

中国脑性瘫痪康复指南(2015):第八部分

中国康复医学会儿童康复专业委员会
中国残疾人康复协会小儿脑性瘫痪康复专业委员会
《中国脑性瘫痪康复指南》编委会

第四章 脑性瘫痪的康复治疗

第四节 药物治疗

近二十年应用于脑瘫患儿的抗痉挛药物主要有:①缓解局灶性痉挛药物:神经肌肉阻滞剂(A型肉毒毒素)和化学去神经支配(苯酚、乙醇);②缓解全面性痉挛药物:口服药物(苯二氮卓类、丹曲林、巴氯芬、替扎尼定)和巴氯芬鞘内注射。此外,脑瘫患儿因负重、营养和抗惊厥药应用等因素,常出现低骨密度和骨质疏松,易造成骨折,故临床上使用维生素D、钙补充剂和双磷酸盐等相应药物以改善脑瘫患儿骨密度^[1]。

一、A型肉毒毒素

证据

1.缓解下肢痉挛

剂量对照研究显示步态动力学与运动学有显著的剂量-效应相关性,高剂量比低剂量在站立位或摆动时有更显著的踝关节背屈活动,且效用更持久^[2](1个I级证据)。随机对照研究采用粗大运动功能测试量表评估结果显示A型肉毒毒素明显提高下肢功能和改善步态^[3-4](1个I级证据、1个II级证据)。病例对照研究中,接受A型肉毒毒素治疗后12周,应用医师评价量表对患儿步态进行分析,步态改善明显,评分高于正常对照组的2倍^[5](1个I级证据)。使用A型肉毒毒素与安慰剂对照研究证明,治疗组的下肢功能较安慰剂组有显著改善^[5-7](3个I级证据)。

2.缓解上肢痉挛

研究表明注射A型肉毒毒素短期内能明显改善上肢功能,但对长期上肢运动功能改善不明显^[8-11](4个I级证据)。联合OT则效果更好,能提升肘部及拇指主动伸展,以及降低腕部、肘部的肌张力,但手抓握的功能测试只有轻微提升,捡硬币测试显示手功能无明显改善^[12](1个II级证据)。随机对照试验观察上肢重复注射A型肉毒毒素联合OT与单用OT治疗结果发现,该方法使痉挛得到持续缓解,父母能感知到患儿的明显进步^[9](1个I级证据)。回顾性研究表明注射A型肉毒毒素的严重不良事件很低^[13](1个I级证据)。

推荐

A型肉毒毒素注射是一种有效、安全的缓解痉挛的治疗技术,缓解下肢痉挛的效果优于缓解上肢痉挛的效果(推荐强度A级)。

二、苯酚、乙醇

证据

乙醇、苯酚局部注射可用于缓解脑瘫患儿的局部痉挛^[14-16](3个IV级证据)。1971年后未见文献报道,但在2014年国际物理医学与康复学会上仍有专家应用乙醇、苯酚和A型肉毒毒素分别局部注射治疗痉挛的报告。

推荐

可配合A型肉毒毒素用于缓解脑瘫患儿的局部痉挛(推荐强度D级)。

三、地西洋

证据

大样本随机对照研究表明地西洋治疗3周后,发现呈剂量依赖性的肌张力减低、被动活动范围增加和自主运动能力提高,但没有明显的功能改善^[17](1个I级证据)。地西洋联合丹曲林的应用与安慰剂组比较痉挛得到显著缓解^[18](1个II级证据)。另有报道地西洋还能提高脑瘫患儿行为与协调能力^[19](1个II级证据)。

推荐

短期应用地西洋可缓解脑瘫患儿的全面痉挛(推荐强度A级),联合丹曲林使用效果明显(推荐强度B级)。

四、丹曲林

证据

丹曲林可改善腱反射和减轻剪刀步^[20](1个II级证据)。有报道丹曲林对痉挛、运动和肌力均无明显影响^[21](1个I级证据);但另一同样剂量(4—12mg/kg·d)的研究表明,丹曲林可减轻痉挛,虽然粗大运动功能无改善,但日常生活活动能力(包括穿衣、饮食方面的协调能力,自主玩耍时的肢体控制,耐力和活动自由度等)有显著提高^[22](1个II级证据)。

