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## Treatment of peripheral vascular malformations – preliminary single-centre experience

### Leczenie obwodowych malformacji naczyniowych – wstępne doświadczenia własne

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#### Key words

vascular malformations, embolisation, minimally invasive treatment

#### Słowa kluczowe

malformacje naczyniowe, embolizacja, leczenie małoinwazyjne

#### Summary

**Introduction.** Vascular malformations present a serious diagnostic and therapeutic problem of modern medicine. They occur in 1.5% of human population and are equally common in both sexes. The symptoms of vascular malformations are diverse and include cosmetic defects, tissue ulcerations, pain, oedema and functional disorders. Vascular malformations are treated with surgical procedures, techniques of interventional radiology, laser therapy or a combination of the methods mentioned above.

**Aim.** To evaluate effectiveness of peripheral vascular malformations treatment with using endovascular techniques.

**Material and methods.** Between 2009-2014, 46 therapeutic procedures were performed in patients with vascular malformations within the lower limbs, shoulder girdle and pelvis in the Department of Interventional Radiology and Neuroradiology, Medical University of Lublin. Various treatment techniques were used – procedures were carried out via arterial access (most commonly – 35/46), venous access (3/46) and by direct puncture of lesions (8/46).

**Results.** The intended technical success was achieved in 82% of procedures; in some patients after several-stage embolisation and subsequent surgical resection. In 8 patients, further embolisation procedures will be performed due to a considerable extent of vascular malformation and its incomplete exclusion from circulation.

**Conclusions.** Minimally invasive endovascular techniques enable effective treatment of vascular malformations; strict cooperation of a multi-disciplinary team of specialists, mainly interventional radiologists, vascular surgeons, plastic surgeons and dermatologists, leads to best outcomes.

#### Streszczenie

**Wstęp.** Malformacje naczyniowe stanowią poważny problem diagnostyczny i terapeutyczny współczesnej medycyny. Dotykają 1,5% populacji ludzkiej i jednakowo często występują u obu płci. Objawy malformacji naczyniowych są bardzo różnorodne, obejmują defekty kosmetyczne, owrzodzenia tkanek, ból, obrzęk oraz zaburzenia czynnościowe. Metody leczenia malformacji naczyniowych obejmują zabiegi chirurgiczne, techniki z zakresu radiologii zabiegowej, laseroterapię czy kombinację wyżej wymienionych metod.

**Cel pracy.** Ocena skuteczności leczenia obwodowych malformacji naczyniowych metodami wewnątrznaczyniowymi.

**Materiał i metody.** W okresie od 2009 roku do 2014 w Zakładzie Radiologii Zabiegowej i Neuroradiologii UM w Lublinie wykonano 46 zabiegów leczniczych u chorych z malformacjami naczyniowymi w obrębie kończyn dolnych, kończyn górnych, obręczy barkowej i miednicy. Stosowano różne techniki leczenia malformacji – zabiegi wykonywano z dostępu przez tętnicę (najczęściej – 35/46), przez żyłę (3/46) oraz z bezpośredniego nakłucia zmiany (8/46).

**Wyniki.** Zamierzony sukces techniczny osiągnięto w 82% zabiegów, u części chorych dopiero po wykonaniu kilkuetapowego zabiegu embolizacji oraz następowej resekcji chirurgicznej. U 8 chorych ze względu na znaczną rozległość malformacji naczyniowej i nadal niepełnej jej wyłączenie z krążenia konieczne będzie wykonanie kolejnych zabiegów embolizacji.

**Wnioski.** Małoinwazyjne techniki wewnątrznaczyniowe dają możliwość skutecznego leczenia malformacji naczyniowych, a najlepsze wyniki można osiągnąć przy ścisłej współpracy multidyscyplinarnego zespołu specjalistów: a przede wszystkim radiologów zabiegowych, chirurgów naczyniowych, chirurgów plastycznych i dermatologów.

