

· 专家共识 ·

Expert Consensus

头颈部血管瘤及脉管畸形手术治疗专家共识

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[摘要] 血管瘤及脉管畸形是头颈部常见的良性脉管源性疾病,随着对非手术方案的深入研究与广泛应用,大量患者在微创或无创情况下得以改善或治愈疾病,但在特定情况下,手术依然是血管瘤及脉管畸形综合序列治疗中不可替代的治疗手段。然而,针对头颈部血管瘤及脉管畸形,如何选择和应用手术诊疗技术,学术界尚未形成规范和共识。为进一步提高国内同行对头颈部血管瘤及脉管畸形的治疗水平,规范和统一手术治疗策略,本文汇集全国多所医学院校及附属医院相关专家的讨论意见,借鉴和参考国内外相关最新成果与诊治经验,从头颈部血管瘤及脉管畸形的临床诊断及术前准备、手术适应证及策略、术后护理及观察、术后常见并发症及处理、随访及社会心理支持等方面,制定此专家共识,以帮助和指导相关专业的医师开展规范化手术治疗。

[关键词] 头颈部;血管瘤;脉管畸形;手术治疗;专家共识

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Expert consensus on surgical treatment of hemangiomas and vascular malformations in the head and neck

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[Summary] Hemangiomas and vascular malformations are common benign vascular diseases in the head and neck region. The evolution and extensive implementation of non-surgical therapeutic modalities have resulted in considerable improvement or even total remission in a significant number of patients in minimally invasive or non-invasive procedures. Nonetheless, under certain circumstances, surgical treatment remains an irreplaceable treatment modality within the comprehensive treatment sequence for hemangiomas and vascular malformations in the head and neck. However, there is no established standard or consensus in the academic community on how to choose and apply surgical treatment. Therefore, to enhance the treatment efficacy of hemangiomas and vascular malformations amongst domestic peers, and to standardize surgical treatment protocols, this consensus brings together the discussion opinions of relevant experts from many medical schools and affiliated hospitals nationwide. Based on the latest domestic and international research results and diagnosis and treatment experience, we formulated this expert consensus from the aspects of preoperative diagnosis and preparation, surgical treatment indications and strategies, postoperative care and observation, common complications and treatment after surgery, follow-up and social psychological support. It is expected to be used in clinical practice and to guide relevant professional doctors carrying out treatment.

[Key words] Head and neck region; Hemangiomas; Vascular malformations; Surgical treatment; Expert consensus

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血管瘤 (hemangioma) 和脉管畸形 (vascular malformation) 是头颈部常见的脉管源性疾病。根据病变的发病机制、生物学特性及远期预后,血管瘤属于良性血管源性肿瘤,其中,婴幼儿血管瘤和先天性血管瘤是最常见的 2 种类型,而脉管畸形则属于良性脉管发育性畸形,常见的脉管畸形类型可细分为静脉畸形、毛细血管畸形、动静脉畸形和淋巴管畸形等,各类型脉管畸形既可独立存在,也可以多类型混合或综合征的形式出现^[1-5]。虽然血管瘤及脉管畸形均属良性病变,但因其高发于头颈部这一特殊部位,轻者可导致容貌损毁或畸形,影响视力、呼吸、发音、进食等生理功能,严重者可导致病变部位反复出血、

疼痛、感染、失明,甚至引发死亡,因疾病导致的社交及心理障碍还会进一步影响患者及家庭的正常生活^[5-8]。因此,针对头颈部血管瘤和脉管畸形开展系统和规范的治疗,具有重要的临床价值和社会意义。根据国际脉管疾病研究学会 (International Society for the Study of Vascular Anomalies, ISSVA) 的分类及相关治疗建议^[1-5],血管瘤及脉管畸形相关综合治疗的总体原则是:根据疾病分类,对症选择和应用不同的治疗手段,而各种手段的组合序列及治疗时机则由病变的致病机制、基本病程、疾病部位、远期危害等多种因素综合决定。随着对非手术治疗方案(局部或全身应用药物、硬化治疗、激光治疗、光动力治

疗、介入栓塞治疗等)的深入研究与广泛应用,针对头颈部血管瘤及脉管畸形的治疗思路已经发生了巨大改变,大量患者在微创或无创情况下得以改善或治愈疾病。但是,对于部分血管瘤及脉管畸形病例,手术仍然是综合序列治疗中最重要的环节,且具有不可替代的地位^[2-4,9-11]。目前,关于手术治疗的适应证和具体策略,学术界尚未形成相关的诊疗共识或规范。

为进一步提高国内同行对头颈部血管瘤及脉管畸形的诊治水平,规范和统一手术治疗方案,我们组织国内多所医学院校及附属医院相关专家,针对头颈部常见血管瘤及脉管畸形类型,参考国内外相关治疗指南、共识及文献资料,共同制定了本专家共识,内容包括临床诊断及术前准备、手术适应证及策略、术后护理及观察、术后常见并发症及处理、随访及社会心理支持等,旨在为头颈部血管瘤及脉管畸形的手术治疗提供全面指导,促进其推广应用。