推荐

- 1.丹曲林可改善腱反射、剪刀步和日常生活活动能力(推荐强度B级)。
- 2.丹曲林可缓解脑瘫的痉挛,但有争议(推荐强度B级)。

五、巴氯芬

证据

口服巴氯芬的研究结果不太一致,使用剂量为每天10—60mg的一个双盲交叉试验结果显示巴氯芬可减轻痉挛,具体表现为被动关节活动度增大,但对能独立行走的患儿没有明显的功能改善^[23](1个II级证据);另一个双盲安慰剂交叉试验,用同样的剂量和年龄分组,用目的达到量表评估发现结果有改善,但应用改良后的Tardieu量表和儿童生活功能量表(pediatric evaluation of disability inventory,PEDI)评估患儿后发现,痉挛和功能并无明显改善^[24](1个II级证据)。

鞘内注射巴氯芬可长期缓解脑瘫患儿的痉挛和改善运动功能^[25—30](1个III级证据,5个IV级证据)。鞘内注射巴氯芬的副作用有脑脊液漏、导管故障和软组织感染等^[31](1个II级证据)。

推荐

- 1.口服巴氯芬可缓解脑瘫患儿的痉挛和被动关节活动度增大,仍有一些争议(推荐强度B级)。
- 2.鞘内注射巴氯芬可缓解脑瘫患儿的痉挛和改善运动功能,同时需注意预防副作用(推荐强度C级)。

六、替扎尼定

证据

小样本安慰剂对照研究使用剂量0.05mg/kg·d的替扎尼定2周,结果发现痉挛减轻、姿势及腱反射改善,但未做功能评估,亦未发现副作用,肝功能正常,该药被认为有可能用于缓解脑瘫患儿痉挛的治疗^[32](1个II级证据)。

推荐

替扎尼定可减轻痉挛(推荐强度B级)。

七、左乙拉西坦

证据

研究显示2例不随意运动型脑瘫患儿应用左乙拉西坦单一治疗,利用视频和视觉类比重量表评估结果,显示令人印象深刻的平衡控制和精细运动有进步,没有发现副作用,而且治疗效果可以维持26个月以上^[33](1个IV级证据)。

推荐

左乙拉西坦可改善不随意运动型脑瘫患儿平衡控制和精细运动(推荐强度D级)。

八、双磷酸盐类药物、维生素D和钙补充剂

证据

随机、安慰剂对照的临床试验提示氨基二磷酸二钠可以提高脑瘫患儿的骨密度,是一种安全和非常有效的方法^[34](1个I级证据),并可降低骨折风险^[35](1个II级证据)。口服阿仑酸钠 1mg/kg/week 可以治疗脑瘫患儿合并的骨质疏松症,且疗效肯定^[36](1个II级证据)。同时服用抗癫痫药的患儿需要摄入高于正常推荐摄入量的维生素D和钙补充剂,以维持骨密度^[37](1个III级证据)。

推荐

- 1.氨基二磷酸二钠可以提高脑瘫患儿的骨密度(推荐强度A级)。
- 2.口服阿仑酸钠可治疗脑瘫患儿合并骨质疏松症(推荐强度B级)。
- 3.服用抗癫痫药的脑瘫患儿需要摄入高于正常推荐摄入量的维生素D和钙补充剂(推荐强度C级)。

九、神经生长因子

证据

神经生长因子具有促进神经元存活、轴突定向再生、髓鞘生成和促进有效连接,恢复感觉、运动和认知功能。据报道鼠神经生长因子对脑卒中、颅脑损伤、脊髓损伤、周围神经病及周围神经损伤、新生儿缺氧缺血性脑病等有效^[38-40](3个II级证据)。有研究神经生长因子可提高婴幼儿脑瘫的运动和智力发育,以及改善肌张力、姿势异常和反射异常^[41-44](4个IV级证据)。但缺少大样本队列对照研究的循证医学依据。

推荐

神经生长因子可用于缺氧缺血性脑病,脊髓和周围神经损伤等的治疗,应用于脑瘫治疗尚缺少大样本研究的循证医学依据(推荐强度D级)。

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第五节 手术治疗

一、矫形手术

证据

脑瘫有多种矫形手术方法,肌腱延长、肌腱转移、旋转截骨术等是进展性脑瘫肌肉骨骼病变常用的矫形手术方法,选择合适的时机进行矫形手术可以缓解肌肉痉挛、平衡肌力、矫正畸形、调整肢体负重力线、改善运动功能,为康复治疗创造有利条件。

