Surgical management of auricular arteriovenous malformations: A literature review

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Abstract

Auricular arteriovenous malformations (AVMs) can cause a variety of symptoms that seriously impact the patient’s appearance, life, and mental well-being. Surgery is the primary management method for auricular AVMs, but there is no consensus on how to surgically manage auricular AVMs. In this article, we document a comprehensive review of the characteristics, classification, and surgical interventions to treat auricular AVMs.

KEYWORDS
Arteriovenous malformation, auricle, endovascular treatment, surgical management

1 | INTRODUCTION

Arteriovenous malformations (AVMs) are high-flow vascular malformations, which have abnormal arteriovenous shunts, commonly referred to as the nidus. These occur between feeding arteries and outflow veins, but they lack a normal capillary network.1 Extracranial AVMs occur primarily in the head and neck region, and the auricle is the second most common site.2,3 Even though auricular AVMs are a benign disease, they often have a serious impact on the appearance, life, and mental status of patients.

The primary goal of auricular AVMs management is to improve function and aesthetics, and maintain long-term efficacy by complete obliteration of the nidus.4–6 There are difficulties in the treatment of auricular AVMs for many reasons, including: (1) the auricle has unique anatomical characteristics (a thin skin envelope covers ear cartilage) that is of great significance to facial aesthetics; (2) there is diversity in the form of clinical manifestations and AVMs are rare; (3) there is a relatively high possibility of AVM recurrence. Although many different techniques or protocols have been used, surgery is still a primary management technique for auricular AVMs.5,7,8 Unfortunately, in the auricular AVMs care community, there are no unified surgical management guidelines. In this article, we document a comprehensive review of the characteristics, classification, and surgical interventions available to treat auricular AVMs, with the goal of providing a reference for future guidelines.

2 | METHOD

This review focused on Pubmed literature up to and inclusive June 2021. PubMed was searched using the following keyword strategy:


**3 | DISCUSSION**

**3.1 | Disease characteristics**

The etiology of auricular AVMs has not been determined, but it is likely that most auricular AVMs are already present at birth, but are not detected.\(^9\) Potential triggers of AVMs include trauma, infection, iatrogenic factors, and hormonal changes (puberty, pregnancy, or hormone therapy), which may stimulate disease progression resulting in a variety of symptoms.\(^4\)\(^5\)\(^7\)\(^8\)\(^10\)\(^11\)\(^16\) For example, arteriovenous shunts and high-flow characteristics of AVMs may not only cause warmth, swelling, erythema, pulsation, fremitus, and ear noise, but also produce ischemic damage to the lesion and surrounding tissues, resulting in ulceration, pain, pruritus, and bleeding.\(^12\)\(^13\) In addition, the abnormal tissue around the cartilage may cause an overgrowth of cartilage and lead to macrotia.\(^14\)

AVMs may involve various anatomical structures of the auricle and extra-auricular tissue, but the external auditory canal (EAC) is usually spared.\(^7\) The “angiosome concept” has been applied to some AVMs of the head and neck, because it is hypothesized that the malformations will occur in the “choke zone” connecting adjacent angiosomes.\(^15\) However, previous studies have found not all auricular AVMs conform to this hypothesis.\(^7\)\(^8\) Auricular AVMs usually have 1–3 arterial feeders and the main feeders are the ipsilateral superficial temporal artery (STA), posterior auricular artery (PAA), and occipital artery (OA).\(^10\)\(^11\)\(^16\)\(^17\) For some large or treated AVMs, the blood supply may also come from branches of maxillary, facial, and vertebral arteries.\(^11\)\(^16\)