1 临床诊断与术前准备

准确的临床诊断与术前评估,是制定治疗计划的基础与前提。血管瘤及脉管畸形的 ISSVA 分类已得到国内外同行及研究人员的广泛认可^[1-5],因此,在本专家共识中,相关疾病的命名、诊断及分类均以最新的 ISSVA 分类(2018 年)为准,不再予以赘述。本共识结合手术治疗适应证、过程及风险,进一步丰富了临床诊断及术前准备的相关细节。

首先,头颈部血管瘤和脉管畸形的诊断高度依赖疾病的自然病程、既往治疗史(治疗方案及效果)及相关体征,相关病史是最终是否选择手术作为治疗方案的重要依据。因此,接诊医师应详细询问和记录患者相关情况,尤其是患者既往针对其所患疾病接受的治疗方案及效果^[6-7]。其次,在综合患者基本情况(年龄、性别、出生情况等)及全身状况的前提下,针对病变部位进行全面且细致的专科检查(病变部位、范围、颜色、质地、与邻近重要解剖结构的关系、皮肤温度、压缩性、搏动感、体位移动试验、穿刺液性状等),评估器官及功能累及情况(视力、吞咽、呼吸、语音、咬合关系、头面颈部相关神经功能等)。再次,各类辅助检查在血管瘤和脉管畸形的临床诊断与术前准备过程中同样发挥着重要作用。通过各类辅助检查,一方面,辅助诊断病变的类型及范围;另一方面,可用于评估上气道区域(口底、舌、口咽及喉部等部位)在治疗前、治疗后的受累情况及治疗风

险^[10,12-17]。常见的辅助检查包括彩色超声多普勒、MRI/MRA、CT/CTA/CTV、数字减影血管造影(digital subtraction angiography, DSA)、电子喉镜、支气管镜等。

除完善上述评估及其他常规术前检验(血常规、尿常规、粪便常规、肝功能、肾功能、肝炎病毒、艾滋病病毒、梅毒等)外,存在下述情况时,还应予以对症处理:①对于罹患巨大血管瘤或脉管畸形、可疑合并相关综合征或存在先天性心脏病史的患者,应在术前通过心肌酶谱、心电图、超声心动图评估有无心功能不全、心脏或主动脉结构异常等情况^[18-20]。②对于罹患巨大血管瘤或脉管畸形、皮肤紫癜或可疑存在严重凝血功能障碍的患者,应予以血栓弹力图、凝血功能、血小板功能、维生素 K、D 二聚体等检查,初步排除局限性血管内凝血(localized intravascular coagulation, LIC)及卡-梅现象(Kasabach-Merritt phenomenon, KMP)^[21-22];对于合并凝血功能障碍的患者,应在术前积极调整患者的凝血功能,对症应用相关药物或血液制品,结合手术方案及出血风险,完善术中及术后输血准备^[10,21,23-25]。③对于病变反复出血、进食障碍的患者,应注意血常规、肝功能、电解质及营养状况^[29]。④对于罹患全身多发血管瘤的患者,术前应完善甲状腺功能检查,排除甲状腺功能低下^[26];同时,还需完善肝脏彩超检测,排除肝脏血管瘤。⑤对于面部红斑沿三叉神经区域分布的患者,应排除 Sturge-Weber 综合征。该综合征可侵及软脑膜并引起婴幼儿惊厥、智力障碍或其他神经功能损害,还需接受眼压、眼底检查以筛查青光眼及脉络膜血管瘤^[27]。⑥对于位于眶部、耳周、咽部、颅底等区域的病变,需请眼科、耳鼻咽喉科、神经外科等相关科室会诊,评估相关器官及功能受损情况;同时,术前需由麻醉科会诊,评估手术的麻醉风险并制定相应的插管方案、麻醉用药方案、完善紧急处理预案等^[12,14-16,28-30]。⑦对于孕妇、婴幼儿、老年人等特殊人群,手术适应证和禁忌证也有一定差异,需结合患者基础身体状况、相关科室会诊意见、麻醉科评估结果等,综合制定个性化围术期治疗及护理方案^[16,21]。

最后,在完成全部评估并最终决定手术治疗后,应与患者及家属进行详细的术前谈话和解释,了解患者及家属的预期,让其充分知晓手术目的、过程、可能出现的风险、预后、术后护理及随访,并签署相关知情同意书。由于头颈部血管瘤及脉管畸形的复杂性,大量患者难以通过一次手术或单一的治疗策

略达到满意的疗效,开展相关治疗前,需充分告知患者及家属相关治疗的长期性和综合性。医患双方的相互理解和全程配合,是实现手术治疗目标的重要基础和前提^[8,31]。

2 手术适应证及策略

2.1 血管瘤

根据疾病的自然病程,血管瘤分为婴幼儿血管瘤(infantile hemangioma, IH)和先天性血管瘤(congenital hemangioma, CH),其中,具有明显增殖期和自行消退特征的婴幼儿血管瘤,是婴幼儿最常罹患的一类血管瘤。根据血管瘤的风险特征等级,相较于其他部位,头面部区域的婴幼儿血管瘤均属于高度或中度风险,即瘤体的快速进展或者不当干预,可引起严重瘢痕、溃疡、畸形,甚至影响邻近重要解剖区域(眼、鼻、气道)的结构及功能,存在较高的容貌畸形风险。目前,婴幼儿血管瘤的主流治疗方案,是以局部或全身用药为主(β 肾上腺素能受体阻滞剂、皮质类固醇激素等),辅以激光或局部注射药物等方案^[32-39],其主要治疗目的和基本原则是,抑制血管瘤增殖、促进瘤体消退、减少瘤体残留,而手术并非婴幼儿血管瘤的早期和首选治疗方案^[32,37-41]。

