·专题讲座·

Chiari畸形外科治疗回顾与进展

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【摘要】 Chiari畸形的外科治疗始于1932年，70余年来随着医学技术的不断进步，其手术方式不断改进。然而，由于发病机制至今尚未明确，致使不能形成一种临床公认的最佳外科手术方法，手术过程中不同层次的操作步骤或具体方法仍存有争议。本文拟就Chiari畸形的概念、发病机制、诊断与神经外科手术治疗方法进行回顾，并对目前的热点问题和新技术应用进行概述。

【关键词】 Arnold-Chiari畸形； 脊髓空洞症； 神经外科手术； 综述

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Surgical treatment of Chiari malformation: review and progress

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【**Abstract**】 The surgical treatment of Chiari malformation (CM) began in 1932. With the advance of medical technology, the surgical technique of CM is also in constant improvement. But due to its pathogenesis has not yet clear, there is no accepted optimal method, and different levels of the operation is still controversial. The author reviewed the concept, pathogenesis, diagnosis and surgical treatment of CM. The hot topics and new technological application were also reviewed in this article.

【**Key words**】 Arnold-Chiari malformation; Syringomyelia; Neurosurgical procedures; Review

自Chiari畸形（CM）的概念提出以来，问题总比答案多[1]，有关其最佳外科手术治疗方法在本专业领域内始终未达成共识。1932年，奥地利医师Cornelis Joachimus首次报告应用颅后窝减压术治疗Chiari Ⅰ型畸形（CM-Ⅰ）[2]，之后的很长时间内颅后窝减压术一直作为Chiari Ⅰ型畸形外科治疗的基本术式，由于该术式术后并发症发生率较高，如小脑下垂、脑脊液漏、假性脑膜膨出、无菌性炎症性发热等，许多学者提出了各种不同的改良术式。这些术式虽然在技术操作上略有改进，但对于减压窗大小、是否剪开硬脑膜、打开蛛网膜、切除下疝的小脑扁桃体、扩大硬脑膜及如何缝合等问题，至今仍未取得共识性结论[3]。近年来，随着神经外科手术技术的不断进步和发展，以及临床经验的积累，Chiari Ⅰ型畸形的外科手术治疗水平也在不断提高，为了更好地了解Chiari Ⅰ型畸形神经外科治疗的临床研究进展，笔者拟对其历史发展过程和研究现状进行概述。

一、Chiari畸形的命名及发病机制

Chiari畸形最早由奥地利医师Hans Chiari在1891年描述并命名，之后亦有学者称之为Arnold畸形或Arnold-Chiari畸形等。近年国外学者趋同于Chiari畸形这一命名，以纪念Hans Chiari教授早期对该病研究所做出的贡献[4]。国内ICD-10疾病编码称之为先天性小脑扁桃体疝。

Hans Chiari医师按照Chiari畸形的解剖形态特征将其分为4型，尽管后来亦有学者出于手术方式的选择提出了不同的分型方法[5-9]，但因争议颇多而未能被临床广泛采纳，目前仍沿用Hans Chiari医师的早期分型，即Ⅰ～Ⅳ型，其中以Ⅰ型最为常见，笔者主要针对Chiari Ⅰ型畸形进行概述。

Chiari畸形的发病机制尚不十分清楚，目前广

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泛接受的假说是：Chiari畸形是由于轴索中胚层发育畸形而致颅后窝发育迟滞和后脑拥挤，或腰池-腹腔过渡分流所致小脑扁桃体下疝[10]。有学者通过对Chiari畸形患者和正常人MRI及CT影像进行测量，发现Chiari畸形患者颅后窝容积较正常人小，这也在一定程度上验证了该假说的可能性[9-11]。西班牙学者Royo-Salvador等[12]则认为，脊髓拴系的牵拉可能是造成小脑扁桃体下疝和脊髓空洞症的原因。Chiari畸形患者大多合并脊髓空洞症，尽管有多种假说可以解释脊髓空洞症的发生机制[13-16]，但目前广泛接受的发病原因是颅后窝狭小和小脑扁桃体下疝导致枕骨大孔区堵塞。因而Chiari畸形的外科治疗旨在解决颅后窝狭小和解除枕骨大孔区堵塞，主要手术方法包括颅后窝减压术（PFD）和颅后窝重建术（PFR），以及之后提出的枕大池重建术均基于这一理念。目前，对于Chiari畸形发病机制的研究仍在继续开展。

二、Chiari畸形的诊断标准和手术适应证

1. 诊断标准　以正中矢状位MRI检查小脑扁桃体下缘超过BO线3～5 mm作为诊断依据[12, 17-20]。BO线系指颅底点（basion）至颅后点（opisthion）的连线，以此线代表枕骨大孔平面。考虑到枕骨大孔后缘的厚度，以选取枕骨大孔后缘最下点为宜。寰枕融合患者需要仔细辨认融合点以判断枕骨大孔后缘的最下点，必要时需借助CT扫描。一般认为，小脑扁桃体下缘超过BO线＜3 mm可不诊断为Chiari畸形，＞5 mm者可确诊为Chiari畸形[17, 20]，BO线位于3～5 mm之间时则应考虑小脑扁桃体是否为旁中线结构，矢状位MRI的扫描层次可能未扫描至小脑扁桃体的最下缘，临床可酌情分析处理。