**3.2 | Classification**

Accurate classification of auricular AVMs may provide guidance for optimal treatment. Currently, classification is based upon clinical and angiographic characteristics. The Schobinger classification is the most used since it divides AVMs into four stages according to the disease progression (Table 1).\(^2\) Using Schobinger staging, the severity of the condition and the urgency of treatment can be established. AVMs can also be classified as focal or diffuse based upon clinical characteristics of the lesion (Table 1).\(^18\)\(^20\) Diffuse lesions are more difficult to manage and have a higher recurrence rate than focal lesions. Additional classifications have been proposed for auricular AVMs. Bulstrode et al. identified four anatomical patterns of auricular AVMs and provided corresponding management protocols for each pattern (Table 2).\(^8\) Vilela Chagas Ferreira et al. classified auricular AVMs into six types by evaluating the extent of damage (partial, total, or extra-auricular) and thickness (only cutaneous or cutaneous + cartilaginous) of the auricle.\(^5\) They suggested that cartilage should be preserved for lesions that do not involve the cartilage and proposed specific reconstruction protocols (Table 2). In addition, due to complex and diverse vascular angioarchitecture in AVMs, it has also been proposed that angiographic classifications be used, so that endovascular treatment approaches can be selected.\(^21\)\(^23\) Further details and diagrams for the classifications are summarized in Table 3 and Figure 1.

**3.3 | Timing of intervention**

There is still no consensus on when to intervene with surgery for auricular AVMs. It has been stated that it is unnecessary to treat small, asymptomatic auricular AVMs.\(^24\)\(^25\) Wu et al. recommended that patients with Schobinger stage I and II lesion, especially in children, be evaluated annually, and interventions should be initiated when they reach stage III.\(^7\) Commonly, however, there is a continuous progression of extracranial AVMs.\(^3\) Children in stage I have an 80–82.6% risk of progression before adulthood, and the majority of patients (96.2–100%) showed progression during their lifetime.\(^3\)\(^26\) It has also been suggested that treatment of lower stage AVMs may promote better long-term control.\(^26\) Therefore, an early intervention plan should be adopted for individuals according to the location and extent of lesions.\(^8\)\(^26\) If the early-stage AVMs are in non-functional or non-esthetically important areas, or if they are expected to progress and lead to complications, then they should be treated more aggressively. In contrast, extensive early-stage AVMs should be managed by observation, as interventions may lead to a more serious deformity, and there is still the possibility of recurrence.

**3.4 | Surgical management**

Surgery is widely used to manage auricular AVMs, but there is no unified optimal surgical management protocol. This section reviews the most common approaches being utilized for auricular AVMs treatment.
3.4.1 | Proximal vascular ligation alone

In early auricular AVMs care, proximal ligation of feeding arteries was used to manage auricular AVMs. However, simply ligating the arteries did not provide effective treatment, and even lead to disease aggravation. Dingman et al. reported three cases of auricular AVMs that underwent ligation of feeding arteries for various times (1, 4, and 8 times, respectively), but all patients eventually had auricular amputations. It is thought that this treatment method failed due to neoangiogenesis and the formation of new collateral circulation. Even if the ipsilateral PAA, STA, OA, and external carotid artery (ECA) are ligated, the lesion can obtain blood supply from the vertebral artery, internal carotid artery, thyroid cervical trunk, or even the contralateral ECA. Importantly, ligation methods may hinder subsequent endovascular treatment. Thus, the current opinion is that simple proximal ligation should not be used to manage auricular AVMs.

3.4.2 | Surgical resection alone

Surgical resection, either partial or total resection, is another common method for the treatment of auricular AVMs. It should be noted that partial or complete resection should only be applied to the AVM lesion, and not the auricle.

In general, the aim of this surgical method is to completely resect the lesion, but partial resections are also used for functional, aesthetic, or safety requirements. Even though partial resection may improve the symptoms and preserve the auricle, it has a much higher risk of recurrence than complete resection. A recent study reported that the recurrence rate following partial resection of auricular AVMs was 66.7% (4/6 patients), and 0% (0/6 patients) in full resection patients over a 9.4 years follow-up. Recurrence or deterioration following partial resection is due to hypoxia around the residual lesions, which can stimulate angiogenesis and new supply vessel formation, causing aggressive growth of the residual lesions. Therefore, partial resection remains controversial as an optimal method for auricular AVMs but can be used in AVMs that cannot be completely resected. In partial resection, an adjuvant method has been proposed where the residual lesion is treated with ethanol embolization.