与婴幼儿血管瘤不同,先天性血管瘤表现为患儿出生时即有明显病灶,出生后无明显增殖期。根据消退特征,先天性血管瘤可进一步分为快速消退型(rapidly involuting congenital hemangioma, RICH)、不消退型(non-involuting congenital hemangioma, NICH)、部分消退型(partially involuting congenital hemangioma, PICH)。其中,RICH 消退迅速,通常无需治疗,若消退后出现局部组织菲薄、脂肪缺失,可采取手术治疗以改善外观。对于 NICH 和 PICH,除了单发、局限、隐蔽、可完整切除是常规的手术指征外,如果瘤体影响正常生理功能或外观,并发严重溃疡、心力衰竭或出血时,也可采取手术切除作为治疗手段^[9,36,38]。

2.1.1 手术适应证 由于手术切除不是血管瘤的首选治疗方案,因此,通常应待血管瘤瘤体经局部或系统用药后,药物效果不佳或患儿出现严重副反应或并发症以致无法继续药物治疗,且外科干预具有必要性、急迫性的前提下,经外科医师严格评估相关指征后,再予以手术治疗^[11,35-40,42-47]。血管瘤的手术适应证如下:①患儿>3岁,瘤体消退完成或自发改善不

再明显,瘤体所在区域仍存在较为严重的赘生组织、皮肤松弛、瘢痕或组织畸形,需改善外观与功能^[48-50]。②血管瘤瘤体发生严重感染、溃疡或反复出血,预计即使远期症状好转或瘤体消退后,仍存在组织畸形及手术风险者,可考虑早期手术治疗^[51-52]。③出现遮蔽视轴、视神经压迫、视力减退、眼球突出或移位等症状,非手术治疗无效或无法耐受的眼眶血管瘤^[53-54]。④血管瘤位于咽部、舌根等上呼吸道区域,经局部及全身用药治疗后无效,仍存在严重的气道梗阻危险^[55]。⑤血管瘤位于唇、鼻等区域,经局部及全身用药治疗无效,远期容貌畸形风险高^[40,56-57]。⑥经局部及全身用药治疗后无效或消退较慢,血管瘤位置隐蔽(如口腔黏膜、头皮毛发等区域),可完整切除。⑦现有检查手段无法明确诊断,需活检以明确病变性质,鉴别并排除其他类型肿瘤或病变。

2.1.2 手术策略 血管瘤的手术治疗属于对症治疗,需在保证不损伤面神经、视神经、眼外肌等重要结构的前提下,根据疾病特点,予以针对性、个性化的手术设计和治疗。一方面,通过手术治疗,防止瘤体进一步侵害周围组织或引起累及全身的症状;另一方面,通过手术对病变区域的软组织畸形予以改善和纠正。因此,手术时机的选择需将血管瘤的自然消退病程考虑在内,在不影响患儿早期心理发育及社交生活的前提下(>3岁或学龄前)^[58],择期手术治疗。手术切口设计,应依照美容、隐蔽和微创的原则,将切口线尽可能隐藏于皮肤张力线或面部美容亚单位的交界处。在不影响器官及功能的前提下,若通过手术切除能够实现完全治愈的目标,可采取在正常组织边界内完整切除的治疗策略;若血管瘤病变范围分布广泛,且无法通过单次切除完整去除全部病变时,手术需基于面部美学亚单位和皮肤张力线的分布规律,有计划地多次、分期、分区进行,辅以激光治疗去除皮肤颜色异常,实现面部轮廓的美学对称性重建,尽可能在外形与功能方面取得满意效果^[41]。对于涉及呼吸道的血管瘤,可在内镜辅助下针对瘤体开展部分切除并辅以药物注射,解除呼吸道梗阻导致的生命危险^[55]。针对个别血管瘤消退或治疗后的组织菲薄、脂肪缺失、色素沉着等现象,可采取手术切除冗余皮肤、局部填充脂肪等方法以改善外观^[49-50,56,59]。

2.2 静脉畸形

静脉畸形(venous malformations, VM)是头颈部最常见的低流速脉管畸形,其内部存在丰富的血窦

结构,除部分界限较为清楚的局限性病灶外,多数病变呈弥漫性改变,边界欠清,对邻近组织的功能和形态影响较大。手术治疗存在创伤大、出血控制难、神经损伤风险高、复发快、无法根治、术后功能障碍、面部瘢痕或畸形等各类风险。因此,目前大范围、弥散型静脉畸形的首选和一线治疗方案是血管内硬化治疗(intravascular sclerotherapy),而非手术治疗^[21,34,60-62]。对于静脉畸形,其手术的目标及基本原则是,在保留头颈部基本外形及生理功能的前提下,尤其是保护重要的神经及肌肉组织,适度切除病变区域,实现外观和功能改善,提高硬化或激光治疗效果,避免医源性损伤或功能障碍。因此,作为多学科、序列化治疗复杂静脉畸形的补充性手段,手术仅在必要情况下实施。