1. 上肢矫形手术 脑瘫上肢矫形手术复杂且具有挑战性,其目的在于恢复手的日常生活活动能力、运动功能,改善外观。脑瘫上肢矫形手术有:拇指内收畸形手术,尺神经运动分支切断术,骨间肌、小指展肌、掌骨骨间肌切断术,腕关节融合术,尺侧腕屈肌转移术,旋前圆肌松解术等。上肢矫形手术疗效均缺乏有效的证据支持^[1-3](3个Ⅳ级证据)。

2. 脊柱矫形手术 脑瘫的脊柱侧凸较为复杂,确定是否手术及采取哪种手术存在一定的困难。当患者脊柱侧凸 Cobb 角达 40°以上时,可以考虑手术治疗,多采用脊柱融合术。患有神经性脊柱侧凸的脑瘫可行脊柱融合术^[4-7](2个Ⅲ级证据,2个Ⅳ级证据),矫正脊柱畸形,纠正骨盆倾斜,手术时建议应用颈/脑干躯体感觉诱发电位进行脊髓监测,避免严重并发症。脊柱融合术后可能会引起消化功能障碍、异位骨化,应避免同时进行髋关节和脊柱手术。

3. 下肢矫形手术:脑瘫在发育过程中常出现异常步态,下肢骨与关节可产生各种挛缩畸形,下肢矫形手术主要原则在于矫正力线,平衡肌力。

(1) 关节矫形手术:髋关节脱位在痉挛型脑瘫中较为常见,但目前手术时机、预后等问题上仍未得到统一。综合性手术治疗痉挛性髋关节脱位是有效的,手术方法为股骨旋转截骨术+髋关节切开复位/骨盆截骨术,骨盆截骨术适用于:年龄1—6岁、髋臼指数小于45°、股骨头大小与髋臼基本适应的患儿。软组织松解手术结合股骨旋转截骨术可以矫正脑瘫骨盆旋转,髋臼成形术结合股骨截骨术及软组织松解术可改善脑瘫患儿股骨头畸形,腰大肌和相关软组织松解手术结合股骨近端缩短和 Chiari 截骨术,对半脱位并伴有疼痛的青少年或成年脑瘫有效^[8-15](2个Ⅲ级证据,6个Ⅳ级证据)。髋关节疾患严重的脑瘫可行全髋关节置换术,痉挛型脑瘫伴发髋关节疼痛和脱位的患者,行近端股骨切除关节置换术可能有一定疗效。单侧髋关节手术对严重痉挛型双瘫或四肢瘫患者是相对禁忌的,不适用于6岁以下或髋关节轻度脱位的患者。

痉挛型脑瘫患儿行软组织手术,包括腰大肌切断术、腰肌-股直肌转移、单纯内收肌切断术、内收肌切断术结合闭孔神经切除,可以减少髋脱位发生率,预防痉挛性髋关节脱位^[16-18](1个Ⅱ级证据,1个Ⅲ级证据,1个Ⅳ级证据)。髋关节不稳定的脑瘫不推荐使用软组织松解术。

传统的软组织手术虽然不能降低高肌张力,但可矫正固定性挛缩和畸形。对于髋关节屈曲畸形,常采用髂腰肌松解术、股直肌松解术。髋内收畸形,常采用内收肌切断术或配合闭孔神经前支切断术。软组织松解手术可改善脑瘫患儿的步态,内收短肌、股薄肌移位术可纠正剪刀步态,股直肌转移术和腘绳肌手术可增加步长^[19-21](1个Ⅲ级证据,2个Ⅳ级证据)。

(2) 膝关节矫形手术:股直肌转移术和腘绳肌手术可用于治疗具有移动能力的痉挛型脑瘫,增加患儿站立位膝关节伸直角度、增加步长。僵直步态的脑瘫可行股直肌转移术^[22-23](2个Ⅲ级证据)。当脑瘫膝关节活动范围小于正常80%时,应进行股直肌转移术,不应进行股直肌远端松解术。近端股直肌松解术不能改善痉挛型脑瘫屈髋肌挛缩和步态异常。GMFCS Ⅳ级的脑瘫行股直肌远端转移术会增加术后膝关节屈曲,GMFCS Ⅳ级的脑瘫不宜行股直肌远端转移术。痉挛型脑瘫行腘绳肌内外侧延长术,可改善腘窝角,增加立位膝关节最大伸直角,改善步行能力及运动功能,但膝过伸的风险高于腘绳肌内侧延长术^[24-27](2个Ⅲ级证据,2个Ⅳ级证据)。重度屈膝畸形的痉挛型脑瘫行软组织矫形加 Ilizarov 外固定支架术,可取得较为满意的效果。