Complete resection is an effective treatment for auricular AVMs to achieve mass elimination. Some scholars believed that resections can be limited to the cartilage level since the EAC is usually not involved, so auricle amputation or partial pinnectomy is used to remove the lesion. This method can provide a positive long-term effect with a relatively minimal risk of recurrence, but the total or partial loss of the auricle may affect the patient’s appearance and mental status. Because cartilage plays an important role in maintaining the shape of the auricle, whether or not cartilage is involved in the AVM should be confirmed before or during operation. Cartilage can be left unremoved and covered with a local flap, skin graft, or temporoparietal fascia flap transfer with a skin graft if it is not directly involved. A recent study reported a case of staged complete resection with preservation of the auricle. First, the patient underwent a partial resection, and the retro auricular residual lesion was excised 2 years later. However, there has been little discussion about the indications and intervals of staged resection.
3.4.3 | Surgical resection combined with endovascular treatment

With the development of interventional radiology and superselective techniques, the endovascular treatment used alone or with surgery is becoming more and more important in the management of auricular AVMs. A variety of agents have been used in the management of auricular AVMs, including ethanol, N-butyl cyanoacrylate (NBCA), Onyx, precipitating hydrophobic injectable liquid (PHIL), polyvinyl alcohol (PVA) particles, coils, and gelatin sponge. The characteristics, drawbacks, and clinical applications of these agents are summarized in Table 4. Among these embolic agents, ethanol is the only agent that can provide permanent embolization of AVMs, but its risk and high technical requirements limit its wide application. Other embolic agents are generally used for hemostasis of acute bleeding and palliative embolization when used alone, due to the possibility of vascular recanalization. In recent years, these agents have been widely used in combination with surgery to manage auricular AVMs, during preoperative embolization and postoperative embolization. Preoperative embolization has been used as a method to reduce intraoperative blood loss and induce lesion fibrosis, which may provide a favorable surgical field and lesion delimitation, thus making the operation easier and more accurate. Due to the risk of the development of collaterals, resection is usually recommended within 1–2 days after embolization, but in some studies, resection has been delayed to 1 week. Preoperative embolization also has risks. Although some of the complications (e.g., skin necrosis) can be solved by subsequent resection, caution is still needed. Therefore,
some authors believed that whether preoperative embolization needs to be performed depends on the condition of auricular AVMs, for example, it may not be used for localized lesions. In addition to preoperative embolization, other methods also can be used to reduce intraoperative bleeding, including hypotension anesthesia, multiple sutures around the lesion, intraoperative ligation of the supply or reflux vessels, and application of epinephrine, coagulant drugs, arterial clips, or electrocautery devices.

Surgical resection combined with postoperative embolization is another application of the combined regimen for the treatment of AVMs. It is suitable for some extensive lesions. First, most of the lesion is surgically removed, and then postoperative embolization is performed on the residual lesion, which is difficult to resect or adjacent to important anatomical structures (e.g., facial nerve). Ethanol is the main embolic agent used for postoperative embolization. This methodology and timing can not only eliminate the lesions but also reduce the use of agents to reduce the risk of complications.

3.4.4 Other surgical treatment

In addition to conventional treatment, high-frequency electrocoagulation has been used to treat AVMs. With this method, an electrocoagulation needle that is non-conductive except for the tip is placed into the lesion and is used to coagulate for 1–3 s with an output power between 10 and 50 W. This method is repeated at various points within the range of the lesion. Results are promising, and one study showed efficacy in 88.9% (8 of 9) of AVM patients, but there was a low cure rate (11.1%), and a higher risk of bleeding. However, the recurrence rate of this method has not been reported.