2.2.1 手术适应证 头颈部静脉畸形的病变性质、部位及累及范围十分复杂,因此,手术需在保留头颈部基本外形及生理功能的前提下实施,尤其是保护重要的神经及肌肉组织。静脉畸形的手术适应证如下^[2-3,6-7,10,12-13,28,46,63-65]:①范围局限、边界清晰、未累及重要解剖结构的静脉畸形,如舌黏膜、颊黏膜、颈部静脉畸形,在不影响美观及器官功能的前提下,通过手术切除可实现完全治愈。②大范围、弥散型静脉畸形,且已出现眼睑下垂、面部软组织萎缩、难以改善的神经或肌肉疼痛等功能障碍^[28,66]。③引发口唇闭合困难、舌体运动受限、严重影响呼吸及吞咽功能的唇部或舌体静脉畸形(巨唇或巨舌畸形)^[13,66-69]。④经过硬化治疗后,头颈部静脉畸形区域出现软组织塌陷、瘢痕或局部皮肤坏死,部分情况下可合并颌骨畸形^[70-72]。⑤软组织区域的静脉畸形病变经治疗消除后,仍存在严重的颌骨畸形及咬合关系紊乱,需行手术治疗针对颌骨畸形及外观缺陷予以整复。⑥硬化或介入治疗效果不佳的颌骨内静脉畸形^[64-65,73]。⑦难以单纯通过硬化治疗取得满意效果、涉及颅内外沟通的静脉畸形^[29]。⑧出现遮蔽视轴、视神经压迫、视力减退、眼球突出或移位等症状,非手术治疗无效或无法耐受的眼眶静脉畸形^[28,53]。⑨局限于舌根、声门上、咽部等特殊部位,存在气道阻塞风险,经内镜辅助下可开展手术切除的静脉畸形^[12,30,66]。⑩位于头颈部解剖结构深面的静脉畸形,通过局部手术进而充分暴露病变、保护神经、提高硬化或激光治疗效果^[74]。⑪现有检查手段无法明确诊断,需活检以明确病变性质,鉴别并排除其他类型肿瘤或病变。

2.2.2 手术策略 对于范围局限、边界清晰、未累及重要解剖结构的静脉畸形,在不影响美观及器官功能的前提下,若通过手术切除能够实现病变的完全治愈,可采取在正常组织边界内手术切除的策略;而对于因多次硬化治疗导致病变区域外界不清时,可在病变边界外的正常组织内设计切口,从而将病变完整切除。而对于大范围、弥散型静脉畸形,可通过分次、逐步切除病变区域,减小病变体积及实性成分,去除静脉石的无菌性刺激,解除神经或肌肉压迫,修整软组织以改善外观和功能,提高远期硬化治疗效果。若硬化治疗后的病变区域出现软组织塌陷、瘢痕或坏死,可通过筋膜瓣或筋膜脂肪瓣予以填充^[75]。合并骨畸形时,可通过截骨或轮廓修整改善外观。对于颌骨静脉畸形,可采取刮治的手术方案^[64-65,73]。对于严重影响患者呼吸道通畅、累及眶部或颅内外的静脉畸形,通常可通过部分切除或联合硬化治疗,改善气道及视力情况,提高后续治疗的安全性^[12-13,15,28]。此外,除应用手术开展对症切除或修复外,手术还可作为其他治疗的辅助手段^[13,76]。①翻瓣激光治疗:局部翻瓣手术能够更充分地暴露头颈部解剖结构深面的病变区域,避免神经损伤,提高激光治疗效果^[74,77]。②网状分隔缝扎手术(图 1):该手术可区间化分隔病变,一方面,可减少术中及术后创面出血,延缓拆线;另一方面,将大范围弥散型静脉畸形分隔后再开展多点硬化治疗,可进一步提高硬化效果^[13]。③气管切开术:对于大范围、弥散型头颈部静脉畸形,接受硬化治疗前/后存在局部肿胀压迫气道的风险,若气管插管难度及风险较大时,可行预防性气管切开术^[13-14,17]。

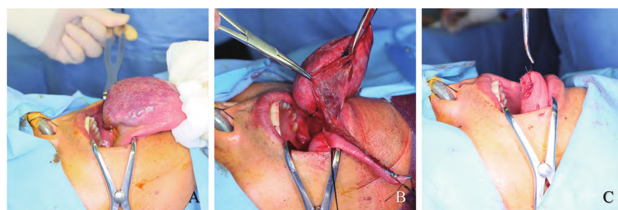


图 1 舌静脉畸形(巨舌畸形)的手术治疗。A.术前舌体外观,舌体前端见一巨大静脉畸形;B.于正常舌体处分隔缝扎,暂时阻断血流供应,于分隔内部完整切除病变;C.术后舌体外观