2. 手术适应证　（1）符合诊断标准且有相关症状。（2）合并大而扩张的脊髓空洞，并伴有相关慢性神经损害症状。（3）排除继发性小脑扁桃体下疝和其他相关疾病如枕寰枢关节失稳、脊髓粘连、脊髓肿瘤、脊髓拴系综合征等。以下情况在制定手术方案时应谨慎处理：（1）单纯Chiari畸形但无脊髓空洞形成，临床症状轻微的患者可以保守观察。（2）小脑扁桃体下疝＜5 mm患者。（3）脊髓空洞较小或萎缩塌陷者，应慎行分流术。（4）合并颅底凹陷、寰枕融合或脊柱侧弯等疾病的患者，应注意进行脊柱关节方面的检查和评价。（5）病程长、体质较弱者应避免实施创伤较大的手术治疗。（6）有难产史的Chiari畸形患者，应考虑枕骨大孔区可能存在粘连。

三、主要手术方法

1. 颅后窝减压术　颅后窝减压术于1932年首次应用于临床[2]，其目的是通过切除部分枕骨以扩大狭小的颅后窝。主要步骤包括骨性减压、硬脑膜（外层）减压。该术式要求骨性减压充分，骨窗大小须达到5 cm×6 cm。目前，对于减压过程中是否应该保持硬脑膜的完整尚存有争议：部分学者认为，在减压过程中保留硬脑膜完整可避免血液进入蛛网膜下隙和形成脑脊液漏；但也有一些学者认为，保留硬脑膜完整虽好但不能达到充分减压之目的，为了有效降低颅内压应该完全敞开硬脑膜。然而，经过不断的临床实践，越来越多的学者认识到“敞开硬脑膜”的缺陷：首先，过大的骨性减压窗可使小脑因失去支撑而下垂，既不能形成有效的脑脊液循环，同时还会诱发难以控制的枕颈部疼痛。其次，硬脑膜完全敞开后，脑组织与肌肉、筋膜等组织直接接触，血液流入蛛网膜下隙形成广泛粘连、脑脊液漏，以及持续性发热等并发症。鉴于该术式的上述缺点，有学者在此基础上对其进行改良，从而建立了“颅后窝重建术”。

2. 颅后窝重建术　颅后窝重建术亦称颅后窝减压并硬脑（脊）膜成形术（PFDD）。该手术方式于1994年首先由Sahuquillo等[21]提出，为颅后窝减压术的改良术式。手术中首先进行广泛的颅后窝减压，保持蛛网膜完整，显微镜下“Y”形切口切开硬脑膜，以同种冻干硬脑膜扩大修补切口，悬吊3～4针形成帐篷状以保持脑脊液通畅；经对10例Chiari畸形患者的观察发现，所有患者均于颅后窝形成枕大池，后脑平均抬高6.20 mm，脊髓空洞缩小，与传统的颅后窝减压术相比，小脑扁桃体下疝还纳程度明显高于对照组。Vanaclocha和Saiz-Sapena等[22]经临床观察发现，采用冻干硬脑膜修补切口易发生脑脊液漏、皮下积液，甚至假性脑膜膨出等并发症，而以自体筋膜作为切口修补材料则能很好地避免上述并发症。Klekamp等[23]通过对比大骨窗和小骨窗减压颅后窝重建术的临床疗效，发现小骨窗减压扩大修补硬脑膜的效果与大骨窗减压效果相当，但前者术后并发症发生率明显降低。经不断改进和试用，颅后窝重建术已在临床广泛推广应用。

3. 枕大池重建术　该术式系由张远征等[24]于2000年首先提出，与颅后窝减压术或颅后窝重建术

相比有较大不同。该项术式的目的不是传统意义上的扩大颅后窝，而是重建被小脑扁桃体阻塞的枕大池，形成有效的脑脊液循环。该术式的过程为小范围骨性减压，即切开硬脑膜和蛛网膜，于软脑膜下切除部分小脑扁桃体，然后修补、缝合硬脑膜。而不要求扩大颅后窝，因此骨性减压范围小，使得手术过程趋向微创化。尤其是近5年来，我国有多位学者报道了改进方案。枕大池重建术经过优化和微创化后其皮肤切口仅2 cm左右，小范围骨性减压为0.50～1.50 cm，保留寰椎后弓。