3.4.5 Reconstruction of the auricle

Following surgical intervention, patients with auricular AVMs often need otoplasty or auricular reconstruction. Macrotia is a common sign of auricular AVMs, which is not only caused by the mass effect of the
TABLE 4 Summary of embolic agents for auricular AVMs

| Characteristics                        | Drawbacks                                                                 | Clinical applications                     |
|----------------------------------------|---------------------------------------------------------------------------|-------------------------------------------|
| **Ethanol**                            | • Provide permanent embolization and low recurrence rate                  | • Effective treatment of auricular AVMs   |
|                                        | • High effective rate                                                     |                                           |
|                                        | • Shrink lesions obviously                                                |                                           |
|                                        | • Without color residue or radiopaque cast                                |                                           |
|                                        | • Relative high risk of complications                                    |                                           |
|                                        | • High requirements for operation skill and experience                   |                                           |
| **NBCA**                               | • Fast-acting                                                             |                                           |
|                                        | • Adhesive                                                                | • Hemostasis of acute bleeding            |
|                                        | • Mechanically block the lumen                                            | • Palliative embolization                |
|                                        | • Recanalization may occur                                                | • Preoperative embolization              |
|                                        | • Need to be mixed with a contrast agent in advance                      |                                           |
|                                        | • Short operating time                                                   |                                           |
|                                        | • Hard after polymerization                                               |                                           |
|                                        | • Cannot shrink lesions obviously                                        |                                           |
|                                        | • Risk of catheter tip adhesion or other complications (e.g. hemorrhaging, tissue ischemia, infarction of other organs) |                                           |
| **Onyx**                               | • Non-adhesive, leading to good penetration without catheter adhesion     |                                           |
|                                        | • Inherent radioactive opacity                                            | • Palliative embolization                |
|                                        | • Soft after solidification                                               | • Preoperative embolization              |
|                                        | • Relatively slow solidification, allows longer injection time            |                                           |
|                                        | • Need to be shaken before injection                                      |                                           |
|                                        | • May cause black skin discoloration                                      |                                           |
|                                        | • Artifacts on CT                                                         |                                           |
|                                        | • Relative high cost and radiation exposure                               |                                           |
|                                        | • Cannot shrink lesions obviously                                        |                                           |
|                                        | • Recanalization can occur                                                |                                           |
| **PHIL**                               | • Non-adhesive, leading to good penetration without catheter adhesion     |                                           |
|                                        | • Soft after solidification                                               |                                           |
|                                        | • Relatively slow solidification, allows longer injection time            |                                           |
|                                        | • Inherent uniform radiopacity, no need to be shaken before injection     |                                           |
|                                        | • Without the risk of skin discoloration                                  |                                           |
|                                        | • Low artifacts on CT and risk of catheter blockage                      |                                           |
| **PVA particles**                      | • Inexpensive                                                             | • Palliative embolization                |
|                                        | • Non-degradable                                                          | • Preoperative embolization              |
|                                        | • Compressible, allows embolization vessels that are larger than catheters |                                           |
|                                        | • Can occlude vessels and cause subsequent thrombosis                    |                                           |
|                                        | • Irregularities cause particles to easily aggregate, which can result in catheter blockage, non-target embolization, and an unpredictable level of embolization | • Palliative embolization                |
|                                        | • Recanalization can occur                                                | • Preoperative embolization              |
|                                        | • Need to be mixed with contrast an agent before injection                |                                           |
| **Coils**                              | • Mechanical embolic devices, work by mechanically occluding blood vessels and subsequent thrombosis |                                           |
|                                        | • Inherent radiopacity                                                    | • Preoperative embolization              |
|                                        | • Recanalization can occur                                                |                                           |
|                                        | • May cause pain or other discomforts                                     |                                           |
|                                        | • Artifacts on CT                                                         |                                           |
| **GS**                                 | • Absorbable                                                              | • Preoperative embolization              |
|                                        | • Rapid embolization                                                     |                                           |
|                                        | • Mechanical obstruction and promote thrombosis                           |                                           |
|                                        | • Low cost and easy to obtain                                             |                                           |
|                                        | • Various shapes and sizes are available, such as sheet or powder         |                                           |
|                                        | • Sheets need a multi-step preparation process                            |                                           |
|                                        | • Need to be mixed contrast medium before injection                      |                                           |
|                                        | • Irregular shape and size may lead to unpredictable behavior during embolization |                                           |

Abbreviations: AVMs, Arteriovenous malformations; GS, gelatin sponge; NBCA, N-butyl cyanoacrylate; PHIL, precipitating hydrophobic injectable liquid; PVA, polyvinyl alcohol.