Figure 1 Surgical resection of tongue venous malformations (macroglossia). A. Appearance of the tongue before surgery. There was a huge venous malformation in the anterior part of the tongue; B. Suture tongue along the normal margin, temporarily block the blood supply, and completely resect the lesion inside the septum; C. Postoperative appearance of the tongue

2.3 毛细血管畸形

毛细血管畸形(capillary malformations, CM)也称为微静脉畸形(venular malformations)、葡萄酒色

斑(port-wine stains, PWS)或鲜红斑痣,病变主要表现为边缘清晰且不规则的红斑,除中线型毛细血管畸形可自行消退外,多数病灶会逐步进展,表现为颜色加深,累及皮肤区域增厚,由粉红型、紫红型发展为增厚型,并伴有结节样增生,部分严重者可伴有局部软硬组织增生或畸形^[78]。脉冲染料激光治疗(pulsed dye laser, PDL)是目前早期 CM 的首选治疗方案^[2,10,79],此外,光动力治疗、非相干光治疗等也已作为辅助手段应用于临床^[80]。手术治疗主要是作为非外科手段治疗无效或无法进一步改善病变时的重要选择。

2.3.1 手术适应证 对于头颈部毛细血管畸形,当非外科手段治疗无效或无法进一步改善病变时,手术清除残留病灶或进一步改善外观畸形是主要治疗目标。因此,毛细血管畸形的手术适应证一般包括以下情况:①接受多次非手术治疗(如激光、光动力治疗)后无效或无法进一步改善病情、病灶增生严重的毛细血管畸形,尤其是病变显著增厚呈深紫色、局部存在鹅卵石样结节、深部病变累及皮肤和皮下血管丛,或已出现巨唇、口唇功能障碍、颌骨发育畸形等严重功能或外观障碍^[46,67,78-79,81-83]。②现有检查手段无法明确诊断,需活检以明确病变性质,鉴别并排除其他类型肿瘤或病变。

2.3.2 手术策略 通过手术清除残留病灶或改善面唇部外观畸形时,应在保存头颈部基本外形及生理功能的前提下,尤其是保护头颈部重要神经结构,依据对称、协调的原则予以适度切除。随后,通过同期修复重建,尽可能实现外观和功能改善,提高患者生活质量;同时,应尽可能避免医源性损伤或功能障碍。由于毛细血管畸形病变范围分布广泛,手术需要基于面部美学亚单位和皮肤张力线的分布规律,有

计划地多次、分期、分区进行,以实现面部轮廓的美学对称性重建^[79,84]。对于唇部的增厚型毛细血管畸形,可采取“三明治”式切除,在保留口轮匝肌闭合功能的基础上,尽可能恢复局部外形,去除冗余组织。根据缺损部位及范围,手术切除后,可采取皮肤组织扩张、预构扩张皮瓣(图 2)等方案予以修复^[85]。若采取中厚或全厚皮片移植,植皮区远期存在术后色素沉着、瘢痕挛缩严重等风险和弊端^[86-87]。而对于部分无法通过单次切除去除全部病变的患者,可采用分次、分区切除结合激光/光动力治疗的序列治疗方案,逐步恢复面部外形,改善病变区域皮肤颜色。对于合并严重颌骨畸形的患者,在软组织矫正的基础上,联合正畸与正颌手术治疗(骨皮质修整术、Le Fort I 型截骨术、下颌骨矢状劈开术、下颌角修整术、鼻畸形矫正术、颏成形术等),能进一步改善患者面容及咬合功能^[79,82]。

2.4 动静脉畸形

动静脉畸形(arteriovenous malformations, AVM)是一类高流速、临床危害较大的先天性血管发育畸形。根据动静脉畸形的进展及特征变化,Schobinger 将其分为 I~IV 期^[88],随着疾病逐步进展,病变累及部位可出现疼痛、溃疡和反复出血,严重者可因血流动力学异常而出现心功能不全或心力衰竭。动静脉畸形治疗困难,复发率高,病情进展迅速且凶险。目前,介入栓塞治疗是颅外动静脉畸形的一线治疗方案^[10,89-91]。但在特定情况下,完整、彻底手术切除,能够实现病灶长期根治,解除因反复出血和血流动力学异常导致的生命危险;而介入联合手术的综合治疗方案,能有效减少术中出血,提高病灶切除率,从而减少病变复发的可能^[89-90,92]。因此,手术在头颈部