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## INTRODUCTION

Vascular malformations are a serious diagnostic and therapeutic problem of modern medicine. They result from improper formation of the vascular system during foetal life. Although malformations are congenital, they can manifest themselves several weeks or even years after birth. In general, malformations grow proportionally with the child. They affect 1.5% of human population and are equally common in both sexes (F:M = 1:1) (1).

The symptoms of vascular malformations are very diverse and include cosmetic defects, tissue ulcerations, pain, oedema and functional disorders. The treatment methods for vascular malformations involve surgical procedures, techniques of interventional radiology, laser therapy or a combination of the methods mentioned above. Generally, only symptomatic malformations are treated.

There are many classifications of vascular malformations, which are based on anatomy or histological structure of lesions, their appearance or development. In 1982, Mulliken and Glowacki published the division of malformations based primarily on features of biological activity of angiogenic lesions, regardless of their external appearance. This classification divided vascular malformations into two groups: haemangiomas and vascular malformations. The former were defined as benign angiogenic lesions, which grow quickly in infancy and undergo spontaneous fibrosis and regression during childhood. Vascular malformations include various lesions composed of dysplastic vessels; in most cases, with the predominant component of one vessel type: capillary, venous, arterial or lymphatic (2).

In 1983, Burrows and co-authors devised a classification based of angiographic differences and nature of the flow in lesions. Another important classification evaluating vascular malformations according to their structure was the Hamburg classification introduced during the 7th International Society for the Study of Vascular anomalies (ISSVA) Congress in Hamburg in 1988 (3).

In 1993, Jackson and co-workers simplified the Burrows classification dividing vascular malformations into slow-flow and fast-flow ones. The former corresponded to venous malformations (VM) whereas the latter to arteriovenous malformations (AVM). In 2006, Chow et al. published the modified angiographic division of peripheral fast-flow arteriovenous malformations. Four categories were suggested according to morphology of a malformation nidus; the division is a modified classification introduced by Houdart et al., who divided intracranial malformations into three types: connections/fistulae:

- arteriovenous,
- arteriovenous,
- arteriovenous.

The classification introduced by Puig in 2003 distinguishes 4 types of slow-flow malformations (4-7).

Clinical evaluation of vascular malformations is most commonly based on the Schobinger classification introduced in 1999 during the ISSVA meeting in Amsterdam. The classification in question is used for treatment planning. The stage I lesions often do not require

treatment and should only be observed as they can be stationary for a long period. The increasing lesions belong to stage II; the stage III lesions are associated with pain, bleedings from lesions, ulcerations or necrosis. The final stage IV includes so advanced lesions that beside the symptoms accompanying stage III malformations they affect the heart and lead to its failure.

Vascular malformations are caused by abnormalities during complex processes of formation of venous and arterial vessels during foetal life. Animal studies demonstrated that the key role is played by vascular endothelial growth factor (VEGF), transforming growth factor (TGF- $\beta$  as well as vascular proteins – angiopoietin-1 and 2, ephrin-B2 and their receptors). The above factors are believed to be responsible for development of vascular malformations in humans (8).

## AIM

To evaluate effectiveness of peripheral vascular malformations treatment with using endovascular techniques.

## MATERIAL AND METHODS

Between 2009 and 2014, 46 therapeutic procedures were performed in patients with vascular malformations within the lower limbs, shoulder girdle and pelvis in the Department of Interventional Radiology and Neuroradiology, Medical University of Lublin. In 20 patients, the minimally invasive procedures preceded surgical resection of the lesion. Various methods of malformation treatment were applied, procedures via arterial access (most commonly – 35/46), venous access (3/46) and by direct puncture of the lesion (8/46). The majority of procedures were performed under local anaesthesia (36/46), the remaining ones under general anaesthesia (10/46). General anaesthesia was required due to the use of absolute alcohol during interventions. Various embolisation techniques were applied, most commonly vascular malformations were embolized with liquid embolizing substances (cyanoacrylate glue or Glubran combined with different concentrations of Lipiodol and Onyx) (fig. 1A, B, C; 2); in some cases, embolisation spirals were additionally used. In transarterial procedures, typical femoral artery accesses in the groin were applied. After introducing a short introducer 5 Fr to the femoral artery, selectively using the system of coaxial catheters and microcatheter, their optimal position within the malformation was achieved for administration of the substance closing the malformation or its part.