(3) 踝关节矫形手术:痉挛型脑瘫马蹄足可行跟腱延长术,矫正畸形,改善痉挛^[28-32](1个Ⅱ级证据,2个Ⅲ级证据,2个Ⅳ级证据)。脑瘫跟腱延长术疗效好,但小腿三头肌力会减弱,需要使用地面反作用力支具。偏瘫、需单侧手术非偏瘫型脑瘫、不需要后期外科手术的脑瘫,跟腱延长术疗效最好。对有固定和动态马蹄内翻足的脑瘫,需要进行腓肠肌筋膜延长术、腓肠肌-比目鱼肌延长术、小腿三头肌延长术。患有痉挛性马蹄内翻足畸形的偏瘫型脑瘫患儿可行胫后肌部分转移、肌腱延长术,8岁以下或不能独立在社区内步行的双瘫和四肢瘫型脑瘫不应进行胫后肌手术。

脑瘫性外翻足,6岁之前一般考虑保守治疗和软组织手术,年长患儿应采用骨性手术来矫正足外翻。关节外距下关节融合术可矫正脑瘫后足外翻^[33-38](3个Ⅲ级证据,3个Ⅳ级证据),改善脑瘫痉挛性扁平外翻足畸形,矫正中足外翻,但不能矫正足外翻畸形合并严重的前足外展畸形,不能矫正前足旋后、跟骨跖屈。改良距下关节融合术对脑瘫患儿背侧距舟关节半脱位有一

定疗效。

(4)一次麻醉下的多部位手术(single-event multilevel surgery, SEMS):SEMS也称为多部位手术或改善步态手术,是指在一次麻醉下矫正多个部位的软组织和骨性畸形。有移动能力的痉挛型脑瘫建议SEMS治疗,改善静态挛缩和膝关节运动功能,提高患儿的运动功能、步态、移动能力、粗大运动功能和生活质量,术后患者家长满意度高^[39-48](5个Ⅱ级证据,2个Ⅲ级证据,3个Ⅳ级证据)。大龄脑瘫患儿行SEMS长期疗效好。步态严重异常的痉挛型双瘫SEMS只能短期改善患儿的步态,很多患者需要进行其他手术治疗。与未行手术患儿相比,行上肢SEMS的脑瘫患儿抓握-伸展能力并没有显著提高^[49](1个Ⅲ级证据)。

推荐

- 1.脊柱融合术是神经肌肉性脊柱侧凸脑瘫的治疗方法(推荐强度C级)。
- 2.综合性手术方案(软组织手术结合股骨旋转截骨术、股骨内翻旋转截骨术结合髋关节切开复位/骨盆截骨术)是痉挛性髋脱位脑瘫患者治疗的一种选择(推荐强度C级)。
- 3.软组织手术是预防痉挛型脑瘫髋关节脱位的一种治疗选择(推荐强度C级)。
- 4.软组织手术是矫正下肢固定性挛缩和畸形、改善异常步态的一种治疗选择(推荐强度C级)。
- 5.股直肌转移术是僵直步态脑瘫治疗的一种选择(推荐强度C级)。
- 6.腓绳肌延长术是改善痉挛型脑瘫膝关节活动度的一种治疗选择(推荐强度C级)。
- 7.跟腱延长术是痉挛性马蹄足畸形脑瘫治疗的一种选择(推荐强度C级)。
- 8.关节外距下关节融合术是足外翻畸形脑瘫治疗的一种选择(推荐强度C级)。
- 9.SEMS是有移动能力的痉挛型脑瘫改善步态的一种选择(推荐强度B级)。

