the surgeon's movements. Moreover, according to Fontaine, vascular surgeons are constantly placed between excessively radical surgery, which may lead to severe postoperative ischemia, and excessive caution which may result in a relapse. That is why for a long time, treatment was mainly conservative and symptomatic.

**With the development of modern, non-surgical and surgical techniques, combined treatment was developed and today, it is the latest therapeutic concept in modern vascular surgery.**

In predominantly av-shunting defects, the treatment of choice is a combined treatment: interventional embolization therapy and perhaps surgery in addition. It does not consist of alternative or competitive techniques, rather non-surgical and surgical forms of treatment are viewed as complementary.

The indications for non-surgical treatment include the following:
- **Laser therapy:** All microvascular av-lesions, where no feeder artery for embolization treatment is present.

**Fig. 2:**
Operation to remove the vascular defect

Extirpation of extratruncular limited vascular malformations:
a) Phlebography of venous malformation of the right shank.
b) Surgical technique according to BELOV.
Sclerotherapy: Adjunctive treatment with different agents for malformed veins.

Embolization treatment: Hypervascular lesions with av-shunting (dependent on the morphological form and site of the malformation).

Before the combined treatment starts, the type of therapy must be planned. One of the main questions is whether a predominant embolization therapy or a predominant surgical therapy should be applied. Although each individual case has its own indications for embolization or surgical therapy, there are some principal guidelines. It is important to know the risk of a combined treatment depending on the location of the vascular defect. In the upper extremities, embolization therapy generally should not extend distally beyond the wrist joint. Otherwise, severe peripheral spasms or digital necrosis can occur. For the lower extremities, embolization therapy generally should not extend distally beyond the proximal shank region. Should it become necessary to perform a treatment distal to these limits, embolization and sclerotherapy could be combined by venous or by arterial access.

Unconventional surgical methods

Unconventional surgical methods are applied in conventionally inoperable forms of vascular malformations. These inoperable forms are the extratruncular vascular defects that infiltrate neighbouring tissues and organs, destroying their form and function in making their dissection and resection difficult, hazardous or even impossible. Another form is the frequent reduction of vascular truncusses (mainly aplasia or hypoplasia of long segments of deep veins), greatly impairing venous drainage and complete haemodynamic of the affected limb, making reconstructive surgery impossible.

To date, there have been nine unconventional operative techniques which have achieved a significant improvement of the described local haemodynamic disturbances and a decrease in the unpleasant symptoms in patients previously considered inoperable.

Multidisciplinary treatment

Multidisciplinary treatment is indicated in vascular malformations where tissue structures are involved, which do not belong to the vascular system. Here, an interdisciplinary treatment is necessary. The specialists and disciplines whose involvement is required are the most: plastic surgeons, orthopaedic surgeons, hand surgeons, paediatric surgeons and urologic surgeons.

Conclusion

The modern surgical management of peripheral vascular malformations proves that the old, simple technique of arterial ligation, removing only limited portions of vascular malformations or amputation, must be completely abandoned. The treatment should conform to the indicated strategy, surgical strategies and operative methods as therapeutic guidelines. The surgical technique must be individualized in each case and can often be unconventional, in accordance with the polymorphism and haemodynamic peculiarities of the vascular defect in each patient.

Current surgical management in the framework of multidisciplinary surgical combined treatment should be performed in treatment centres for vascular malformations, directed by specialists in congenital vascular pathology. A vascular surgeon and a radiologist are preferred, and they should be familiar with the indications, possibilities and limits of the different therapeutic methods. Our long-term experiences demonstrate that good will, patience and specialized knowledge, combined with operative proficiency and therapeutic creativity and flexibility, can produce good results in this very difficult and extremely challenging field of angiography.