In 20 patients, complete surgical resection of the lesion was performed after embolisation. Surgical procedures were carried out by the team of vascular surgeons. In 1/3 of patients, the closure of vascular malformation was possible only after two or more endovascular procedures.

## RESULTS

The intended technical success was achieved in 82% of procedures; in some patients only after two-stage embolisation and subsequent surgical resection.

In 8 patients, further embolisation procedures will have to be performed due to a considerable extent of vascular malformation and its incomplete exclusion from circulation. A severe complication observed during 2 procedures was pulmonary microembolism manifesting with typical cough fits in two patients, in one of them accompanied by thoracic pain. Slight glue deposits in the pulmonary arterial bed were visible during X-ray performed immediately after the procedure. Microembolism resulted from the permeation of slight amounts of embolisation material through the malformation nidus to the venous bed. In both cases, an iatrogenic episode of pulmonary microembolism did not affect the length of hospitalization and was not associated with permanent changes. The other intra-procedure complications were noted in 3 patients – haematomas at the site of femoral artery puncture in the groin. No other post-embolisation complications were observed.

The follow-up imaging examinations performed 1 to 12 months after endovascular procedures (and subsequent surgical resection in some cases) revealed that 7 patients initially evaluated as full recovery cases (of 38 individuals considered effectively cured) had recurrences of symptoms; only in one of them, the recurrence was observed after embolisation combined with surgery.

### DISCUSSION

Proper diagnosis and treatment of vascular malformations is an extremely difficult and complex problem of modern medicine. The lesions are rare and therefore physicians facing this problem often do not know how to help and where to refer patients. In Poland, there are no centres specializing in the treatment of vascular malformations, which unfortunately results in the lack of suitable experience in dealing with complex vascular lesions. Moreover, it should be remembered that each vascular malformation is different and requires individual diagnostic-therapeutic approach. The choice of an appropriate method of treatment depends on the body part containing the lesion, the place of branching and diameter of vessels supplying and carrying out the blood from malformations, size/volume of lesion nidus, vascular connections as well as extent and nature of blood flow (slow- and fast-flow malformations). The most common type of interventional radiology procedure used for the treatment of vascular malformations is selective embolisation of lesions using liquid embolizing substances.

The complications developing during procedures are divided into severe, i.e. acute pulmonary hypertension, tissue necrosis, haemoglobinuria or acute renal failure, which occur in 0.2-13% of cases, and less severe developing during procedures or after them are observed in 3-48% of cases. They include neurological deficits, transient symptoms of ischaemia, pain, sensory disorders, bleedings, infections or local lesions, such as haematomas, pseudoaneurysms or arteriovenous fistulas at the site of vascular access.

In most cases, treatment of vascular malformations takes many years and sometimes does not result in fully

satisfactory outcomes. According to different literature data, the percentage of fully cured lesions ranges from 30 to 75%, while the percentage of recurrences of symptoms is between 25-60%. Moreover, both patients and physicians should be aware of a relatively high percentage of post-treatment complications. The main rule that should be followed is to initiate the treatment only in symptomatic patients. Patients and their families should be informed in detail about the treatment method, anticipated outcomes and possible complications (9-11).

### CONCLUSIONS

Minimally invasive endovascular techniques enable effective treatment of vacuolar malformations; nevertheless, it should be remembered that treatment success depends on appropriate diagnostic-therapeutic procedures. The best results can be obtained when a multi-disciplinary approach is applied (involving interventional radiologists, vascular surgeons, plastic surgeons, dermatologists).