四、相关争议、共识及发展趋势

1. 有争议的问题　Chiari畸形的最佳外科手术方法一直存在分歧，主要争论焦点为手术方式的安全性和有效性，目前尚缺乏多中心随机对照临床试验结果。张远征等[24]认为，可从缺陷分析的角度评价哪种手术方式能更好地减轻术后反应、降低术后并发症发生率，例如无菌性炎症、脑脊液漏、假性脑膜膨出、脑积水、小脑下垂、脊髓空洞不缩小需再次手术等。一项新的手术技术唯有在不增加并发症且能够达到手术目标时，方能被临床所接受。Chiari畸形术后最常见的并发症为无菌性炎症，主要表现为长期反复发热，其与硬脑膜、蛛网膜开放，缝合不严密形成漏口或缺口造成术区积液，以及血性脑脊液刺激，生物胶、骨蜡和人工硬脑膜等生物替代品吸收反应等因素有关。所有这些并发症均涉及到术中骨性减压窗大小、硬脑膜蛛网膜处理方法、小脑扁桃体下疝切除与否、是否应用人工硬脑膜、术后腰椎穿刺治疗等问题。(1)减压骨窗的大小：尽管大多数学者认为小范围骨性减压效果与大范围间无明显差异且术后不良反应轻微、并发症少[23,25-30]，但是仍有一部分学者坚持大范围骨性减压是必要的[31]。检索我国近5年神经外科核心期刊发表的文献，共检出与Chiari畸形治疗相关文献37篇，其中81.08%(30/37)的文献支持采用小骨窗减压。(2)硬脑膜的处理：部分学者认为硬脑膜扩大修补术可导致脑脊液漏和术后无菌性炎症发生率升高[32-33]，但大多数学者认为，硬脑膜扩大修补术可以保证和维持颅后窝重建术的效果，减少症状复发和再次手术率[28,32,34-35]。目前，对于自体筋膜和人工材料对手术效果的影响尚未取得共识，有学者认为自体筋膜可使术后并发症发生率降低[36]，也有学者认为人工材料具有较好的生物相容性，并不增加并发症的发生率[37]。检出的37篇文献中，83.78%(31/37)采用硬脑膜扩大修补术，而修补材料则认为自体筋膜优于人工修补材料。(3)小脑扁桃体下疝的处理：尽管早在1995年Fisher[38]即报告了小脑扁桃体下疝切除的神经外科手术经验，但国外学者对此项技术始终持谨慎的态度，近年来鲜见相关文献报道。我国自提出枕大池重建术以来，一直有对该项手术技术进行改进的文献报道，在检出的37篇文献中有26篇文献(70.27%)共报告900余例Chiari畸形患者经神经外科手术实施小脑扁桃体下疝切除，而且经软脑膜下切除小脑扁桃体下疝并未造成术后无菌性炎症和脑脊液漏等并发症发生率升高，患者术后亦极少出现神经功能损害或异常[34,39-41]。根据我国学者总结的微创枕大池重建术的特点[42]，术后反应轻，并发症如无菌性炎症、脑脊液漏、脑积水罕见，亦无小脑下垂或再次手术等事件发生，特别是通过术中B超监测实时量化评价手术效果，判断切除范围，对提高手术质量具有较高的临床实用价值。(4)近期研究特点：通过复习近期文献，我们还发现以下特点，即术中超声和多普勒超声的应用对手术方式的选择具有良好的判断价值，近年来临床应用逐渐增多[43-47]，而且术中超声监测对判断是否需要施行硬脑膜扩大修补术亦有帮助[43,48-49]；重视Chiari畸形合并其他相关病症的处理，尤其是脊柱侧弯、枕寰枢关节失稳和脊髓拴系综合征等[10-11,50-55]；对颅后窝由径线测量转向容积测量对Chiari畸形的机制和治疗方向具有参考价值[10,12]。

2. 对分流术的看法和认识　分流术包括脊髓空洞腹腔或胸腔分流术和脊髓空洞蛛网膜下隙分流术，但这些分流术均不作为Chiari畸形合并脊髓空洞症患者的Ⅰ期首选术式。一般认为，在主流术式难以实施或不奏效的情况下可以考虑采用分流术，但分流术的作用仅是平衡空洞和蛛网膜下隙之间的压力。

3. 有关术后疗效的评价　评价方法仍然沿用Tator等[56]所报告的方法，即症状稳定和改善为有效、症状加重为无效。对Chiari畸形合并脊髓空洞症的患者，若术后症状无改善或空洞不缩小，以及对枕大池形态不满意，原则上不建议再次手术进行修复；少数小脑扁桃体下疝明显、症状突出的患者，可于确定枕骨大孔区无粘连发生时实施枕大池重建术。一般情况下，应于术前对患者进行全面系统

地评价，确定是否合并脊髓拴系综合征、枕寰枢关节失稳、脊柱侧弯等情况，有合并症者可以考虑实施脊髓终丝切断术或脊髓空洞分流术。近年来，采用脊髓终丝切断术治疗Chiari畸形、脊髓空洞症、脊柱侧弯等病症的文献[3,11,53]值得关注。

值得强调的是：Chiari畸形常可同时合并或伴发其他脑脊髓先天性疾患，因此在诊断过程中，除了需关注颅颈交界区的多种发育不良状态外，尚应对胸腰椎甚至骶椎伴随的畸形加以全面评价和系统分析，选择针对责任病因的外科手术治疗方法，对提高手术安全性和有效性具有重要意义。

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