二、脊神经后根切断术

证据

脊神经后根切断术(selective posterior rhizotomy, SPR /selective dorsal rhizotomy, SDR)需根据患儿具体情况、痉挛部位,选择L2—L5、S1后根节段,配合电刺激监测,结合个人经验选择性切断马尾神经,这是决定疗效的重要因素,选择合适的患者对于手术成功至关重要。脊神经后根切断术可有效减轻中度到重度痉挛型脑瘫的痉挛程度,改善功能,提高步行能力,对脑瘫身体机构和功能领域有积极的长期影响^[50-57](2个Ⅰ级证据,5个Ⅲ级证据,1个Ⅳ级证据)。脊神经后根切断术可能对3—8岁、GMFCS Ⅲ—Ⅳ级的脑瘫最有效^[58](1个Ⅰ级证据),但对GMFCSⅡ-Ⅲ级的患儿长期改善作用微弱,对GMFCS Ⅳ-V级的患儿无长期持续改善作用^[59](1个Ⅲ级证据)。痉挛型双瘫、轻度四肢瘫、不能进行巴氯芬鞘内注射或药物治疗无反应的脑瘫可行脊神经后根切断术,需要轮椅移动和智力发育迟缓的痉挛型四肢瘫患儿、10岁以上的脑瘫、肌张力障碍、手足徐动、共济失调的患儿不宜行脊神经后根切断术。选择性颈神经后根切断术可解除上肢痉挛,改善肢体功能,但部分患者术后仍有肘关节屈曲、前臂旋前、腕关节屈曲等畸形。行脊神经后根切断术时应进行术中电生理监测。脊神经后根切断术对脑瘫患儿腰椎稳定性有一定影响,术后会出现腰椎过度前凸、椎骨脱离和脊椎滑脱等脊柱畸形,可有支气管痉挛、肺炎、尿潴留和感觉丧失等并发症^[60-61](2个Ⅲ级证据)。

推荐

脊神经后根切断术是3—8岁、GMFCS Ⅲ—Ⅳ级下肢痉挛型脑瘫治疗的一种选择,但应严格掌握适应证(推荐强度A级)。

三、巴氯芬鞘内注射

证据

巴氯芬鞘内注射(intrathecal baclofen therapy, IBT)对顽固性痉挛型脑瘫有效,可以抗痉挛,改善言语、交流和流涎控制能力,减少排便次数,对疼痛和运动障碍有积极影响,改善步态,改善常规抗痉挛治疗困难的脑瘫患儿坐轮椅时舒适程度,改善护理难度。巴氯芬鞘内注射治疗严重痉挛的患者、常规口服治疗难治的患者、混合型脑瘫患者是安全有效的^[62-67](3个Ⅱ级证据,3个Ⅳ级证据)。巴氯芬鞘内注射治疗产生并发症的发病率为44%、继发感染率为73%,发生并发症并进行手术治疗率为31%。

推荐

巴氯芬鞘内注射是严重痉挛型脑瘫儿童治疗的一种选择(推荐强度C级)。

四、周围神经微创手术

证据

选择性周围神经切断术(胫神经、坐骨神经、肌皮神经、正中神经、尺神经、副神经、颈段和腰骶段脊神经前、后根)是治疗痉挛型脑瘫安全有效的手术方法,可降低肌张力、纠正痉挛性畸形、改善运动功能^[68-69](2个IV级证据)。保守治疗无效的痉挛型脑瘫,选择性周围神经切断术可以缓解痉挛、改善功能^[70-73](4个IV级证据)。选择性胫神经肌支切断术治疗脑瘫痉挛性马蹄内翻足,可降低肌张力。选择性股神经切断术可以改善股四头肌痉挛引起的膝关节僵硬,增加膝关节活动度。周围神经选择性切断术治疗下肢痉挛,部分患者出现肌力下降、肢体麻木。

肩外旋肌选择性神经切断术、C7神经根切断术、C8神经根切断术、背根神经节经皮射频毁损手术等周围神经微创手术的疗效缺乏有效的证据支持^[74](1个IV级证据)。肩外旋肌选择性神经切断术可以缓解脑瘫患儿的肌肉痉挛;C7神经根切断、对侧健康C7神经根转移到患侧臂丛中干可以部分缓解脑瘫屈肌痉挛,增强伸肌力量;C8神经根切断术不能长期缓解脑瘫手部痉挛,痉挛治疗效果差^[75](1个IV级证据)。严重髋屈曲/内收痉挛疼痛的脑瘫行背侧神经节经皮射频毁损手术,可以改善痉挛、疼痛,使护理更为容易^[76](1个IV级证据)。

推荐

选择性周围神经部分切断术是保守治疗无效痉挛型脑瘫的治疗选择(推荐强度D级)。

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