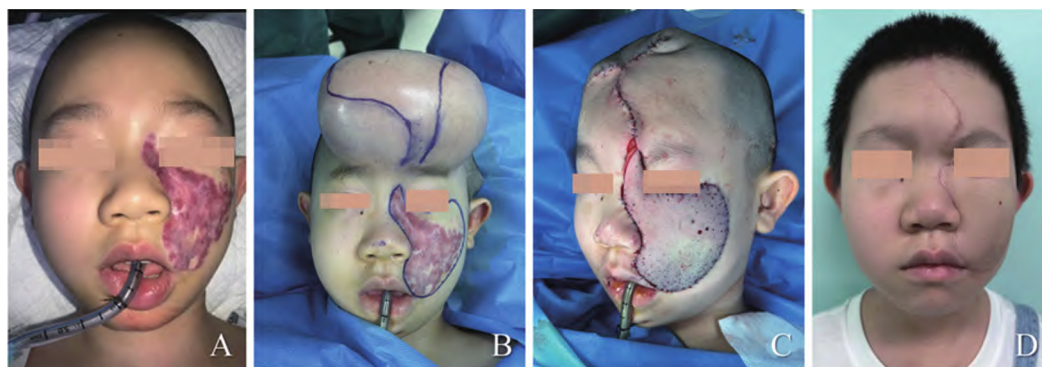


图 2 面部毛细血管畸形的手术治疗。A.术前外观;B.根据美学亚单位设计面部切口线,利用额部预构扩张皮瓣转移修复面部缺损;C.病变切除及修复后即刻外观;D.术后复查外观

Figure 2 Surgical resection of capillary malformations located at facial area. A. Appearance before surgery; B. Incision line was designed according to subunits of aesthetics. The expanded frontal flap was used to repair facial defect; C. Appearance after lesion excision and immediate repair; D. Postoperative appearance

动静脉畸形的治疗中占有举足轻重的地位。

2.4.1 手术适应证 头颈部动静脉畸形手术治疗的相关适应证,一般包括以下情况^[2,10,46,89-91,93-97]:①对于 Schobinger I 期或 II 期动静脉畸形,若范围局限、边界清晰、未累及重要解剖结构,在不影响器官功能的前提下,手术切除可实现完全治愈。②对于 Schobinger III 期或 IV 期动静脉畸形,若全身基础情况(如患有充血性心力衰竭)可控,并已行介入栓塞(术前 1 周内),在控制出血、局部严重症状得以缓解的前提下,手术切除可实现完全治愈^[98]。③介入治疗远期效果不佳、介入栓塞后发生严重并发症(如组织缺损或坏死、栓塞材料外露)、病变反复发生严重感染或溃疡、神经或肌肉压迫导致剧烈疼痛无法改善的动静脉畸形^[99]。④经介入治疗后,动静脉畸形区域内的畸形血管团消失,局部组织存在畸形,需改善外观与功能;⑤引发视神经压迫、视力减退或眼球突出等症状的眼眶动静脉畸形^[28,100]。⑥现有检查手段无法明确诊断,需活检以明确病变性质,鉴别并排除其他类型肿瘤或病变。

2.4.2 手术策略 手术治疗头颈部动静脉畸形的核心原则是,对动静脉畸形病理生理机制的正确理解,错误地通过近心端栓塞、手术结扎供血血管、牺牲静脉引流通路而不治疗病灶或动静脉瘘区域,已被证明会导致疾病的快速复发和局部坏死^[101-102]。切除不彻底是早期快速复发的重要原因,因此,要尽可能完整切除病变,尤其是通过手术切除可实现完全治愈时,手术切缘不应过于保守,可采取正常组织边界内切除的策略。对于暂时无法实现一期完整切除的大范围动静脉畸形,可通过介入治疗实现动静脉畸形内部的去血管化,随后再通过手术进一步实现病变区域功能与外形的改善和恢复。

对于符合手术指征的 III 期或 IV 期动静脉畸形,若全身基础情况(如患有充血性心力衰竭)可控,可在介入栓塞(术前 1 周内)后,在控制出血和缓解局部症状的前提下开展手术治疗,实现病灶的完全切除和患者安全利益的最大化。而针对颌骨动静脉畸形,目前通过介入栓塞治疗可实现控制出血和保存颌骨连续性的目标,因此,针对颌骨动静脉畸形,下颌骨节段性切除术已不作为常规推荐的治疗方案。而对于符合上述手术指征的颌骨动静脉畸形,尤其是继发感染或者存在栓塞材料外露者,可通过颌骨病灶刮除手术,实现病灶清除^[91,95-97]。由于颌骨动静

脉畸形好发于儿童及青少年,手术时机和颌骨发育之间的平衡仍需予以慎重把握和评估。当病灶切除后,依据手术切除部位及范围,同期通过自体骨移植(腓骨、髂骨等)、皮肤组织扩张、预构扩张皮瓣、游离皮瓣等方式,利用远处健康组织予以颌骨重建或软组织修复,恢复咬合关系。通常不建议皮片移植(图 3)。由于动静脉畸形相关病理及生理机制非常复杂,长期随访十分必要,早期发现,及时干预,避免因病情快速进展导致后续治疗难度增加。



图 3 下唇、下颌及颊部软组织动静脉畸形的手术治疗。A.术前外观; B.术后外观(背阔肌皮瓣修复)

Figure 3 Surgical resection of arteriovenous malformations located at soft tissues of the lower lip, mandibular and buccal area. A. Appearance before surgery; B. Postoperative appearance (surgical defects were reconstructed with free latissimus dorsi myocutaneous flap)