Lesion but also the enlargement of cartilage. For macrotia, otoplasty can be performed during resection surgery or after ethanol embolization. However, for macrotia caused by AVMs, the literature on how to perform otoplasty is limited. Fortunately, many surgical procedures for congenital macrotia have been reported in the literature, such as simple wedge resection, helical chondrocutaneous...
advancement flap with a crescent scaphal resection, or crescent scaphal resection with a wedge resection of the helical rim. These methods may provide references for the otoplasty of macrotia caused by AVMs.

Partial or total auricular reconstruction done during surgery (immediate), or post-surgery (delayed) is also commonly used in patients to improve appearance. Immediate reconstruction has been used for patients whose cartilage was intact or largely preserved during the resection, and this was accomplished by direct suture, local flap, skin graft, or temporoparietal fascia flap transfer combined with skin graft to cover the cartilage. In patients with less cartilage exposure after resection, immediate reconstruction can provide a satisfactory auricle shape. However, for those patients whose skin and subcutaneous tissue of the entire auricle was excised, the outcomes after the immediate reconstruction are less satisfactory because of the possible loss or deformation of cartilage. Delayed reconstructions can be used in patients who have undergone partial pinnectomy or auricle amputation. In delayed reconstruction, costal cartilage or a prosthesis is used as a framework and covered by appropriate tissue flaps and skin grafts. Due to the possibility of recurrence, it is important to wait and monitor the patient for a period after resection before reconstruction. Whereas, there is no consensus on the optimal interval, ranging from 6 months to 5 years in previous studies.

### 3.5 Post-treatment management

Due to the complexity of treatment options and the possibility of recurrence, post-treatment management of auricular AVMs is essential. First, it is necessary to evaluate the patient’s treatment outcomes to determine the subsequent management protocol. Outcome measures that have been used are changes in symptoms or signs, degree of devascularization, complications, and recurrence rate. However, the outcome measures and/or measurement methods used in different studies are heterogeneous, which make it difficult to comprehensively evaluate treatment outcomes and directly compare various treatment methods. Recently, the Outcome measures for Vascular MAIformations (OVAMA) project proposed a core domain set (CDS), and recommended outcome measures that should be used in clinical studies of peripheral vascular malformations. The CDS for AVMs is shown in Table 5. It is worth noting that there is no “recurrence” in CDS because participants argued that it was not a single domain but a reflection of other domains. “Radiological characteristics” are also not included in CDS because imaging follow-up is not mandatory, but it should be reported if the examination was performed. In addition, although the OVAMA Steering Group has conducted studies to explore how to measure these core domains, there is no final core outcome measurement set.

After evaluating treatment outcomes, follow-up is necessary for patients who no longer need special intervention. When conditions permit, both clinical and imaging follow-up should be used because imaging can detect lesion changes more accurately and earlier than clinical follow-up. Since the vast majority of recurrences occur within 5 years after treatment, a minimum 5 years follow-up is recommended.

### 4 CONCLUSION

This review comprehensively documented characteristics, classifications, and surgical management options for auricular AVM patients. Despite the introduction of diverse surgical techniques, the management of auricular AVMs is challenging. When formulating treatment protocols for patients, methods should be selected and applied alone or in combination according to the patient's condition and requirements, to obtain the best outcome between function, aesthetics, and efficacy.

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### CONFLICT OF INTEREST

The authors declare no conflict of interest.
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