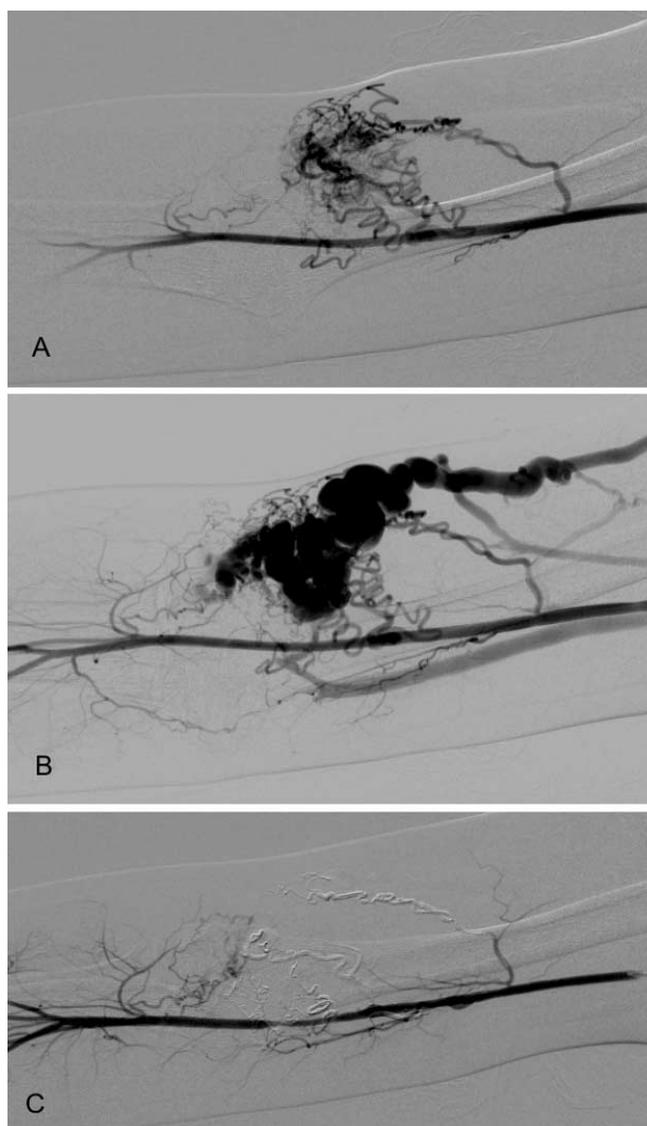


Fig. 1. A vascular malformation of the right forearm before and after embolisation with Onyx.

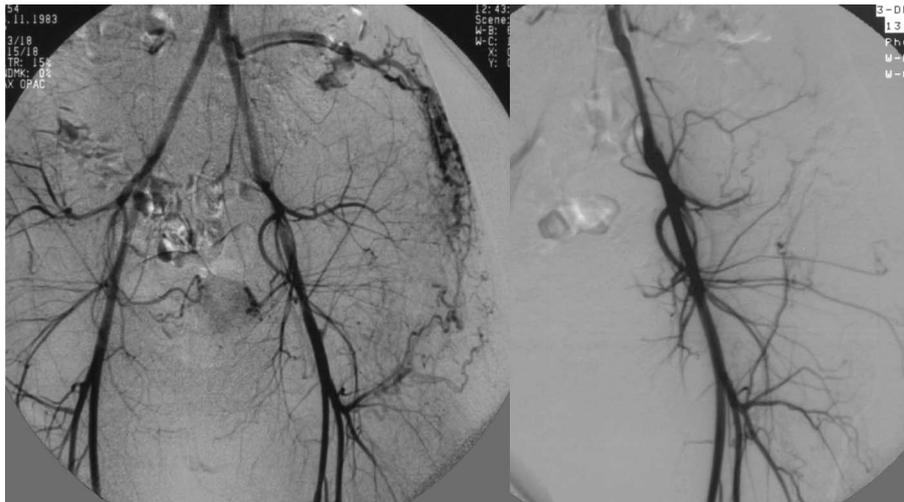


Fig. 2. A vascular malformation of the left buttock before and after embolisation with cyanoacrylate glue.

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