2.5 淋巴管畸形

淋巴管畸形(lymphatic malformations, LM)是先天性淋巴管发育畸形,根据淋巴管畸形内部囊腔大小,分为巨囊型、微囊型和混合型。随着对淋巴管畸形遗传学背景及致病机制理解的逐渐深入,淋巴管畸形的治疗手段也在逐步多样化。目前,主流和常用的治疗手段包括硬化治疗、激光治疗、手术切除、靶向药物治疗等^[2,7,9-10,103-104]。虽然对于巨囊型和混合型淋巴管畸形,硬化治疗通常可取得较为满意的远期疗效,但对于范围较大的弥散性微囊型淋巴管畸形,

硬化治疗及靶向药物治疗的疗效仍不太理想^[103,105-107]。此外,不同于其他类型的脉管畸形,淋巴管畸形的囊腔内部为淋巴液,在感染或炎症刺激下,淋巴管畸形区域极易发生病灶内出血或快速增大,导致病变进展或复发,进一步影响相关治疗的远期疗效。因此,手术仍是目前淋巴管畸形的重要治疗选择之一。

2.5.1 手术适应证 头颈部淋巴管畸形的手术治疗基本原则是,在保存头面部基本外形及生理功能的前提下,尤其是保留神经结构,可予以适度切除,改善患者外观和功能,提高生活质量;同时,应尽可能避免医源性损伤或功能障碍。头颈部淋巴管畸形手术治疗的相关适应证,一般包括以下情况^[2,10,46,106,108-109]:①范围局限、边界清晰、未累及重要解剖结构的淋巴管畸形,在不影响美观及器官功能的前提下,通过切除可实现完全治愈目标^[109]。②引发口唇闭合困难、舌体运动受限、严重影响呼吸及吞咽功能的舌淋巴管畸形(巨舌畸形)^[68,107]。③引发口唇闭合困难、颜面部严重畸形的颊部或唇部淋巴管畸形(图4)^[67,83]。④硬化治疗或口服靶向药物治疗后,反复感染、出血、肿胀且(或)病变消退效果不理想的微囊型淋巴管畸形^[105-106]。⑤引发视神经压迫、视力减退或眼球突出等症状的眼眶微囊型淋巴管畸形^[28]。⑥颈部、口底等区域的淋巴管畸形,存在危及生命的并发症风险(如呼吸道梗阻、病变压迫重要的解剖结构等),硬化治疗或口服靶向药物治疗后,病灶消退不理想^[110-112]。⑦拟接受靶向药物治疗的患者,需通过活检明确病变具体组织学类型及突变情况,用于筛选敏感的靶向药物;⑧现有检查手段无法明确诊断,需活检以明确病变性质,鉴别并排除其他类型肿瘤或病变。

2.5.2 手术策略 依照上述适应证,在不影响美观及器官功能的前提下,手术切除可完全治愈时,可采取正常组织边界内切除的治疗策略。对于具有手术治疗指征、范围较广、无法一次完全切除的弥漫性微囊型淋巴管畸形,可采取分次、部分切除的治疗策略,以达到降低手术致畸可能性、恢复器官功能、改善外观的目的^[107,112];同时,可将切除的病灶进一步行基因检测以筛选合适的靶向药物,以便于设计综合治疗方案。相比单纯手术或硬化治疗,手术联合硬化治疗、手术联合靶向药物治疗的综合治疗策略,能够有效减少病变区域术后淋巴液渗出量,进一步提高治疗效果^[111-112]。此外,还可通过淋巴管-静脉吻合

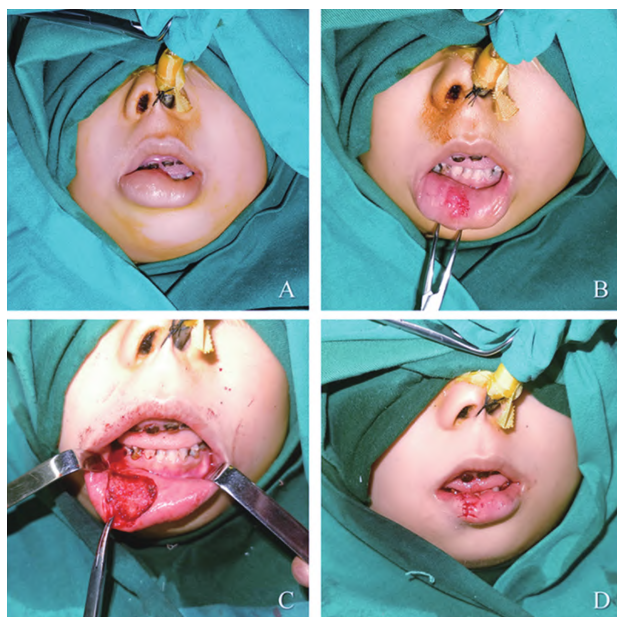


图4 下唇淋巴管畸形(唇畸形)手术治疗。A.右侧下唇肿胀畸形;B.右侧下唇内侧黏膜见粟粒状淋巴水疱;C.病变切除后创面;D.术后下唇外观

Figure 4 Surgical resection of lymphatic malformations in the lower lip (lip malformations). A. Swelling and malformations of the right lower lip; B. Miliary change was observed in the inner mucosa of the right lower lip; C. Wound appearance after lesion resection; D. Postoperative appearance of the lower lip

术,重建淋巴管畸形病变区域的静脉回流通道,促进和改善淋巴回流,进而缩小囊腔体积^[113-115]。发生于舌腹及口底部位的巨大巨囊型淋巴管畸形,常伴言语障碍和气道阻塞,接受硬化治疗或手术治疗前/后存在局部肿胀压迫气道的风险,当气管插管难度及风险较大时,可行气管切开术^[116]。

3 术后护理与观察

头颈部血管瘤和脉管畸形患者术后出血风险较高,部分患儿年龄小、配合差等,更增加了护理及观察的难度^[12-13,76,108,117-118]。因此,在围术期,应对患者的基本生命体征(如血压、心率、呼吸、脉搏、血氧饱和度等)予以密切观察,做好凝血功能调整、血液及血液制品的准备^[10,21,23]。

对于头颈部Ⅰ类切口,需注意日常换药,保持局部创面干燥、清洁,促进伤口恢复。对于口腔内Ⅱ类切口,需引导患者术后早期漱口、刷牙,严格保持口腔卫生,防止局部感染。术后应密切观察手术区域局部症状的改善与变化,如疼痛、肿胀、引流量及性质、皮肤颜色等,早期发现症状变化,并积极预防切口感染和对症治疗。尤其要注意的是,相比其他类型的脉管畸形,在感染或炎症刺激下,淋巴管畸形区域更加

容易发生病灶内出血或快速增大,导致局部进展或复发,进一步影响疗效。因此,淋巴管畸形患者接受手术后,更应注意和加强围术期抗感染、抗肿胀治疗。

对于存在呼吸道梗阻风险的患者,尤其是手术区涉及舌、口底、咽旁等部位时,气道通畅与否是整个围术期都要密切关注的核心问题,术后应密切观察有无气促、紫绀、“三凹征”等症状,术后应考虑采取健侧卧位,以防止误吸或舌后坠;定时更换体位,防止产生压力性损伤,必要时可予以低流量吸氧^[10,12,15-16]。对于行气管切开术的患者,术后应密切观察患者基础生命体征,做好气管切开伤口及套管的日常维护,待本阶段治疗结束且确保患者自主呼吸通畅后,再行试堵管和拔除气管插管^[15-17]。

此外,若患者术前存在进食或营养障碍,术后要积极给予营养支持治疗,制定个性化营养护理方案,密切关注电解质平衡变化,及时对症处理^[118]。对于接受游离皮瓣移植修复的患者,还要密切注意受区皮瓣情况(颜色、质地、肿胀、皮肤纹路、皮温等)及供区的康复情况^[94]。

4 术后常见并发症及处理

对于头颈部血管瘤和脉管畸形患者,术后可能出现的并发症主要包括疼痛、出血、感染、暂时性/永久性神经损伤、瘢痕、颌面畸形、肺栓塞、失血性休克、皮肤及皮瓣坏死、肺炎等^[10,12-13,21,23-24,40,44,76,79,94,108-119]。患者年龄越小,相关手术并发症的发生率通常越高^[44]。适当和及时的镇痛治疗,可极大提高患者舒适度。为防止出血和感染,术中应遵守无菌操作原则,做到严格止血,术后给予适当的抗感染及抗肿胀治疗,加强伤口换药与护理。为避免重要神经损伤,术中应尽可能保留和保护神经组织,避免过度牵拉和损伤。一旦术中损伤神经,应在术后积极予以营养神经类药物支持。为防止休克或弥散性血管内凝血(disseminated intravascular coagulation, DIC)的发生,要根据患者术前及术后的凝血状况,制定个性化的抗凝治疗方案,尤其是要预防深静脉血栓和肺栓塞发生^[23-25,118]。

5 随访及社会心理支持

手术治疗后,应对患者进行定期随访,监测病情变化,评估治疗效果,如症状缓解程度、病灶缩小程度、生活质量有无改善等^[2-3,10,79]。在随访频率方面,建议分别于术后第 1、3、6 个月随访评估。通常而言,孤

立型病损的手术效果较好,复发率较低。复合型和弥散型病变的手术效果较差,易出现术后并发症和复发。而在各类脉管畸形中,淋巴管畸形因全身或局部感染后的复发最为常见^[3,7,10,12]。因此,在随访过程中,若发现残留病灶或复发迹象,需及时采取干预措施,以避免病情进展。同时,需对患者或患儿家长进行健康教育和心理疏导,增强其信心和勇气,提高生活质量^[31,120]。

6 结语

本共识汇集了全国多所医学院校及附属医院相关专家的讨论意见,从临床诊断及术前准备、手术适应证及策略、术后护理及观察、术后常见并发症及处理、随访及社会心理支持等方面提出观点并达成共识,期望能帮助和指导相关专业的医师开展手术治疗。随着相关疾病诊疗方案的进展与更新,本共识也将及时予以修订,以期及时、有效、可靠地指导头颈部血管瘤及脉管畸形的规范化手术治疗。

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